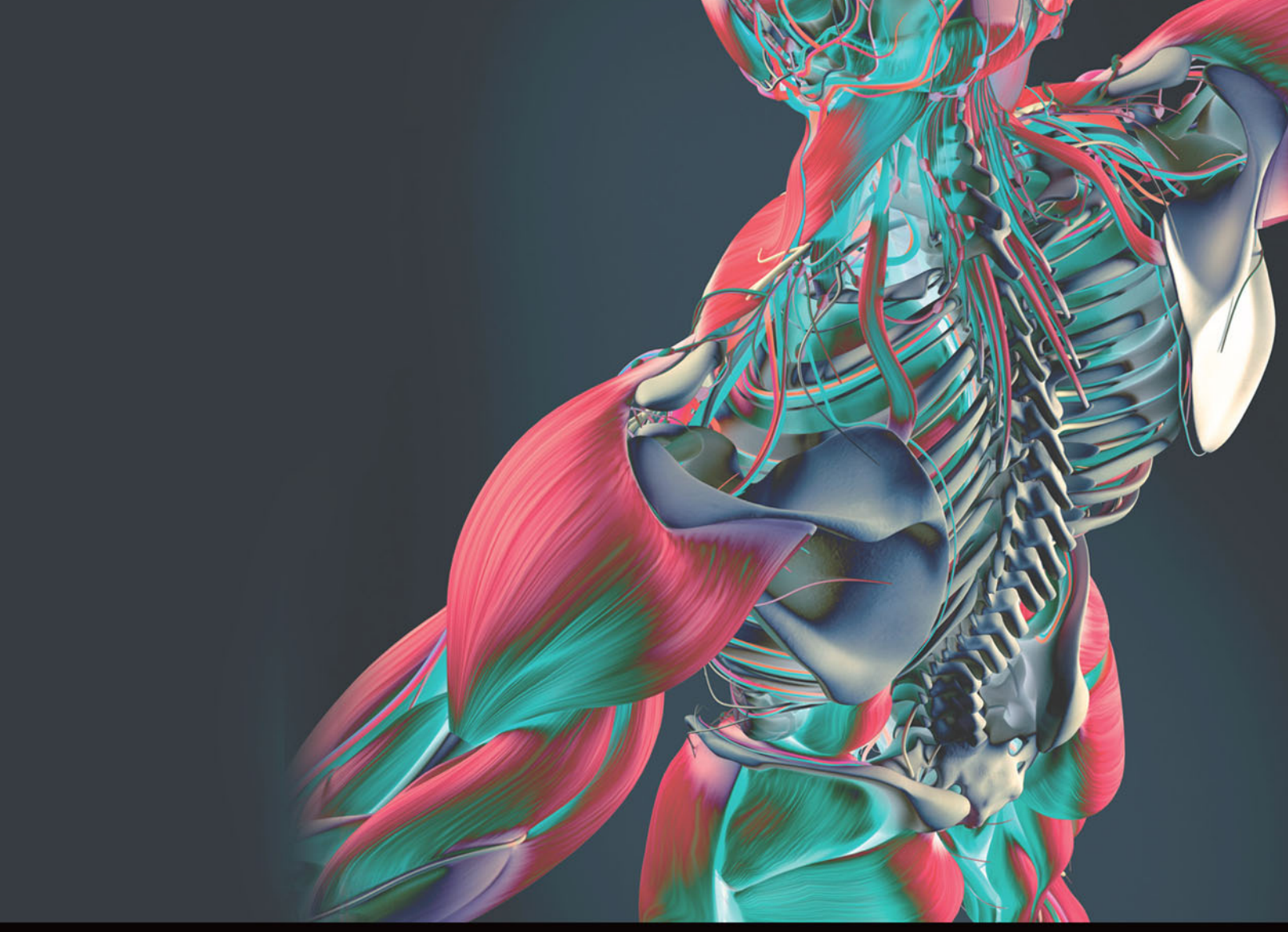

APLEY & SOLOMON'S

System of Orthopaedics and Trauma

Tenth Edition

EDITED BY

Ashley Blom, David Warwick, Michael R. Whitehouse

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CRC Press
Taylor & Francis Group

Apley and Solomon's System of Orthopaedics and Trauma

Alan Graham Apley 1914–1996

Louis Solomon 1928–2014

Inspired teachers, wise mentors and joyful friends

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Apley and Solomon's System of Orthopaedics and Trauma

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CRC Press
Taylor & Francis Group
Boca Raton London New York

CRC Press is an imprint of the
Taylor & Francis Group, an **informa** business

CRC Press
Taylor & Francis Group
6000 Broken Sound Parkway NW, Suite 300
Boca Raton, FL 33487-2742

Printed on acid-free paper

International Standard Book Number-13: 978-1-4987-5167-4 (Pack – Book and eBook)
International Standard Book Number-13: 978-1-4987-5177-3 (Paperback; restricted territorial availability)

Library of Congress Cataloging-in-Publication Data

Names: Blom, Ashley, editor. | Warwick, David, 1962- editor. | Whitehouse, Michael (Michael R.), editor. | Preceded by (work): Solomon, Louis. Apley's system of orthopaedics and fractures.
Title: Apley & Solomon's system of orthopaedics and trauma / [edited by] Ashley Blom, David Warwick, Michael Whitehouse.
Other titles: Apley and Solomon's system of orthopaedics and trauma | System of orthopaedics and trauma.
Description: Tenth edition. | Boca Raton : CRC Press, [2017] | Preceded by Apley's system of orthopaedics and fractures / Louis Solomon, David Warwick, Selvadurai Nayagam. 9th ed. 2010.
Identifiers: LCCN 2016059350 (print) | LCCN 2016059955 (ebook) | ISBN 9781498751674 (hardback bundle : alk. paper) | ISBN 9781498751773 (pbk. : alk. paper) | ISBN 9781498751711 (eBook VitalSource) | ISBN 9781498751704 (eBook PDF).
Subjects: | MESH: Orthopedic Procedures | Musculoskeletal System--injuries | Fracture Fixation—methods.
Classification: LCC RD731 (print) | LCC RD731 (ebook) | NLM WE 168 | DDC 616.7--dc23
LC record available at https://lccn.loc.gov/2016059350

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DEDICATION

To Louis
from your friends and colleagues on behalf of the thousands of patients
who have benefitted from your lifetime's work

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PREFACE

Orthopaedics in a changing world

Since Alan Apley published the first edition of this book the world has changed considerably and so has the practice of orthopaedic surgery. In 1959, hip replacement was rare and had high failure rates, knee replacement and arthroscopy did not exist and fractures were primarily treated in traction.

The last edition of this book commented on the projected impact of the HIV/AIDS epidemic. The epidemic has largely been brought under control, with effective treatment resulting in normal life expectancy for sufferers. However, in untreated individuals, the incidence of secondary infection such as tuberculosis is high and the prognosis is still dire. It is interesting and encouraging to note that both the National Joint Registry for England and Wales and the Malawian Joint Registry have shown that hip replacement is an effective treatment for patients who have multimorbidity which includes AIDS with no increased risk of early postoperative mortality compared with patients who do not have AIDS.

Over the lifetime of this book many treatments have been invented, extensively used, found to be ineffective or suboptimal and subsequently have declined dramatically in popularity. Examples of this include arthroscopic debridement for knee osteoarthritis, metal-on-metal hip replacement and excision arthroplasty of the distal ulna. It is important that we continue to challenge the efficacy of existing and novel treatments. In a world of increasing global need orthopaedics has to be proven to be efficacious and cost-effective.

Since 1959, the world's population has more than doubled to over 7 billion people and has aged considerably. Life expectancy at birth is now 80 years in Europe and 74 years in Asia. There are still marked disparities – for instance Japan has a life expectancy at birth of 83 years compared to 57 years in South Africa – but these differences are narrowing. It is projected that by 2050 4% of the world's population (but 16% of Japan's population) will be over 80 years of age. Between 2010 and 2050 the proportion of the population aged over 65 years will double in most countries, and it is predicted to increase from 5% to 11% in South Africa, 5% to 13% in India and 17% to 36% in Spain.

Orthopaedics remains as relevant a speciality as ever, treating a large burden of the world's morbidity. However, the nature of care has changed, with a much lower burden of chronic musculoskeletal infections today and a steeply rising incidence of joint replacement for primarily degenerative conditions. The World Health Organization estimates that 10% of men and 18% of women aged over 60 years have symptomatic osteoarthritis. Total knee and total hip replacement are now the second and third commonest elective operative procedures performed in developed countries. For example, in England and Wales, which have a combined population of approximately 55 million people, over 170 000 hip and knee replacements are performed annually. The provision of arthroplasty varies greatly, with 226 knee replacements per 100 000 population performed annually in the United States of America compared to only 3 per 100 000 population in neighbouring Mexico. Increasingly the outcomes of common procedures, such as arthroplasty and fractured neck of femur fixation, are being monitored by national registries in a wide range of countries and healthcare settings. It is heartening that even low-income countries such as Malawi have established implant registries which are providing clinically important data. As the prevalence of infectious diseases declines in low-income countries and people live longer, more health resources will be spent on treating long-term conditions of the elderly such as osteoarthritis.

Accidents and emergencies still represent a major healthcare burden. Over 1.25 million people die worldwide annually as a result of road traffic accidents. The majority of these occur in Asia. Millions more are seriously injured. Injuries from road traffic accidents are the third largest cause of morbidity among adult males. Orthopaedic care remains of paramount importance for effectively and quickly returning patients as closely as possible to their pre-injury state and thereby allowing them to participate fully in society.

The provision of health care and resources varies considerably between countries: Greece has 6.3 doctors per 1000 population, South Africa has 0.8 and India only 0.7. While the number of doctors practising in some countries has remained relatively static, in Australia and the United Kingdom there has been an increase of over 50% in the number of registered doctors in the past decade. Part of this is due to migration of doctors, which may exacerbate shortages in low-income countries. More than 40 000 foreign-trained doctors, including an author of this preface, work in the United Kingdom, nearly half of whom come from India and Pakistan. In Israel, New Zealand, Norway and Ireland over a third of practising doctors are foreign-trained. Movement of doctors between countries promotes the spread of ideas and innovation and improves training. However, there is a natural gravitation of expertise towards countries that offer higher remuneration and better working conditions at the expense of low- and middle-income countries. The United States of America spends $8713 per capita on health care, while China spend $649 and India $215.

With rapidly increasing per capita GDP in countries such as China and India, the demographics of health care will change markedly over the next decade. The relative need to treat infection and injury will hopefully decline, but this will inevitably be coupled with an increase in treatments for longer-term musculoskeletal conditions.

Ashley W. Blom
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Bristol and Southampton, 2017

Data are publically available from the OECD at:
http://www.oecd-ilibrary.org/social-issues-migration-health/health-at-a-glance_19991312#

PREFACE TO THE NINTH EDITION

When Alan Apley produced the first edition of his System of Orthopaedics and Fractures 50 years ago he saw it as an aid to accompany the courses that he conducted for aspiring surgeons who were preparing for the FRCS exams. With characteristic humour, he called the book 'a prophylactic against writer's cramp'. Pictures were unnecessary: if you had any sense (and were quick enough to get on the heavily oversubscribed Apley Course), you would be treated to an unforgettable display of clinical signs by one of the most gifted of teachers.

You also learnt how to elicit those signs by using a methodical clinical approach – the Apley System. The Fellowship exam was heavily weighted towards clinical skills. Miss an important sign or stumble over how to examine a knee or a finger and you could fail outright. What Apley taught you was how to order the steps in physical examination in a way that could be applied to every part of the musculoskeletal system. 'Look, Feel, Move' was the mantra. He liked to say that he had a preference for four-letter words. And always in that order! Deviate from the System by grasping a patient's leg before you look at it minutely, or by testing the movements in a joint before you feel its contours and establish the exact site of tenderness and you risked becoming an unwilling participant in a theatrical comedy.

Much has changed since then. With each new edition the System has been expanded to accommodate new tests and physical manoeuvres developed in the tide of super-specialization. Laboratory investigations have become more important and imaging techniques have advanced out of all recognition. Clinical classifications have sprung up and attempts are now made to find a numerical slot for every imaginable fracture. No medical textbook is complete without its 'basic science' component, and advances are so rapid that changes become necessary within the period of writing a single edition. The present volume is no exception: new bits were still being added right up to the time of proofreading.

For all that, we have retained the familiar structure of the Apley System. As in earlier editions, the book is divided into three sections: General Orthopaedics, covering the main types of musculoskeletal disorder; Regional Orthopaedics, where we engage with these disorders in specific parts of the body; and thirdly Fractures and Joint Injuries. In a major departure from previous editions, we have enlisted the help of colleagues who have particular experience of conditions with which we as principal authors are less familiar. Their contributions are gratefully acknowledged. Even here, though, we have sought their permission to 'edit' their material into the Apley mould so that the book still has the sound and 'feel' of a single authorial voice.

For the second edition of the book, in 1963, Apley added a new chapter: 'The Management of Major Accidents'. Typically frank, he described the current arrangements for dealing with serious accidents as 'woefully inadequate' and offered suggestions based on the government's Interim Report on Accident Services in Great Britain and Ireland (1961). There has been a vast improvement since then and the number of road accident deaths today is half of what it was in the 1960s (Department of Transport statistics). So important is this subject that the relevant section has now been rewritten by two highly experienced Emergency and Intensive Care Physicians and is by far the longest chapter in the present edition.

Elsewhere the text has been brought completely up to date and new pictures have been added. In most cases the illustrations appear as composites – a series of images that tell a story rather than a single 'typical' picture at one moment in the development of some disorder. At the beginning of each Regional chapter, in a run of pictures we show the method of examining that region: where to stand, how to confront the patient and where to place our hands. For the experienced reader this may seem like old hat; but then we have designed this book for orthopaedic surgeons of all ages and all levels of experience. We all have something to learn from each other.

As before, operations are described only in outline, emphasizing the principles that govern the choice of treatment, the indications for surgery, the design of the operation, its known complications and the likely outcome. Technical procedures are learnt in

simulation courses and, ultimately, in the operating theatre. Written instructions can only ever be a guide. Drawings are usually too idealized and 'in theatre' photographs are usually intelligible only to someone who has already performed that operation. Textbooks that grapple with these impediments tend to run to several volumes.

The emphasis throughout is on clinical orthopaedics. We acknowledge the value of a more academic approach that starts with embryology, anatomy, biomechanics, molecular biology, physiology and pathology before introducing any patient to the reader. Instead we have chosen to present these 'basic' subjects in small portions where they are relevant to the clinical disorder under discussion: bone growth and metabolism in the chapter on metabolic bone disease, genetics in the chapter on osteodystrophies, and so forth.

In the preface to the last edition we admitted our doubts about the value of exhaustive lists of references at the end of each chapter. We are even more divided about this now, what with the plethora of 'search engines' that have come to dominate the internet. We can merely bow our heads and say we still have those doubts and have given references only where it seems appropriate to acknowledge where an old idea started or where something new is being said that might at first sight be questioned.

More than ever we are aware that there is a dwindling number of orthopaedic surgeons who grew up in the Apley era, even fewer who experienced his thrilling teaching displays, and fewer still who worked with him. Wherever they are, we trust that they will recognize the Apley flavour in this new edition. Our chief concern, however, is for the new readers who – we hope – will glean something that helps them become the next generation of teachers and mentors.

LS
SN
DJW

ACKNOWLEDGEMENTS

This textbook is an iterative process and for this current edition new authors have been asked to revise and refresh the existing text. The editors and new authors thoroughly acknowledge the contribution of those who have gone before them, much of whose work remains in this updated text.

Chapter 2, *Infection*, contains some material from 'Infection' by Louis Solomon, H. Srinivasan, Surendar Tuli & Shunmugam Govender. The material has been revised and updated by the current author.

Chapter 4, *Crystal deposition disorders*, contains some material from 'Crystal deposition disorders' by Louis Solomon. The material has been revised and updated by the current authors.

Chapter 5, *Osteoarthritis*, contains some material from 'Osteoarthritis and related disorders' by Louis Solomon. The material has been revised and updated by the current authors.

Chapter 6, *Osteonecrosis and osteochondritis*, contains some material from 'Osteonecrosis and osteochondritis' by Louis Solomon. The material has been revised and updated by the current authors.

Chapter 7, *Metabolic and endocrine bone disorders*, contains some material from 'Metabolic and endocrine bone disorders' by Louis Solomon. The material has been revised and updated by the current authors.

Chapter 8, *Genetic disorders, skeletal dysplasias and malformations*, contains some material from 'Genetic disorders, skeletal dysplasias and malformations' by Louis Solomon & Deborah Eastwood. The material has been revised and updated by the current authors.

Chapter 9, *Tumours*, contains some material from 'Tumours' by Will Aston, Timothy Briggs & Louis Solomon. The material has been revised and updated by the current authors.

Chapter 10, *Neuromuscular disorders*, contains some material from 'Neuromuscular disorders' by Deborah Eastwood, Thomas Staunton & Louis Solomon. The material has been revised and updated by the current author.

Chapter 11, *Peripheral nerve disorders*, contains some material from 'Peripheral nerve injuries' by David Warwick, H. Srinivasan & Louis Solomon. The material has been revised and updated by the new contributor Michael Fox.

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Chapter 18, *The back*, contains some material from 'The back' by Stephen Eisenstein, Surendar Tuli & Shunmugam Govender. The material has been revised and updated by the current authors.

Chapter 19, *The hip*, contains some material from 'The hip' by Louis Solomon, Reinhold Ganz, Michael Leunig, Fergal Monsell & Ian Learmonth. The material has been revised and updated by the current authors.

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Chapter 26 The wrist Contains some material from 'Injuries of the Forearm and Wrist' by David Warwick with updates from the new contributors Adam Watts, Mike Uglow and Joanna Thomas.

Chapter 28, *Injuries of the spine*, contains some material from 'Injuries of the spine' by Stephen Eisenstein & Wagih El Masry. The material has been revised and updated by the current authors.

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Chapter 31, *Injuries of the knee and leg*, contains some material from 'Injuries of the knee and leg' by Selvadurai Nayagam. The material has been revised and updated by the current author.

LIST OF ABBREVIATIONS USED

AAS	atlantoaxial subluxation
ABC	aneurysmal bone cyst
ABPI	ankle brachial pressure index
ACA	angulation correction axis
ACDF	anterior cervical discectomy and fusion
ACE	angiotensin-converting enzyme
ACEI	angiotensin-converting enzyme inhibitor
ACL	anterior cruciate ligament
ACLR	anterior cruciate ligament reconstruction
ACPA	anti-citrullinated peptide antibodies
ACTH	adrenocorticotropic hormone
ADH	antidiuretic hormone
ADHD	attention deficit hyperactivity disorder
ADHR	autosomal dominant hypophosphataemic rickets
ADI	atlantodental interval
ADL	activity of daily living
AFO	ankle–foot orthosis
AFP	alpha-fetoprotein
AIDP	acute inflammatory demyelinating polyneuropathy
AIDS	acquired immune deficiency syndrome
AJCC	American Joint Committee on Cancer
AL	anterolateral
ALI	acute lung injury
ALIF	anterior lumbar interbody fusion
ALP	alkaline phosphatase
ALS	amyotrophic lateral sclerosis
AM	anteromedial
AMC	arthrogryposis multiplex congenita
ANA	antinuclear antibodies
anti-CCP	anti-cyclic citrullinated peptide antibodies
AO/ASIF	Arbeitsgemeinschaft für Osteosynthesefragen/Association for the Study of Internal Fixation
AP	anteroposterior
APACHE	Acute Physiology and Chronic Health Evaluation (model)
APC	antigen-presenting cell *and* anteroposterior compression (injuries)
ARCO	Association Research Circulation Osseous
ARDS	acute respiratory distress syndrome
ARHR	autosomal recessive hypophosphatemic rickets
ARM	awareness, recognition, management
ARMD	adverse reaction to metal debris
AS	ankylosing spondylitis
ASCT	autologous stem-cell transplantation
ASIS	anterior superior iliac spine
ATFL	anterior talofibular ligament
ATLS	Advanced Trauma Life Support
AUSCAN	Australian–Canadian Hand Osteoarthritis Index
AVN	avascular necrosis
AVPU	aware, verbally responsive, pain responsive, and unresponsive
BAPRAS	British Association of Plastic, Reconstructive and Aesthetic Surgeons
BASICS	British Association for Immediate Care
BCIS	bone cement implantation syndrome
BCP	bicalcium phosphate
BMD	bone mineral density
BMI	body mass index
BMP	bone morphogenetic protein
BOA	British Orthopaedic Association
BOAST	BOA Standards for Trauma
BSA	body surface area
BSR	British Society for Rheumatology
BUN	blood urea nitrogen
BVM	bag–valve–mask
CaSR	calcium-sensing receptor
C-A-T™	Combat Application Tourniquet
CC	cartilage calcification
CCP	cyclic citrullinated peptide
CDH	congenital dislocation of the hip
CDR	cervical disc replacement
4CF	four-corner fusion
CIMT	constraint-induced movement therapy
CKD-MBD	chronic kidney disease mineral bone disorder
CMAP	compound muscle action potential

CMC	carpometacarpal
CMI	cell-mediated immunity
CNS	central nervous system
COC	ceramic on ceramic (THA bearing)
COMP	cartilage oligomeric matrix protein
COP	ceramic on polyethylene (THA bearing)
CORA	centre of rotation of angulation
COX-2	cyclooxygenase-2
CPM	continuous passive motion
CPPD	calcium pyrophosphate dihydrate
CR	cruciate retaining
CRP	C-reactive protein
CRPS	complex regional pain syndrome
CSF	cerebrospinal fluid
CT	computed tomography
CTX	serum type I collagen C-terminal cross-linking telopeptide
CVP	central venous pressure
DDD	degenerative disc disease
DDH	developmental dysplasia of the hip
DIC	disseminated intravascular coagulation
DIP(J)	distal interphalangeal (joint)
DISH	diffuse idiopathic skeletal hyperostosis
DISI	dorsal intercalated segment instability
DLC	*discoligamentous complex*
DLIF	direct lateral interbody fusion
DMARDs	disease-modifying antirheumatic drugs
DMD	Duchenne muscular dystrophy
DNA	deoxyribonucleic acid
DRUJ	distal radioulnar joint
DTH	delayed type hypersensitivity
DVT	deep vein thrombosis
DXA	dual-energy X-ray absorptiometry
ECRB	extensor carpi radialis brevis
ECRL	extensor carpi radialis longus
ECU	extensor carpi ulnaris
EDF	elongation–derotation–flexion
EEG	electroencephalography
eFAST	extended focused assessment sonography in trauma
eGFR	estimated glomerular filtration rate
EMG	electromyography
EMS	emergency medical service
EMT	emergency medical technician
ENL	erythema nodosum leprosum
ENT	ear, nose and throat
EPL	extensor pollicis longus
ESR	erythrocyte sedimentation rate
ETA	estimated time of arrival
$EtCO_2$	end-tidal carbon dioxide
EULAR	European League Against Rheumatism
FAB	foot abduction brace
FABER	Flexion, ABduction, and External Rotation test
FABS	flexion, abduction, supination
FAI	femoroacetabular impingement
FAST	focused assessment sonography in trauma
FBC	full blood count
FDP	flexor digitorum profundus
FDS	flexor digitorum superficialis
FFF-STA	Flat foot associated with a short tendo Achilles
FFO	functional foot orthoses
FGF	fibroblast growth factor
FGFR	fibroblast growth receptor
FHH	familial hypocalciuric hypercalcaemia
FHON	femoral head osteonecrosis
FISH	fluorescence in situ hybridization
FLS	Fracture Liaison Services
fMRI	functional magnetic resonance imaging
FMS	fibromyalgia syndrome
FNCLCC	Federation Nationale des Centres de Lutte Contre le Cancer
FPB	flexor pollicis brevis
FPE	fatal pulmonary embolism
FPL	flexor pollicis longus
GABA	gamma-aminobutryic acid
GAGs	glycosaminoglycans
GCS	Glasgow Coma Scale
GCT	giant cell tumour
GCTTS	giant cell tumour of tendon sheath
GMFCS	gross motor function classification system
GPI	general paralysis of the insane
GGT	gamma-glutamyl transferase
GH	*growth hormone*
GRF	ground reaction force
HA	hydroxyapatite
HEMS	helicopter emergency medical service
HHR	humeral head replacement
HIE	hypoxic–ischaemic encephalopathy
HIV	human immunodeficiency virus
HLA	human leucocyte antigen
HMSN	hereditary motor and sensory neuropathy
HNPP	hereditary neuropathy with liability to pressure palsies
HO	heterotopic ossification
HOOS	Hip Dysfunction and Osteoarthritis Outcome Score
HR	hip resurfacing
HRT	hormone replacement therapy
IASP	International Association for the Study of Pain
ICF	International Classification of Functioning, Disability and Health
ICP	intracerebral pressure
ICS	intercostal space
ICU	intensive care unit

IDH isocitrate dehydrogenase
IFSSH International Federation of Societies for Surgery of the Hand
IGRA interferon-gamma release assay
IL interleukin
IM intramuscular
IMRT intensity-modulated radiotherapy
INR international normalized ratio
IP(J) interphalangeal joint
IRIS immune reconstitution inflammatory syndrome
IRMER Ionising Radiation Medical Exposure Regulations
ISS injury severity score
ITB intrathecal baclofen
IV intervertebral *and* intravenous
IVF *in vitro* fertilization
IVH intraventricular haemorrhage
JIA juvenile idiopathic arthritis
JOAMEQ Japanese Orthopaedic Association Cervical Myelopathy Evaluation Questionnaire
KAFO knee–ankle–foot orthosis
KOOS Knee Dysfunction and Osteoarthritis Outcome Score
LBP lower back pain
LC lateral compression
LCH Langerhans cell histiocytosis
LCL lateral collateral ligament
LCPD Legg–Calvé–Perthes disease
LDH lactate dehydrogenase
LHB long head of biceps
LLD leg length discrepancy
LMA laryngeal mask airway
LMN lower motor neuron
LMWH low molecular weight heparin
MAP mean arterial pressure
MARS metal artifact reduction sequences (MRI)
MB multibacillary
MCL medial collateral ligament
MCP(J) metacarpophalangeal (joint)
M-CSF macrophage colony-stimulating factor
MCV mean corpuscular volume
MDM2 murine double minute-2
MED multiple epiphyseal dysplasia
MEN multiple endocrine neoplasia
MGUS monoclonal gammopathy of undetermined significance
MHC major histocompatibility complex
MIC minimal inhibitory concentration
MIPO minimally invasive percutaneous osteosynthesis
MND motor neuron disease
MO multiple osteochondromas
MODS multiple organ failure or dysfunction syndrome
MOM metal on metal (THA bearing)
MOP metal on polyethylene (THA bearing)
MP migration percentage
MPFL medial patellofemoral ligament
MPM mortality prediction model
MPNST malignant peripheral nerve sheath tumour
MPS mucopolysaccharidoses
MRC Medical Research Council
MRA magnetic resonance arthrography or angiography
MRI magnetic resonance imaging
MRSA methicillin-resistant *Staphylococcus aureus*
MSSA methicillin-sensitive *Staphylococcus aureus*
MTC Major Trauma Centre
MTP(J) metatarsophalangeal (joint)
NARU National Ambulance Resilience Unit
NCIN *National Cancer Intelligence Network*
NCTH non-compressible torso haemorrhage
NCV nerve conduction velocity
NDI Neck Disability Index
NF neurofibromatosis
NIBP non-invasive blood pressure
NICE National Institute for Health and Care Excellence
NOF non-ossifying fibroma
NP nasopharyngeal
NPS Nail–patella syndrome
NSAIDs non-steroidal anti-inflammatory drugs
OA osteoarthritis
OCD osteochondritis dissecans
OFD osteofibrous dysplasia
OI osteogenesis imperfecta
OMT Oberg, Manske and Tonkin (classification)
ONJ osteonecrosis of the jaw
OP oropharyngeal
OPG osteoprotegerin
OPLL ossification of the posterior longitudinal ligament
P1NP serum type I collagen extension propeptide
PA posteroanterior
PACS Picture Archiving and Communication System
PAFC pulmonary artery flotation catheterization
PAO periacetabular osteotomy
PAOP pulmonary artery occlusion pressure
PB paucibacillary
PCA patient-controlled analgesia
PCL posterior cruciate ligament
PCR polymerase chain reaction
PD proton density
PDB Paget's disease of bone

PE pulmonary embolism
PEA pulseless electrical activity
PEEP positive end-expiratory pressure
PET positron emission tomography
PH Pavlik harness
PHEM pre-hospital emergency medicine
Pi inorganic phosphate
PIP(J) proximal interphalangeal (joint)
PJI periprosthetic infection
PL posterolateral
PLC posterior ligamentous complex *and* posterolateral corner
PLL posterior longitudinal ligament
PLRI posterolateral rotatory instability
PM posteromedial
PMMA polymethylmethacrylate
PNS peripheral nervous system
PPE personal protective equipment
PPS post-polio syndrome
pQCT peripheral quantitative computer tomography
PRC proximal row carpectomy
PRICE protection, rest, ice, compression and elevation
PRICER protection, rest, ice, compression, elevation and rehabilitation
PRP platelet rich plasma
PS posterior stabilized
PSA prostate-specific antigen
PsA psoriatic arthritis
PTH parathyroid hormone
PTHrP parathyroid hormone-related peptide
PTS post-thrombotic syndrome
PVL periventricular leucomalacia
PVNS pigmented villonodular synovitis
QCT quantitative computed tomography
QoL quality of life
QUS quantitative ultrasonometry
RA radiographic absorptiometry *and* rheumatoid arthritis
RANKL receptor activator of nuclear factor-$\acute{\kappa}\beta$ ligand
REBOA resuscitative endovascular balloon occlusion of the aorta
RF rheumatoid factor
RGO reciprocating gait orthoses
RICE rest, ice, compression and elevation
RNA ribonucleic acid
RR reversal reaction
RSD reflex sympathetic dystrophy
RSI rapid sequence induction
RTC road traffic crash
SAC space available for spinal cord
SACE serum angiotensin converting enzyme
SAMU Services de l'Aide Medical Urgente
SAPHO synovitis, acne, pustulosis, hyperostosis and osteitis
SAPS simplified acute physiology score
SAS subaxial subluxation
SBC simple bone cyst
SCFE slipped capital femoral epiphysis
SCI spinal cord injury
SCIWORA spinal cord injury without obvious radiographic abnormality
SCM sternocleidomastoid muscle
SDD selective digestive tract decontamination
SDR selective dorsal rhizotomy
SE spin echo
SED spondyloepiphyseal dysplasia
SEMLS single event multi-level surgery
SERM selective oestrogen receptor modulator
SIJ sacroiliac joint
SIRS systemic inflammatory response
SLAP superior labrum, anterior and posterior (tear)
SLE systemic lupus erythematosus
SLIC Subaxial Cervical Spine Injury Classification
SMR standardized mortality ratio
SMUR Services Mobile d'Urgence et de Reamination
SNAP sensory nerve action potential
SNPs single nucleotide polymorphisms
SOFA sequential organ failure assessment
SONK 'spontaneous' osteonecrosis of the knee
SOP standard operating procedure
SPA spondyloarthropathy
SpA spondyloarthritis
SPECT single photon emission computed tomography
SPORT Spine Patient Outcomes Research Trial
STIR short-tau inversion recovery
STT scaphoid–trapezium–trapezoid arthritis *and* soft-tissue tumour
SUA serum uric acid
SUFE slipped upper femoral epiphysis
TAR thrombocytopaenia with absent radius syndrome
TARN Trauma Audit and Research Network
TB tuberculosis
TBI total body involvement
TBS Trabecular Bone Score
^{99m}Tc-HDP technetium(^{99m}Tc)-labelled hydroxymethylene diphosphonate
^{99m}Tc-MDP technetium(^{99m}Tc)-labelled methyl diphosphonate
TDR total disc replacement
TE time to echo
TFCC triangular fibrocartilage complex
THA total hip arthroplasty
TIP terminal interphalangeal (joint)

TISS therapeutic intervention scoring system
TKR total knee replacement
TLIF transforaminal lumbar interbody fusion
TMT tarsometatarsal
TNF tumour necrosis factor
TNM tumour–node–metastasis
TOE transoesophageal echocardiogram
TSF Taylor spatial frame
TSH thyroid-stimulating hormone
TSR total shoulder replacement
TU Trauma Unit
UCP unilateral cerebral palsy
UFD unifacet dislocation
UHMWPE ultra-high molecular weight polyethylene
UICC Union for International Cancer Control
ULT urate-lowering therapy
UMN upper motor neuron
UPS undifferentiated pleomorphic sarcoma
US ultrasound
VAC vacuum-assisted closure
VACTERLS refers to the systems involved and the defects identified: *v*ertebral, *a*nal, *c*ardiac, *t*racheal, *e*sophageal, *r*enal, *l*imb and *s*ingle umbilical artery
VCR vertebral column resection
VCT voluntary counselling and testing
VFA Vertebral Fracture Assessment
VISI volar intercalated segment instability
VMO vastus medialis oblique
VP ventriculoperitoneal
VQ ventilation–perfusion
VS vertical shear *and* vertical subluxation
VTE venous thromboembolism
WALANT wide awake local anaesthetic no tourniquet
WBC white blood cell
WHO World Health Organization
XLH sex-linked hypophosphataemic rickets

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Section 1

General Orthopaedics

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Diagnosis in orthopaedics

1

Louis Solomon & Charles Wakeley

Orthopaedics is concerned with bones, joints, muscles, tendons and nerves – the skeletal system and all that makes it move. Conditions that affect these structures fall into seven easily remembered pairs:

1 Congenital and developmental abnormalities
2 Infection and inflammation
3 Arthritis and rheumatic disorders
4 Metabolic and endocrine disorders
5 Tumours and lesions that mimic them
6 Neurological disorders and muscle weakness
7 Injury and mechanical derangement

Diagnosis in orthopaedics, as in all of medicine, is the identification of disease. It begins from the very first encounter with the patient and is gradually modified and fine-tuned until we have a picture, not only of a pathological process but also of the functional loss and the disability that goes with it. Understanding evolves from the systematic gathering of information from the history, the physical examination, tissue and organ imaging and special investigations. Systematic, but never mechanical; behind the enquiring mind there should also be what D. H. Lawrence has called 'the intelligent heart'. It must never be forgotten that the patient has a unique personality, a job and hobbies, a family and a home; all have a bearing upon, and are in turn affected by, the disorder and its treatment.

HISTORY

'Taking a history' is a misnomer. The patient tells a story; it is we the listeners who construct a history. The story may be maddeningly disorganized; the history has to be systematic. Carefully and patiently compiled, it can be every bit as informative as examination or laboratory tests.

As we record it, certain key words and phrases will inevitably stand out: injury, pain, stiffness, swelling, deformity, instability, weakness, altered sensibility and loss of function or inability to do certain things that were easily accomplished before.

Each symptom is pursued for more detail: we need to know when it began, whether suddenly or gradually, spontaneously or after some specific event; how it has changed or progressed; what makes it worse; what makes it better.

While listening, we consider whether the story fits some pattern that we recognize, for we are already thinking of a diagnosis. Every piece of information should be thought of as part of a larger picture which gradually unfolds in our understanding. The surgeon-philosopher Wilfred Trotter (1870–1939) put it well: 'Disease reveals itself in casual parentheses.'

Symptoms

Pain

Pain is the most common symptom in orthopaedics. It is usually described in metaphors that range from inexpressively bland to unbelievably bizarre – descriptions that tell us more about the patient's state of mind than about the physical disorder. Yet there are clearly differences between the throbbing pain of an abscess and the aching pain of chronic arthritis, between the 'burning pain' of neuralgia and the 'stabbing pain' of a ruptured tendon.

Severity is even more subjective. High and low pain thresholds undoubtedly exist, but pain is as bad as it feels to the patient, and any system of 'pain grading' must take this into account. The main value of estimating severity is in assessing the progress of the disorder or the response to treatment. The commonest method is to invite the patient to mark the severity on an analogue scale of 1–10, with 1 being mild and easily ignored, and 10 being totally unbearable. The problem about this type of grading is that patients who have never experienced very severe pain simply do not know what 8 or 9 or

10 would feel like. The following is suggested as a simpler system:

Grade I (mild) Pain that can easily be ignored
Grade II (moderate) Pain that cannot be ignored, interferes with function and needs attention or treatment from time to time
Grade III (severe) Pain that is present most of the time, demanding constant attention or treatment
Grade IV (excruciating) Totally incapacitating pain

Identifying the site of pain may be equally vague. Yet its precise location is important, and in orthopaedics it is useful to ask the patient to point to – rather than to say – where it hurts. Even then, do not assume that the site of pain is necessarily the site of pathology; 'referred' pain and 'autonomic' pain can be very deceptive.

Referred pain Pain arising in or near the skin is usually localized accurately. Pain arising in deep structures is more diffuse and is sometimes of unexpected distribution; thus, hip disease may manifest with pain in the knee (so might an obturator hernia). This is not because sensory nerves connect the two sites; it is due to inability of the cerebral cortex to differentiate clearly between sensory messages from separate but embryologically related sites. A common example is 'sciatica' – pain at various points in the buttock, thigh and leg, supposedly following the course of the sciatic nerve. Such pain is not necessarily due to pressure on the sciatic nerve or the lumbar nerve roots; it may be 'referred' from any one of a number of structures in the lumbar spine, the pelvis and the posterior capsule of the hip joint. See Figure 1.1.

Autonomic pain We are so accustomed to matching pain with some discrete anatomical structure and its known sensory nerve supply that we are apt to dismiss any pain that does not fit the usual pattern as 'atypical' or 'inappropriate' (i.e. psychologically determined).

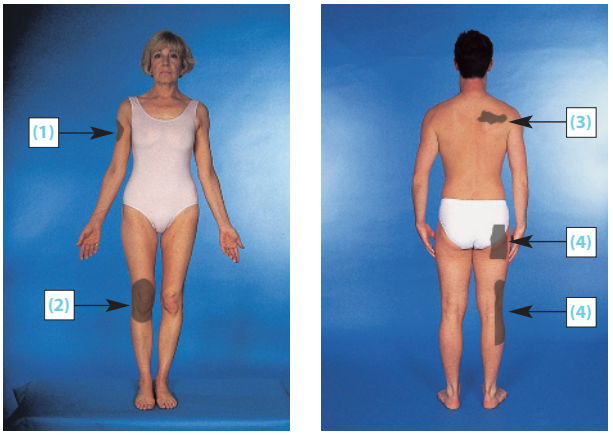


Figure 1.1 Referred pain Common sites of referred pain: (1) from the shoulder; (2) from the hip; (3) from the neck; (4) from the lumbar spine.

But pain can also affect the autonomic nerves that accompany the peripheral blood vessels and this is much more vague, more widespread and often associated with vasomotor and trophic changes. It is poorly understood, often doubted, but nonetheless real.

Stiffness

Stiffness may be generalized (typically in systemic disorders such as rheumatoid arthritis and ankylosing spondylitis) or localized to a particular joint. Patients often have difficulty in distinguishing localized stiffness from painful movement; limitation of movement should never be assumed until verified by examination.

Ask when it occurs: regular early morning stiffness of many joints is one of the cardinal symptoms of rheumatoid arthritis, whereas transient stiffness of one or two joints after periods of inactivity is typical of osteoarthritis.

Locking 'Locking' is the term applied to the sudden inability to complete a particular movement. It suggests a mechanical block – for example, due to a loose body or a torn meniscus becoming trapped between the articular surfaces of the knee. Unfortunately, patients tend to use the term for any painful limitation of movement; much more reliable is a history of 'unlocking', when the offending body slips out of the way.

Swelling

Swelling may be in the soft tissues, the joint or the bone; to the patient they are all the same. It is important to establish whether it followed an injury, whether it appeared rapidly (think of a haematoma or a haemarthrosis) or slowly (due to inflammation, a joint effusion, infection or a tumour), whether it is painful (suggestive of acute inflammation, infection or a tumour), whether it is constant or comes and goes, and whether it is increasing in size.

Deformity

The common deformities are described by patients in terms such as round shoulders, spinal curvature, knock knees, bow legs, pigeon toes and flat feet. Deformity of a single bone or joint is less easily described and the patient may simply declare that the limb is 'crooked'.

Some 'deformities' are merely variations of the normal (e.g. short stature or wide hips); others disappear spontaneously with growth (e.g. flat feet or bandy legs in an infant). However, if the deformity is progressive, or if it affects only one side of the body while the opposite joint or limb is normal, it may be serious (Figure 1.2).

Weakness

Generalized weakness is a feature of all chronic illness, and any prolonged joint dysfunction will inevitably

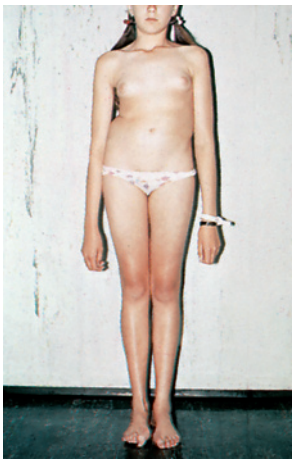



Figure 1.2 Deformity This young girl complained of a prominent right hip; the real deformity was scoliosis.

lead to weakness of the associated muscles. However, pure muscular weakness – especially if it is confined to one limb or to a single muscle group – is more specific and suggests some neurological or muscle disorder. Patients sometimes say that the limb is 'dead' when it is actually weak, and this can be a source of confusion. Questions should be framed to discover precisely which movements are affected, for this may give important clues, if not to the exact diagnosis at least to the site of the lesion.

Instability

The patient may complain that the joint 'gives way' or 'jumps out of place'. If this happens repeatedly, it suggests abnormal joint laxity, capsular or ligamentous deficiency, or some type of internal derangement such as a torn meniscus or a loose body in the joint. If there is a history of injury, its precise nature is important.

Change in sensibility

Tingling or numbness signifies interference with nerve function – pressure from a neighbouring structure (e.g. a prolapsed intervertebral disc), local ischaemia (e.g. nerve entrapment in a fibro-osseous tunnel) or a peripheral neuropathy. It is important to establish its exact distribution; from this we can tell whether the fault lies in a peripheral nerve or in a nerve root. We should also ask what makes it worse or better; a change in posture might be the trigger, thus focusing attention on a particular site.

Loss of function

Functional disability is more than the sum of individual symptoms and its expression depends upon the needs of that particular patient. The patient may say, 'I can't stand for long' rather than 'I have backache'; or 'I can't put my socks on' rather than 'My hip is stiff.' Moreover, what to one patient is merely inconvenient may, to another, be incapacitating. Thus a lawyer or a teacher may readily tolerate a stiff knee provided it is painless, but to a plumber or a parson the same disorder might spell economic or spiritual disaster. One question should elicit the important information: 'What can't you do now that you used to be able to do?'

Past history

Patients often forget to mention previous illnesses or accidents, or they may simply not appreciate their relevance to the present complaint. They should be asked specifically about childhood disorders, periods of incapacity and old injuries. A 'twisted ankle' many years ago may be the clue to the onset of osteoarthritis in what is otherwise an unusual site for this condition. Gastrointestinal disease, which in the patient's mind has nothing to do with bones, may be important in the later development of ankylosing spondylitis or osteoporosis. Similarly, certain rheumatic disorders may be suggested by a history of conjunctivitis, iritis, psoriasis or urogenital disease. Metastatic bone disease may erupt many years after a mastectomy for breast cancer. Patients should also be asked about previous medication: many drugs, and especially corticosteroids, have long-term effects on bone. Alcohol and drug abuse are important, and we must not be afraid to ask about them.

Family history

Patients often wonder (and worry) about inheriting a disease or passing it on to their children. To the doctor, information about musculoskeletal disorders in the patient's family may help with both diagnosis and counselling. When dealing with a suspected case of bone or joint infection, ask about communicable diseases, such as tuberculosis or sexually transmitted disease, in other members of the family.

Social background

No history is complete without enquiry about the patient's background. There are the obvious things such as the level of care and nutrition in children; dietary constraints which may cause specific deficiencies; and, in certain cases, questions about smoking habits, alcohol consumption and drug abuse, all of which call for a special degree of tact and non-judgemental enquiry.

Find out details about the patient's work practices, travel and recreation: could the disorder be due to

a particular repetitive activity in the home, at work or on the sports field? Is the patient subject to any unusual occupational strain? Has he or she travelled to another country where tuberculosis is common?

Finally, it is important to assess the patient's home circumstances and the level of support by family and friends. This will help to answer the question: 'What has the patient lost and what is he or she hoping to regain?'

EXAMINATION

In *A Case of Identity*, Sherlock Holmes has the following conversation with Dr Watson.

Watson: You appeared to read a good deal upon [your client] which was quite invisible to me.

Holmes: Not invisible but unnoticed, Watson.

Some disorders can be diagnosed at a glance: who would mistake the facial appearance of acromegaly or the hand deformities of rheumatoid arthritis for anything else? Nevertheless, even in these cases systematic examination is rewarding: it provides information about the patient's particular disability, as distinct from the clinicopathological diagnosis; it keeps reinforcing good habits; and, never to be forgotten, it lets the patient know that he or she has been thoroughly attended to.

The examination actually begins from the moment we set eyes on the patient. We observe his or her general appearance, posture and gait. Can you spot any distinctive feature: Knock-knees? Spinal curvature? A short limb? A paralysed arm? Does he or she appear to be in pain? Do their movements look natural? Do they walk with a limp, or use a stick? A telltale gait may suggest a painful hip, an unstable knee or a foot-drop. The clues are endless and the game is played by everyone (qualified or lay) at each new encounter throughout life. In the clinical setting the assessment needs to be more focused.

When we proceed to the structured examination, the patient must be suitably undressed; no mere rolling up of a trouser leg is sufficient. If one limb is affected, both must be exposed so that they can be compared.

We examine the good limb (for comparison), then the bad. There is a great temptation to rush in with both hands – a temptation that must be resisted. Only by proceeding in a purposeful, orderly way can we avoid missing important signs.

Alan Apley, who developed and taught the system used here, shied away from using long words where short ones would do the job. (He also used to say, 'I'm neither an inspector nor a manipulator, and I am definitely not a palpator.') Thus the traditional clinical routine, inspection, palpation, manipulation, was replaced by *look*, *feel*, *move*. With time, his teaching has been extended and we now add *test*, to include the special manoeuvres we employ in assessing neurological integrity and complex functional attributes.

Look

Abnormalities are not always obvious at first sight. A systematic, step-by-step process helps to avoid mistakes.

Shape and posture The first things to catch one's attention are the shape and posture of the limb or the body or the entire person who is being examined. Is the patient unusually thin or obese? Does the overall posture look normal? Is the spine straight or unusually curved? Are the shoulders level? Are the limbs normally positioned? It is important to look for deformity in three planes, and always compare the affected part with the normal side. In many joint disorders and in most nerve lesions the limb assumes a characteristic posture. In spinal disorders the entire torso may be deformed. Now look more closely for swelling or wasting – one often enhances the appearance of the other! Or is there a definite lump?

Skin Careful attention is paid to the colour, quality and markings of the skin. Look for bruising, wounds and ulceration. Scars are an informative record of the past – surgical archaeology, so to speak (see Figure 1.3). Colour reflects vascular status or pigmentation – for example, the pallor of ischaemia, the blueness of cyanosis, the redness of inflammation, or the dusky purple of an old bruise. Abnormal creases, unless due to fibrosis, suggest underlying deformity which is not always obvious; tight, shiny skin with no creases is typical of oedema or trophic change.

General survey Attention is initially focused on the symptomatic or most obviously abnormal area, but we

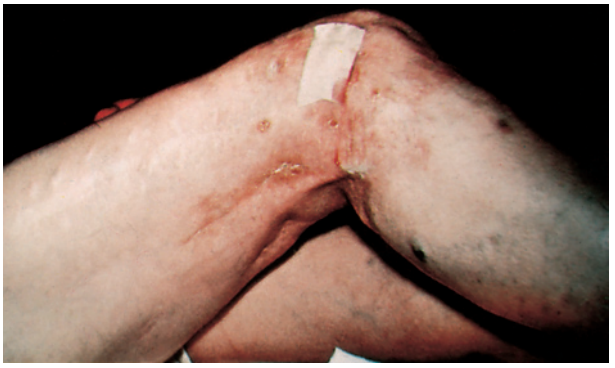


Figure 1.3 Look Scars often give clues to the previous history. The faded scar on this patient's thigh is an old operation wound – internal fixation of a femoral fracture. The other scars are due to postoperative infection; one of the sinuses is still draining.

must also look further afield. The patient complains of the joint that is hurting now, but we may see at a glance that several other joints are affected as well.

Feel

Feeling is exploring, not groping aimlessly. Know your anatomy and you will know where to feel for the landmarks; find the landmarks and you can construct a virtual anatomical picture in your mind's eye.

The skin Is it warm or cold; moist or dry; and is sensation normal?

The soft tissues Can you feel a lump; if so, what are its characteristics? Are the pulses normal?

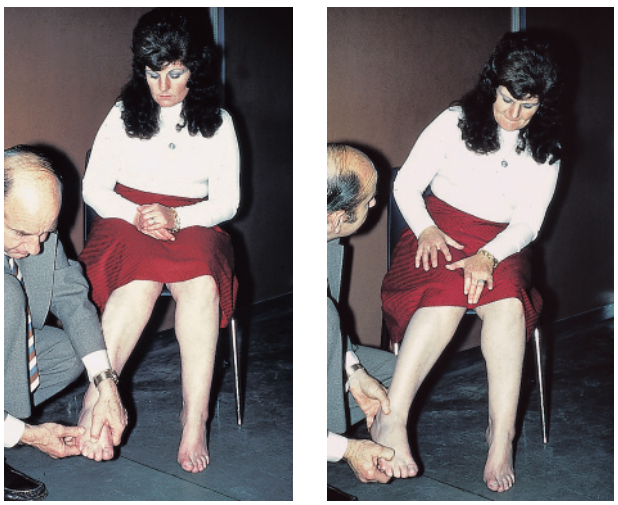
The bones and joints Are the outlines normal? Is the synovium thickened? Is there excessive joint fluid?

Tenderness Once you have a clear idea of the structural features in the affected area, feel gently for tenderness (Figure 1.4). Keep your eyes on the patient's face; a grimace will tell you as much as a grunt. Try to localize any tenderness to a particular structure; if you know precisely *where* the trouble is, you are halfway to knowing *what* it is.

Move

'Movement' covers several different activities: active movement, passive movement, abnormal or unstable movement, and provocative movement (see Figures 1.5 and 1.6).

Active movement Ask the patient to move without your assistance. This will give you an idea of the degree of mobility and whether it is painful or not. Active movement is also used to assess muscle power.



(a) (b)

Figure 1.4 Feeling for tenderness (a) The wrong way – there is no need to look at your fingers, you should know where they are. (b) It is much more informative to look at the patient's face!

Passive movement Here it is the examiner who moves the joint in each anatomical plane. Note whether there is any difference between the range of active and passive movement.

Range of movement is recorded in degrees, starting from zero which, by convention, is the neutral or anatomical position of the joint, and finishing where movement stops, due either to pain or to anatomical limitation. Describing the range of movement is often made to seem difficult. Words such as 'full', 'good', 'limited' and 'poor' are misleading. Always cite the range or span, from start to finish, in degrees. For example, 'knee flexion 0–140 degrees' means that the range of flexion is from zero (the knee absolutely straight) through an arc of 140 degrees (the leg making an acute angle with the thigh). Similarly, 'knee flexion 20–90 degrees' means that flexion begins at 20 degrees (i.e. the joint cannot extend fully) and continues only to 90 degrees.

For accuracy you can measure the range of movement with a goniometer, but with practice you will learn to estimate the angles by eye. Normal ranges of movement are shown in chapters dealing with individual joints. What is important is always to compare the symptomatic with the asymptomatic or normal side.

While testing movement, feel for crepitus. Joint crepitus is usually coarse and fairly diffuse; tenosynovial crepitus is fine and precisely localized to the affected tendon sheath.

Unstable movement This is movement which is inherently unphysiological. You may be able to shift or angulate a joint out of its normal plane of movement, thus demonstrating that the joint is unstable. Such abnormal movement may be obvious (e.g. a wobbly knee); often, though, you have to use special manoeuvres to pick up minor degrees of instability.

Provocative movement One of the most telling clues to diagnosis is reproducing the patient's symptoms by applying a specific, provocative movement. Shoulder pain due to impingement of the subacromial structures may be 'provoked' by moving the joint in a way that is calculated to produce such impingement; the patient recognizes the similarity between this pain and his or her daily symptoms. Likewise, a patient who has had a previous dislocation or subluxation can be vividly reminded of that event by stressing the joint in such a way that it again threatens to dislocate; indeed, merely starting the movement may be so distressing that the patient goes rigid with anxiety at the anticipated result – this is aptly called the *apprehension test.*

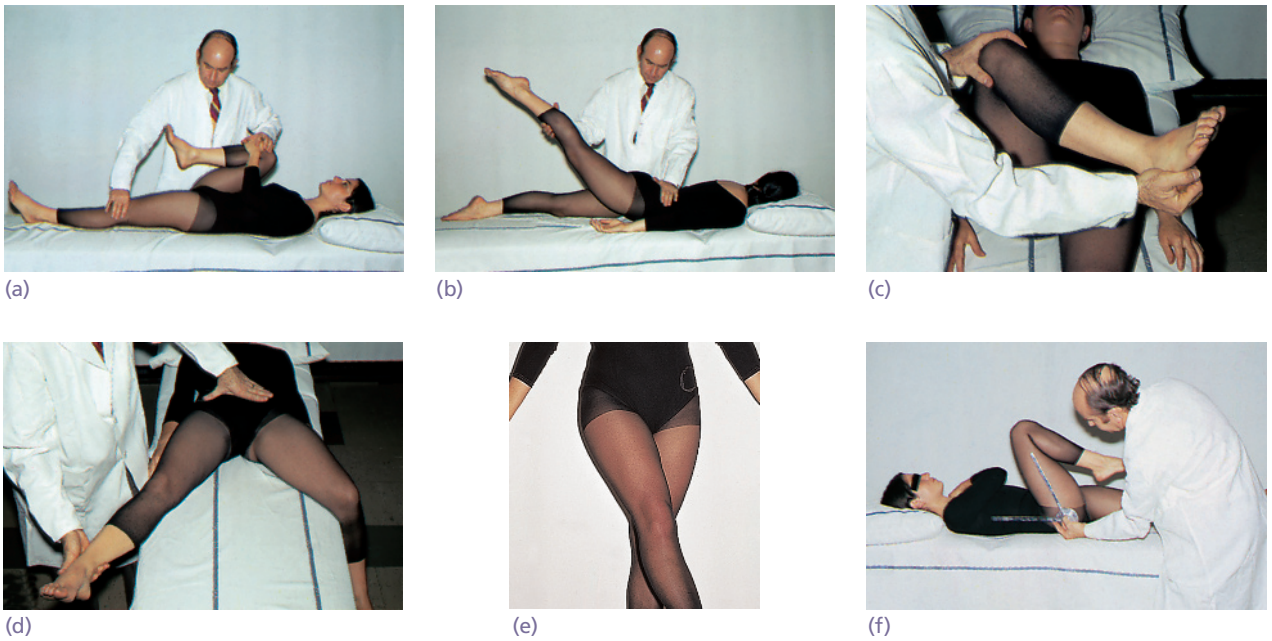


Figure 1.5 Testing for movement (a) Flexion, (b) extension, (c) rotation, (d) abduction, (e) adduction. The range of movement can be estimated by eye or measured accurately using a goniometer (f).

Test

The apprehension test referred to in the previous paragraph is one of several clinical tests that are used to elicit suspected abnormalities: some examples are *Thomas' test* for flexion deformity of the hip, *Trendelenburg's test* for instability of the hip, *McMurray's test* for a torn meniscus of the knee, *Lachman's test* for cruciate ligament instability and various tests for intra-articular fluid. These and others are described in the relevant chapters in Section 2. Tests for muscle tone, motor power, reflexes and various modes of sensibility are part and parcel of neurological examination, which is discussed later in this chapter.

Caveat

We recognize that the sequence set out here may sometimes have to be modified. We may need to 'move' before we 'look': an early scoliotic deformity of the spine often becomes apparent only when the patient bends forwards. The sequence may also have to be altered because a patient is in severe pain or disabled: you would not try to move a limb at all in someone with a suspected fracture when an X-ray can provide the answer. When examining a child, you may have to take your chances with look or feel or move whenever you can!

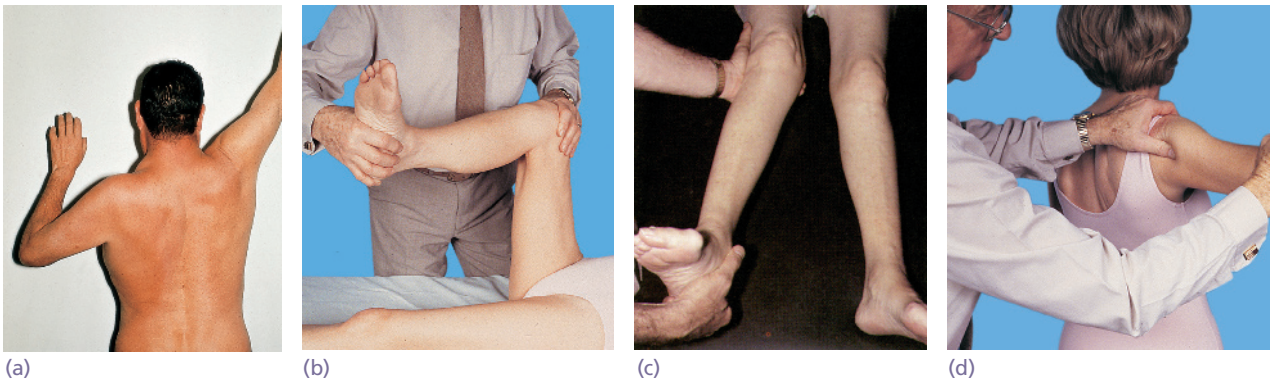


Figure 1.6 Move (a) Active movement – the patient moves the joint. The right shoulder is normal; the left has restricted active movement. (b) Passive movement – the examiner moves the joint. (c) Unstable movement – the joint can be moved across the normal planes of action, in this case demonstrating valgus instability of the right knee. (d) Provocative movement – the examiner moves (or manipulates) the joint so as to provoke the symptoms of impending pain or dislocation. Here he is reproducing the position in which an unstable shoulder is likely to dislocate.

Terminology

Colloquial terms such as front, back, upper, lower, inner aspect, outer aspect, bow legs, knock knees have the advantage of familiarity but are not applicable to every situation. Universally acceptable anatomical definitions are therefore necessary in describing physical attributes.

Bodily surfaces, planes and positions are always described in relation to the **anatomical position** – as if the person were standing erect, facing the viewer, legs together with the knees pointing directly forwards, and arms held by the sides with the palms facing forwards.

The principal **planes** of the body (Figure 1.7) are named **sagittal, coronal and transverse**; they define the direction across which the body (or body part) is viewed in any description. **Sagittal planes**, parallel to each other, pass vertically through the body from front to back; the **midsagittal** or **median plane** divides the body into right and left halves. **Coronal planes** are also orientated vertically, corresponding to a frontal view, at right angles to the sagittal planes; **transverse planes** pass horizontally across the body.

Anterior signifies the frontal aspect and **posterior** the rear aspect of the body or a body part. The terms **ventral** and **dorsal** are also used for the front and the back respectively. Note, though, that the use of these terms is somewhat confusing when it comes to the foot: here the upper surface is called the **dorsum** and the sole is called the **plantar surface**.

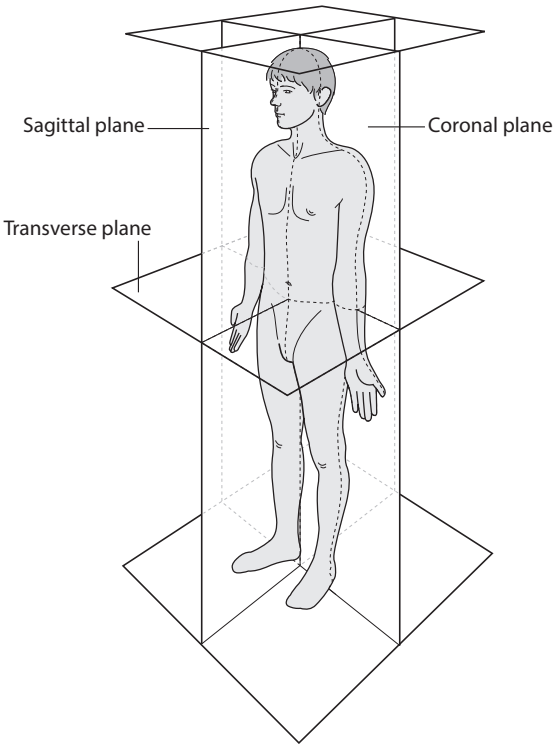


FIgure 1.7 Planes The principal planes of the body, as viewed in the anatomical position: sagittal, coronal and transverse.

Medial means facing towards the median plane or midline of the body, and **lateral** away from the median plane. These terms are usually applied to a limb, the clavicle or one half of the pelvis. Thus the inner aspect of the thigh lies on the medial side of the limb and the outer part of the thigh lies on the lateral side. We could also say that the little finger lies on the medial or **ulnar side** of the hand and the thumb on the lateral or **radial side** of the hand.

Proximal and **distal** are used mainly for parts of the limbs, meaning respectively the upper end and the lower end as they appear in the anatomical position. Thus the knee joint is formed by the distal end of the femur and the proximal end of the tibia.

Axial alignment describes the longitudinal arrangement of adjacent limb segments or parts of a single bone. The knees and elbows, for example, are normally angulated slightly outwards (**valgus**) while the opposite – 'bow legs' – is more correctly described as **varus** (see 'Physical variations and deformities' later in this chapter). Angulation in the middle of a long bone would always be regarded as abnormal.

Rotational alignment refers to the tortile arrangement of segments of a long bone (or an entire limb) around a single longitudinal axis. For example, in the anatomical position the patellae face forwards while the feet are turned slightly outwards; a marked difference in rotational alignment of the two legs is abnormal.

Flexion and extension are joint movements in the sagittal plane, most easily imagined in hinge joints like the knee, elbow and the joints of the fingers and toes. In elbows, knees, wrists and fingers, flexion means bending the joint and extension means straightening it. In shoulders and hips, flexion is movement in an anterior direction and extension is movement posteriorwards. In the ankle, flexion is also called **plantarflexion** (pointing the foot downwards) and extension is called **dorsiflexion** (drawing the foot upwards). Thumb movements are the most complicated and are described in Chapter 16.

Abduction and adduction are movements in the coronal plane, away from or towards the median plane. Not quite for the fingers and toes, though: here abduction and adduction mean away from and towards the longitudinal midline of the hand or foot!

Lateral rotation and medial rotation are twisting movements, outwards and inwards, around a longitudinal axis.

Pronation and supination are also rotatory movements, but the terms are applied only to movements of the forearm and the foot.

Circumduction is a composite movement made up of a rhythmic sequence of all the other movements. It is possible only for ball-and-socket joints such as the hip and shoulder.

Specialized movements such as opposition of the thumb, lateral flexion and rotation of the spine, and inversion or eversion of the foot, will be described in the relevant chapters.

NEUROLOGICAL EXAMINATION

If the symptoms include weakness or incoordination or a change in sensibility, or if they point to any disorder of the neck or back, a complete neurological examination of the related part is mandatory. Once again we follow a systematic routine, first looking at the general appearance, then assessing motor function (muscle tone, power and reflexes) and finally testing for sensory function (both skin sensibility and deep sensibility) (see Table 1.1 and Figure 1.8).

Table 1.1 Nerve root supply and actions of main muscle groups

Muscles/Muscle action	Nerve root supply
Sternomastoids	Spinal accessory C2, 3, 4
Trapezius	Spinal accessory C3, 4
Diaphragm	C3, 4, 5
Deltoid	C5, 6
Supra- and infraspinatus	C5, 6
Serratus anterior	C5, 6, 7
Pectoralis major	C5, 6, 7, 8
Elbow flexion extension	C5, 6 C7
Supination	C5, 6
Pronation	C6
Wrist extension flexion	C6, (7) C7, (8)
Finger extension flexion ab- and adduction	C7 C7, 8, T1 C8, T1
Hip flexion extension adduction abduction	L1, 2, 3 L5, S1 L2, 3, 4 L4, 5, S1
Knee extension flexion	L(2), 3, 4 L5, S1
Ankle dorsiflexion plantarflexion inversion eversion	L4, 5 S1, 2 L4, 5 L5, S1
Toe extension flexion abduction	L5 S1 S1, 2

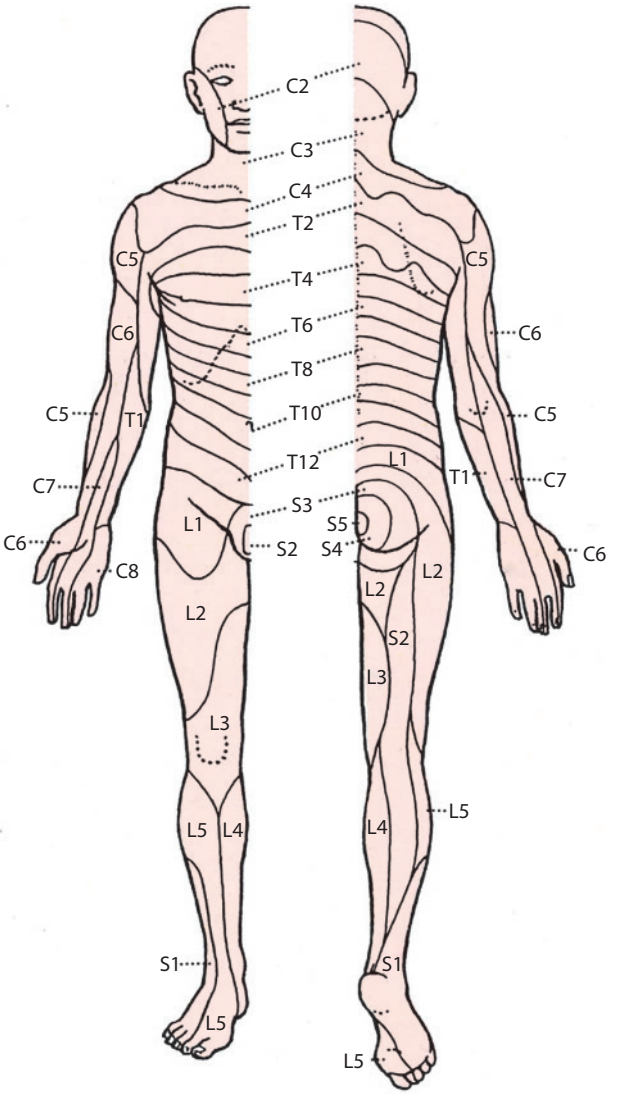


FIgure 1.8 Examination Dermatomes supplied by the spinal nerve roots.

Appearance

Some neurological disorders result in postures that are so characteristic as to be diagnostic at a glance: the claw hand of an ulnar nerve lesion; 'drop wrist' following radial nerve palsy (Figure 1.9); or the 'waiter's tip' deformity of the arm in brachial plexus injury. Usually, however, it is when the patient moves that we can best appreciate the type and extent of motor disorder: the dangling arm following a brachial plexus injury; the flail lower limb of poliomyelitis; the symmetrical paralysis of spinal cord lesions; the characteristic drop-foot gait following sciatic or peroneal nerve damage; and the jerky, 'spastic' movements of cerebral palsy.

Concentrating on the affected part, we look for trophic changes that signify loss of sensibility: the smooth, hairless skin that seems to be stretched too tight; atrophy of the fingertips and the nails; scars

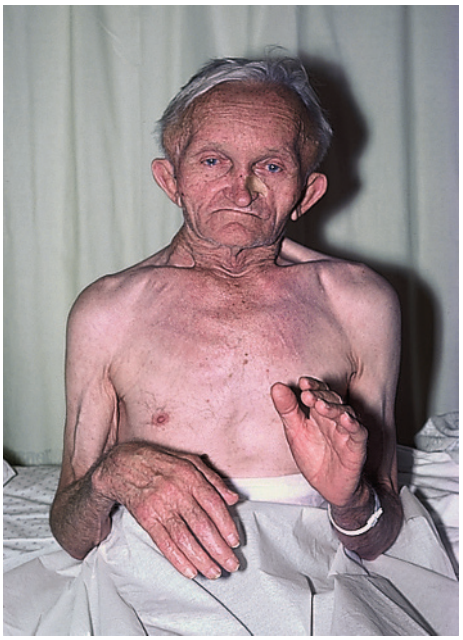


FIgure 1.9 Posture Posture is often diagnostic. This patient's 'drop wrist' – typical of a radial nerve palsy – is due to carcinomatous infiltration of the supraclavicular lymph nodes on the right.

that tell of accidental burns; and ulcers that refuse to heal. Muscle wasting is important: if localized and asymmetrical, it may suggest dysfunction of a specific motor nerve.

Muscle tone

Tone in individual muscle groups is tested by moving the nearby joint to stretch the muscle. Increased tone (spasticity) is characteristic of upper motor neuron disorders such as cerebral palsy and stroke. It must not be confused with rigidity (the 'lead-pipe' or 'cogwheel' effect) which is seen in Parkinson's disease. Decreased tone (flaccidity) is found in lower motor neuron lesions; for example, poliomyelitis. Muscle power is diminished in all three states; it is important to recognize that a 'spastic' muscle may still be weak.

Power

Motor function is tested by having the patient perform movements that are normally activated by specific nerves. We may learn even more about composite movements by asking the patient to perform specific tasks, such as holding a pen, gripping a rod, doing up a button or picking up a pin.

Testing for power is not as easy as it sounds; the difficulty is making ourselves understood. The simplest way is to place the limb in the 'test' position, then ask the patient to hold it there as firmly as possible and resist any attempt to change that position. The normal limb is examined first, then the affected limb, and the two are compared. Finer muscle actions, such as those of the thumb and fingers, may be reproduced by first demonstrating the movement yourself, then testing it in the unaffected limb, and then in the affected one.

Muscle power is usually graded on the Medical Research Council scale:

Grade 0	No movement
Grade 1	Only a flicker of movement
Grade 2	Movement with gravity eliminated
Grade 3	Movement against gravity
Grade 4	Movement against resistance
Grade 5	Normal power

It is important to recognize that muscle weakness may be due to muscle disease rather than nerve disease. In muscle disorders the weakness is usually more widespread and symmetrical, and sensation is normal.

Tendon reflexes

A deep tendon reflex is elicited by rapidly stretching the tendon near its insertion. A sharp tap with the tendon hammer does this well; but all too often this is performed with a flourish and with such force that the finer gradations of response are missed. It is better to employ a series of taps, starting with the most forceful and reducing the force with each successive tap until there is no response. Comparing the two sides in this way, we can pick up fine differences showing that a reflex is 'diminished' rather than 'absent'. In the upper limb we test biceps, triceps and brachioradialis; and in the lower limb the patellar and Achilles tendons.

The tendon reflexes are monosynaptic segmental reflexes; that is, the reflex pathway takes a 'short cut' through the spinal cord at the segmental level. Depression or absence of the reflex signifies interruption of the pathway at the posterior nerve root, the anterior horn cell, the motor nerve root or the peripheral nerve. It is a reliable pointer to the segmental level of dysfunction: thus, a depressed biceps jerk suggests pressure on the fifth or sixth cervical (C5 or C6) nerve roots while a depressed ankle jerk signifies a similar abnormality at the first sacral level (S1). An unusually brisk reflex, on the other hand, is characteristic of an upper motor neuron disorder (e.g. cerebral palsy, a stroke or injury to the spinal cord); the lower motor neuron is released from the normal central inhibition and there is an exaggerated response to tendon stimulation. This may manifest as ankle clonus: a sharp upward jerk on the foot (dorsiflexion) causes a repetitive, 'clonic' movement of the foot; similarly, a sharp downward push on the patella may elicit patellar clonus.

Superficial reflexes

The superficial reflexes are elicited by stroking the skin at various sites to produce a specific muscle contraction; the best known are the abdominal (T7–T12), cremasteric (L1, 2) and anal (S4, 5) reflexes. These are corticospinal (upper motor neuron) reflexes. Absence of the reflex indicates an upper motor neuron lesion (usually in the spinal cord) above that level.

The plantar reflex

Forceful stroking of the sole normally produces flexion of the toes (or no response at all). An extensor response (the big toe extends while the others remain in flexion) is characteristic of upper motor neuron disorders. This is the *Babinski sign* – a type of withdrawal reflex which is present in young infants and normally disappears after the age of 18 months.

Sensibility

Sensibility to touch and to pinprick may be increased (hyperaesthesia) or unpleasant (dysaesthesia) in certain irritative nerve lesions. More often, though, it is diminished (hypoaesthesia) or absent (anaesthesia), signifying pressure on or interruption of a peripheral nerve, a nerve root or the sensory pathways in the spinal cord. The area of sensory change can be mapped out on the skin and compared with the known segmental or dermatomal pattern of innervation. If the abnormality is well defined, it is an easy matter to establish the level of the lesion, even if the precise cause remains unknown.

Brisk percussion along the course of an injured nerve may elicit a tingling sensation in the distal distribution of the nerve (*Tinel's sign*). The point of hypersensitivity marks the site of abnormal nerve sprouting: if it progresses distally at successive visits, this signifies regeneration; if it remains unchanged, this suggests a local neuroma.

Tests for temperature recognition and two-point discrimination (the ability to recognize two touch-points a few millimetres apart) are also used in the assessment of peripheral nerve injuries.

Deep sensibility can be examined in several ways. In the vibration test a sounded tuning fork is placed over a peripheral bony point (e.g. the medial malleolus or the head of the ulna); the patient is asked if he or she can feel the vibrations and to say when they disappear. By comparing the two sides, differences can be noted. Position sense is tested by asking the patient to find certain points on the body with the eyes closed – for example, touching the tip of the nose with the forefinger. The sense of joint posture is tested by grasping the big toe and placing it in different positions of flexion and extension. The patient (whose eyes are closed) is asked to say whether it is 'up' or 'down'. Stereognosis, the ability to recognize shape and texture by feel alone, is tested by giving the patient (again with eyes closed) a variety of familiar objects to hold and asking him or her to name each object.

The pathways for deep sensibility run in the posterior columns of the spinal cord. Disturbances are therefore found in peripheral neuropathies and in spinal cord lesions such as posterior column injuries or tabes dorsalis. The sense of balance is also carried in the posterior columns. This can be tested by asking the patient to stand upright with his or her eyes closed; excessive body sway is abnormal (*Romberg's sign*).

Cortical and cerebellar function

A staggering gait may imply an unstable knee – or a disorder of the spinal cord or cerebellum. If there is no musculoskeletal abnormality to account for the sign, a full examination of the central nervous system will be necessary.

EXAMINING INFANTS AND CHILDREN

Paediatric practice requires special skills. You may have no first-hand account of the symptoms; a baby screaming with pain will tell you very little, and over-anxious parents will probably tell you too much. When examining the child, be flexible. If he or she is moving a particular joint, take your opportunity to examine movement then and there. You will learn much more by adopting methods of play than by applying a rigid system of examination. And leave any test for tenderness until last!

Infants and small children

The baby should be undressed, in a warm room, and placed on the examining couch. Look carefully for birthmarks, deformities and abnormal movements – or absence of movement. If there is no urgency or distress, take time to examine the head and neck, including facial features which may be characteristic of specific dysplastic syndromes. The back and limbs are then examined for abnormalities of position or shape.

Examining for joint movement can be difficult. Active movements can often be stimulated by gently stroking the limb. When testing for passive mobility, be careful to avoid frightening or hurting the child. In the neonate, and throughout the first two years of life, examination of the hips is mandatory, even if the child appears to be normal. This is to avoid missing the subtle signs of developmental dysplasia of the hips (DDH) at the early stage when treatment is most

Table 1.2 Normal developmental milestones

Age	Normal developmental milestone(s)
Newborn	Grasp reflex present Morrow reflex present
3–6 months	Holds head up unsupported
6–9 months	Able to sit up
9–12 months	Crawling Standing up
9–18 months	Walking
18–24 months	Running

effective. It is also important to assess the child's general development by testing for the normal milestones which are expected to appear during the first two years of life (Table 1.2).

Older children

Most children can be examined in the same way as adults, though with different emphasis on particular physical features. Posture and gait are very important; subtle deviations from the norm may herald the appearance of serious abnormalities such as scoliosis or neuromuscular disorders, while more obvious 'deformities' such as knock knees and bow legs may be no more than transient stages in normal development; similarly with mild degrees of 'flat feet' and 'pigeon toes'. More complex variations in posture and gait patterns, when the child sits and walks with the knees turned inwards (medially rotated) or outwards (laterally rotated) are usually due to anteversion or retroversion of the femoral necks, sometimes associated with compensatory rotational 'deformities' of the femora and tibiae. Seldom need anything be done about this; the condition usually improves as the child approaches puberty and only if the gait is very awkward would one consider performing corrective osteotomies of the femora.

PHYSICAL VARIATIONS AND DEFORMITIES

Joint laxity

Children's joints are much more mobile than those of most adults, allowing them to adopt postures that would be impossible for their parents. An unusual degree of joint mobility can also be attained by adults willing to submit to rigorous exercise and practice, as witness the performances of professional dancers and athletes, but in most cases, when the exercises stop, mobility gradually reverts to the normal range.

Persistent generalized joint hypermobility occurs in about 5% of the population and is inherited as a simple Mendelian dominant (Figure 1.10). Those affected

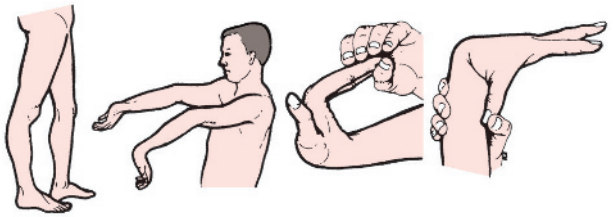


Figure 1.10 Tests for joint hypermobility
Hyperextension of knees and elbows; metacarpophalangeal joints extending to 90 degrees'; thumb able to touch forearm.

describe themselves as being 'double-jointed': they can hyperextend their metacarpophalangeal joints beyond a right angle, hyperextend their elbows and knees and bend over with knees straight to place their hands flat on the ground; some can even 'do the splits' or place their feet behind their neck!

It is doubtful whether these individuals should be considered 'abnormal'. However, epidemiological studies have shown that they do have a greater than usual tendency to recurrent dislocation (e.g. of the shoulder or patella). Some experience recurrent episodes of aching around the larger joints; however, there is no convincing evidence that hypermobility by itself predisposes to osteoarthritis.

Generalized hypermobility is not usually associated with any obvious disease, but severe laxity is a feature of certain rare connective tissue disorders such as Marfan's syndrome, Ehlers–Danlos syndrome, Larsen's disease and osteogenesis imperfecta.

Deformity

The boundary between variations of the normal and physical deformity is blurred. Indeed, in the development of species, what at one point of time might have been seen as a deformity could over the ages have turned out to be so advantageous as to become essential for survival. So too in humans. The word 'deformity' is derived from the Latin for 'misshapen', but the range of 'normal shape' is so wide that variations should not automatically be designated as deformities, and some undoubted 'deformities' are not necessarily pathological; for example, the generally accepted cut-off points for 'abnormal' shortness or tallness are arbitrary and people who in one population might be considered abnormally short or abnormally tall could, in other populations, be seen as quite ordinary. However, if one leg is short and the other long, no one would quibble with the use of the word 'deformity'!

Specific terms are used to describe the 'position' and 'shape' of the bones and joints. Whether, in any particular case, these amount to 'deformity' will be determined by additional factors such as the extent

to which they deviate from the norm, symptoms to which they give rise, the presence or absence of instability and the degree to which they interfere with function.

Varus and valgus It seems pedantic to replace 'bow legs' and 'knock knees' with 'genu varum' and 'genu valgum', but comparable colloquialisms are not available for deformities of the elbow, hip or big toe; and, besides, the formality is justified by the need for clarity and consistency. Varus means that the part distal to the joint in question is displaced towards the median plane, valgus away from it (Figure 1.11).

Kyphosis and lordosis Seen from the side, the normal spine has a series of curves: convex posteriorly in the thoracic region (kyphosis), and convex anteriorly in the cervical and lumbar regions (lordosis). Excessive curvature constitutes kyphotic or lordotic deformity (also sometimes referred to as hyperkyphosis and hyperlordosis). Colloquially speaking, excessive thoracic kyphosis is referred to as 'round-shouldered'.

Scoliosis Seen from behind, the spine is straight. Any curvature in the coronal plane is called scoliosis. The position and direction of the curve are specified by terms such as thoracic scoliosis, lumbar scoliosis, convex to the right, concave to the left, etc.

Postural deformity A postural deformity is one which the patient can, if properly instructed, correct voluntarily: e.g. thoracic 'kyphosis' due to slumped shoulders. Postural deformity may also be caused by temporary muscle spasm.

Structural deformity A deformity which results from a permanent change in anatomical structure cannot be voluntarily corrected. It is important to distinguish postural scoliosis from structural (fixed) scoliosis. The former is non-progressive and benign; the latter is usually progressive and may require treatment.

'Fixed deformity' This term is ambiguous. It seems to mean that a joint is deformed and unable to move but this is not so. It means that one particular movement cannot be completed. Thus the knee may be able to flex fully but not extend fully – at the limit of its extension it is still 'fixed' in a certain amount of flexion. This would be called a 'fixed flexion deformity'.

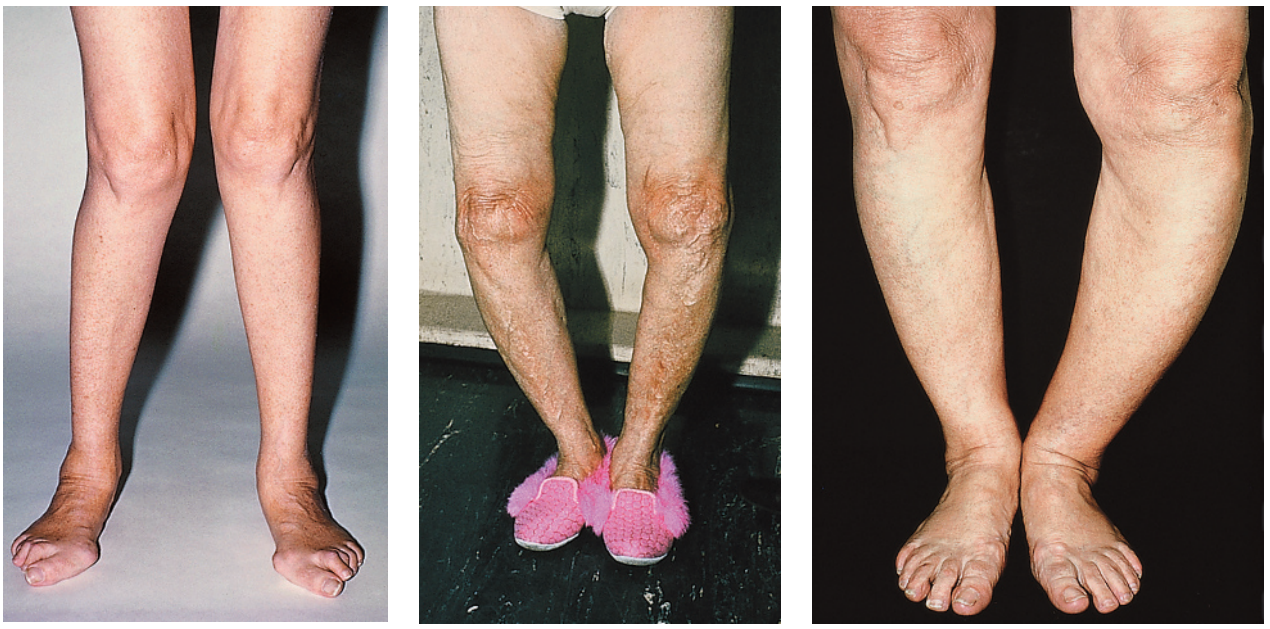
CAUSES OF JOINT DEFORMITY

There are six basic causes of joint deformity.

Contracture of the overlying skin This is seen typically when there is severe scarring across the flexor aspect of a joint, e.g. due to a burn or following surgery.

Contracture of the subcutaneous fascia The classical example is Dupuytren's contracture in the palm of the hand.

Muscle contracture Fibrosis and contracture of muscles that cross a joint will cause a fixed deformity of the joint. This may be due to deep infection or fibrosis following ischaemic necrosis (Volkmann's ischaemic contracture).



(a) (b) (c)

FIgure 1.11 Varus and valgus (a) Valgus knees in a patient with rheumatoid arthritis. The toe joints are also valgus. (b) Varus knees due to osteoarthritis. (c) Another varus knee? No – the deformity here is in the left tibia due to Paget's disease.

Muscle imbalance Unbalanced muscle weakness or spasticity will result in joint deformity which, if not corrected, will eventually become fixed. This is seen most typically in poliomyelitis and cerebral palsy. Tendon rupture, likewise, may cause deformity.

Joint instability Any unstable joint will assume a 'deformed' position when subjected to force.

Joint destruction Trauma, infection or arthritis may destroy the joint and lead to severe deformity.

CAUSES OF BONE DEFORMITY

Bone deformities in small children are usually due to genetic or developmental disorders of cartilage and bone growth; some can be diagnosed *in utero* by special imaging techniques (e.g. achondroplasia); some become apparent when the child starts to walk, or later still during one of the growth spurts (e.g. hereditary multiple exostosis); and some only in early adulthood (e.g. multiple epiphyseal dysplasia). There are a myriad genetic disorders affecting the skeleton, yet any one of these conditions is rare. The least unusual of them are described in Chapter 8.

Acquired deformities in children may be due to fractures involving the physis (growth plate); ask about previous injuries. Other causes include rickets, endocrine disorders, malunited diaphyseal fractures and tumours.

Acquired deformities of bone in adults are usually the result of previous malunited fractures. However, causes such as osteomalacia, bone tumours and Paget's disease should always be considered.

Bony lumps

A bony lump may be due to faulty development, injury, inflammation or a tumour. Although X-ray examination is essential, the clinical features can be highly informative (for example, see Figure 1.12).

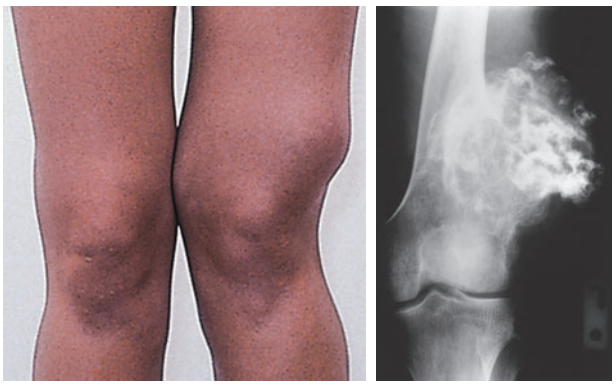


FIgure 1.12 Bony lumps The lump above the left knee is hard, well defined and not increasing in size. The clinical diagnosis of cartilage-capped exostosis (osteochondroma) is confirmed by the X-rays.

Size A large lump attached to bone, or a lump that is getting bigger, is nearly always a tumour.

Site A lump near a joint is most likely to be a tumour (benign or malignant); a lump in the shaft may be fracture callus, inflammatory new bone or a tumour. A benign tumour has a well-defined margin; malignant tumours, inflammatory lumps and callus have a vague edge.

Consistency A benign tumour feels bony and hard; malignant tumours often give the impression that they can be indented.

Tenderness Lumps due to active inflammation, recent callus or a rapidly growing sarcoma are tender.

Multiplicity Multiple bony lumps are uncommon: they occur in hereditary multiple exostosis and in Ollier's disease.

Joint stiffness

The term 'stiffness' covers a variety of limitations. We consider three types of stiffness in particular: (1) all movements absent; (2) all movements limited; (3) one or two movements limited.

All movements absent Surprisingly, although movement is completely blocked, the patient may retain such good function that the restriction goes unnoticed until the joint is examined. Surgical fusion is called 'arthrodesis'; pathological fusion is called 'ankylosis'. Acute suppurative arthritis typically ends in bony ankylosis; tuberculous arthritis heals by fibrosis and causes fibrous ankylosis – not strictly a 'fusion' because there may still be a small jog of movement.

All movements limited After severe injury, movement may be limited as a result of oedema and bruising. Later, adhesions and loss of muscle extensibility may perpetuate the stiffness.

With active inflammation all movements are restricted and painful and the joint is said to be 'irritable'. In acute arthritis spasm may prevent all but a few degrees of movement.

In osteoarthritis the capsule fibroses and movements become increasingly restricted, but pain occurs only at the extremes of motion.

Some movements limited When one particular movement suddenly becomes blocked, the cause is usually mechanical. Thus a torn and displaced meniscus may prevent extension of the knee but not flexion.

Bone deformity may alter the arc of movement, such that it is limited in one direction (loss of abduction in coxa vara is an example) but movement in the opposite direction is full or even increased.

These are all examples of 'fixed deformity'.

DIAGNOSTIC IMAGING

The map is not the territory

Alfred Korzybski

Plain film radiography

Plain film X-ray examination is over 100 years old. Notwithstanding the extraordinary technical advances of the last few decades, it remains the most useful method of diagnostic imaging. Whereas other methods may define an inaccessible anatomical structure more accurately, or may reveal some localized tissue change, the plain film provides information simultaneously on the size, shape, tissue 'density' and bone architecture – characteristics which, taken together, will usually suggest a diagnosis, or at least a range of possible diagnoses.

The radiographic image

X-rays are produced by firing electrons at high speed onto a rotating anode. The resulting beam of X-rays is attenuated by the patient's soft tissues and bones, casting what are effectively 'shadows' which are displayed as images on an appropriately sensitized plate or stored as digital information which is then available to be transferred throughout the local information technology (IT) network. See Figure 1.13.

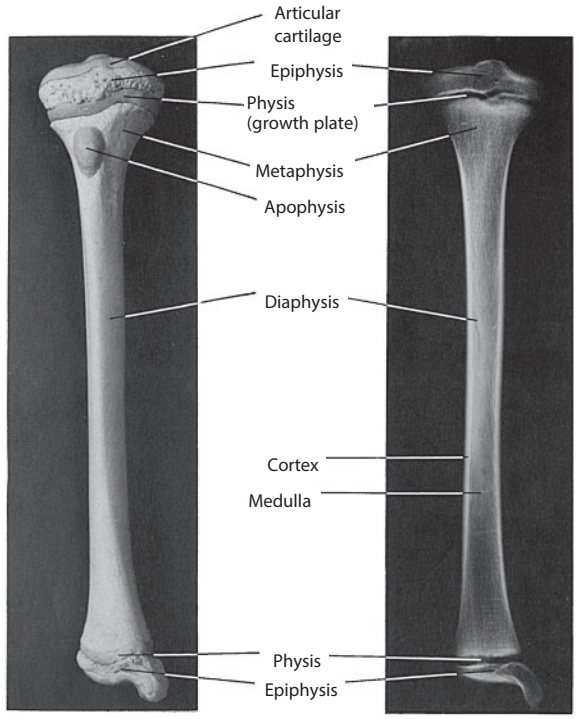


FIgure 1.13 The radiographic image X-ray of an anatomical specimen to show the appearance of various parts of the bone in the X-ray image.

The more dense and impenetrable the tissue, the greater the X-ray attenuation and therefore the more blank, or white, the image that is captured. Thus, a metal implant appears intensely white, bone less so and soft tissues in varying shades of grey depending on their 'density'. Cartilage, which causes little attenuation, appears as a dark area between adjacent bone ends; this 'gap' is usually called the joint space, though of course it is not a space at all, merely a radiolucent zone filled with cartilage. Other 'radiolucent' areas are produced by fluid-filled cysts in bone.

One bone overlying another (e.g. the femoral head inside the acetabular socket) produces superimposed images; any abnormality seen in the resulting combined image could be in either bone, so it is important to obtain several images from different projections in order to separate the anatomical outlines. Similarly, the bright image of a metallic foreign body superimposed upon that of, say, the femoral condyles could mean that the foreign body is in front of, inside or behind the bone. A second projection, at right angles to the first, will give the answer.

Picture Archiving and Communication System (PACS) This is the system whereby all digitally coded images are filed, stored and retrieved to enable the images to be sent to work stations throughout the hospital, to other hospitals or to the Consultant's personal computer.

Radiographic interpretation

Although *radiograph* is the correct word for the plain image which we address, in the present book we have chosen to retain the old-fashioned term 'X-ray', which has become entrenched by long usage. The process of interpreting this image should be as methodical as clinical examination. It is seductively easy to be led astray by some flagrant anomaly; systematic study is the only safeguard. A convenient sequence for examination is: *the patient – the soft tissues – the bones – the joints.*

THE PATIENT

Make sure that the name on the film is that of your patient; mistaken identity is a potent source of error. The clinical details are important; it is surprising how much more you can see on the X-ray when you know the background. Similarly, when requesting an X-ray examination, give the radiologist enough information from the patient's history and the clinical findings to help in guiding his or her thoughts towards the diagnostic possibilities and options. For example, when considering a malignant bone lesion, simply knowing the patient's age may provide an important clue: under the age of 10 it is most likely to be a Ewing's sarcoma; between 10 and 20 years it is more likely to

be an osteosarcoma; and over the age of 50 years it is likely to be a metastatic deposit.

THE SOFT TISSUES

Generalized change Muscle planes are often visible and may reveal wasting or swelling. Bulging outlines around a hip, for example, may suggest a joint effusion; and soft-tissue swelling around interphalangeal joints may be the first radiographic sign of rheumatoid arthritis. Tumours tend to displace fascial planes, whereas infection tends to obliterate them.

Localized change Is there a mass, soft tissue calcification, ossification, gas (from penetrating wound or gas-forming organism) or the presence of a radio-opaque foreign body?

THE BONES

Shape The bones are well enough defined to allow one to check their general anatomy and individual shape (Figure 1.14). For example, for the spine, look at the overall vertebral alignment, then at the disc spaces, and then at each vertebra separately, moving from the body to the pedicles, the facet joints and finally the spinous appendages. For the pelvis, see if the shape is symmetrical with the bones in their normal positions, then look at the sacrum, the two innominate bones, the pubic rami and the ischial tuberosities, then the femoral heads and the upper ends of the femora, always comparing the two sides.

Generalized change Take note of changes in bone 'density' (osteopaenia or osteosclerosis). Is there abnormal trabeculation, as in Paget's disease (Figure 1.15a)? Are there features suggestive of diffuse metastatic infiltration, either sclerotic or lytic? Other polyostotic lesions include fibrous dysplasia, histiocyotis, multiple exostosis and Paget's disease. With aggressive-looking polyostotic lesions, think of metastases (including myeloma and lymphoma) and also multifocal infection. By contrast, most primary tumours are monostotic.

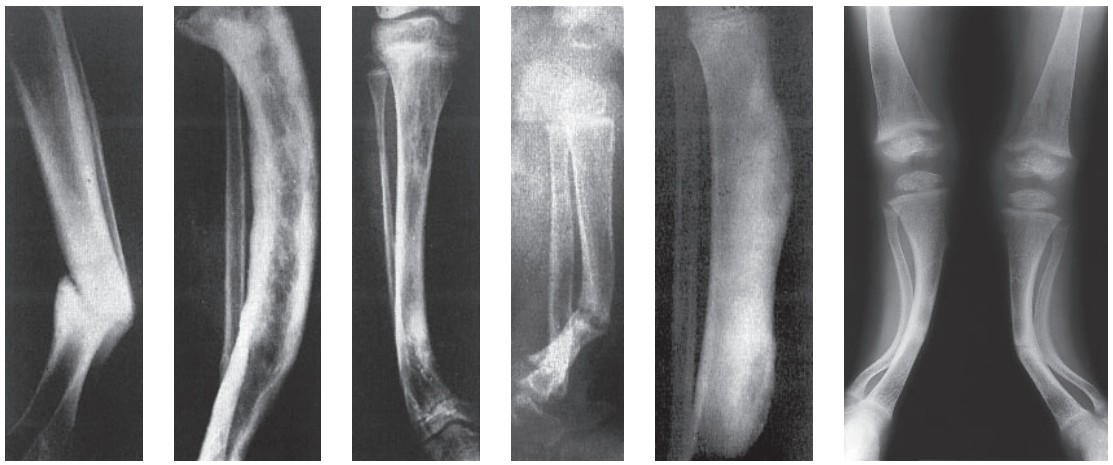
Localized change Focal abnormalities should be approached in the same way as one would conduct a clinical analysis of a soft tissue abnormality. Start describing the abnormality from the centre and move outwards. Determine the lesion's size, site, shape, density and margins, as well as adjacent periosteal changes and any surrounding soft tissue changes. Remember that benign lesions are usually well defined with sclerotic margins (Figure 1.15b) and a smooth periosteal reaction. Ill-defined areas with permeative bone destruction (Figure 1.15c) and irregular or speculated periosteal reactions (Figure 1.15d) suggest an aggressive lesion such as infection or a malignant tumour.

THE JOINTS

The radiographic 'joint' consists of the articulating bones and the 'space' between them.

The 'joint space' The joint space is, of course, illusory; it is occupied by a film of synovial fluid plus radiolucent articular cartilage which varies in thickness from 1 mm or less (the carpal joints) to 6 mm (the knee). It looks much wider in children than in adults because much of the epiphysis is still cartilaginous and therefore radiolucent. Lines of increased density within the radiographic articular 'space' may be due to calcification of the cartilage or menisci (chondrocalcinosis). Loose bodies, if they are radio-opaque, appear as rounded patches overlying the normal structures.

Shape Note the general orientation of the joint and the congruity of the bone ends (actually the subarticular bone plates), if necessary comparing the



(a) (b) (c) (d) (e) (f)

FIgure 1.14 X-rays – bent bones (a) Malunited fracture; (b) Paget's disease; (c) dyschondroplasia; (d) congenital pseudarthrosis; (e) syphilitic sabre tibia; (f) osteogenesis imperfecta.

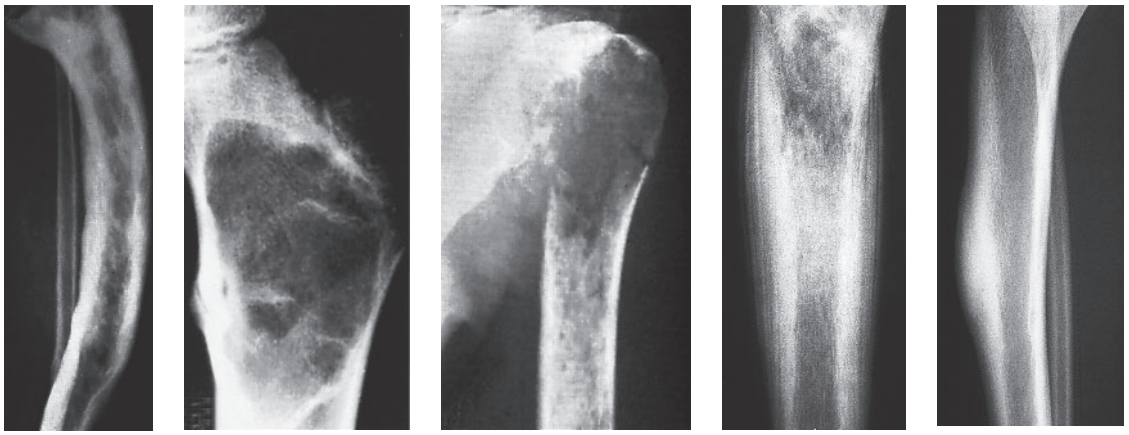


FIgure 1.15 X-rays – important features to look for (a) *General shape and appearance*, in this case the cortices are thickened and the bone is bent (Paget's disease). (b,c) *Interior density*, a vacant area may represent a true cyst (b), or radiolucent material infiltrating the bone, like the metastatic tumour in (c). (d) *Periosteal reaction*, typically seen in healing fractures, bone infection and malignant bone tumours – as in this example of Ewing's sarcoma. Compare this with the smooth periosteal new bone formation shown in (e).

abnormal with the normal opposite side. Then look for narrowing or asymmetry of the joint 'space': narrowing signifies loss of hyaline cartilage and is typical of infection, inflammatory arthropathies and osteoarthritis. Further stages of joint destruction are revealed by irregularity of the radiographically visible bone ends and radiolucent cysts in the subchondral bone. Bony excrescences at the joint margins (osteophytes) are typical of osteoarthritis.

Erosions Look for associated bone erosions. The position of erosions and symmetry help to define various types of arthropathy. In rheumatoid arthritis and psoriasis the erosions are periarticular (at the bare area where the hyaline cartilage covering the joint has ended and the intracapsular bone is exposed to joint fluid). In gout the erosions are further away from the articular surfaces and are described as juxta-articular. Rheumatoid arthritis is classically symmetrical and predominantly involves the metacarpophalangeal and proximal interphalangeal joints in both hands. The erosions in psoriasis are usually more feathery with ill-defined new bone at their margins. Ill-defined erosions suggest active synovitis whereas corticated erosions indicate healing and chronicity.

Diagnostic associations

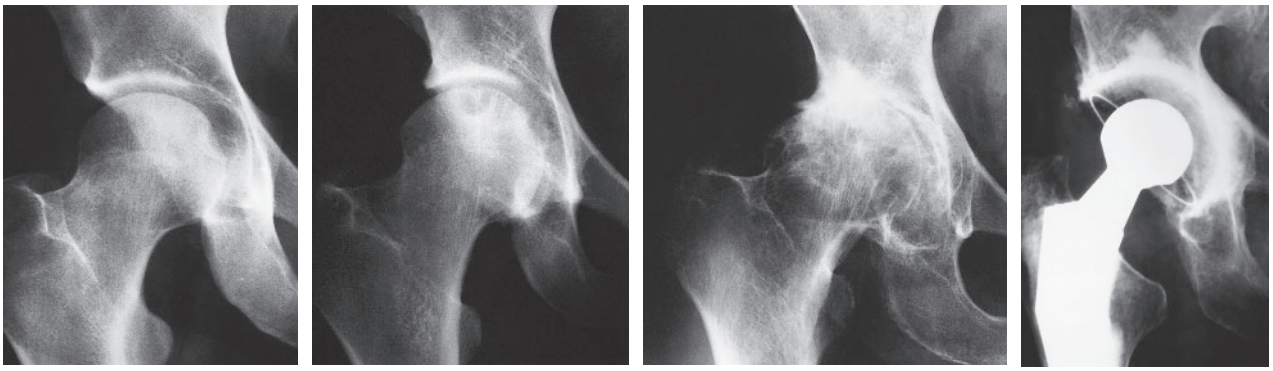
However carefully the individual X-ray features are observed, the diagnosis will not leap ready-made off the X-ray plate. Even a fracture is not always obvious. It is the pattern of abnormalities that counts: if you see one feature that is suggestive, look for others that are commonly associated.

- Narrowing of the joint space + subchondral sclerosis and cysts + osteophytes = osteoarthritis (Figure 1.16).
- Narrowing of the joint space + osteoporosis + periarticular erosions = inflammatory arthritis. Add to this the typical distribution, more or less symmetrically in the proximal joints of both hands, and you must think of rheumatoid arthritis.
- Bone destruction + periosteal new bone formation = infection or malignancy until proven otherwise.
- Remember: the next best investigation is either the previous radiograph or the subsequent follow-up radiograph. Sequential films demonstrate either progression of changes in active pathology or status quo in long-standing conditions.

Limitations of conventional radiography

Conventional radiography involves exposure of the patient to ionizing radiation, which under certain circumstances can lead to radiation-induced cancer. The Ionizing Radiation Medical Exposure Regulations (IRMER) 2000 are embedded in European Law, requiring all clinicians to justify any exposure of the patient to ionizing radiation. It is a criminal offence to breach these regulations. Ionizing radiation can also damage a developing fetus, especially in the first trimester.

As a diagnostic tool, conventional radiography provides poor soft-tissue contrast: for example, it cannot distinguish between muscles, tendons, ligaments and hyaline cartilage. Ultrasound (US), computed tomography (CT) and magnetic resonance imaging (MRI) are now employed to complement plain X-ray examination. However, in parts of the world where these techniques are not available, some modifications of plain radiography still have a useful role.



(a) (b) (c) (d)

Figure 1.16 Plain X-rays of the hip Stages in the development of osteoarthritis (OA). (a) Normal hip: anatomical shape and position, with joint 'space' (articular cartilage) fully preserved. (b) Early OA, showing joint space slightly decreased and a subarticular cyst in the femoral head. (c) Advanced OA: joint space markedly decreased; osteophytes at the joint margin. (d) Hip replacement: the cup is radiolucent but its position is shown by a circumferential wire marker. Note the differing image 'densities': (1) the metal femoral implant; (2) the polyethylene cup (radiolucent); (3) acrylic cement impacted into the adjacent bone.

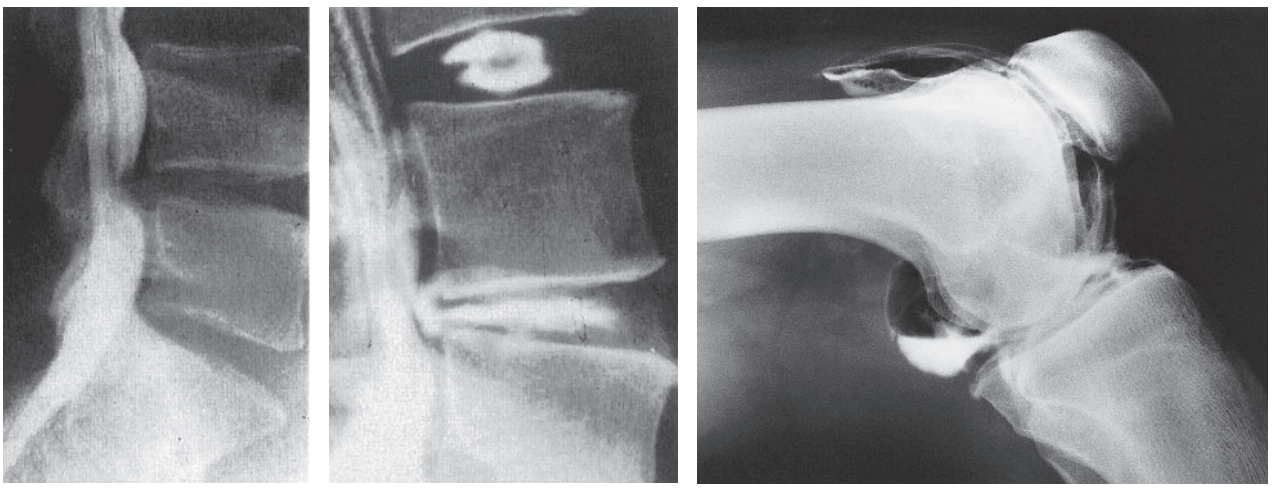
X-RAYS USING CONTRAST MEDIA

Substances that alter X-ray attenuation characteristics can be used to produce images which contrast with those of the normal tissues. The contrast media used in orthopaedics are mostly iodine-based liquids which can be injected into sinuses, joint cavities or the spinal theca (Figure 1.17). Air or gas also can be injected into joints to produce a 'negative image' outlining the joint cavity.

Oily iodides are not absorbed and maintain maximum concentration after injection. However, because they are non-miscible, they do not penetrate well into all the nooks and crannies. They are also tissue irritants, especially if used intrathecally and are now rarely used as they have been shown to cause adhesive arachnoiditis. Ionic, water-soluble iodides permit much more detailed imaging and, although also somewhat irritant and neurotoxic, are rapidly absorbed and excreted.

Sinography

Sinography is the simplest form of contrast radiography. The medium (usually one of the ionic water-soluble compounds) is injected into an open sinus; the film shows the track and whether or not it leads to the underlying bone or joint.



(a) (b) (c)

Figure 1.17 Contrast radiography (a) Myelography shows the outline of the spinal theca. Where facilities are available, myelography has been largely replaced by CT and MRI. (b) Discography is sometimes useful: note the difference between a normal intervertebral disc (upper level) and a degenerate disc (lower level). (c) Contrast arthrography of the knee shows a small popliteal herniation.

Arthrography

Arthrography is a particularly useful form of contrast radiography. Intra-articular loose bodies will produce filling defects in the opaque contrast medium. In the knee, torn menisci, ligament tears and capsular ruptures can be shown. In children's hips, arthrography is a useful method of outlining the cartilaginous (and therefore radiolucent) femoral head. In adults with avascular necrosis of the femoral head, arthrography may show up torn flaps of cartilage. After hip replacement, loosening of a prosthesis may be revealed by seepage of the contrast medium into the cement/bone interface. In the hip, ankle, wrist and shoulder, the injected contrast medium may disclose labral tears or defects in the capsular structures. In the spine, contrast radiography can be used to diagnose disc degeneration (discography) and abnormalities of the small facet joints (facetography).

Myelography

Myelography was used extensively in the past for the diagnosis of disc prolapse and other spinal canal lesions. It has been largely replaced by non-invasive methods such as CT and MRI. However, it still has a place in the investigation of nerve root lesions and as an adjunct to other methods in patients with back pain.

The oily media are no longer used, and even with the ionic water-soluble iodides there is a considerable incidence of complications, such as low-pressure headache (due to the lumbar puncture), muscular spasms or convulsions (due to neurotoxicity, especially if the chemical is allowed to flow above the mid-dorsal region) and arachnoiditis (which is attributed to the hyperosmolality of these compounds in relation to cerebrospinal fluid). Precautions, such as keeping the patient sitting upright after myelography, must be strictly observed.

Metrizamide has low neurotoxicity and at working concentrations it is more or less isotonic with cerebrospinal fluid. It can therefore be used throughout the length of the spinal canal; the nerve roots are also well delineated (radiculography). A bulging disc, an intrathecal tumour or narrowing of the bony canal will produce characteristic distortions of the opaque column in the myelogram.

Plain tomography

Tomography provides an image 'focused' on a selected plane. By moving the tube and the X-ray film in opposite directions around the patient during the exposure, images on either side of the pivotal plane are deliberately blurred out. When several 'cuts' are studied, lesions obscured in conventional X-rays may be revealed. The method is useful for diagnosing segmental bone necrosis and depressed fractures in cancellous bone (e.g. of the vertebral body or the tibial plateau); these defects are often obscured in the plain X-ray by the surrounding intact mass of bone. Small radiolucent lesions, such as osteoid osteomas and bone abscesses, can also be revealed.

A useful procedure in former years, conventional tomography has been largely supplanted by CT and MRI.

Computed tomography (CT)

Like plain tomography, CT produces sectional images through selected tissue planes – but with much greater resolution (Figure 1.18). A further advance over conventional tomography is that the images are trans-axial (like transverse anatomical sections), thus exposing anatomical planes that are never viewed in plain film X-rays. A general (or 'localization') view is obtained, the region of interest is selected and a series of cross-sectional images is produced and digitally recorded. 'Slices' through the larger joints or tissue masses may be 3–5 mm apart; those through the small joints or intervertebral discs have to be much thinner.

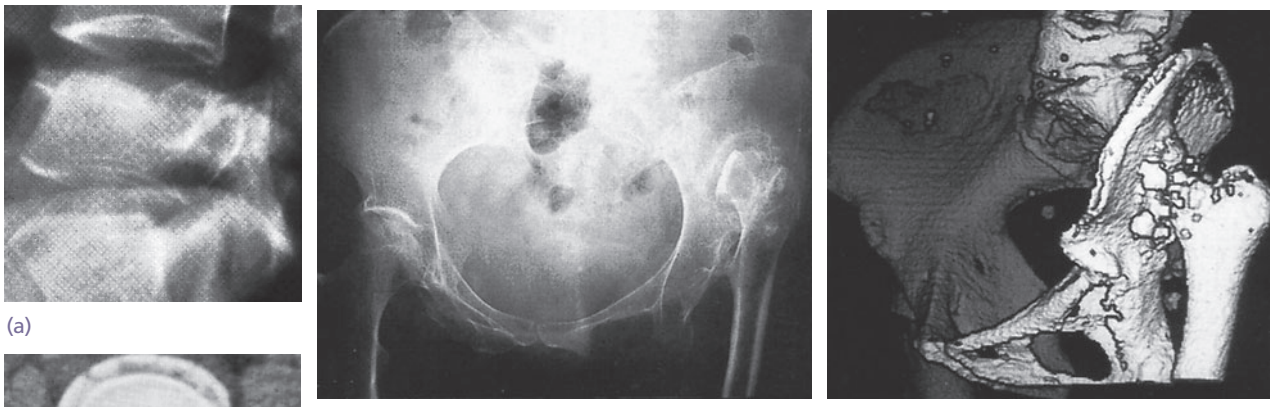
New multislice CT scanners provide images of high quality from which multiplanar reconstructions in all three orthogonal planes can be produced. Three-dimensional surface rendered reconstructions and volume rendered reconstructions may help in demonstrating anatomical contours, but fine detail is lost in this process.

Clinical applications

Because CT achieves excellent contrast resolution and spatial localization, it is able to display the size, shape and position of bone and soft-tissue masses in transverse planes. Image acquisition is extremely fast. The technique is therefore ideal for evaluating acute trauma to the head, spine, chest, abdomen and pelvis. It is better than MRI for demonstrating fine bone detail and soft-tissue calcification or ossification.

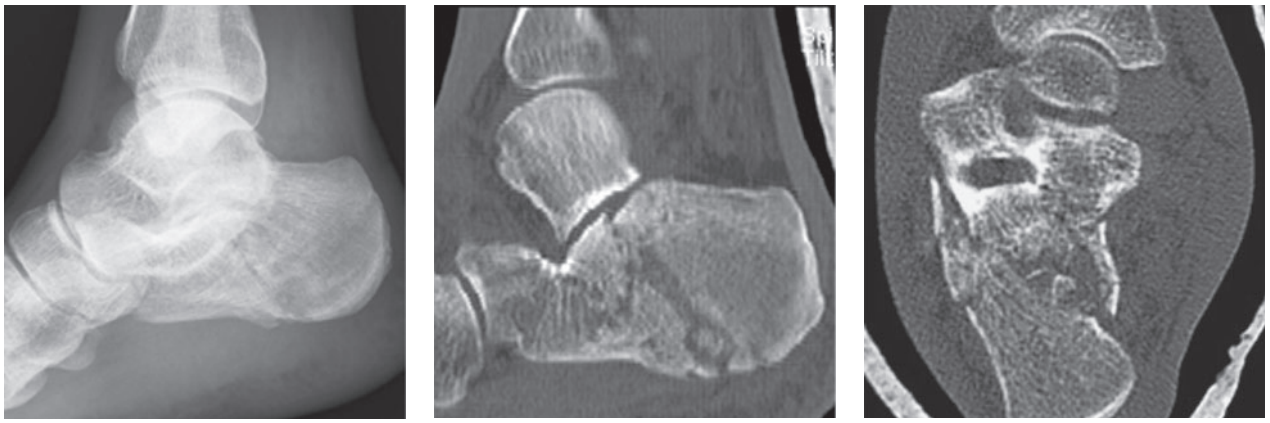
Computed tomography is also an invaluable tool for assisting with preoperative planning in secondary fracture management. It is routinely used for assessing injuries of the vertebrae, acetabulum, proximal tibial plateau, ankle and foot – indeed complex fractures and fracture-dislocations at any site (Figure 1.19).

It is also useful in the assessment of bone tumour size and spread, even if it is unable to characterize the tumour type. It can be employed for guiding soft-tissue and bone biopsies.



(a) (b) (c) (d)

Figure 1.18 Computed tomography (CT) The plain X-ray (a) shows a fracture of the vertebral body but one cannot tell precisely how the bone fragments are displaced. The CT (b) shows clearly that they are dangerously close to the cauda equina. (c) Congenital hip dislocation, defined more clearly by (d) three-dimensional CT reconstruction.



(a) (b) (c)

Figure 1.19 CT for complex fractures (a) A plain X-ray shows a fracture of the calcaneum but the details are obscure. CT sagittal and axial views (b,c) give a much clearer idea of the seriousness of this fracture.

Limitations

An important limitation of CT is that it provides relatively poor soft-tissue contrast when compared with MRI.

A major disadvantage of this technique is the relatively high radiation exposure to which the patient is subjected. It should, therefore, be used with discretion.

Magnetic resonance imaging (MRI)

Magnetic resonance imaging produces cross-sectional images of any body part in any plane. It yields superb soft-tissue contrast, allowing different soft tissues to be clearly distinguished, e.g. ligaments, tendons, muscle and hyaline cartilage. Another big advantage of MRI is that it does not use ionizing radiation. It is, however, contraindicated in patients with pacemakers and possible metallic foreign bodies in the eye or brain, as these could potentially move when the patient is introduced into the scanner's strong magnetic field. Approximately 5% of patients cannot tolerate the scan due to claustrophobia, but newer scanners are being developed to be more 'open'.

MRI physics

The patient's body is placed in a strong magnetic field (between 5 and 30 000 times the strength of the Earth's magnetic field). The body's protons have a positive charge and align themselves along this strong

external magnetic field. The protons are spinning and can be further excited by radiofrequency pulses, rather like whipping a spinning top. These spinning positive charges will not only induce a small magnetic field of their own, but will produce a signal as they relax (slow down) at different rates.

A proton density map is recorded from these signals and plotted in *x*, *y* and *z* coordinates. Different speeds of tissue excitation with radiofrequency pulses (repetition times, or TR) and different intervals between recording these signals (time to echo, or TE) will yield anatomical pictures with varying 'weighting' and characteristics. T1 weighted (T1W) images have a high spatial resolution and provide good anatomical-looking pictures. T2 weighted (T2W) images give more information about the physiological characteristics of the tissue. Proton density (PD) images are also described as 'balanced' or 'intermediate' as they are essentially a combination of T1 and T2 weighting and yield excellent anatomical detail for orthopaedic imaging. Fat suppression sequences allow highlighting of abnormal water, which is particularly useful in orthopaedics when assessing both soft-tissue and bone marrow oedema.

Intravenous contrast

Just as in CT, enhancement by intravenous contrast relies on an active blood supply and leaky cell membranes. Areas of inflammation and active tumour tissue will be highlighted. Gadolinium compounds are employed as they have seven unpaired electrons and work by creating local magnetic field disturbances at their sites of accumulation.

Indirect arthrography

Gadolinium compounds administered intravenously will be secreted through joint synovium into joint effusions resulting in indirect arthrography. However, there is no additional distension of the joint, which limits its effect.

Direct arthrography

Direct puncture of joints under image guidance with a solution containing dilute gadolinium (1:200 concentration) is routinely performed. This provides a positive contrast within the joint and distension of the joint capsule, thereby separating many of the closely applied soft-tissue structures that can be demonstrated on the subsequent MRI scan.

Clinical applications

Magnetic resonance imaging is becoming cheaper and more widely available. Its excellent anatomical detail, soft-tissue contrast and multiplanar capability make it ideal for non-invasive imaging of the musculoskeletal system (Figures 1.20 and 1.21). The multiplanar capability provides accurate cross-sectional information and the axial images in particular will reveal detailed limb compartmental anatomy. The excellent soft-tissue contrast allows identification of similar density soft tissues, for example in distinguishing between tendons, cartilage and ligaments. By using combinations of T1W, T2W and fat-suppressed sequences, specific abnormalities can be further characterized with tissue specificity, so further extending the diagnostic possibilities.

In orthopaedic surgery, MRI of the hip, knee, ankle, shoulder and wrist is now fairly commonplace. It can detect the early changes of bone marrow oedema and osteonecrosis before any other imaging

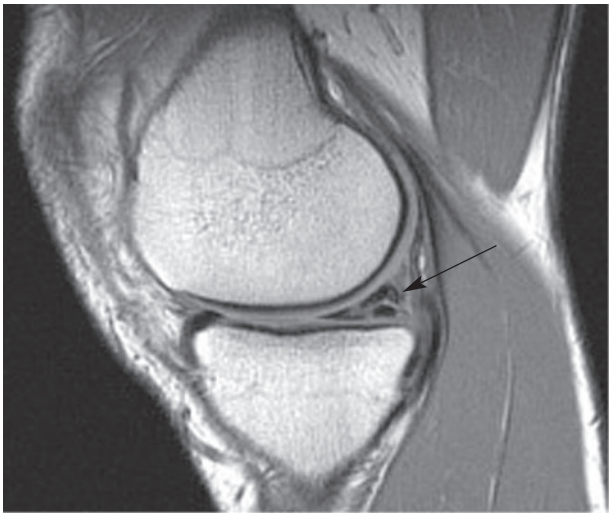
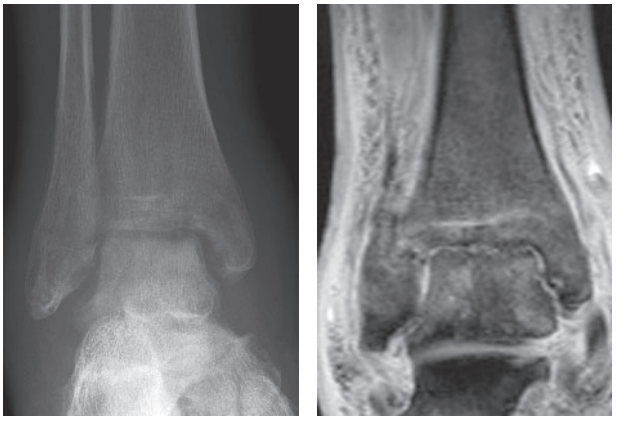


Figure 1.20 Magnetic resonance imaging MRI is ideal for displaying soft-tissue injuries, particularly tears of the menisci of the knee; this common injury is clearly shown in the picture.



(a) (b)

Figure 1.21 MRI A case of septic arthritis of the ankle, suspected from the plain X-ray (a) and confirmed by MRI (b).

modality. In the knee, MRI is as accurate as arthroscopy in diagnosing meniscal tears and cruciate ligament injuries. Bone and soft-tissue tumours should be routinely examined by MRI as the intraosseous and extraosseous extent and spread of disease, as well as the compartmental anatomy, can be accurately assessed. Additional use of fat-suppression sequences determines the extent of perilesional oedema and intravenous contrast will demonstrate the active part of the tumour.

Intravenous contrast is used to distinguish vascularized from avascular tissue (e.g. following a scaphoid fracture) or in defining active necrotic areas of tumour, or in demonstrating areas of active inflammation.

Direct MRI arthrography is used to distend the joint capsule and outline labral tears in the shoulder and the hip. In the ankle, it provides the way to demonstrate anterolateral impingement and assess the integrity of the capsular ligaments.

New generation MRI scanners

Many new scanners are being developed in the clinical setting using more powerful magnetic fields. Previously, the field strength was commonly between 0.5 and 1.5 Tesla. More recently, scanners using 3 Tesla have started being introduced. The increased field strength yields improved contrast and definition, but it is also more susceptible to artefacts.

Dedicated small-part scanners are also being introduced to assess limbs, for example for occult scaphoid fractures in the Emergency Department. Upright scanners have been developed to assess pathology that is apparent only when the patient is weight-bearing.

Limitations

Despite its undoubted value, MRI (like all singular methods of investigation) has its limitations and it must be seen as one of a group of imaging modalities, none of which by itself is appropriate in every situation. Conventional radiographs and CT are more sensitive to soft-tissue calcification and ossification, changes which can easily be easily overlooked on MRI. Conventional radiographs should therefore be used in combination with MRI to prevent such errors.

Diagnostic ultrasound

High-frequency sound waves, generated by a transducer, can penetrate several centimetres into the soft tissues; as they pass through the tissue interfaces, some of these waves are reflected back (like echoes) to the transducer, where they are registered as electrical signals and displayed as images on a screen. Unlike X-rays, the image does not depend on tissue density but rather on reflective surfaces and soft-tissue interfaces. This is the same principle as applies in sonar detection for ships or submarines.

Depending on their structure, different tissues are referred to as highly echogenic, mildly echogenic or echo-free. Fluid-filled cysts are echo-free; fat is highly echogenic; and semi-solid organs manifest varying degrees of 'echogenicity', which makes it possible to differentiate between them.

Real-time display on a monitor gives a dynamic image, which is more useful than the usual static images. A big advantage of this technique is that the equipment is simple and portable and can be used almost anywhere; another is that it is entirely harmless.

Clinical applications

Because of the marked echogenic contrast between cystic and solid masses, ultrasonography is particularly useful for identifying hidden 'cystic' lesions such as haematomas, abscesses, popliteal cysts and arterial aneurysms. It is also capable of detecting intra-articular fluid and may be used to diagnose a synovial effusion or to monitor the progress of an 'irritable hip'.

Ultrasound is commonly used for assessing tendons and diagnosing conditions such as tendinitis and partial or complete tears. The rotator cuff, patellar ligament, quadriceps tendon, Achilles tendon, flexor tendons and peroneal tendons are typical examples. The same technique is used extensively for guiding needle placement in diagnostic and therapeutic joint and soft-tissue injections. Another important application is in the screening of newborn babies for congenital dislocation (or dysplasia) of the hip; the cartilaginous femoral head and acetabulum (which are, of course, 'invisible' on X-ray) can be clearly identified, and their relationship to each other shows whether the hip is normal or abnormal.

Ultrasound imaging is quick, cheap, simple and readily available. However, the information obtained is highly operator-dependent, relying on the experience and interpretation of the technician.

Doppler ultrasound

Blood flow can be detected by using the principle of a change in frequency of sound when material is moving towards or away from the ultrasound transducer. This is the same principle as the change in frequency of the noise from a passing fire engine when travelling towards and then away from an observer. Abnormal increased blood flow can be observed in areas of inflammation or in aggressive tumours. Different flow rates can be shown by different colour representations ('colour Doppler').

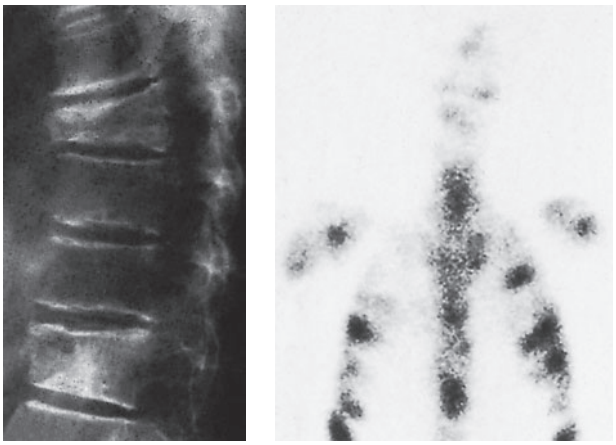
Radionuclide imaging

Photon emission by radionuclides taken up in specific tissues can be recorded by a gamma camera to produce an image which reflects physiological activity in that tissue or organ. The radiopharmaceutical used for radionuclide imaging has two components: a chemical compound that is chosen for its metabolic uptake in the target tissue or organ, and a radioisotope tracer that will emit photons for detection.

Isotope bone scans

For bone imaging, the ideal isotope is technetium-99m (^{99m}Tc): it has the appropriate energy characteristics for gamma camera imaging, it has a relatively short half-life (6 hours) and it is rapidly excreted in the urine. A bone-seeking phosphate compound is used as the substrate as it is selectively taken up and concentrated in bone. The low background radioactivity means that any site of increased uptake is readily visible (Figure 1.22).

Technetium-labelled hydroxymethylene diphosphonate (^{99m}Tc-HDP) is injected intravenously and its activity is recorded at two stages: (1) the early perfusion phase, shortly after injection, while the isotope is still in the blood stream or the perivascular space thus reflecting local blood flow difference; and (2) the delayed bone phase, 3 hours later, when the isotope has been taken up in bone tissue. Normally, in the early perfusion phase the vascular soft tissues around the joints produce the sharpest (most active) image; 3 hours later this activity has faded and the bone outlines are shown more clearly, the greatest activity appearing in the cancellous tissue at the ends of the long bones.



(a) (b)

Figure 1.22 Radionuclide scanning (a) The plain X-ray showed a pathological fracture, probably through a metastatic tumour. (b) The bone scan revealed generalized secondaries, here involving the spine and ribs.

Changes in radioactivity are most significant when they are localized or asymmetrical. The following four types of abnormality are seen.

Increased activity in the perfusion phase This is due to increased soft-tissue blood flow, suggesting inflammation (e.g. acute or chronic synovitis), a fracture, a highly vascular tumour or regional sympathetic dystrophy.

Decreased activity in the perfusion phase This is much less common and signifies local vascular insufficiency.

Increased activity in the delayed bone phase This could be due either to excessive isotope uptake in the osseous extracellular fluid or to more avid incorporation into newly forming bone tissue; either would be likely in a fracture, implant loosening, infection, a local tumour or healing after necrosis, and nothing in the bone scan itself distinguishes between these conditions.

Diminished activity in the bone phase This is due to an absent blood supply (e.g. in the femoral head after a fracture of the femoral neck) or to replacement of bone by pathological tissue.

CLINICAL APPLICATIONS

Radionuclide imaging is useful in many situations: (1) the diagnosis of stress fractures or other undisplaced fractures that are not detectable on the plain X-ray; (2) the detection of a small bone abscess, or an osteoid osteoma; (3) the investigation of loosening or infection around prostheses; (4) the diagnosis of femoral head ischaemia in Perthes' disease or avascular necrosis in adults; (5) the early detection of bone metastases. The scintigraphic appearances in these conditions are described in the relevant chapters. In most cases the isotope scan serves chiefly to pinpoint the site of abnormality and it should always be viewed in conjunction with other modes of imaging.

Bone scintigraphy is relatively sensitive but nonspecific. One advantage is that the whole body can be imaged to look for multiple sites of pathology (occult metastases, multifocal infection and multiple occult fractures). It is also one of the only techniques to give information about physiological activity in the tissues being examined (essentially osteoblastic activity). However, the technique carries a significant radiation burden (equivalent to approximately 200 chest X-rays) and the images yielded make anatomical localization difficult (poor spatial resolution). For localized problems MRI has superseded bone scintigraphy as it yields much greater specificity due to its superior anatomical depiction and tissue specificity.

Other radionuclide compounds

Gallium-67 (^{67}Ga) Gallium-67 concentrates in inflammatory cells and has been used to identify sites of hidden infection: for example, in the investigation of prosthetic loosening after joint replacement. However, it is arguable whether it gives any more reliable information than the ^{99m}Tc bone scan.

Indium-111-labelled leucocytes (^{111}I) The patient's own white blood cells are removed and labelled with indium-111 before being re-injected into the patient's bloodstream. Preferential uptake in areas of infection is expected, thereby hoping to distinguish sites of active infection from chronic inflammation. For example, white cell uptake is more likely to be seen with an infected total hip replacement as opposed to mechanical loosening. However, as this technique is expensive and still not completely specific, it is seldom performed.

Single-photon emission computed tomography (SPECT)

Single-photon emission computed tomography (SPECT) is essentially a bone scan in which images are recorded and displayed in all three orthogonal planes. Coronal, sagittal and axial images at multiple levels make spatial localization of pathology possible: for example, activity in one side of a lumbar vertebra on the planar images can be further localized to the body, pedicle or lamina of the vertebra on the SPECT images.

Positron emission tomography (PET)

Positron emission tomography (PET) is an advanced nuclear medicine technique that allows functional imaging of disease processes. Positron-emitting isotopes with short half-lives are produced on site at specialist centres using a cyclotron. Various radiopharmaceuticals can be employed, but currently the most commonly used is 18-fluoro-2-deoxy-D-glucose (^{18}FDG). The ^{18}FDG is accumulated in different parts of the body where it can effectively measure the rate of consumption of glucose. Malignant tumours metabolize glucose at a faster rate than benign tumours and PET scanners are extremely useful in looking for occult sites of disease around the body on this basis.

PET/CT is a hybrid examination performing both PET and CT on the patient in order to superimpose the two images produced. The combination of these two techniques uses the sensitivity of PET for functional tissue changes and the cross-sectional anatomy detail of CT to localize the position of this activity.

PET is useful in oncology to identify occult malignant tumours and metastases and more accurately 'stage' the disease. Furthermore, activity levels at known sites of disease can be used to assess treatment and distinguish 'active' residual tumour or tumour recurrence from 'inactive' post-surgical scarring and necrotic tumour.

Bone mineral densitometry

Bone mineral density (BMD) measurement is now widely used in identifying patients with osteoporosis and an increased risk of osteoporotic fractures. Various techniques have been developed, including radiographic absorptiometry (RA), quantitative computed tomography (QCT) and quantitative ultrasonometry (QUS). However, the most widely used technique is dual energy X-ray absorptiometry (DXA) (Figure 1.23).

RA uses conventional radiographic equipment and measures bone density in the phalanges. QCT measures trabecular bone density in vertebral bodies, but is not widely available and involves a higher dose of ionizing radiation than DXA. QUS assesses bone mineral density in the peripheral skeleton (e.g. the wrist and calcaneus) by measuring both the attenuation of ultrasound and the variation of speed of sound through the bone.

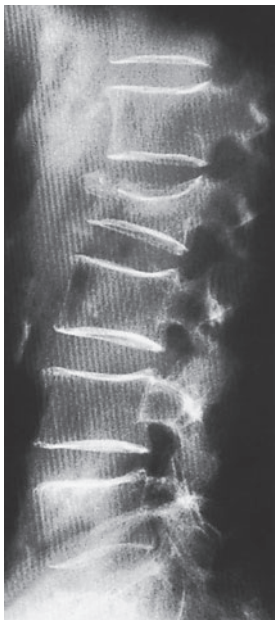
DXA employs columnated low-dose X-ray beams of two different energy levels in order to distinguish the density of bone from that of soft tissue. Although this involves the use of ionizing radiation, it is an extremely low dose. A further advantage of DXA is the development of a huge international database that allows expression of bone mineral density values in comparison both to an age- and sex-matched population (Z score) and also to the peak adult bone mass (T score). The T score in particular allows calculation of relative fracture risk. Individual values for both the lumbar spine and hips are obtained as there is often a discrepancy between these two sites and the fracture risk is more directly related to the value at the target area. By World Health Organization (WHO) criteria, T scores of < -1.0 indicate 'osteopenia' and T scores of < -2.5 indicate 'osteoporosis'.

BLOOD TESTS

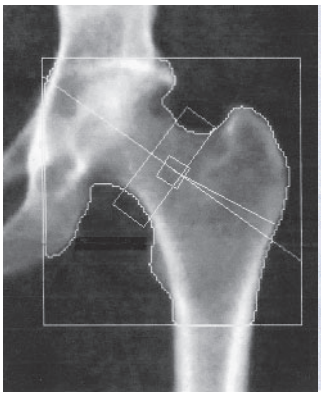
Non-specific blood tests

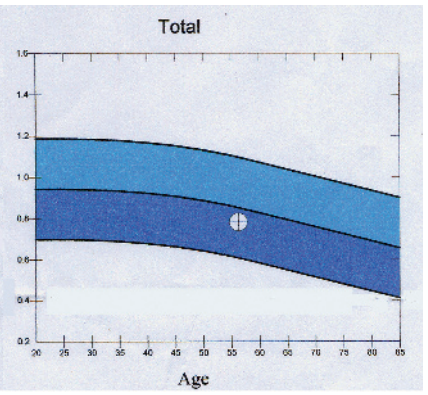
Non-specific blood abnormalities are common in bone and joint disorders; their interpretation hinges on the clinical and X-ray findings.

Hypochromic anaemia This is usual in rheumatoid arthritis, but it may also be a consequence of



(a)





DXA Results Summary:

Region	Area (cm^2)	BMC (g)	BMD (g/cm^2)	T score	PR(%)	Z score	AM(%)
Neck	5.37	3.62	0.675	–1.6	79	–0.5	93
Troch	12.48	7.10	0.569	–1.3	81	–0.6	90
Inter	20.07	18.92	0.943	–1.0	86	–0.5	92
Total	**37.92**	**29.64**	**0.782**	**–1.3**	**83**	**–0.6**	**92**
Ward's	1.03	0.54	0.527	–1.8	72	0.0	100

Total BMD CV 1.)%
WHO Classification: Osteopenia
Fracture Risk: Increased

(b)

Figure 1.23 Measurement of bone mass (a) X-ray of the lumbar spine shows a compression fracture of L2. The general loss of bone density accentuates the cortical outlines of the vertebral body end-plates. These features are characteristic of diminished bone mass, which can be measured accurately by dual energy X-ray absorptiometry. (b) DXA scan from another woman who attended for monitoring at the onset of the menopause.

gastrointestinal bleeding due to the anti-inflammatory drugs.

Leucocytosis Although generally associated with infection, a mild leucocytosis is not uncommon in rheumatoid arthritis and during an attack of gout.

The erythrocyte sedimentation rate (ESR) ESR is usually increased in acute and chronic inflammatory disorders and after tissue injury. However, patients with low-grade infection may have a normal ESR and this should not be taken as a reassuring sign. The ESR is strongly affected by the presence of monoclonal immunoglobulins; a high ESR is almost mandatory in the diagnosis of myelomatosis.

C-reactive protein (and other acute phase proteins) These may be abnormally increased in chronic inflammatory arthritis and (temporarily) after injury or operation. The test is often used to monitor the progress and activity of rheumatoid arthritis and chronic infection.

Plasma gamma globulins Measured by protein electrophoresis. Their precise characterization is helpful in the assessment of certain rheumatic disorders, and more particularly in the diagnosis of myelomatosis.

Rheumatoid factor tests

Rheumatoid factor, an IgM autoantibody, is present in about 75% of adults with rheumatoid arthritis. However, it is not pathognomonic: some patients with undoubted rheumatoid arthritis remain 'seronegative', while rheumatoid factor is found in some patients with other disorders such as systemic lupus erythematosus and scleroderma.

Ankylosing spondylitis, Reiter's disease and psoriatic arthritis characteristically test negative for rheumatoid factor; they have been grouped together as the 'seronegative spondarthritides'.

Tissue typing

Human leucocyte antigens (HLA) can be detected in white blood cells and they are used to characterize individual tissue types. The seronegative spondarthritides are closely associated with the presence of HLAB27 on chromosome 6; this is frequently used as a confirmatory test in patients suspected of having ankylosing spondylitis or Reiter's disease, but it should not be regarded as a specific test because it is positive in about 8% of normal Western Europeans.

Biochemistry

Biochemical tests are essential in monitoring patients after any serious injury. They are also used routinely in the investigation of rheumatic disorders and abnormalities of bone metabolism. Their significance is discussed under the relevant conditions.

SYNOVIAL FLUID ANALYSIS

Arthrocentesis and synovial fluid analysis is a much neglected diagnostic procedure; given the correct indications, it can yield valuable information. It should be considered in the following conditions:

Acute joint swelling after injury The distinction between synovitis and bleeding may not be obvious; aspiration will settle the question immediately.

Acute atraumatic synovitis in adults Synovial fluid analysis may be the only way to distinguish between infection, gout and pseudogout. Characteristic crystals can be identified on polarized light microscopy.

Suspected infection Careful examination and laboratory investigations may provide the answer, but they take time. Joint aspiration is essential for early diagnosis.

Chronic synovitis Here joint aspiration is less urgent, and is only one of many diagnostic procedures in the investigation of suspected tuberculosis or atypical rheumatic disorders.

Technique

Joint aspiration should always be performed under strict aseptic conditions. After infiltrating the skin with a local anaesthetic, a 20-gauge needle is introduced and a sample of joint fluid is aspirated; even a small quantity of fluid (less than 0.5 mL) is enough for diagnostic analysis.

The volume of fluid and its appearance are immediately noted. Normal synovial fluid is clear and slightly yellow. A cloudy or turbid fluid is due to the presence of cells, usually a sign of inflammation. Blood-stained fluid may be found after injury, but is also seen in acute inflammatory disorders and in pigmented villonodular synovitis.

A single drop of fresh synovial fluid is placed on a glass slide and examined through the microscope. Blood cells are easily identified; abundant leucocytes may suggest infection. Crystals may be seen, though this usually requires a careful search; they are better characterized by polarized light microscopy (see Chapter 4).

Dry smears are prepared with heparinized fluid; more concentrated specimens can be obtained if the fluid is centrifuged. After suitable staining (Wright's and Gram's), the smear is examined for pus cells and organisms. Remember, though, that negative findings do not exclude infection.

Laboratory tests

If enough fluid is available, it is sent for full laboratory investigation (cells, biochemistry and bacteriological culture; see Table 1.3). A simultaneous blood specimen allows comparison of synovial and blood glucose concentration; a marked reduction of synovial glucose suggests infection.

A high white cell count (more than 10 000/mm^3) is usually indicative of infection, but a moderate leucocytosis is also seen in gout and other types of inflammatory arthritis.

Bacteriological culture and tests for antibiotic sensitivity are essential in any case of suspected infection.

BONE BIOPSY

Bone biopsy is often the crucial means of making a diagnosis or distinguishing between local conditions that closely resemble one another. Confusion is most likely to occur when the X-ray or MRI discloses an area of bone destruction that could be due to a compression fracture, a bone tumour or infection (e.g. a collapsed vertebral body). In other cases it is obvious that the lesion is a tumour – but what type of tumour? Benign or malignant? Primary or metastatic? Radical surgery should never be undertaken for a suspected neoplasm without first confirming the diagnosis histologically, no matter how 'typical' or 'obvious' the X-ray appearances may be.

In bone infection, the biopsy permits not only histological proof of acute inflammation but also bacteriological typing of the organism and tests for antibiotic sensitivity.

Table 1.3 Examination of synovial fluid

Suspected condition	Appearance	Viscosity	White cells	Crystals	Biochemistry	Bacteriology
Normal	Clear yellow	High	Few	–	As for plasma	–
Septic arthritis	Purulent	Low	+	–	Glucose low	+
Tuberculous arthritis	Turbid	Low	+	–	Glucose low	+
Rheumatoid arthritis	Cloudy	Low	++	–	–	–
Gout	Cloudy	Normal	++	Urate	–	–
Pseudogout	Cloudy	Normal	+	Pyrophosphate	–	–
Osteoarthritis	Clear yellow	High	Few	Often +	–	–

The investigation of metabolic bone disease sometimes calls for a tetracycline-labelled bone biopsy to show: (a) the type of abnormality (osteoporosis, osteomalacia, hyperparathyroidism), and (b) the severity of the disorder.

Open or closed?

Open biopsy, with exposure of the lesion and excision of a sizeable portion of the bone, seems preferable, but it has several drawbacks:

- It requires an operation, with the attendant risks of anaesthesia and infection.
- New tissue planes are opened up, predisposing to spread of infection or tumour.
- The biopsy incision may jeopardize subsequent wide excision of the lesion.
- The more inaccessible lesions (e.g. a tumour of the acetabular floor) can be reached only by dissecting widely through healthy tissue.

A carefully performed 'closed' biopsy, using a needle or trephine of appropriate size to ensure the removal of an adequate sample of tissue, is the procedure of choice except when the lesion cannot be accurately localized or when the tissue consistency is such that a sufficient sample cannot be obtained. Solid or semi-solid tissue is removed intact by the cutting needle or trephine; fluid material can be aspirated through the biopsy needle.

Precautions

- The biopsy site and approach should be carefully planned with the aid of X-rays or other imaging techniques.
- If there is any possibility of the lesion being malignant, the approach should be sited so that the wound and biopsy track can be excised if later radical surgery proves to be necessary.
- The procedure should be carried out in an operating theatre, under anaesthesia (local or general) and with full aseptic technique.
- For deep-seated lesions, fluoroscopic control of the needle insertion is essential.
- The appropriate size of biopsy needle or cutting trephine should be selected.
- A knowledge of the local anatomy and of the likely consistency of the lesion is important. Large blood vessels and nerves must be avoided; potentially vascular tumours may bleed profusely and the means to control haemorrhage should be readily to hand. More than one surgeon has set out to aspirate an 'abscess' only to plunge a wide-bore needle into an aneurysm!
- Clear instructions should be given to ensure that the tissue obtained at the biopsy is suitably processed. If infection is suspected, the material should go into a culture tube and be sent to the laboratory as soon as possible. A smear may also be useful. Whole tissue is transferred to a jar containing formalin, without damaging the specimen or losing any material. Aspirated blood should be allowed to clot and can then be preserved in formalin for later paraffin embedding and sectioning. Tissue thought to contain crystals should not be placed in formalin as this may destroy the crystals; it should be either kept unaltered for immediate examination or stored in saline.
- No matter how careful the biopsy, there is always the risk that the tissue will be too scanty or too unrepresentative for accurate diagnosis. Close consultation with the radiologist and pathologist beforehand will minimize this possibility. In the best hands, needle biopsy has an accuracy rate of over 95%.

DIAGNOSTIC ARTHROSCOPY

Arthroscopy is performed for both diagnostic and therapeutic reasons. Almost any joint can be reached but the procedure is most usefully employed in the knee, shoulder, wrist, ankle and hip. If the suspect lesion is amenable to surgery, it can often be dealt with at the same sitting without the need for an open operation. However, arthroscopy is an invasive procedure and its mastery requires skill and practice; it should not be used simply as an alternative to clinical examination and imaging.

Technique

The instrument is basically a rigid telescope fitted with fibreoptic illumination. Tube diameter ranges from about 2 mm (for small joints) to 4–5 mm (for the knee). It carries a lens system that gives a magnified image. The eyepiece allows direct viewing by the arthroscopist, but it is far more convenient to fit a small, sterilizable solid-state television camera which produces a picture of the joint interior on a television monitor.

The procedure is best carried out under general anaesthesia; this gives good muscle relaxation and permits manipulation and opening of the joint compartments. The joint is distended with fluid and the arthroscope is introduced percutaneously. Various instruments (probes, curettes and forceps) can be inserted through other skin portals; they are used to help expose the less accessible parts of the joint, or to obtain biopsies for further examination. Guided by the image on the monitor, the arthroscopist explores the joint in a systematic fashion, manipulating the arthroscope with one hand and the probe or forceps

with the other. At the end of the procedure the joint is washed out and the small skin wounds are sutured. The patient is usually able to return home later the same day.

Diagnosis

The knee is the most accessible joint. The appearance of the synovium and the articular surfaces usually allows differentiation between inflammatory and non-inflammatory, destructive and non-destructive lesions. Meniscal tears can be diagnosed and treated immediately by repair or removal of partially detached segments. Cruciate ligament deficiency, osteocartilaginous fractures, cartilaginous loose bodies and synovial 'tumours' are also readily visualized.

Arthroscopy of the shoulder is more difficult, but the articular surfaces and glenoid labrum can be adequately explored. Rotator cuff lesions can often be diagnosed and treated at the same time. Arthroscopy of the wrist is useful for diagnosing torn triangular fibrocartilage and interosseous ligament ruptures.

Arthroscopy of the hip is becoming more common and is proving to be useful in the diagnosis of unexplained hip pain. Labral tears, synovial lesions, loose bodies and articular cartilage damage (all of which are difficult to detect by conventional imaging techniques) have been diagnosed with a reported accuracy rate of over 50%.

Complications

Diagnostic arthroscopy is safe but not entirely free of complications, the commonest of which are haemarthosis, thrombophlebitis, infection and joint stiffness (particularly contracture of the anterior capsule). There is also a significant incidence of algodystrophy following arthroscopy.

FURTHER READING

Apley AG, Solomon L. *Physical Examination in Orthopaedics.* Oxford: Butterworth Heinemann, 1997.

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Infection

2

Enrique Gómez-Barrena

Infection – as distinct from mere residence of microorganisms – is a condition in which pathogenic microorganisms multiply and spread within the body tissues. Microorganisms may reach the musculoskeletal tissues by:

- *direct introduction* through the skin (a pinprick, an injection, a stab wound, a laceration, an open fracture or an operation, particularly when biomaterials are implanted),
- *direct spread from a contiguous focus of infection*, or
- *indirect spread via the bloodstream* from a distant site such as the nose or mouth, the respiratory tract, the bowel or the genitourinary tract.

Depending on the type of invader, the site of infection and the host response, the result may be a pyogenic osteomyelitis, a septic arthritis, a chronic granulomatous reaction (classically seen in tuberculosis of either bone or joint), or an indolent response to a less aggressive organism (as in low-grade periprosthetic infections) or to an unusual organism (e.g. a fungal infection). Soft-tissue infections range from superficial wound sepsis to widespread cellulitis and life-threatening necrotizing fasciitis. Parasitic lesions such as hydatid disease also are considered in this chapter, although these are infestations rather than infections.

Clinical aspects of infection will be particularly developed in this chapter. The team approach, including microbiologists, infectious disease and internal medicine doctors, is certainly the basis for success in infection. However, many cases of musculoskeletal infection need the leadership of the orthopaedic surgeon to ensure timely diagnosis and treatment of the patient with musculoskeletal symptoms and signs that suggest infection.

GENERAL ASPECTS OF INFECTION

Infection usually gives rise to an acute or chronic *inflammatory reaction*, which is the body's way of combating the invaders by destroying them, or at least immobilizing and confining them to a restricted area. The classical signs of inflammation are frequently present (*redness*, *swelling*, *heat*, *pain* and *loss of function*) and offer clinical clues about the infection and the patient's reaction.

Bone infection differs from soft-tissue infection since bone consists of a collection of rigid compartments. Bone is thus more susceptible than soft tissues to vascular damage and cell death due to pressure in acute inflammation. Unless it is rapidly suppressed, bone infection will inevitably lead to necrosis. *Osteomyelitis* is infection of bone and frequently seeds in trabecular areas affecting both bone and bone marrow. Soft-tissue infection depends on the main affected tissue, but of special interest to the orthopaedic surgeon is joint infection or *infectious arthritis*, whether *septic arthritis* or *granulomatous arthritis*. All these forms of infection will be addressed below.

Host susceptibility to infection is increased by (a) *local factors* such as trauma, scar tissue, poor circulation, diminished sensibility, chronic bone or joint disease and the presence of foreign bodies including implants, as well as (b) *systemic factors* such as malnutrition, general illness, debility, diabetes, rheumatoid disease, corticosteroid administration and all forms of immunosuppression, either acquired or induced.

BOX 2.1 FACTORS PREDISPOSING TO BONE INFECTION

Malnutrition and general debility
Diabetes mellitus
Corticosteroid administration
Immune deficiency
Immunosuppressive drugs
Venous stasis in the limb
Peripheral vascular disease
Loss of sensibility
Iatrogenic invasive measures
Trauma

Resistance is also diminished in the very young and the very old.

Bacterial colonization and resistance to antibiotics is enhanced by the ability of certain microorganisms (including *Staphylococcus*) to adhere to avascular bone surfaces and foreign implants, protected from both host defences and antibiotics by a protein–polysaccharide slime (*glycocalyx* or *biofilm*). Biofilm formation aids the development of a complex bacterial community that protects microorganisms adherent to biomaterials. Biofilm maturation with microorganism release further expands this colonization, and eradication of biofilm-forming microorganisms becomes impossible without implant removal or exchange. Thus, bacterial adherence to biomaterials and biofilm formation are crucial aspects to consider when treating musculoskeletal infections in the presence of implants.

Acute pyogenic bone infections are characterized by the formation of pus – a concentrate of defunct leucocytes, dead and dying bacteria and tissue debris – which is often localized in an abscess. Pressure builds up within the abscess and infection may then extend into a contiguous joint or through the cortex and along adjacent tissue planes. It may also spread further afield via lymphatics (causing lymphangitis and lymphadenopathy) or via the bloodstream (bacteraemia and septicaemia). An accompanying systemic reaction varies from a vague feeling of lassitude with mild pyrexia to severe illness, fever, toxaemia and shock. The generalized effects are due to the release of bacterial enzymes and endotoxins as well as cellular breakdown products from the host tissues.

Chronic pyogenic infection may follow unresolved acute infection and is characterized by persistence of the infecting organism (or, more frequently, multiple microorganisms) in pockets of necrotic tissue. Purulent material accumulates and may be discharged through sinuses at the skin or a poorly healed wound. Factors which predispose to this outcome are the presence of damaged muscle, dead bone (*sequestrum*) or a foreign implant, diminished local blood supply and a weak host response. Resistance is likely to be depressed in the very young and the very old, in states of malnutrition or immunosuppression, and in certain diseases such as diabetes and leukaemia.

Chronic non-pyogenic infection may result from invasion by organisms that produce a cellular reaction leading to the formation of granulomas consisting largely of lymphocytes, modified macrophages and multinucleated giant cells; this type of granulomatous infection is seen most typically in tuberculosis. Systemic effects are less acute but may ultimately be very debilitating, with lymphadenopathy, splenomegaly and tissue wasting.

Treatment

1 Identify the infecting organism and administer effective antibiotic treatment or chemotherapy.
2 Provide analgesia and general supportive measures, including rest of the affected part or splintage of the affected joint.
3 Release pus as soon as it is detected.
4 Eradicate avascular and necrotic tissue.
5 Stabilize the bone if it has fractured and restore continuity if there is a gap in the bone.
6 Maintain or regain soft-tissue and skin cover.

If treated early with effective antibiotics, acute infections can usually be cured. Once there is pus and bone necrosis, operative drainage will be needed.

When treating patients with bone or joint infection, it is wise to maintain continuous collaboration with a specialist in microbiology.

ACUTE HAEMATOGENOUS OSTEOMYELITIS

Aetiology and pathogenesis

Acute haematogenous osteomyelitis is mainly a disease of children. When adults are affected, it is usually because their resistance is lowered. Trauma may determine the site of infection, possibly by causing a small haematoma or fluid collection in a bone, in patients with concurrent bacteraemia. The incidence of acute haematogenous osteomyelitis in Western European children is thought to have declined in recent years, probably a reflection of improving social conditions. Different studies from the United Kingdom confirm a low incidence (less than 1 case per 100 000) in most recent periods although it is almost certainly much higher among less affluent populations. Also, a decrease in surgical treatment of those cases has been identified, possibly related to earlier and more effective antibiotic treatment.

The causal organism in both adults and children is usually *Staphylococcus aureus* (found in over 70% of cases), and less often one of the other Gram-positive cocci, such as the Group A beta-haemolytic streptococcus (*Streptococcus pyogenes*) which is found in chronic skin infections, as well as Group B streptococcus (especially in newborn babies) or the alphahaemolytic diplococcus *S. pneumoniae*. In children between 1 and 4 years of age, the Gram-negative *Haemophilus influenzae* used to be a fairly common pathogen for osteomyelitis and septic arthritis, but the introduction of *H. influenzae* type B vaccination in the 1990s has been followed by a much reduced incidence of this infection in many countries. In recent years its place has been taken by the increasing presence of *Kingella kingae*, mainly following upper respiratory infection in young children. Other Gram-negative organisms (e.g. *Escherichia coli*, *Pseudomonas aeruginosa*, *Proteus mirabilis* and the anaerobic *Bacteroides fragilis*) occasionally cause acute bone infection.

Curiously, patients with sickle-cell disease are prone to infection by *Salmonella typhi*. Anaerobic organisms (particularly *Peptococcus magnus*) have been found in patients with osteomyelitis, usually as part of a mixed infection. Unusual organisms are more likely to be found in heroin addicts and as opportunistic pathogens in patients with compromised immune defence mechanisms.

The bloodstream is invaded, perhaps from a minor skin abrasion, treading on a sharp object, an injection point, a boil, a septic tooth or – in the newborn – from an infected umbilical cord. *In adults*, the source of infection may be a urethral catheter, an indwelling arterial line or a contaminated needle and syringe. *In children*, the infection usually starts in the vascular metaphysis of a long bone, most often in the proximal tibia or in the distal or proximal ends of the femur. Predilection for this site has traditionally been attributed to the peculiar arrangement of the blood vessels in that area: the non-anastomosing terminal branches of the nutrient artery twist back in hairpin loops before entering the large network of sinusoidal veins; the relative vascular stasis and consequent lowered oxygen tension are believed to favour bacterial colonization. The structure of the fine vessels in the hypertrophic zone of the physis may more easily allow bacteria to pass through and adhere to type 1 collagen in that area. *In infants*, in whom there are still anastomoses between metaphyseal and epiphyseal blood vessels, infection can also reach the epiphysis (Figure 2.1). *In adults*, haematogenous infection accounts for only about 20% of cases of osteomyelitis. *Staphylococcus aureus* is the commonest organism but *Pseudomonas aeruginosa* often appears in patients using intravenous drugs. Adults with diabetes and vascular disease, who are prone to soft-tissue infections of the foot, may develop contiguous bone infection involving a variety of organisms.

Pathology

Acute haematogenous osteomyelitis shows a characteristic progression marked by *inflammation*, *suppuration*, *bone necrosis*, *reactive new bone formation* and, ultimately, *resolution and healing* or else intractable *chronicity*. However, the pathological picture varies considerably, depending on the patient's age, the site of infection, the virulence of the organism and the host response.

ACUTE OSTEOMYELITIS IN CHILDREN

The 'classical' picture is seen in children between 2 and 6 years. The earliest change in the metaphysis is an acute inflammatory reaction with vascular congestion, exudation of fluid and infiltration by polymorphonuclear leucocytes. The intraosseous pressure rises rapidly, causing intense pain, obstruction to blood flow and intravascular thrombosis. Even at an early stage, the bone tissue is threatened by impending ischaemia and resorption due to a combination of phagocytic activity and the local accumulation of cytokines, growth factors, prostaglandin and bacterial enzymes. By the second or third day, pus forms within the bone and forces its way along the Volkmann canals to the surface where it produces a subperiosteal abscess. This is much more evident in children, because of the relatively loose attachment of the periosteum, than in adults. From the subperiosteal abscess, pus can spread along the shaft, to re-enter the bone at another level or burst into the surrounding soft tissues. The developing physis acts as a barrier to direct spread towards the epiphysis, but where the metaphysis is partly

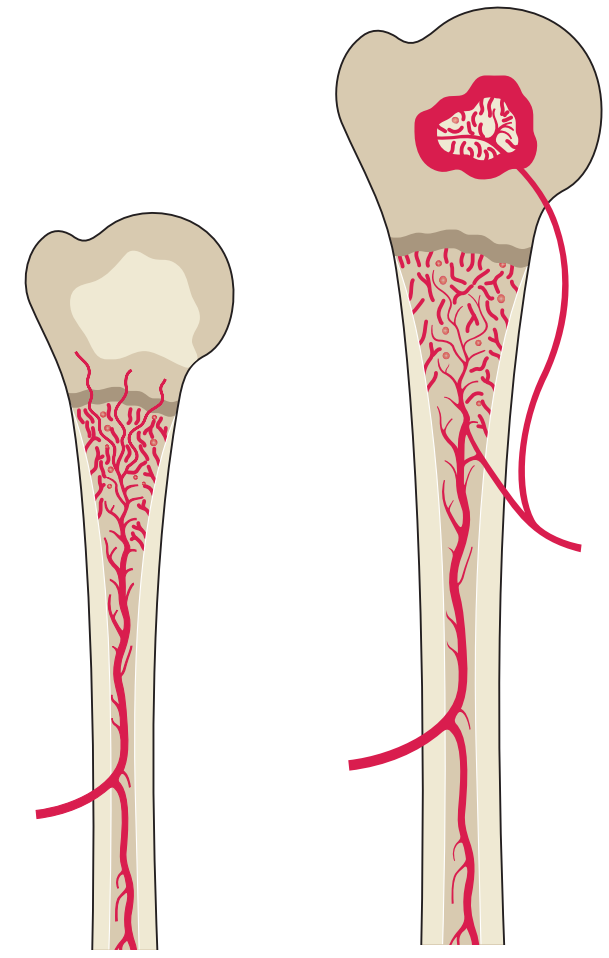


Figure 2.1 Epiphyseal and metaphyseal blood supply (a) In newborn infants some metaphyseal arterioles from the nutrient artery penetrate the physis and may carry infection directly from the metaphysis to the epiphysis. (b) In older children the physis acts as a barrier and the developing epiphysis receives a separate blood supply from the epiphyseal and periarticular blood vessels.

intracapsular (e.g. at the hip, shoulder or elbow) pus may discharge through the periosteum into the joint.

The rising intraosseous pressure, vascular stasis, small-vessel thrombosis and periosteal stripping increasingly compromise the blood supply; by the end of a week there is usually microscopic evidence of bone death. Bacterial toxins and leucocytic enzymes also may play their part in the advancing tissue destruction. With the gradual ingrowth of granulation tissue the boundary between living and devitalized bone becomes defined. Pieces of dead bone may separate as *sequestra* varying in size from mere spicules to large necrotic segments of the cortex in neglected cases.

Macrophages and lymphocytes arrive in increasing numbers and the debris is slowly removed by a combination of phagocytosis and osteoclastic resorption. A small focus in cancellous bone may be completely resorbed, leaving a tiny cavity, but a large cortical or cortico-cancellous sequestrum will remain entombed, inaccessible to either final destruction or repair.

Another feature of advancing acute osteomyelitis is new bone formation. Initially, the area around the infected zone is porotic (probably due to hyperaemia and osteoclastic activity) but if the pus is not released, either spontaneously or by surgical decompression, new bone starts forming on viable surfaces in the bone and from the deep layers of the stripped periosteum. This is typical of pyogenic infection, and fine streaks of subperiosteal new bone usually become apparent on X-ray by the end of the second week. With time, this new bone thickens to form a casement, or *involucrum*, enclosing the sequestrum and infected tissue. If the infection persists, pus and tiny sequestrated spicules of bone may discharge through perforations (*cloacae*) in the involucrum and track by sinuses to the skin surface (Figure 2.2).

If the infection is controlled and intraosseous pressure released at an early stage, this dire progress can be halted. The bone around the zone of infection becomes increasingly dense; this, together with the periosteal reaction, results in thickening of the bone. In some cases the normal anatomy may eventually be reconstituted; in others, though healing is sound, the bone is left permanently deformed.

If healing does not occur, a nidus of infection may remain locked inside the bone, causing pus and sometimes bone debris to be discharged intermittently through a persistent sinus (or several sinuses). The infection has now lapsed into *chronic osteomyelitis*, which may last for many years.

ACUTE OSTEOMYELITIS IN INFANTS

The early features of acute osteomyelitis in infants are much the same as those in older children. However, a significant difference, during the first year of life,

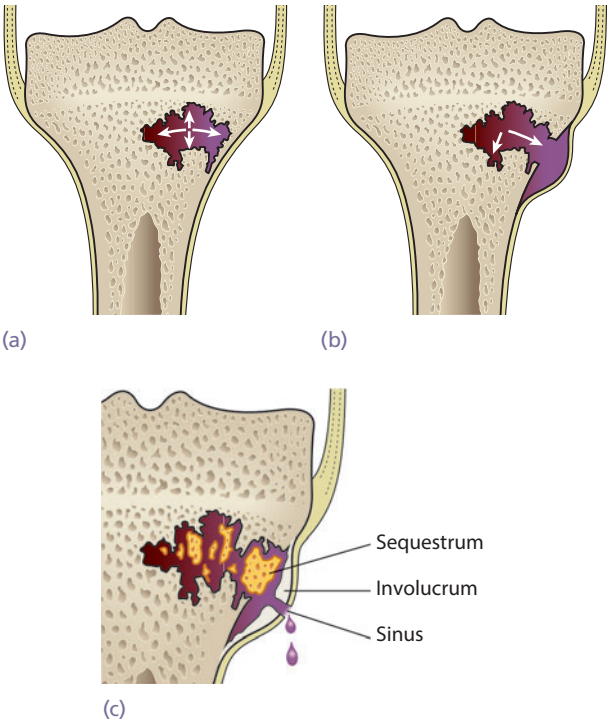


Figure 2.2 Acute osteomyelitis (a) Infection in the metaphysis may spread towards the surface, to form a subperiosteal abscess (b). Some of the bone may die and is encased in periosteal new bone as a sequestrum (c). The encasing involucrum is sometimes perforated by sinuses.

is the frequency with which the metaphyseal infection spreads to the epiphysis and from there into the adjacent joint. In the process, the physeal anlage may be irreparably damaged, further growth at that site is severely retarded and the joint will be permanently deformed. How this comes about is still argued over. It has long been held that, during the first 6–9 months of life, small metaphyseal vessels penetrate the physeal cartilage and this may permit the infection to spread into the cartilaginous epiphysis, although definitive proof of this mechanism has not been shown. Whatever the mechanism, what is indisputable is that osteomyelitis and septic arthritis often go together during infancy. Another feature in infants is an unusually exuberant periosteal reaction resulting in sometimes bizarre new bone formation along the diaphysis; fortunately, with longitudinal growth and remodelling, the diaphyseal anatomy is gradually restored.

ACUTE OSTEOMYELITIS IN ADULTS

Bone infection in the adult usually follows an open injury, an operation or spread from a contiguous focus of infection (e.g. a neuropathic ulcer or an infected diabetic foot) in over 70% of the cases. True haematogenous osteomyelitis is uncommon and, when it

does occur, it usually affects one of the vertebrae (e.g. following a pelvic infection), a metaphyseal region of a long bone or a small cuboidal bone. A vertebral infection may spread through the end plate and the intervertebral disc into an adjacent vertebral body. If a long bone is infected, the abscess is likely to spread within the medullary cavity, eroding the cortex and extending into the surrounding soft tissues. Periosteal new bone formation is less obvious than in childhood and the weakened cortex may fracture. If the bone end becomes involved, there is also a risk of the infection spreading into an adjacent joint.

Clinical features

Clinical features differ in the three described groups.

IN CHILDREN

The patient, usually a child over 4 years, presents with severe pain, malaise and a fever; in neglected cases, toxaemia may be marked. The parents will have noticed that he or she refuses to use one limb or to allow it to be handled or even touched. There may be a recent history of infection: a septic toe, a boil, a sore throat or a discharge from the ear. Typically the child looks ill and feverish; the pulse rate is likely to be over 100 and the temperature is raised. The limb is held still and there is acute tenderness near one of the larger joints (e.g. above or below the knee, in the popliteal fossa or in the groin). Even the gentlest manipulation is painful and joint movement is restricted ('pseudoparalysis'). Local redness, swelling, warmth and oedema are later signs and signify that pus has escaped from the interior of the bone. Lymphadenopathy is common but non-specific. *It is important to remember that all these features may be attenuated if antibiotics have been administered.*

IN INFANTS

In children under 1 year old, and especially in the newborn, the constitutional disturbance can be misleadingly mild; the baby simply fails to thrive and is drowsy but irritable. Suspicion should be aroused by a history of birth difficulties, umbilical artery catheterization or a site of infection (however mild) such as an inflamed intravenous infusion point or even a heel puncture. Metaphyseal tenderness and resistance to joint movement can signify either osteomyelitis or septic arthritis; indeed, both may be present, so the distinction hardly matters. Look for other sites – multiple infection is not uncommon, especially in babies who acquire the infection in hospital. Radionuclide bone scans may help to discover additional sites.

BOX 2.2 CARDINAL FEATURES OF ACUTE OSTEOMYELITIS IN CHILDREN

Pain
Fever
Refusal to bear weight
Elevated white blood cell count
Elevated ESR
Elevated CRP

IN ADULTS

A common site for haematogenous infection is the thoracolumbar spine. There may be a history of some urological procedure followed by a mild fever and backache. Local tenderness is not very marked and it may take weeks before X-ray signs appear; when they do appear, the diagnosis may still need to be confirmed by fine-needle aspiration and bacteriological culture. Other imaging may be required such as MRI, CT or SPECT-CT. In particular, deep abscess will need to be ruled out in case of suspicion, as surgical drainage may be required. Other bones are occasionally involved, especially if there is a background of diabetes, malnutrition, drug addiction, leukaemia, immunosuppressive therapy or debility. *In the very elderly, and in those with immune deficiency*, systemic features are mild and the diagnosis is easily missed.

Diagnostic imaging

PLAIN X-RAY

During the first week after the onset of symptoms, the plain radiograph shows no abnormality of the bone. Displacement of the fat planes signifies soft-tissue swelling, but this could as well be due to a haematoma or soft-tissue infection. By the second week there may be a faint extracortical outline due to periosteal new bone formation; this is the classic X-ray sign of early pyogenic osteomyelitis, but treatment should not be delayed while waiting for it to appear. Later, the periosteal thickening becomes more obvious and there is patchy rarefaction of the metaphysis; later still, the ragged features of bone destruction appear (Figure 2.3). An important late sign is the combination of regional osteoporosis with a localized segment of apparently increased density. Osteoporosis is a feature of metabolically active, and thus living, bone; the segment that fails to become osteoporotic is metabolically inactive and possibly dead.

ULTRASONOGRAPHY

Ultrasonography may detect a subperiosteal collection of fluid in the early stages of osteomyelitis, but it cannot distinguish between a haematoma and pus.

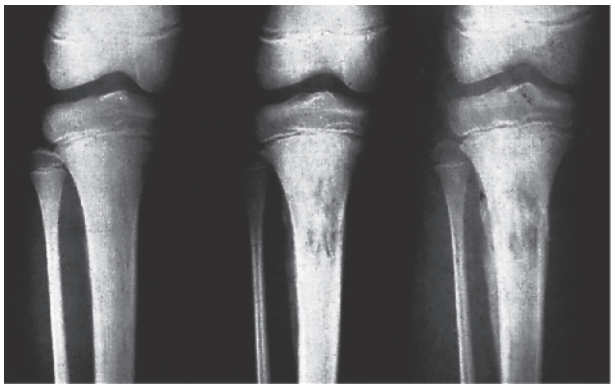


Figure 2.3 Acute osteomyelitis The first X-ray, 2 days after symptoms began, is normal – it always is; metaphyseal mottling and periosteal changes were not obvious until the second film, taken 14 days later; eventually much of the shaft was involved.

CT

Computed (or computerized) tomography offers the advantage of planar bone definition, including bone destruction and soft tissue mass, such as an abscess, within or surrounding bone. Disadvantages include high radiation dose. The excellent anatomical definition obtained from CT often justifies the higher radiation exposure.

RADIONUCLIDE SCANNING

Radioscintigraphy with ^{99}mTc-HDP reveals increased activity in both the perfusion phase and the bone phase. This is a highly sensitive investigation, even in the very early stages, but it has relatively low specificity and other inflammatory lesions can show similar changes. In doubtful cases, scanning with ^{67}Ga-citrate or ^{111}In-labelled leucocytes has been considered, but its use is decreasing in favour of other modalities.

SPECT/CT

As an alternative to radionuclide scanning, hybrid single-photon emission computed tomography/computer tomography (SPECT/CT) imaging is increasingly used in musculoskeletal infections. SPECT/CT imaging, compared with conventional planar study and SPECT alone, provides improved anatomic localization of infection and more accurate definition of its extent. Advantages of this modality include excellent differentiation between soft-tissue and bone infections, assessment of suspected infected sites with underlying structural bone alterations, and clear definition of infective foci within complex anatomical locations.

MAGNETIC RESONANCE IMAGING (MRI)

Magnetic resonance imaging can be helpful in cases of doubtful diagnosis, and particularly in suspected infection of the axial skeleton. It is also the best method of demonstrating bone marrow inflammation. It is extremely sensitive, even in the early phase of bone infection, and it can therefore assist in differentiating between soft-tissue infection and osteomyelitis. However, specificity is too low to exclude other local inflammatory lesions.

Laboratory investigations

The most certain way to confirm the clinical diagnosis is to aspirate pus or fluid from the metaphyseal subperiosteal abscess, the extraosseous soft tissues or an adjacent joint. This is best done using a 16- or 18-gauge trocar needle. Even if no pus is found, a smear of the aspirate is sent for detailed microbiological examination and tests for sensitivity to antibiotics. Immediate examination for cells and organisms through a simple Gram stain may help to identify the type of infection initially and assist with the early choice of antibiotic, but only until microbiological diagnosis through culture and antibiogram (the true etiological diagnosis to define specific treatment) is established. *Aspiration* will give a positive result in over 60% of cases that could be improved in case of open surgery by culture of tissue samples.

Blood cultures should be obtained if fever above 38 °C is detected, even though positive culture is obtained in less than half the cases of proven infection. The *C-reactive protein (CRP)* values are usually elevated within 12–24 hours and the *erythrocyte sedimentation rate (ESR)* within 24–48 hours after the onset of symptoms; both reactants (CRP+ESR) offer more information if simultaneously elevated. The *white blood cell (WBC) count* rises and the haemoglobin concentration may be diminished. *In the very young and the very old, these tests are less reliable and may show values within the range of normal.*

Anti-staphylococcal antibody titres may be raised. This test is useful in atypical cases where the diagnosis is in doubt.

Osteomyelitis in an unusual site or with an unusual organism should alert one to the possibility of heroin addiction, sickle-cell disease (*Salmonella* may be cultured from the faeces) or deficient host defence mechanisms including HIV infection.

Other tests such as IL-6 and alpha-defensin immunoassay are under evaluation, but their role is yet to be established.

Differential diagnosis

Cellulitis This is often mistaken for osteomyelitis. There is widespread superficial redness, with a clear demarcation between infected and normal skin, and lymphangitis. The source of skin infection may not be obvious and should be searched for (e.g. on the sole or between the toes). If doubt remains about the

diagnosis, MRI will help to distinguish between bone infection and soft-tissue infection. The organism is usually a staphylococcus or streptococcus. Mild cases will respond to high dosage oral antibiotics; severe cases need intravenous antibiotic treatment.

Acute suppurative arthritis Tenderness is diffuse, and movement at the joint is completely abolished by muscle spasm. In infants, the distinction between metaphyseal osteomyelitis and septic arthritis of the adjacent joint is somewhat theoretical, as both often coexist. A progressive rise in C-reactive protein values over 24–48 hours is considered suggestive of concurrent septic arthritis.

Streptococcal necrotizing myositis Group A beta-haemolytic streptococci (the same organisms which are responsible for the common 'sore throat') occasionally invade muscles and cause an acute myositis which, in its early stages, may be mistaken for cellulitis or osteomyelitis. Although the condition is rare, it should be kept well to the foreground in the differential diagnosis because it may rapidly spiral out of control towards muscle necrosis, septicaemia and death. Intense pain and board-like swelling of the limb in a patient with fever and a general feeling of illness are warning signs of a medical emergency. MRI will reveal muscle swelling and possibly signs of tissue breakdown. Immediate treatment with intravenous antibiotics is essential. Surgical debridement of necrotic tissue – and sometimes even amputation – may be needed to save a life.

Acute rheumatism The pain is less severe and it tends to flit from one joint to another. There may also be signs of carditis, rheumatic nodules or *erythema marginatum*.

Sickle-cell crisis The patient may present with features indistinguishable from those of acute osteomyelitis. In areas where *Salmonella* is endemic, it would be wise to treat such patients with suitable antibiotics until infection is definitely excluded.

Gaucher's disease 'Pseudo-osteitis' may occur with features closely resembling those of osteomyelitis. The diagnosis is made by finding other stigmata of the disease, especially enlargement of the spleen and liver.

Treatment

If osteomyelitis is suspected on clinical grounds, blood and fluid samples should be taken for laboratory investigation and then treatment started immediately without waiting for final confirmation of the diagnosis.

There are four important aspects to the management of the patient:

- appropriate antimicrobial therapy (first empirical, then specific)
- surgical drainage if required
- splintage and rest of the affected part
- supportive treatment for pain and dehydration.

ANTIBIOTICS

Blood and aspiration material are sent immediately for examination and culture, but the prompt intravenous administration of antibiotics is so vital that treatment should not await the result.

Initially, the choice of antibiotics is based on the findings from direct examination of the pus smear and the clinician's experience of local conditions – in other words, early empirical antibiotic administration, a 'best guess' at the most likely pathogen. *Staphylococcus aureus* is the most common at all ages, but treatment should provide cover also for other bacteria that are likely to be encountered in each age group; a more appropriate drug which is also capable of good bone penetration can be substituted, if necessary, once the infecting organism is identified and its antibiotic sensitivity is known. Factors such as the patient's age, general state of resistance, renal function, degree of toxaemia and previous history of allergy must be taken into account. The following classical recommendations are offered as a guide.

- *Neonates and infants up to 6 months of age*
  Initial antibiotic treatment should be effective against penicillin-resistant *Staphylococcus aureus*, Group B streptococcus and Gram-negative organisms. Drugs of choice are flucloxacillin plus a third-generation cephalosporin such as cefotaxime. Alternatively, effective empirical treatment can be provided by a combination of flucloxacillin (for penicillin-resistant staphylococci), benzylpenicillin (for Group B streptococci) and gentamicin (for Gram-negative organisms).
- *Children 6 months to 6 years of age*
  Empirical treatment in this age group should include cover against *Haemophilus influenzae*, unless it is known for certain that the child has had an anti-haemophilus vaccination. This is best provided by a combination of intravenous flucloxacillin and cefotaxime or cefuroxime.
- *Older children and previously fit adults*
  The vast majority in this group will have a staphylococcal infection and can be started on intravenous flucloxacillin and fusidic acid. Fusidic acid is preferred to benzylpenicillin partly because of the high prevalence of penicillin-resistant staphylococci and because it is particularly well concentrated in bone. However, for a known streptococcal infection benzylpenicillin is better. Patients who are allergic to penicillin should be treated with a polypeptide.

- *Elderly and previously unfit patients*
  In this group there is a greater than usual risk of Gram-negative infections, due to respiratory, gastrointestinal, or urinary disorders and the likelihood of the patient needing invasive procedures. The antibiotic of choice would be a combination of flucloxacillin and a second- or third-generation cephalosporin.
- *Patients with sickle-cell disease*
  These patients are prone to osteomyelitis, which may be caused by a staphylococcal infection but in many cases is due to *Salmonella* and/or other Gram-negative organisms. Chloramphenicol, which is effective against Gram-positive, Gram-negative and anaerobic organisms, used to be the preferred antibiotic, though there were always worries about the rare complication of aplastic anaemia. The current antibiotic of choice is a third-generation cephalosporin or a fluoroquinolone such as ciprofloxacin.
- *Heroin addicts and immunocompromised patients*
  Unusual infections (e.g. with *Pseudomonas aeruginosa*, *Proteus mirabilis* or anaerobic *Bacteroides* species) are likely in these patients. Infants with human immunodeficiency virus (HIV) infection may also have picked up other sexually transmitted organisms during birth. All patients with this type of background are therefore best treated empirically with a broad-spectrum antibiotic such as one of the third-generation cephalosporins or a fluoroquinolone preparation, depending on the results of sensitivity tests.
- *Patients considered to be at risk of methicillin-resistant Staphylococcus aureus (MRSA) infection*
  Patients admitted with acute haematogenous osteomyelitis and who have a previous history of MRSA infection, or any patient with a bone infection admitted to a hospital or a ward where MRSA is endemic, should be treated with intravenous vancomycin (or other glucopeptide such as teicoplanin) together with a third-generation cephalosporin. The usual programme is to administer the drugs intravenously (if necessary adjusting the choice of antibiotic once the results of antimicrobial sensitivity become available, and to the antibiotic trough and peak blood levels adjusted for the patient's kidney function and metabolism) until the patient's condition begins to improve and the CRP values return to normal levels – which usually takes 2–4 weeks depending on the virulence of the infection and the patient's general degree of fitness. By that time the most appropriate antibiotic would have been prescribed, on the basis of sensitivity tests; this can then be administered orally for another 3–6 weeks although, if bone destruction is marked, the period of treatment may have to be longer. While patients are on oral antibiotics, it is also important to track the serum antibiotic levels in order to ensure that the minimal inhibitory concentration (MIC) is maintained or exceeded. CRP, ESR and WBC values are also checked at regular intervals and treatment can be discontinued when these are seen to remain normal.

SURGICAL DRAINAGE

If antibiotics are given early (within the first 48 hours after the onset of symptoms), drainage is often unnecessary. However, if the clinical features do not improve within 36 hours of starting treatment, or even earlier, if there are signs of deep pus (swelling, oedema, fluctuation), and most certainly if pus is aspirated, the abscess should be drained by open surgery under general anaesthesia. If pus is found – and released – there is little to be gained by drilling into the medullary cavity. If there is no obvious abscess, it is reasonable to drill a few holes into the bone in various directions. There is no evidence that widespread drilling has any advantage and it may do more harm than good; if there is an extensive intramedullary abscess, drainage can be better achieved by cutting a small window in the cortex. The wound is closed without a drain and the splint (or traction) is reapplied. Once the signs of infection subside, movements are encouraged and the child is allowed to walk with the aid of crutches. Full weight-bearing is usually possible after 3–4 weeks.

At present, not more than one-third of patients with confirmed osteomyelitis are likely to need an operation and the percentage is decreasing; adults with vertebral infection seldom do.

SPLINTAGE

Some type of splintage is desirable, partly for comfort but also to prevent joint contractures. Simple skin traction may suffice and, if the hip is involved, this also helps to prevent dislocation. At other sites a plaster slab or half-cylinder may be used, but it should not obscure the affected area.

GENERAL SUPPORTIVE TREATMENT

The distressed child needs to be comforted and treated for pain. Analgesics should be given at repeated intervals without waiting for the patient to ask for them. Septicaemia and fever can cause severe dehydration and it may be necessary to give fluid intravenously.

Complications

A lethal outcome from septicaemia is nowadays extremely rare; with antibiotics the child nearly always recovers and the bone may return to normal. But morbidity and sequelae are common, especially if treatment is delayed or the organism is insensitive to the chosen antibiotic.

Epiphyseal damage and altered bone growth In neonates and infants whose epiphyses are still entirely cartilaginous, metaphyseal vessels penetrate the physis and may carry the infection into the epiphysis. If this happens, the physeal growth plate can be irrevocably damaged and the cartilaginous epiphysis may be destroyed, leading to arrest of growth and shortening of the bone. At the hip joint, the proximal end of the femur may be so badly damaged as to result in a pseudarthrosis.

Suppurative arthritis This may occur: (1) in very young infants, in whom the growth plate is not an impenetrable barrier; (2) where the metaphysis is intracapsular, as in the upper femur; or (3) from metastatic infection. In infants, it is so common as almost to be taken for granted, especially with osteomyelitis of the femoral neck. Ultrasound will help to demonstrate an effusion, but the definitive diagnosis is obtained by joint aspiration.

Metastatic infection This is sometimes seen – generally in infants – and may involve other bones, joints, serous cavities, the brain or lung. In some cases, the infection may be multifocal from the outset. Secondary infection sites are easily missed when attention is focused on one particular area; it is important to be alert to this complication and to repeatedly examine the child all over.

Pathological fracture Fracture is uncommon, but it may occur if treatment is delayed and the bone is weakened, either by erosion at the site of infection or by overzealous debridement.

Chronic osteomyelitis Despite improved methods of diagnosis and treatment, acute osteomyelitis sometimes fails to resolve. Weeks or months after the onset of acute infection, a sequestrum may appear in the follow-up X-ray and the patient may develop a chronic infection and a draining sinus. This may be related to late or inadequate treatment but is also seen in debilitated patients and in those with compromised defence mechanisms.

SUBACUTE HAEMATOGENOUS OSTEOMYELITIS

This condition is no longer rare, and in some countries the incidence is equal to that of acute osteomyelitis. Its relative mildness is presumably due to the organism being less virulent or the patient more resistant (or both). Its skeletal distribution is more variable than in acute osteomyelitis, but the distal femur and the proximal and distal tibia are the frequent sites. The anatomical classification (metaphyseal with or without cortical erosion, diaphyseal cortical or periosteal, epiphyseal, and vertebral) suggested by Roberts and colleagues in the 1980s is still helpful.

Pathology

Typically, there is a well-defined cavity in cancellous bone – usually in the tibial metaphysis – containing glairy seropurulent fluid (rarely pus). The cavity is lined by granulation tissue containing a mixture of acute and chronic inflammatory cells. The surrounding bone trabeculae are often thickened. The lesion sometimes encroaches on and erodes the bony cortex. Occasionally it appears in the epiphysis and, in adults, in one of the vertebral bodies.

Clinical features

The patient is usually a child or adolescent who has had pain near one of the larger joints for several weeks or even months. He or she may have a limp and often there is slight swelling, muscle wasting and local tenderness. The temperature is usually normal and there is little to suggest an infection. The WBC count and blood cultures usually show no abnormality but the ESR is sometimes elevated.

Imaging

The typical radiographic lesion is a circumscribed, round or oval radiolucent 'cavity' 1–2 cm in diameter. Most often it is seen in the tibial or femoral metaphysis, but it may occur in the epiphysis or in one of the cuboidal bones (e.g. the calcaneum). Sometimes the 'cavity' is surrounded by a halo of sclerosis (the classic *Brodie's abscess*); occasionally it is less well defined, extending into the diaphysis (Figure 2.4).

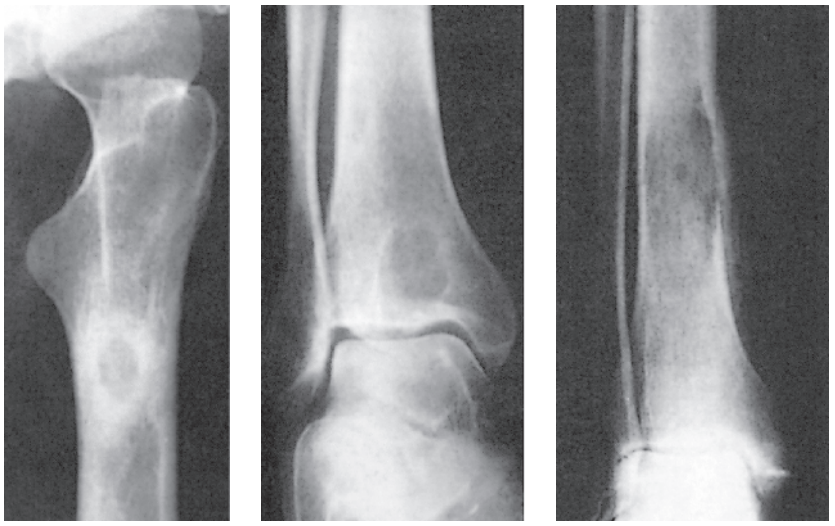
Metaphyseal lesions cause little or no periosteal reaction; diaphyseal lesions may be associated with periosteal new bone formation and marked cortical thickening. If the cortex is eroded, the lesion may be mistaken for a malignant tumour.

The radioisotope scan shows markedly increased activity.

Diagnosis

The clinical and X-ray appearances may resemble those of cystic tuberculosis, eosinophilic granuloma or osteoid osteoma; occasionally they mimic a malignant bone tumour such as Ewing's sarcoma. Epiphyseal lesions are easily mistaken for chondroblastoma. The diagnosis often remains in doubt until a biopsy is performed.

If fluid is encountered, it should be sent for bacteriological culture; this is positive in about half the cases and the organism is almost invariably *Staphylococcus aureus*.



(a) (b) (c)

Figure 2.4 Subacute osteomyelitis (a,b) The classic Brodie's abscess looks like a small walled-off cavity in the bone with little or no periosteal reaction. (c) Sometimes rarefaction is more diffuse and there may be cortical erosion and periosteal reaction.

Treatment

Treatment may be conservative if the diagnosis is not in doubt. Immobilization and antibiotics (flucloxacillin and fusidic acid) intravenously for 4 or 5 days and then orally for another 6 weeks usually result in healing, though this may take up to 12 months. If the diagnosis is in doubt, an open biopsy is needed and the lesion may be curetted at the same time. Curettage is also indicated if the X-ray shows that there is no healing after conservative treatment; this is always followed by a further course of antibiotics.

POST-TRAUMATIC OSTEOMYELITIS

Open fractures are always contaminated and are therefore prone to infection. The combination of tissue injury, vascular damage, oedema, haematoma, dead bone fragments and an open pathway to the atmosphere must invite bacterial invasion even if the wound is not contaminated with visible particulate dirt. *This is the most common cause of osteomyelitis in adults.*

Staphylococcus aureus is the usual pathogen, but other organisms such as *Escherichia coli*, *Proteus mirabilis* and *Pseudomonas aeruginosa* are sometimes involved. Occasionally, anaerobic organisms (clostridia, anaerobic streptococci or *Bacteroides*) appear in contaminated wounds.

Clinical features

The patient becomes feverish and develops pain and swelling over the fracture site; the wound is inflamed and there may be a seropurulent discharge. Blood tests reveal leucocytosis, increased CRP levels, and an elevated ESR; it should be remembered, though, that these inflammatory markers are non-specific and may be affected by tissue trauma.

X-ray appearances may be more difficult than usual to interpret because of bone fragmentation. *MRI* can be helpful in differentiating between bone and soft-tissue infection, but it is less reliable in distinguishing between long-standing infection and bone destruction due to trauma.

Microbiological investigation

If the wound is infected, a wound swab should be examined and cultured for organisms which can be tested for antibiotic sensitivity. Unfortunately, though, standard laboratory methods still yield negative results in about 20% of cases of overt infection. Routine wound swabs of open fracture wounds in the absence of infection is not recommended as cultured organisms are very unlikely to be the same as the organism causing any subsequent infection. Multiple tissue samples taken with clean, sterile instruments are preferred for microbiological investigations.

Treatment

The essence of treatment of open fractures is prophylaxis of infection: thorough cleansing and debridement of open fractures, the provision of drainage by leaving the wound open, immobilization of the fracture and antibiotics. In most cases a combination of flucloxacillin and benzylpenicillin (or sodium fusidate), given 6-hourly for 48 hours, will suffice. If the wound is clearly contaminated, it is wise also to give metronidazole for 4 or 5 days to control both aerobic and anaerobic organisms. Recent developments include the treatment of open fractures in one stage. This is a viable treatment option and can lead to good results if the soft tissue and bone debridement is meticulous and complete and adequate vascularized and tension free soft tissue closure can be obtained; this may require advance soft tissue procedures such as local or free flaps.

Pyogenic wound infection, once it has taken root, is difficult to eradicate. The presence of necrotic soft tissue and dead bone, together with a mixed bacterial flora, conspire against effective antibiotic control. Treatment requires soft tissue management and repeat debridement is required if there is evidence of inadequate debridement or infection.

Traditionally it was recommended that stable implants (fixation plates and intramedullary nails) should be left in place until the fracture had united, and this advice is still respected in recognition of the adage that even worse than an infected fracture is an *infected unstable* fracture. However, advances in external fixation techniques have meant that almost all fractures can, if necessary, be securely fixed by that method, with the added advantage that the wound remains accessible for dressings and superficial debridement. If these measures fail, the management is essentially that of chronic osteomyelitis.

CHRONIC OSTEOMYELITIS

This used to be the dreaded sequel to acute haematogenous osteomyelitis; nowadays, it more frequently follows an open fracture or an operation. The usual organisms (and with time there is always a mixed infection) are *Staphylococcus aureus*, *Escherichia coli*, *Streptococcus pyogenes*, *Proteus mirabilis* and *Pseudomonas aeruginosa*; in the presence of foreign implants *Staphylococcus epidermidis* (frequently coagulase negative staphylococcus), which is normally non-pathogenic, is the commonest of all.

Predisposing factors

Acute haematogenous osteomyelitis, if left untreated – and provided the patient does not succumb to septicaemia – will subside into a chronic bone infection which lingers indefinitely, perhaps with alternating 'flare-ups' and spells of apparent quiescence. The host defences are inevitably compromised by the presence of scar formation, dead and dying bone around the focus of infection, poor penetration of new blood vessels and non-collapsing cavities in which microbes can thrive. Bacteria covered in a protein–polysaccharide slime (*glycocalyx*) that protects them from both the host defences and antibiotics have the ability to adhere to inert surfaces such as bone sequestra and metal implants, where they multiply and colonize the area. There is also evidence that bacteria can survive inside osteoblasts and osteocytes and be released when the cells die.

These processes are evident in patients who have been inadequately treated ('too little too late'), but in any event certain patients are at greater risk than others: those who are very old or debilitated, those suffering from substance abuse and those with diabetes, peripheral vascular disease, skin infections, malnutrition, lupus erythematosus or any type of immune deficiency. The commonest of all predisposing factors is local trauma, such as an open fracture or a prolonged bone operation, especially if this involves the use of a foreign implant. Periprosthetic infection may evolve to chronic osteomyelitis and, due to its clinical relevance, will be addressed separately.

Pathology

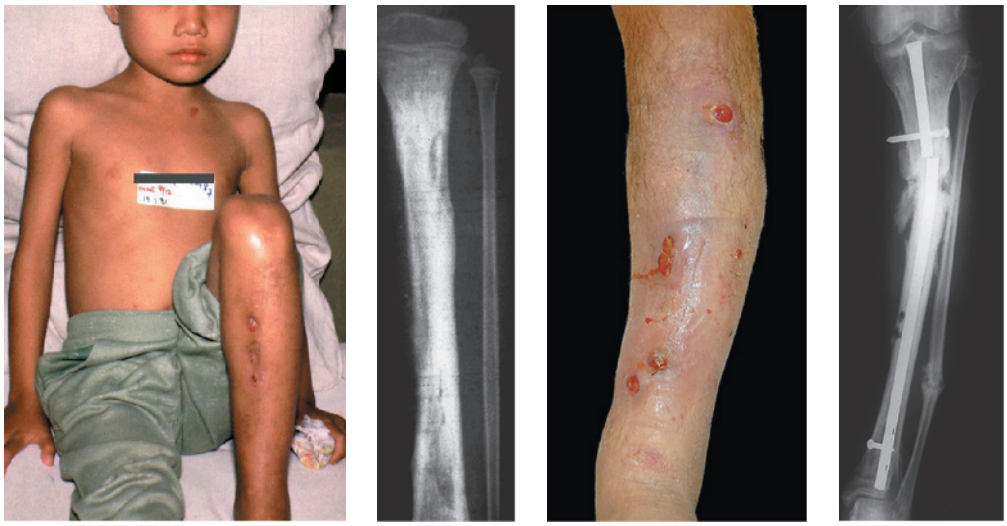
Bone is destroyed or devitalized, either in a discrete area around the focus of infection or more diffusely along the surface of an implant. Cavities containing pus and pieces of dead bone (sequestra) are surrounded by vascular tissue, and beyond that by areas of sclerosis – the result of chronic reactive new bone formation – which may take the form of a distinct bony sheath (involucrum). In the worst cases a sizeable length of the diaphysis may be devitalized and encased in a thick involucrum. Sequestra act as substrates for bacterial adhesion in much the same way as foreign implants, ensuring the persistence of infection until they are removed or discharged through perforations in the involucrum and sinuses that drain to the skin. A sinus may seal off for weeks or even months, giving the appearance of healing, only to reopen (or appear somewhere else) when the tissue tension rises. Bone destruction, and the increasingly brittle sclerosis, sometimes results in a pathological fracture. The histological picture is one of chronic inflammatory cell infiltration around areas of acellular bone or microscopic sequestra.

Clinical features

The patient presents because pain, pyrexia, redness and tenderness have recurred (a 'flare'), or with a discharging sinus. In long-standing cases, the tissues are thickened and often puckered or folded inwards where a scar or sinus adheres to the underlying bone. There may be a seropurulent discharge and excoriation of the surrounding skin. In post-traumatic osteomyelitis the bone may be deformed or ununited. See Figure 2.5.

Imaging

X-ray examination will usually show bone resorption – either as a patchy loss of density or as frank excavation around an implant – with thickening and sclerosis of the surrounding bone. However, there are marked variations: there may be no more than localized loss of trabeculation, or an area of osteoporosis, or periosteal thickening; sequestra show up as unnaturally dense fragments, in contrast to the surrounding osteopaenic bone; sometimes the bone is crudely thickened and misshapen, resembling a tumour. A *sinogram* may help to localize the site of infection.



(a) (b)

Figure 2.5 Chronic osteomyelitis
Chronic osteomyelitis may follow acute. The young boy (a) presented with draining sinuses at the site of a previous acute infection. The X-ray shows densely sclerotic bone. (b) In adults, chronic osteomyelitis is usually a sequel to open trauma or operation.

Radioisotope scintigraphy is sensitive but not specific. ^{99m}Tc-HDP scans show increased activity in both the perfusion phase and the bone phase. Scanning with ^{67}Ga-citrate or ^{111}In-labelled leucocytes is said to be more specific for osteomyelitis; such scans could be useful for showing up hidden foci of infection, although its low specificity has led to limited use.

CT and *MRI* are invaluable in planning operative treatment: together they will show the extent of bone destruction and reactive oedema, hidden abscesses and sequestra. *SPECT/CT* may provide advantages of sensitivity and local definition, and its use may increase in complex cases.

Investigations

During acute flares the CSR, ESR and WBC levels may be increased; these non-specific signs are helpful in assessing the progress of bone infection but they are not diagnostic.

Organisms cultured from discharging sinuses should be tested repeatedly for antibiotic sensitivity; with time, they often change their characteristics and become resistant to treatment. Note, however, that a superficial swab sample may not reflect the really persistent infection in the deeper tissues or may suffer from contamination; sampling from deeper tissues is crucial to understand the bone infection.

The most effective antibiotic treatment can be applied only if the pathogenic organism is identified and tested for sensitivity. Unfortunately, standard bacterial cultures still give negative results in about 20% of cases of overt infection. In recent years, more sophisticated molecular techniques have been developed, based on the amplification of bacterial DNA or RNA fragments (the polymerase chain reaction or PCR) and their subsequent identification by gel electrophoresis. However, although this has been shown to reveal unusual and otherwise undetected organisms in a significant percentage of cases, the technique is not widely available for routine testing.

A range of other investigations may also be needed to confirm or exclude suspected systemic disorders (such as diabetes) that could influence the outcome.

Table 2.1 Staging for adult chronic osteomyelitis

Lesion type	
Stage 1	Medullary
Stage 2	Superficial
Stage 3	Localized
Stage 4	Diffuse
Host category	
Type A	Normal
Type B	Compromised by local or systemic conditions
Type C	Severely compromised by local and systemic conditions

Staging of chronic osteomyelitis in long bones

'Staging' the condition helps in risk–benefit assessment and has some predictive value concerning the outcome of treatment. The system popularized by Cierny and colleagues in 2003 is based on both the local pathological anatomy and the host background (Table 2.1). The least serious, and most likely to benefit, are patients classified as Stage 1 or 2, Type A, i.e. those with localized infection and free of compromising disorders. Type B patients are somewhat compromised by a few local or systemic factors, but if the infection is localized and the bone still in continuity and stable (Stage 1–3) they have a reasonable chance of recovery. Type C patients are so severely

compromised that the prognosis is considered to be poor. If the lesion is also classified as Stage 4 (e.g. intractable diffuse infection in a non-united fracture), operative treatment may be contraindicated and the best option may be long-term palliative treatment known as suppression treatment. Occasionally one may have to advise amputation.

Treatment

ANTIBIOTICS

Chronic infection is seldom eradicated by antibiotics alone. Yet bactericidal drugs are important (a) to suppress the infection and prevent its spread to healthy bone and (b) to control acute flares. The choice of antibiotic depends on microbiological studies, but the drug must be capable of penetrating sclerotic bone and should be non-toxic with long-term use. Fusidic acid, clindamycin and the cephalosporins are good examples. Vancomycin and teicoplanin are effective in most cases of methicillin-resistant *Staphylococcus aureus* infection (MRSA).

Antibiotics are administered for 4–6 weeks (starting from the beginning of treatment or the last debridement) before considering operative treatment. During this time, serum antibiotic concentrations should be measured at regular intervals to ensure that they are kept at several times the minimal bactericidal concentration. *Continuous collaboration with a specialist in microbiology is important.* If surgical clearance fails, antibiotics should be continued for another 4 weeks before considering another attempt at full debridement.

LOCAL TREATMENT

A sinus may be painless and need dressing simply to protect the clothing. Colostomy paste can be used to stop excoriation of the skin. An acute abscess may need urgent incision and drainage, but this is only a temporary measure.

OPERATION

A waiting policy, punctuated by spells of bed rest and antibiotics to control flares, may have to be patiently endured until there is a clear indication for radical surgery: for *chronic haematogenous infections* this means intrusive symptoms, failure of adequate antibiotic treatment, and/or clear evidence of a sequestrum or dead bone; for *post-traumatic infections*, an intractable wound and/or an infected ununited fracture; for *postoperative infection*, similar criteria and evidence of bone erosion.

The presence of a *foreign implant* may prompt surgical intervention to remove the implant, whether in case of internal fixation (plates, screws and intramedullary nails that may be substituted by external fixation until infection control) or substitution, as discussed below. When undertaking operative treatment, collaboration with a plastic surgeon is strongly recommended.

Debridement At operation all infected soft tissue and dead or devitalized bone, as well as any infected implant, must be excised. The wound is inspected after 3 or 4 days and, if there are renewed signs of tissue death, the debridement may have to be repeated – several times if necessary. Antibiotic cover is continued for at least 4 weeks after the last debridement.

Dealing with the 'dead space' There are several ways of dealing with the resulting 'dead space'. *Porous antibiotic- impregnated beads* can be laid in the cavity and left for 2 or 3 weeks and then replaced with *cancellous bone grafts.* Bone grafts have also been used on their own; in the *Papineau technique* the entire cavity is packed with small cancellous chips (preferably autogenous) mixed with an antibiotic and a fibrin sealant. Where possible, the area is covered by adjacent muscle and the skin wound is sutured without tension. An alternative approach is to employ a *muscle flap transfer*: in suitable sites a large wad of muscle, with its blood supply intact, can be mobilized and laid into the cavity; the surface is later covered with a split-skin graft. In areas with too little adjacent muscle (e.g. the distal part of the leg), the same objective can be achieved by transferring a myocutaneous island flap on a long vascular pedicle. A free vascularized bone graft is considered to be a better option, provided the site is suitable and the appropriate facilities for microvascular surgery are available.

A different technique is the *Lautenbach approach*, involving radical excision of all avascular and infected tissue followed by closed irrigation and suction drainage, and an appropriate antibiotic solution in high concentration to allow the 'dead space' to be filled by vascular granulation tissue.

In refractory cases it may be possible to excise the infected and/or devitalized segment of bone completely and then close the gap by the *Ilizarov method* of 'transporting' a viable segment from the remaining diaphysis. This is especially useful if infection is associated with non-union after fracture.

Soft-tissue cover Last but not least, the bone must be adequately covered with skin. For small defects, split thickness skin grafts may suffice; for larger wounds local musculocutaneous flaps, or free vascularized flaps, are needed. Vacuum-assisted closure (VAC) may help when the deep infection is solved, not before.

Aftercare Success is difficult to measure; a minute focus of infection might escape the therapeutic onslaught, only to flare into full-blown osteomyelitis many years later. Prognosis should always be guarded; local trauma must be avoided and any recurrence of symptoms, however slight, should be taken seriously and investigated. The watchword is 'cautious optimism' – a 'probable cure' is better than no cure at all.

PERIPROSTHETIC INFECTION

Periprosthetic joint infection (PJI) is a specific type of infection related to joint replacement and a dreadful complication, potentially chronic, with significant clinical relevance for the affected patient, the treating surgeon and the health system. With an incidence of about 1–2% in hip arthroplasty, 2–3% in knee arthroplasty, 1–2% in the shoulder and even 3–5% in the elbow, the economic impact may represent a 5- to 10-fold cost increase compared to a primary arthroplasty. Patient risk factors include obesity, diabetes, rheumatoid arthritis and immunosuppressive treatments. Other risk factors include previous surgery, perioperative infection at a distant site, allogeneic blood transfusion, prolonged operative time and postoperative complications, including hematoma, superficial surgical site infection, wound drainage, and wound dehiscence.

A simple classification based on clinical manifestations differentiates early-onset PJI (if under 3 months after surgery), commonly initiated at operation through intraoperative contamination by relatively virulent microorganisms; delayed-onset PJI (after 3 months but before 12–24 months after surgery), by less virulent microorganisms but with same origin at operation; and late-onset PJI (more than 12–24 months after surgery), frequently due to haematogenous infection but occasionally due to very low-grade microorganisms with an extremely indolent infection initiated at the time of surgery. Tsukayama and colleagues popularized a classification that included early postoperative infection, haematogenous infection, and late chronic infection, adding a fourth type with positive intraoperative culture in a patient with revision for presumed aseptic failure, although some of these may not be true infections.

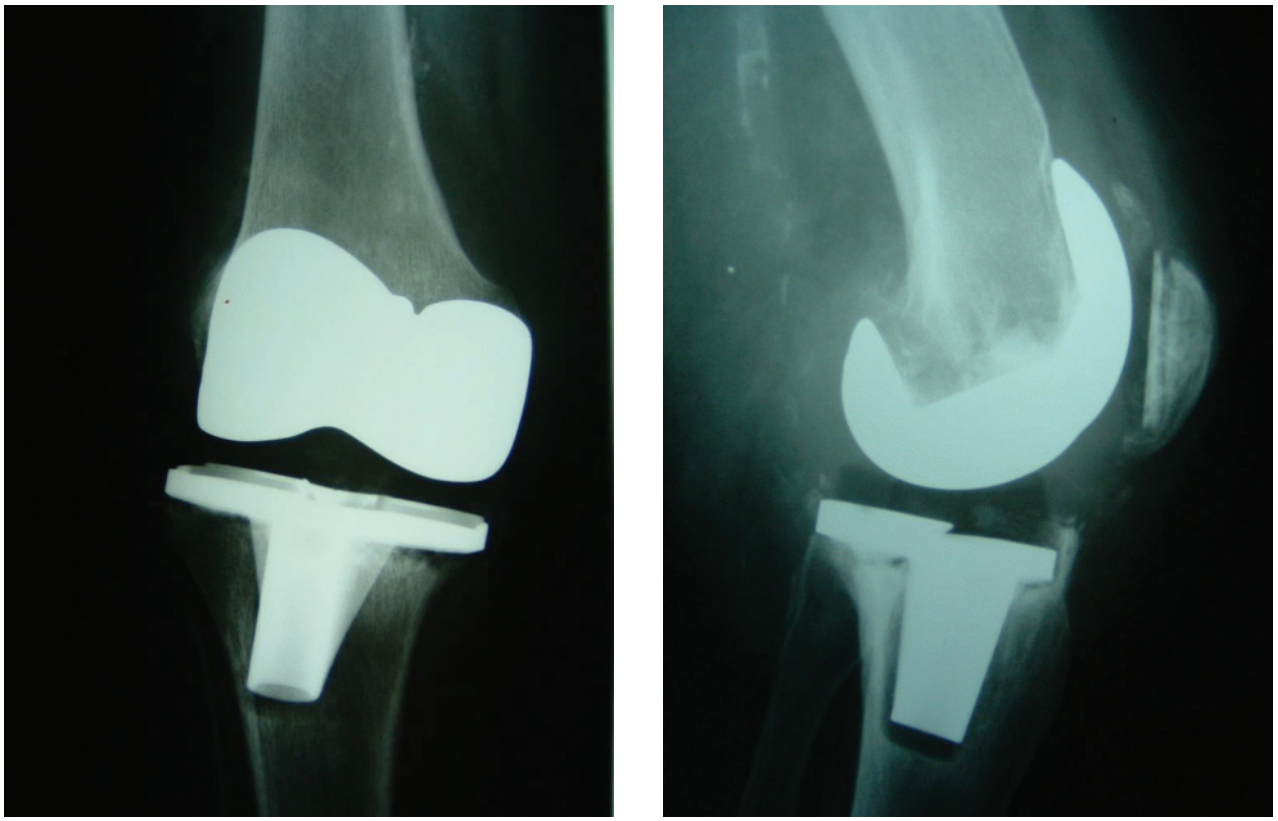
Adherence and biofilms Once in contact with the surface of the implant, microorganisms colonize the surface of the implant in competition with the host cells, in the so-called 'race for the surface'. After the initial adherence, the microorganisms colonize the implant through biofilm formation. Biofilms are complex communities of microorganisms embedded in an extracellular matrix formed on surfaces. From the attachment of microbial cells to a surface, the biofilm grows and matures until detachment and propagation, protecting microorganisms in a multicellular non-homogeneous structure where microbial cells communicate with one another (e.g. through quorum sensing) as in a multicellular organism protected from antibiotics and the host immune system. Clearing the biofilm requires surgical treatment, frequently with implant removal together with radical debridement of all infected tissues, followed by specific intravenous antibiotics.

The causative microorganism of PJI is most frequently *Staphylococcus aureus*, followed by coagulase-negative *Staphylococcus*. Together, these represent more than 50% of PJIs. *Streptococcus* species, *Enterococcus* species, aerobic Gram-negative bacilli, and some anaerobic (such as *Propionibacterium acnes* in the shoulder) account for 20–30%, while polymicrobial infections occur in 10–20%, the rest being culture-negative or other infrequent microorganisms. The increasing presence of multiresistant microorganisms requires carefully individualized antibiotic treatments.

The general diagnosis of PJI requires assessment of whether the joint is infected and, if so, determination of the causative microorganisms and their antimicrobial susceptibility. Thus, the diagnosis of PJI results from a combination of clinical findings, radiographic results (including early osteolysis and intraosseous abscesses, Figure 2.6), laboratory results (particularly CRP and ESR, WBC, but also IL-6 and procalcitonin) from peripheral blood and synovial fluid, microbiological data, intraoperative inspection, and histological evaluation of periprosthetic tissue. No single test offers sufficient accuracy alone. Clinically, definitive evidence of periprosthetic joint infection is obtained only when a sinus tract in communication with the prosthesis or an identical pathogen found in two separate periprosthetic tissue or fluid samples is confirmed. Other minor criteria also provide supportive (although not definitive) evidence, such as purulence surrounding the prosthesis, acute inflammation in periprosthetic histology, single virulent organism, elevated WBC, CRP and ESR.

Sonication of retrieved implants has been introduced to culture the dislodged biofilm and microorganisms from the surface of infected implants. Microorganisms were similarly obtained from all the different materials in the infected, retrieved prosthesis, proving that any remaining part of the implant may retain microorganisms, unless removed in revision surgery.

Treatment of periprosthetic joint infection usually requires both surgery and medical therapy, including prolonged antibiotic therapy after hospital discharge. The team approach, including surgeons, microbiologists, infectious disease physicians, nursing staff and other health professionals, is strictly required. Surgical treatment options oscillate from debridement with prosthesis retention (in early infections, particularly of haematogenous origin), one-stage arthroplasty exchange, two-stage arthroplasty exchange with or without antibiotic-loaded polymethylmethacrylate spacer, arthroplasty resection without reimplantation, or even suppression treatment consisting of long-term antibiotic treatment alone. Rarely, amputation may be required in case of vital risk for the patient.



(a) (b)

Figure 2.6 Periprosthetic joint infection Septic loosening surrounding the tibial stem in this case with PJI 3 years after total knee arthroplasty associated with immunodepression due to chemotherapy in the treatment of severe malignancy in (a) anteroposterior and (b) lateral radiographic views.

But besides the complex, multidisciplinary treatment of these infections, perioperative and postoperative prevention of PJI is a major aspect in the control of these severe entities.

GARRÉ'S SCLEROSING OSTEOMYELITIS

In 1893 Garré described a rare form of non-suppurative osteomyelitis which is characterized by marked sclerosis and cortical thickening. There is no abscess, only a diffuse enlargement of the bone at the affected site – usually the diaphysis of one of the tubular bones or the mandible. The patient is typically an adolescent or young adult with a long history of aching and slight swelling over the bone. Occasionally there are recurrent attacks of more acute pain accompanied by malaise and slight fever.

X-rays show increased bone density and cortical thickening; in some cases the marrow cavity is completely obliterated. There is no abscess cavity.

Diagnosis can be difficult. If a small segment of bone is involved, it may be mistaken for an osteoid osteoma. If there is marked periosteal layering of new bone, the lesion resembles a Ewing's sarcoma. The biopsy will disclose a low-grade inflammatory lesion with reactive bone formation. Microorganisms are seldom cultured but the condition is usually ascribed to a staphylococcal infection.

Treatment is by operation: the abnormal area is excised and the exposed surface thoroughly curetted. Bone grafts, bone transport or free bone transfer may be needed.

MULTIFOCAL NON-SUPPURATIVE OSTEOMYELITIS

This obscure disorder – it is not even certain that it is an infection – was first described in isolated cases in the 1960s and 1970s, and later in a more comprehensive report on 14 patients of mixed age and sex. It is now recognized that: (1) it is not as rare as initially suggested; (2) it comprises several different syndromes which have certain features in common; and (3) there is an association with chronic skin infection, especially pustular lesions of the palms

and soles (palmo-plantar pustulosis) and pustular psoriasis.

In children the condition usually takes the form of multifocal (often symmetrical), recurrent lesions in the long-bone metaphyses, clavicles and anterior ribcage; in adults the changes appear predominantly in the sterno-costo-clavicular complex and the vertebrae. In recent years the various syndromes have been drawn together under the convenient acronym SAPHO – standing for synovitis, acne, pustulosis, hyperostosis and osteitis.

Early osteolytic lesions show histological features suggesting a subacute inflammatory condition; in long-standing cases there may be bone thickening and round cell infiltration. The aetiology is unknown. Despite the local and systemic signs of inflammation, there is no purulent discharge and microorganisms have seldom been isolated. The two most characteristic clinical syndromes will be described.

Subacute recurrent multifocal osteomyelitis

This appears as an inflammatory bone disorder affecting mainly children and adolescents. Patients develop recurrent attacks of pain, swelling and tenderness around one or other of the long-bone metaphyses (usually the distal femur or the proximal or distal tibia), the medial ends of the clavicles or a vertebral segment. Over the course of several years multiple sites are affected, sometimes symmetrically and sometimes simultaneously; with each exacerbation, the child is slightly feverish and may have a raised ESR.

X-ray changes are characteristic. There are small lytic lesions in the metaphysis, usually closely adjacent to the physis. Some of these 'cavities' are surrounded by sclerosis; others show varying stages of healing. The clavicle may become markedly thickened. If the spine is affected, it may lead to collapse of a vertebral body. *Radioscintigraphy* shows increased activity around the lesions.

Biopsy of the lytic focus is likely to show the typical histological features of acute or subacute inflammation. In long-standing lesions there is a chronic inflammatory reaction with lymphocyte infiltration. Bacteriological cultures are almost invariably negative.

Treatment is entirely palliative; antibiotics have no effect on the disease. Although the condition may run a protracted course, the prognosis is good and the lesions eventually heal without complications.

Sterno-costo-clavicular hyperostosis

Patients are usually in their forties or fifties, and men are affected more often than women. Clinical and radiological changes are usually confined to the sternum and adjacent bones and the vertebral column. As with recurrent multifocal osteomyelitis, there is a curious association with cutaneous pustulosis. The usual complaint is of pain, swelling and tenderness around the sternoclavicular joints; sometimes there is also a slight fever and the ESR may be elevated. Patients with vertebral column involvement may develop back pain and stiffness.

X-rays show hyperostosis of the medial ends of the clavicles, the adjacent sternum and the anterior ends of the upper ribs, as well as ossification of the sternoclavicular and costoclavicular ligaments. Vertebral changes include sclerosis of individual vertebral bodies, ossification of the anterior longitudinal ligament, anterior intervertebral bridging, end-plate erosions, disc space narrowing and vertebral collapse. *Radioscintigraphy* shows increased activity around the sternoclavicular joints and affected vertebrae.

The condition usually runs a protracted course with recurrent 'flares'. There is no effective treatment but symptoms tend to diminish or disappear in the long term; however, the patient may be left with ankylosis of the affected joints.

INFANTILE CORTICAL HYPEROSTOSIS (CAFFEY'S DISEASE)

Infantile cortical hyperostosis, also known as Caffey's disease, is a rare disease of infants and young children. It usually starts during the first few months of life with painful swelling over the tubular bones and/or the mandible (see Figure 2.7). The child may be feverish and irritable, refusing to move the affected limb. Infection may be suspected but, apart from the swelling, there are no local signs of inflammation. The ESR, though, is usually elevated.

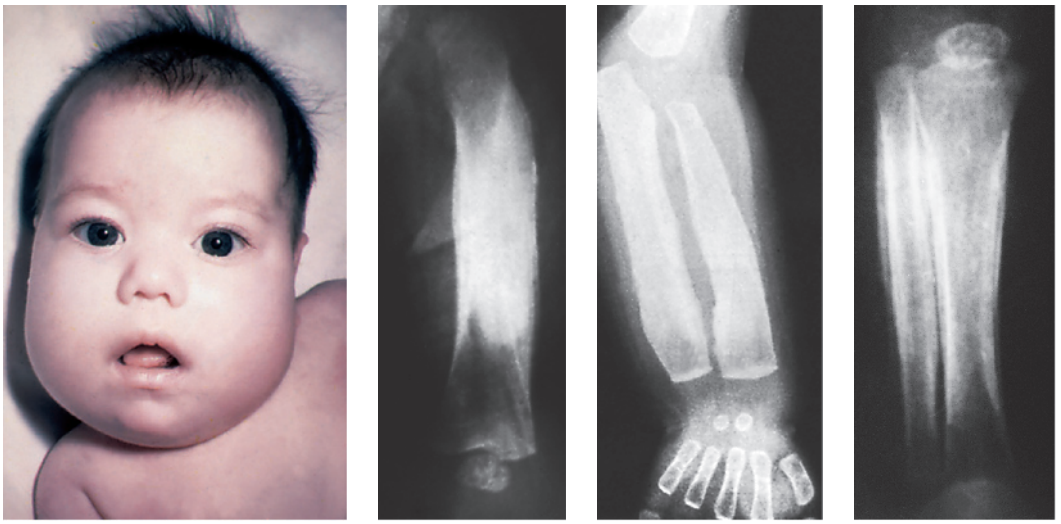
X-rays characteristically show periosteal new bone formation resulting in thickening of the affected bone.

After a few months the local features may resolve spontaneously, only to reappear somewhere else. Flat bones, such as the scapula and cranial vault, may also be affected.

Other causes of hyperostosis (osteomyelitis, scurvy) must be excluded. The cause of Caffey's disease is unknown but a virus infection has been suggested. Antibiotics are sometimes employed; it is doubtful whether they have any effect.

ACUTE SUPPURATIVE ARTHRITIS

A joint can become infected by: (1) direct invasion through a penetrating wound, intra-articular injection



(a) (b) (c) (d)

Figure 2.7 Caffey's disease This infant with Caffey's disease developed marked thickening of the mandible (a) and long bones. The lesions gradually cleared up, leaving little or no trace of their former ominous appearance.

or arthroscopy; (2) direct spread from an adjacent bone abscess; or (3) blood spread from a distant site. In infants it is often difficult to tell whether the infection started in the metaphyseal bone and spread to the joint or vice versa. In practice it hardly matters and in advanced cases it should be assumed that the entire joint and the adjacent bone ends are involved.

The causal organism is usually *Staphylococcus aureus*; however, in children between 1 and 4 years old, *Haemophilus influenzae* is an important pathogen unless they have been vaccinated against this organism. Occasionally other microbes, such as *Streptococcus*, *Escherichia coli* and *Proteus*, are encountered.

Predisposing conditions are rheumatoid arthritis, chronic debilitating disorders, intravenous drug abuse, immunosuppressive drug therapy and acquired immune deficiency syndrome (AIDS).

Pathology

The usual trigger is a haematogenous infection which settles in the synovial membrane; there is an acute inflammatory reaction with a serous or seropurulent exudate and an increase in synovial fluid. As pus appears in the joint, articular cartilage is eroded and destroyed, partly by bacterial enzymes and partly by proteolytic enzymes released from synovial cells, inflammatory cells and pus (Figure 2.8). In infants the entire epiphysis, which is still largely cartilaginous, may be severely damaged; in older children, vascular occlusion may lead to necrosis of the epiphyseal bone. In adults the effects are usually confined to the articular cartilage, but in late cases there may be extensive erosion due to synovial proliferation and ingrowth.

If the infection goes untreated, it will spread to the underlying bone or burst out of the joint to form abscesses and sinuses.

With healing there may be: (1) complete resolution and a return to normal; (2) partial loss of articular cartilage and fibrosis of the joint; (3) loss of articular cartilage and bony ankylosis; or (4) bone destruction and permanent deformity of the joint.

Clinical features

The clinical features differ somewhat according to the age of the patient.

In newborn infants the emphasis is on septicaemia rather than joint pain. The baby is irritable and refuses to feed; there is a rapid pulse and sometimes a fever. Infection is often suspected, but it could be anywhere! The joints should be carefully felt and moved to elicit the local signs of warmth, tenderness and resistance to movement. The umbilical cord should be examined for a source of infection. An inflamed intravenous infusion site should always excite suspicion. The baby's chest, spine and abdomen should be carefully examined to exclude other sites of infection. *Special care should be taken not to miss a concomitant osteomyelitis in an adjacent bone end.*

In children the usual features are acute pain in a single large joint (commonly the hip or the knee) and reluctance to move the limb ('pseudoparesis'). The child is ill, with a rapid pulse and a swinging fever. The overlying skin looks red and in a superficial joint swelling may be obvious. There is local warmth and marked tenderness. All movements are restricted, and often completely abolished, by pain and spasm. It is essential to look for a source of infection – a septic toe, a boil or a discharge from the ear.

In adults it is often a superficial joint (knee, wrist, a finger, ankle or toe) that is painful, swollen and inflamed. There is warmth and marked local tenderness, and movements are restricted. The patient

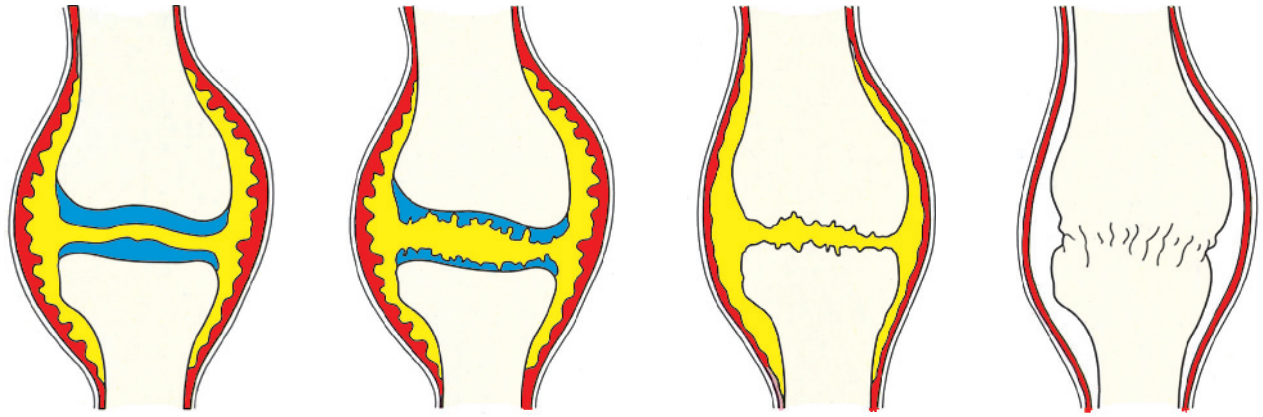


Figure 2.8 Acute suppurative arthritis – pathology In the early stage (a), there is an acute synovitis with a purulent joint effusion. (b) Soon the articular cartilage is attacked by bacterial and cellular enzymes. If the infection is not arrested, the cartilage may be completely destroyed (c). Healing then leads to bony ankylosis (d).

should be questioned and examined for evidence of gonococcal infection or drug abuse. Patients with rheumatoid arthritis, and especially those on corticosteroid treatment, may develop a 'silent' joint infection. Suspicion may be aroused by an unexplained deterioration in the patient's general condition; every joint should be carefully examined.

Imaging

Ultrasonography is the most reliable method for revealing a joint effusion in early cases. Both hips should be examined for comparison. Widening of the space between capsule and bone of more than 2 mm is indicative of an effusion, which may be echo-free (perhaps a transient synovitis) or positively echogenic (more likely septic arthritis).

X-ray examination is usually normal early on but signs to be watched for are soft-tissue swelling, loss of tissue planes, widening of the radiographic 'joint space' and slight subluxation (because of fluid in the joint) (Figure 2.9). With some infections there is sometimes gas in the joint. Narrowing and irregularity of the joint space are late features.

MRI and radionuclide imaging are helpful in diagnosing arthritis in obscure sites such as the sacroiliac and sternoclavicular joints.

Investigations

The white blood cell count, CRP and ESR are raised and blood culture may be positive. However, special investigations take time and it is much quicker (and usually more reliable) to aspirate the joint and examine the fluid. It may be frankly purulent but beware! – in early cases the fluid may look clear. A white cell count and Gram stain should be carried out immediately: the normal synovial fluid leucocyte count is under 300 per mL; it may be over 10 000 per mL in non-infective inflammatory disorders, but counts of over 50 000 per mL are highly suggestive of sepsis. Gram-positive cocci are probably *Staphylococcus aureus*; Gram-negative cocci are either *Haemophilus influenzae* or *Kingella kingae* (in children) or *Gonococcus* (in adults). Samples of fluid are also sent for full microbiological examination and tests for antibiotic sensitivity.

Differential diagnosis

Acute osteomyelitis In young children, osteomyelitis may be indistinguishable from septic arthritis; often one must assume that both are present.

Other types of infection *Psoas abscess* and local *infection of the pelvis* must be kept in mind. Systemic features will obviously be the same as those of septic arthritis.

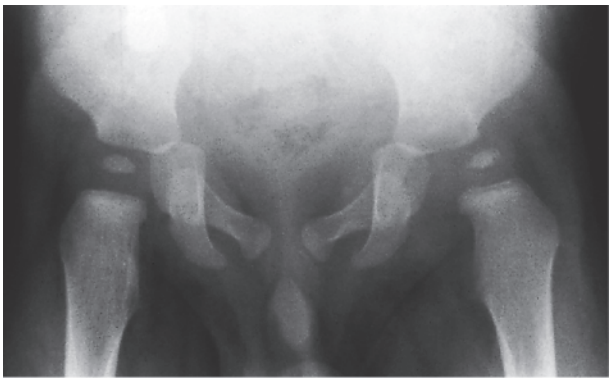
Trauma Traumatic synovitis or haemarthrosis may be associated with acute pain and swelling. A history of injury does not exclude infection. Diagnosis may remain in doubt until the joint is aspirated.

Irritable joint At the onset the joint is painful and lacks some movement, but the child is not really ill and there are no signs of infection. Ultrasonography may help to distinguish septic arthritis from transient synovitis.

Haemophilic bleed An acute haemarthrosis closely resembles septic arthritis. The history is usually conclusive, but aspiration will resolve any doubt.

Rheumatic fever Typically the pain flits from joint to joint, but at the onset one joint may be misleadingly inflamed. However, there are no signs of septicaemia.

Juvenile rheumatoid arthritis This may start with pain and swelling of a single joint, but the onset is



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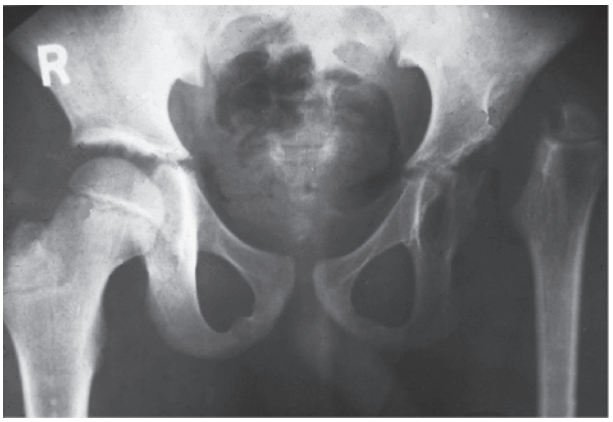

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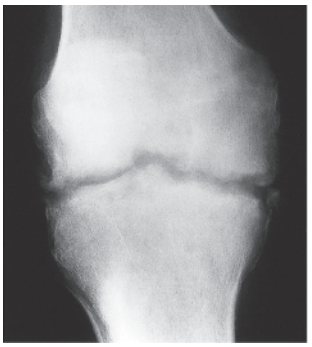


(c)

Figure 2.9 Suppurative arthritis – X-ray (a) In this child the left hip is subluxated and the soft tissues are swollen. (b) If the infection persists untreated, the cartilaginous epiphysis may be entirely destroyed, leaving a permanent pseudarthrosis. (c) Septic arthritis in an adult knee joint.

usually more gradual and systemic symptoms less severe than in septic arthritis.

Sickle-cell disease The clinical picture may closely resemble that of septic arthritis – and indeed the bone nearby may actually be infected! – so this condition should always be excluded in communities where the disease is common.

Gaucher's disease In this rare condition acute joint pain and fever can occur without any organism being found ('pseudo-osteitis'). Because of the predisposition to true infection, antibiotics should be given.

Gout and pseudogout *In adults*, acute crystal-induced synovitis may closely resemble infection. On aspiration the joint fluid is often turbid, with a high white blood cell count; however, microscopic examination by polarized light will show the characteristic crystals.

Treatment

The first priority is to aspirate the joint and examine the fluid. Treatment is then started without further delay and follows the same lines as for acute osteomyelitis. Once the blood and tissue samples have been obtained, there is no need to wait for detailed results before giving antibiotics. If the aspirate looks purulent, the joint should be drained without waiting for laboratory results (see below).

DRAINAGE

Under anaesthesia the joint is opened through a small incision, drained and washed out with physiological saline. A small catheter is left in place and the wound is closed; suction–irrigation is continued for another 2 or 3 days. This is the safest policy and is certainly advisable (1) in very young infants, (2) when the hip is involved, and (3) if the aspirated pus is very thick. For the knee, arthroscopic debridement and copious irrigation may be equally effective. Older children with early septic arthritis (symptoms for less than 3 days) involving any joint except the hip can often be treated successfully by repeated closed aspiration of the joint; however, if there is no improvement within 48 hours, open drainage will be necessary.

ANTIBIOTICS

Antibiotic treatment follows the same guidelines as presented for acute haematogenous osteomyelitis. The initial choice of antibiotics is based on judgement of the most likely pathogens.

Neonates and infants up to the age of 6 months should be protected against staphylococcus and Gram-negative streptococci with one of the penicillinase-resistant penicillins (e.g. flucloxacillin) plus a third-generation cephalosporin.

Children from 6 months to puberty can be treated similarly. There is a risk of *Haemophilus* infection if they have not been immunized.

Older teenagers and adults can be started on flucloxacillin and fusidic acid. If the initial examination shows Gram-negative organisms a third-generation cephalosporin is added. More appropriate drugs can be substituted after full microbiological investigation. Antibiotics should be given intravenously for 4–7 days and then orally for another 3 weeks.

SPLINTAGE

The joint should be rested, and for neonates and infants this may mean light splintage; with hip infection, the

joint should be held abducted and 30 degrees flexed, on traction to prevent dislocation.

GENERAL SUPPORTIVE CARE

Analgesics are given for pain and intravenous fluids for dehydration.

AFTERCARE

Once the patient's general condition is satisfactory and the joint is no longer painful or warm, further damage is unlikely. If articular cartilage has been preserved, gentle and gradually increasing active movements are encouraged. If articular cartilage has been destroyed, the aim is to keep the joint immobile while ankylosis is awaited. Splintage in the optimum position is therefore continuously maintained, usually by plaster, until ankylosis is sound.

Complications

Infants under 6 months of age have the highest incidence of complications, most of which affect the hip. The most obvious risk factors are a delay in diagnosis and treatment (more than 4 days) and concomitant osteomyelitis of the proximal femur.

Subluxation and dislocation of the hip, or instability of the knee should be prevented by appropriate posturing or splintage.

Damage to the cartilaginous physis or the epiphysis in the growing child is the most serious complication. Sequelae include *retarded growth*, *partial or complete destruction of the epiphysis*, *deformity of the joint*, *epiphyseal osteonecrosis*, *acetabular dysplasia* and *pseudarthrosis of the hip.*

Articular cartilage erosion (chondrolysis) is seen in older patients and this may result in restricted movement or complete *ankylosis of the joint.*

GONOCOCCAL ARTHRITIS

Neisseria gonorrhoeae is the commonest cause of septic arthritis in sexually active adults, especially among poorer populations. Even in affluent communities the incidence of sexually transmitted diseases has increased (probably related to the increased use of non-barrier contraception) and with it the risk of gonococcal and syphilitic bone and joint diseases and their sequelae. The infection is acquired only by direct mucosal contact with an infected person – carrying a risk of greater than 50% after a single contact!

Clinical features

Two types of clinical disorder are recognized: (a) *disseminated gonococcal infection* – a triad of polyarthritis, tenosynovitis and dermatitis – and (b) *septic arthritis of a single joint* (usually the knee, ankle, shoulder, wrist or hand). Both syndromes may occur in the same patient. There may be a slight pyrexia and the ESR and WBC count will be raised. If the condition is suspected, the patient should be questioned about possible contacts during the previous days or weeks and they should be examined for other signs of genitourinary infection (e.g. a urethral discharge or cervicitis).

Joint aspiration may reveal a high white blood cell count and typical Gram-negative organisms, but bacteriological investigations are often disappointing. Samples should also be taken from the various mucosal surfaces and tests should be performed for other sexually transmitted infections.

Treatment

Treatment is similar to that of other types of pyogenic arthritis. Patients will usually respond quite quickly to a third-generation cephalosporin given intravenously or intramuscularly. However, bear in mind that many patients with gonococcal infection also have chlamydial infection, which is resistant to cephalosporins; both are sensitive to quinolone antibiotics such as ciprofloxacin and ofloxacin. If the organism is found to be sensitive to penicillin (and the patient is not allergic), treatment with ampicillin or amoxicillin and clavulanic acid is also effective.

SEPTIC ARTHRITIS AND HIV-1 INFECTION

Septic arthritis has been encountered quite frequently in HIV-positive intravenous drug users, HIV-positive haemophiliacs and other patients with AIDS. The usual organisms are *Staphylococcus aureus* and *Streptococcus*; however, opportunistic infection by unusual organisms is not uncommon.

The patient may present with an acutely painful, inflamed joint and marked systemic features of bacteraemia or septicaemia. In some cases the infection is confined to a single, unusual site such as the sacroiliac joint; in others several joints may be affected simultaneously. Opportunistic infection by unusual organisms may produce a more indolent clinical picture.

Treatment follows the general principles outlined before. Patients with staphylococcal and streptococcal infections usually respond well to antibiotic treatment and joint drainage; opportunistic infections may be more difficult to control.

SPIROCHAETAL INFECTIONS

Two conditions which are likely to be encountered by the orthopaedic surgeon are dealt with here: *syphilis*

and *yaws*. *Lyme disease*, which also originates with a spirochaetal infection, is better regarded as due to a systemic autoimmune response and is discussed in Chapter 3.

Syphilis

Syphilis is caused by the spirochaete *Treponema pallidum*, generally acquired during sexual activity by direct contact with infectious lesions of the skin or mucous membranes. The infection spreads to the regional lymph nodes and thence to the bloodstream. The organism can also cross the placental barrier and enter the fetal blood stream directly during the latter half of pregnancy, giving rise to congenital syphilis.

In acquired syphilis a *primary* ulcerous lesion, or *chancre*, appears at the site of inoculation about a month after initial infection. This usually heals without treatment but, a month or more after that, the disease enters a *secondary phase* characterized by the appearance of a maculopapular rash and bone and joint changes due to periostitis, osteitis and osteochondritis. After a variable length of time, this phase is followed by a *latent period* which may continue for many years. The term is somewhat deceptive because in about half the cases pathological lesions continue to appear in various organs and 10–30 years later the patient may present again with tertiary syphilis, which takes various forms including the appearance of large granulomatous gummata in bones and joints and neuropathic disorders in which the loss of sensibility gives rise to joint breakdown (*Charcot joints*).

In congenital syphilis, the primary infection may be so severe that the fetus is either stillborn or the infant dies shortly after birth. The ones who survive manifest pathological changes similar to those described above, though with modified clinical appearances and a contracted timescale. See Figure 2.10.

Clinical features of acquired syphilis

Early features The patient usually presents with pain, swelling and tenderness of the bones, especially those with little soft-tissue covering, such as the frontal bones of the skull, the anterior surface of the tibia, the sternum and the ribs. *X-rays* may show typical features of *periostitis* and *thickening of the cortex* in these bones, as well as others that are not necessarily symptomatic. *Osteitis* and *septic arthritis* are less common. Occasionally these patients develop polyarthralgia or polyarthritis. Enquiry may reveal a history of sexually transmitted disease.

Late features The typical late feature, which may appear only after many years, is the syphilitic *gumma*, a dense granulomatous lesion associated with local bone resorption and adjacent areas of sclerosis. Sometimes this results in a pathological fracture. *X-rays* may show thick periosteal new bone formation at other sites, especially the tibia.

The other well-recognized feature of tertiary syphilis is a neuropathic arthropathy due to loss of sensibility in the joint – most characteristically the knee.

Other neurological disorders, the early signs of which may only be discovered on careful examination, are tabes dorsalis and 'general paralysis of the insane' (GPI). With modern treatment, these late sequelae have become rare.

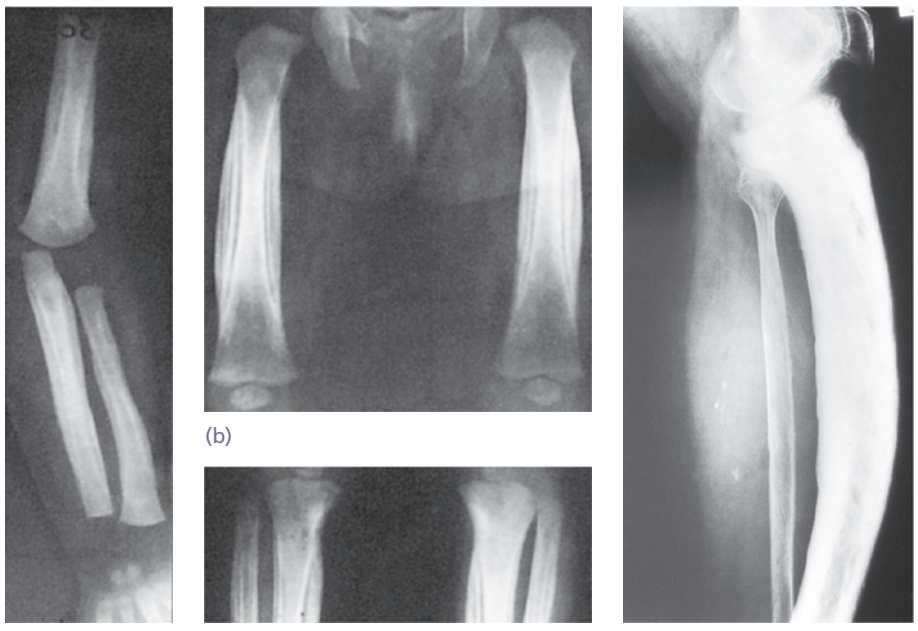


Figure 2.10 Syphilis (a–c) Congenital syphilis, with diffuse periostitis of many bones. (d) Acquired syphilitic periostitis of the tibia.

Clinical features of congenital syphilis

Early congenital syphilis Although the infection is present at birth, bone changes do not usually appear until several weeks afterwards. The baby is sick and irritable and examination may show skin lesions, hepatosplenomegaly and anaemia. Serological tests are usually positive in both mother and child.

The first signs of skeletal involvement may be joint swelling and 'pseudoparalysis' – the child refuses to move a painful limb. Several sites may be involved, often symmetrically, with slight swelling and tenderness at the ends or along the shafts of the tubular bones. The characteristic *X-ray changes* are of two kinds: *osteochondritis* (*'metaphysitis'*) – trabecular erosion in the juxta-epiphyseal regions of tubular bones showing first as a lucent band near the physis and later as frank bone destruction which may result in epiphyseal separation; and, less frequently, *periostitis* – diffuse periosteal new bone formation along the diaphysis, usually of mild degree but sometimes producing an 'onion-peel' effect. The condition must be distinguished from scurvy (rare in the first 6 months of life), multifocal osteomyelitis, the battered baby syndrome and Caffey's disease (see above).

Late congenital syphilis Bone lesions in older children and adolescents resemble those of acquired syphilis and some features occurring 10 or 15 years after birth may be manifestations of tertiary disease, the result of gumma formation and endarteritis. Gummata appear either as discrete, punched-out radiolucent areas in the medulla or as more extensive destructive lesions in the cortex. The surrounding bone is thick and sclerotic. Sometimes the predominant feature is dense endosteal and periosteal new bone formation affecting almost the entire bone (the classic '*sabre tibia*').

Other abnormalities which have come to be regarded as 'classic' features in older children are dental malformations ('Hutchinson's teeth'), erosion of the nasal bones, thickening and expansion of the finger phalanges (dactylitis) and painless effusions in the knees or elbows ('Clutton's joints').

Treatment

Early lesions will usually respond to intramuscular injections of benzylpenicillin given weekly for 3 or 4 doses. Late lesions will require high-dosage intravenous penicillin for a week or 10 days, but some forms of tertiary syphilis will not respond at all. An alternative would be treatment with one of the third-generation cephalosporins.

Yaws

Yaws is a non-venereal spirochaetal infection caused by *Treponema pertenue*. It is seen mainly in the poorer tropical parts of Africa, Asia and South America. Though considered – at least in Europe – to be a 'rare' disease, several thousand cases a year are reported in Indonesia.

The infection is contracted by skin-to-skin contact. A knobbly ulcer covered by a scab (the *primary* or '*mother*' *yaw*), usually develops on the face, hands or feet. Secondary skin lesions appear 1–4 months later and successive lesions may go on to pustular ulceration; as each one heals it leaves a pale tell-tale scar. This *secondary stage* is followed by a long latent period, merging into a *tertiary stage* during which skeletal changes similar to those of syphilis develop – periosteal new bone formation, cortical destruction and osteochondritis.

Clinical features

Children under 10 years old are the usual victims. In areas where the disease is endemic, the typical skin lesions and an associated lymphadenopathy are quickly recognized. Elsewhere, further investigations may be called for – serological tests and dark-field examination of scrapings from one of the skin lesions.

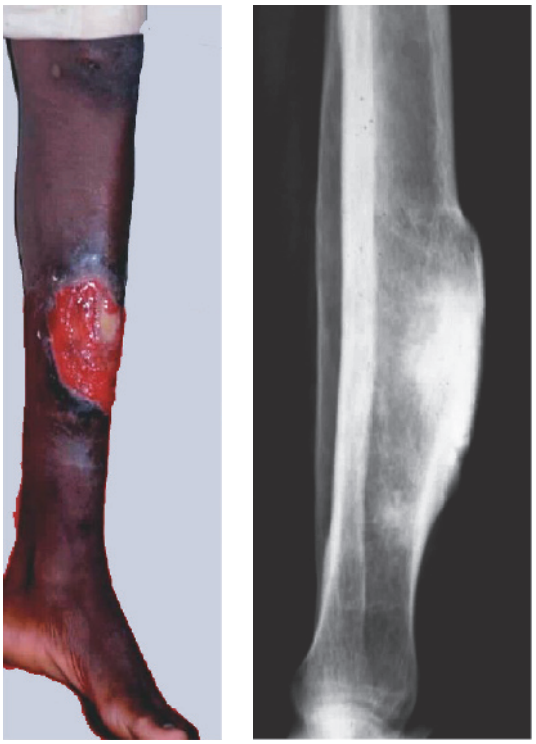
At a later stage deformities and bone tenderness may become apparent. *X-rays* show features such as cortical erosion, joint destruction and periosteal new bone formation; occasionally thickening of a long bone may be so marked as to resemble the 'sabre tibia' of late congenital syphilis.

Treatment

Treatment with benzylpenicillin, preferably given by intramuscular injection, is effective. For those who are hypersensitive to penicillin, erythromycin is a satisfactory alternative.

TROPICAL ULCER

Tropical ulcer, though the name sounds vague and non-specific, is a distinct entity that is seen frequently in tropical and subtropical regions, particularly in parts of Africa, where people walk bare-legged through rough terrain or long grass. It almost always occurs on the leg and men make up the majority of patients. The initial lesion is a small split in the skin (a cut, thorn-scratch, insect bite or other minor abrasion), which is then contaminated with all kinds of dirt or stagnant water. The most likely infecting organisms are *Fusiformis fusiformis* and *Borrelia vincentii* (both common in faeces). This results in an indolent ulcer which defies most forms of topical treatment (and certainly traditional remedies native to those parts of the world)



(a) (b)

Figure 2.11 Tropical ulcer (a) What started as a small ulcer has turned into a large spreading lesion. (b) The X-ray shows the typical marked periosteal reaction in the underlying bone.

(Figure 2.11). The ulcer may eventually bore its way into the soft tissues and the underlying bone; occasionally, after many years, it gives rise to a locally invasive squamous-cell carcinoma.

Clinical features

What starts as a small inflamed scratch or cut develops over a few days into a large pustule. By the time the patient attends for medical treatment, the pustule has usually ruptured, leaving a foul-smelling, discharging ulcer with hard rolled edges on the leg, the ankle or foot. In some cases the ulcer has already started to spread and after 4–6 weeks it may be several centimetres in diameter! Two or three adjacent ulcers may join up to form a large sloughing mass that erodes tendons, ligaments and the underlying bone. Even if the bone is not directly involved, X-ray examination may show a marked periosteal reaction to the overlying infection. With time that segment of the bone may become thickened and sclerotic, or there may be erosion of the cortex. With healing, soft-tissue scarring sometimes causes joint contractures at the knee, the ankle or the foot.

Occasionally an invasive squamous cell carcinoma develops in a chronic ulcer.

Treatment

'*Prevention* is better than cure.' For people living or working in the tropics, the chance of infection can be reduced by wearing shoes and any type of covering for the legs. Scratches and abrasions should be cleaned and kept clean until they heal.

Early cases of tropical ulcer may respond to benzylpenicillin or erythromycin given daily for a week. If this is not effective, a broad-spectrum antibiotic will be needed (e.g. a third-generation cephalosporin). Ulcers should be cleansed every day and kept covered with moist or non-adherent dressings. Topical treatment with metronidazole gel is advisable.

Late cases of ulceration will require painstaking cleansing and de-sloughing together with broad-spectrum antibiotics effective against the causative anaerobic Gram-negative organisms as well as secondary infecting microbes cultured from swab samples. Soft-tissue and bone destruction may be severe enough to require extensive debridement and skin-grafting. Occasionally amputation is the best option.

TUBERCULOSIS

Once common throughout the world, tuberculosis showed a steady decline in its prevalence in developed countries during the latter half of the twentieth century, due mainly to the effectiveness of public health programmes, a general improvement in nutritional status and advances in chemotherapy. In the last two decades, however, the annual incidence (particularly of extrapulmonary tuberculosis) has risen again, a phenomenon which has been attributed variously to a general increase in the proportion of elderly people, changes in population movements, the spread of intravenous drug abuse and the emergence of AIDS.

The skeletal manifestations of the disease are seen chiefly in the spine and the large joints, but the infection may appear in any bone or any synovial or bursal sheath. Predisposing conditions include chronic debilitating disorders, diabetes, drug abuse, prolonged corticosteroid medication, AIDS and other disorders resulting in reduced defence mechanisms.

Pathology

Mycobacterium tuberculosis (usually human, sometimes bovine) enters the body via the lung (droplet infection) or the gut (swallowing infected milk products) or, rarely, through the skin. In contrast to pyogenic infection, it causes a granulomatous reaction which is associated with tissue necrosis and caseation.

Primary complex The initial lesion in lung, pharynx or gut is a small one with lymphatic spread to

regional lymph nodes; this combination is the primary complex. Usually the bacilli are fixed in the nodes and no clinical illness results, but occasionally the response is excessive, with enlargement of glands in the neck or abdomen.

Even though there is often no clinical illness, the initial infection has two important sequels: (1) within nodes which are apparently healed or even calcified, bacilli may survive for many years, so that a reservoir exists; (2) the body has been sensitized to the toxin (a positive Mantoux or Heaf test being an index of sensitization) and, should reinfection occur, the response is quite different, the lesion being a destructive one which spreads by contiguity.

Secondary spread If resistance to the original infection is low, widespread dissemination via the bloodstream may occur, giving rise to miliary tuberculosis, meningitis or multiple tuberculous lesions. More often, blood spread occurs months or years later, perhaps during a period of lowered immunity, and bacilli are deposited in extrapulmonary tissues. Some of these foci develop into destructive lesions to which the term 'tertiary' may be applied.

Tertiary lesion Bones or joints are affected in about 5% of patients with tuberculosis (Figure 2.12). There is a predilection for the vertebral bodies and the large synovial joints. Multiple lesions occur in about one-third of patients. In established cases it is difficult to tell whether the infection started in the joint and then spread to the adjacent bone or vice versa; synovial membrane and subchondral bone have a common blood supply and they may, of course, be infected simultaneously.

Once the bacilli have gained a foothold, they elicit a chronic inflammatory reaction. The characteristic microscopic lesion is the tuberculous granuloma (or 'tubercle') – a collection of epithelioid and multinucleated giant cells surrounding an area of necrosis, with round cells (mainly lymphocytes) around the periphery (Figure 2.13).

Within the affected area, small patches of caseous necrosis appear. These may coalesce into a larger yellowish mass, or the centre may break down to form an abscess containing pus and fragments of necrotic bone.

Bone lesions tend to spread quite rapidly. Epiphyseal cartilage is no barrier to invasion and soon the infection reaches the joint. Only in the vertebral bodies, and more rarely in the greater trochanter of the femur or the metatarsals and metacarpals, does the infection persist as a pure chronic osteomyelitis.

If the synovium is involved, it becomes thick and oedematous, giving rise to a marked effusion. A pannus

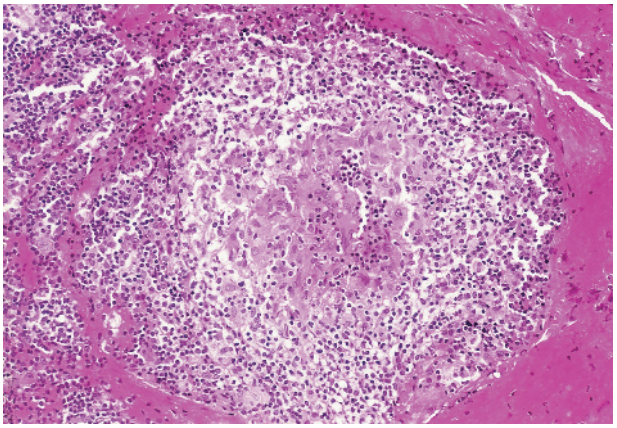
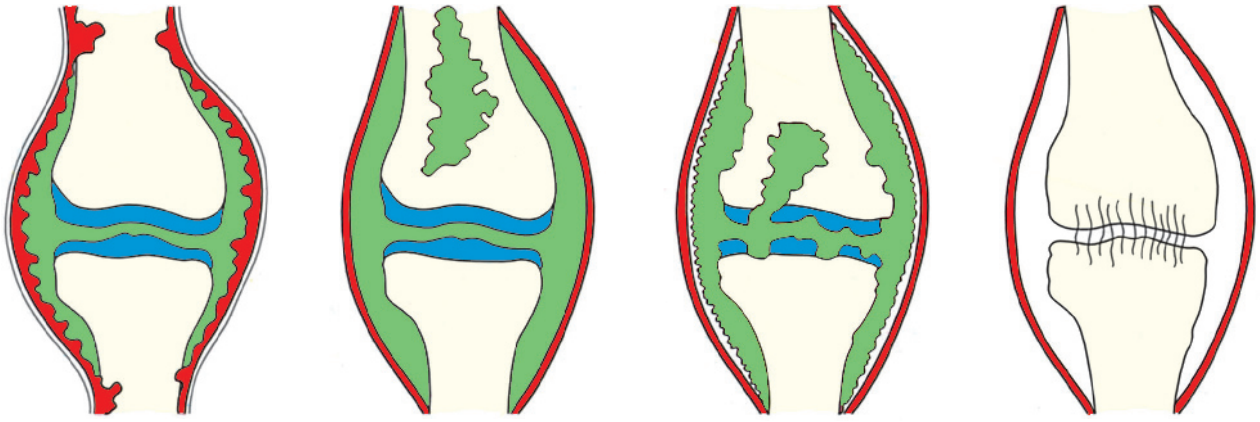


Figure 2.13 Tuberculosis – histology A typical tuberculous granuloma, with central necrosis and scattered giant cells surrounded by lymphocytes and histiocytes.



(a) (b) (c) (d)

Figure 2.12 Tuberculous arthritis – pathology The disease may begin as synovitis (a) or osteomyelitis (b). From either, it can extend to become a true arthritis (c); not all the cartilage is destroyed, and healing is usually by fibrous ankylosis (d).

of granulation tissue may extend from the synovial reflections across the joint; articular cartilage is slowly destroyed, though the rapid and complete destruction elicited by pyogenic organisms does not occur in the absence of secondary infection. At the edges of the joint, along the synovial reflections, there may be active bone erosion. In addition, the increased vascularity causes local osteoporosis.

If unchecked, caseation and infection extend into the surrounding soft tissues to produce a 'cold' abscess ('cold' only in comparison to a pyogenic abscess). This may burst through the skin, forming a sinus or tuberculous ulcer, or it may track along the tissue planes to point at some distant site. Secondary infection by pyogenic organisms is common. If the disease is arrested at an early stage, healing may be by resolution to apparent normality. If articular cartilage has been severely damaged, healing is by fibrosis and incomplete ankylosis, with progressive joint deformity. Within the fibrocaseous mass, mycobacteria may remain imprisoned, retaining the potential to flare up into active disease many years later.

Clinical features

There may be a history of previous infection or recent contact with tuberculosis. The patient, usually a child or young adult, complains of pain and (in a superficial joint) swelling. In advanced cases there may be attacks of fever, night sweats, lassitude and loss of weight. Relatives tell of 'night cries': the joint, splinted by muscle spasm during the waking hours, relaxes with sleep and the inflamed or damaged tissues are stretched or compressed, causing sudden episodes of intense pain. Muscle wasting is characteristic and synovial thickening is often striking (Figure 2.14). Regional lymph nodes may be enlarged and tender. Movements are limited in all directions. As articular erosion progresses the joint becomes stiff and deformed.

In tuberculosis of the spine, pain may be deceptively slight – often no more than an ache when the spine is jarred. Consequently, the patient may not present until there is a visible abscess (usually in the groin or the lumbar region to one side of the midline) or until collapse causes a localized kyphosis. Occasionally, the presenting feature is weakness or instability in the lower limbs.

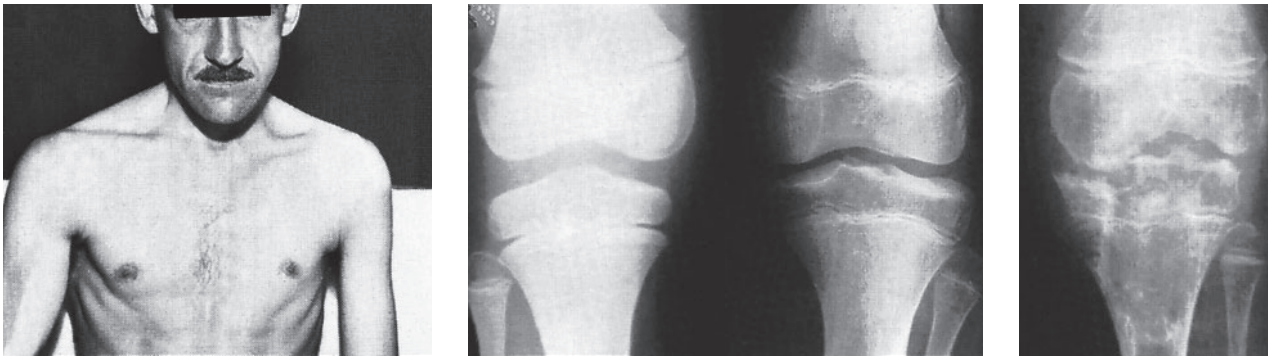
Multiple foci of infection are sometimes found, with bone and joint lesions at different stages of development. This is more likely in people with lowered resistance.

X-ray

Soft-tissue swelling and periarticular osteoporosis are characteristic. The bone ends take on a 'washed-out' appearance and the articular space is narrowed. In children the epiphyses may be enlarged, probably the result of long-continued hyperaemia. Later on there is erosion of the subarticular bone; characteristically this is seen *on both sides of the joint*, indicating an inflammatory process starting in the synovium. Cystic lesions may appear in the adjacent bone ends but there is little or no periosteal reaction. In the spine the characteristic appearance is one of bone erosion and collapse around a diminished intervertebral disc space; the soft-tissue shadows may define a paravertebral abscess.

Investigations

The ESR is usually increased and there may be a relative lymphocytosis. The Mantoux or Heaf test will be positive: these are sensitive but not specific tests; i.e. a negative Mantoux virtually excludes the diagnosis,



(a) (b) (c)

Figure 2.14 Tuberculosis – clinical and X-ray features (a) Generalized wasting used to be a common feature of all forms of tuberculosis. Nowadays, skeletal tuberculosis occurs in deceptively healthy-looking individuals. An early feature is periarticular osteoporosis due to synovitis – the left knee in (b). This often resolves with treatment, but if cartilage and bone are destroyed (c), healing occurs by fibrosis and the joint retains a 'jog' of painful movement.

but a positive test merely indicates tuberculous infection, now or at some time in the past.

If synovial fluid is aspirated, it may be cloudy, the protein concentration is increased and the white cell count is elevated. Acid-fast bacilli are identified in synovial fluid in 10–20% of cases, and cultures are positive in over half. A synovial biopsy is more reliable: sections will show the characteristic histological features and acid-fast bacilli may be identified; cultures are positive in about 80% of patients who have not received antimicrobial treatment.

Diagnosis

Except in areas where tuberculosis is common, diagnosis is often delayed simply because the disease is not suspected. Features that should trigger more active investigation are:

- a long history of pain or swelling
- involvement of only one joint
- marked synovial thickening
- severe muscle wasting
- enlarged and matted regional lymph nodes
- periarticular osteoporosis on X-ray
- a positive Mantoux test.

Synovial biopsy for histological examination and culture is often necessary. Joint tuberculosis must be differentiated from the following.

Transient synovitis This is fairly common in children. At first it seems no different from any other low-grade inflammatory arthritis; however, it always settles down after a few weeks' rest in bed. If the synovitis recurs, further investigation (even a biopsy) may be necessary.

Monarticular rheumatoid arthritis Occasionally, rheumatoid arthritis starts in a single large joint. This is clinically indistinguishable from tuberculosis and the diagnosis may have to await the results of synovial biopsy.

Subacute arthritis Diseases such as amoebic dysentery or brucellosis are sometimes complicated by arthritis. The history, clinical features and pathological investigations usually enable a diagnosis to be made.

Haemorrhagic arthritis The physical signs of blood in a joint may resemble those of tuberculous arthritis. If the bleeding has followed a single recent injury, the history and absence of marked wasting are diagnostic. Following repeated bleeding, as in haemophilia, the clinical resemblance to tuberculosis is closer, but there is also a history of bleeding elsewhere.

Pyogenic arthritis In long-standing cases it may be difficult to exclude an old septic arthritis.

Treatment

REST

Hugh Owen Thomas long ago urged that tuberculosis should be treated by rest – which had to be 'prolonged, uninterrupted, rigid and enforced'. This often involved splintage of the joint and traction to overcome muscle spasm and prevent collapse of the articular surfaces. With modern chemotherapy this is no longer mandatory; rest and splintage are varied according to the needs of the individual patient. Those who are diagnosed and treated early are kept in bed only until pain and systemic symptoms subside, and thereafter are allowed restricted activity until the joint changes resolve (usually 6 months to a year). Those with progressive joint destruction may need a longer period of rest and splintage to prevent ankylosis in a bad position; however, as soon as symptoms permit, movements are again encouraged.

CHEMOTHERAPY

The most effective treatment is a combination of antituberculous drugs, which should always include rifampicin and isoniazid. During the last decade the incidence of drug resistance has increased and this has led to the addition of various 'potentiating' drugs to the list. The following is one of several recommended regimens.

Initial, '*intensive phase treatment*', consists of isoniazid 300–400 mg, rifampicin 450–600 mg and fluoroquinolones 400–600 mg daily for 5–6 months. All replicating sensitive bacteria are likely to be killed by this bactericidal attack. This is followed by a '*continuation phase treatment*' lasting 9 months, the purpose of which is to eliminate the 'persisters', slow-growing, intermittently-growing, dormant or intracellular mycobacteria. This involves the use of isoniazid and pyrazinamide 1500 mg per day for 4½ months and isoniazid and rifampicin for another 4½ months. Then a '*prophylactic phase*', consisting of isoniazid and ethambutol 1200 mg per day for a further 3–4 months.

During the entire treatment period, drugs and dosage may have to be adjusted and modified, depending on the individual patient's age, size, general health and drug reactions.

OPERATION

Operative drainage or clearance of a tuberculous focus is seldom necessary nowadays. However, a cold abscess may need immediate aspiration or draining.

Once the condition is controlled and arthritis has completely subsided, normal activity can be resumed, though the patient must report any renewed symptoms. If, however, the joint is painful and the articular surface is destroyed, arthrodesis or replacement arthroplasty may be considered. The longer the period

of quiescence, the less the risk of reactivation of the disease; there is always some risk and it is essential to give chemotherapy for 3 months before and after the operation.

BRUCELLOSIS

Brucellosis is an unusual but nonetheless important cause of subacute or chronic granulomatous infection in bones and joints. Three species of organism are seen in humans: *Brucella melitensis*, *Brucella abortus* (from cattle) and *Brucella suis* (from pigs). Infection usually occurs from drinking unpasteurized milk or from coming into contact with infected meat (e.g. among farmers and meat packers). In the past it has been more common in countries around the Mediterranean and in certain parts of Africa and India. About 50% of patients with chronic brucellosis develop arthritis.

Pathology

The organism enters the body with infected milk products or, occasionally, directly through the skin or mucosal surfaces. It is taken up by the lymphatics and then carried by the blood stream to distant sites. Foci of infection may occur in bones (usually the vertebral bodies) or in the synovium of the larger joints. The characteristic lesion is a chronic inflammatory granuloma with round-cell infiltration and giant cells. There may be central necrosis and caseation leading to abscess formation and invasion of the surrounding tissues.

Clinical features

The patient usually presents with fever, headache and generalized weakness, followed by joint pains and backache. The initial illness may be acute and alarming; more often it begins insidiously and progresses until the symptoms localize in a single large joint (usually the hip or knee) or in the spine. The joint becomes painful, swollen and tender; movements are restricted in all directions. If the spine is affected, there is usually local tenderness and back movements are restricted.

The systemic illness follows a fluctuating course, with alternating periods of fever and apparent improvement (hence the older term 'undulant fever'). Diagnosis is often long delayed and may not be resolved until destructive changes are advanced.

X-rays

The picture is that of a subacute arthritis, with loss of articular space, slowly progressive bone erosion and periarticular osteoporosis. In the spine there may be destruction and collapse of adjacent vertebral bodies with obliteration of the disc.

Investigations

A positive agglutination test (titre above 1/80) is diagnostic. Joint aspiration or biopsy may allow the organism to be cultured and identified.

Diagnosis

Diagnosis is usually delayed while other types of subacute arthritis are excluded.

Tuberculosis and brucellosis have similar clinical and radiological features. The distinction is often difficult and may have to await the results of agglutination tests, synovial biopsy and bacteriological investigation.

Reiter's disease and other forms of reactive arthritis often follow an initial systemic illness. However, fever is not so marked and joint erosion is usually late and mild.

Treatment

ANTIBIOTICS

The infection usually responds to a combined onslaught with tetracycline and streptomycin for 3–4 weeks. Alternative drugs, which are equally effective and which may be used as 'combination therapy', are rifampicin and the newer cephalosporins.

Operation An abscess will need drainage, and necrotic bone and cartilage should be meticulously excised. If the joint is destroyed, arthrodesis or arthroplasty may be necessary once the infection is completely controlled.

LEPROSY

Leprosy is a mildly infectious chronic inflammatory disease caused by acid-fast *Mycobacterium leprae*. It is characterized by granulomatous lesions in the peripheral nerves, the skin and the mucosa of the upper respiratory tract.

Leprosy was once common throughout the world. Today it is rarely seen outside parts of South Asia, Africa, Latin America and some of the Pacific Islands. While the disease is easily cured with drugs, its crippling effects persist in a cumulative number of people.

The infection is acquired mainly by respiratory transmission; unbroken skin to skin contact is thought not to be dangerous. Several years may elapse before clinical features appear.

Pathology

Most people infected with *Mycobacterium leprae* develop protective immunity and get rid of the infection.

Some develop a few skin lesions, appearing as vague hypopigmented macules (*indeterminate leprosy*), that recover spontaneously. If the condition progresses, it takes one of several forms, depending on the host's immune response.

Tuberculoid leprosy occurs where there is delayed type hypersensitivity (DTH) to *Mycobacterium leprae* antigens, combined with some decrease in cell-mediated immunity (CMI). The granuloma in tuberculoid leprosy is focal and circumscribed and is made up of epithelioid cells, with a few scattered giant cells and a cuff of lymphocytes, very similar to tuberculosis.

Lepromatous leprosy is seen in patients who are unable to mount effective CMI against *Mycobacterium leprae*. Here the granuloma is diffuse and extensive and it consists of macrophages, many loaded with acid-fast bacilli. There may be a sprinkling of round cells in the lepromatous granuloma. The entire body skin may thus be affected.

Borderline types are intermediate forms that show some features of both of the above conditions. Without treatment, they tend to progress increasingly towards the lepromatous form.

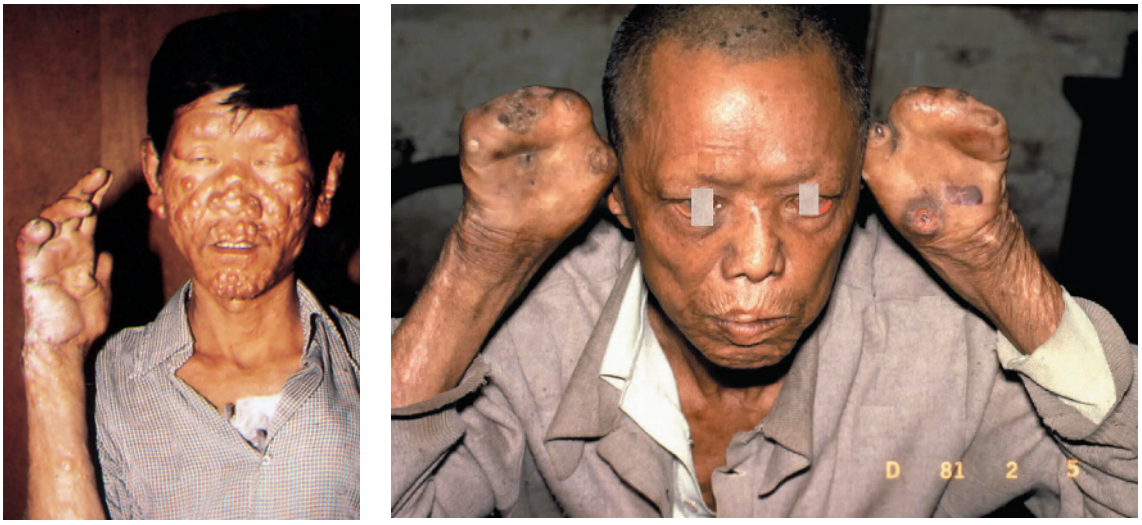
Peripheral nerves are always affected in leprosy. Dermal nerve twigs, cutaneous nerves as well as major nerve trunks may thus be involved. The affected nerves become thickened. Besides the granuloma there is hypertrophy of the epineurium and perineurium, demyelination, axonal degeneration and endoneurial fibrosis. A thickened nerve trunk may be strangulated by its own sheath or by the rigid walls of a fibro-osseous tunnel through which it passes (e.g. the ulnar nerve at the elbow). Sometimes, a tuberculoid granuloma in a nerve undergoes caseation. An important factor contributing to nerve damage is that medication is less likely to reach the segment of the nerve thus rendered ischaemic.

The chronic course of leprosy is often punctuated by acute inflammatory episodes – so-called '*reactions*' – which are due to the deposition of immune complexes (erythema nodosum leprosum or ENL or Type II reaction) or due to an increase in CMI and DTH levels (reversal reaction or RR or Type I reaction). Reactions occurring in the nerves (acute neuritis) greatly increase the risk of nerve damage.

Clinical features

Hypopigmented skin patches with impaired sensibility develop in all types of leprosy. Thickened cutaneous nerves may be seen and thickened nerve trunks may be felt where they are superficial, especially where they cross a bone (typically behind the medial condyle of the humerus at the elbow). Irrecoverable nerve damage with characteristic patterns of muscle weakness and deformities of the hands and feet may also be seen (Figure 2.15). Trophic ulcers, causing progressive destruction of the affected part, appear in the hands and feet.

Skin lesions in *tuberculoid leprosy* are sparse, well-demarcated, hypopigmented and anaesthetic. In contrast, in *lepromatous leprosy*, the skin is affected diffusely and extensively and the lesions present as multiple, symmetrically distributed macular patches with some sensory impairment. Plaques and nodules develop in advanced stages. Coarsening of the facial skin and loss of eyebrows may produce typical leonine features. Lepromatous ulceration of the nasal mucosa leads to destruction of the nasal septum and nasal deformity.



(a) (b)

Figure 2.15 Leprosy – late features (a) Patient showing typical ulnar claw-hand deformity. (b) This patient was even worse off, having lost all the fingers of both hands.

Peripheral nerves are affected extensively in *lepromatous leprosy* whereas in *tuberculoid leprosy* the neural lesions are few and focal in distribution. Cutaneous nerves as well as major nerve trunks of the upper and lower limbs are usually involved. Except for the Vth and VIIth nerves, the cranial nerves are not affected. Clinical defects in nerve function appear early in *tuberculoid leprosy* but much later in *lepromatous leprosy.*

Nerve lesions in tuberculoid leprosy may undergo caseation and liquefaction resulting in an intraneural 'cold abscess' mimicking an intraneural tumour, or the pus may break through the epineurium to present as a chronic collar-stud abscess.

Diagnosis

In countries where the disease is common the clinical diagnosis is seldom in doubt. Suggestive signs are the appearance of skin lesions with loss of sensibility, palpably or even visibly thickened nerves which may also be tender, areas of anaesthesia, chronic ulcers of the feet and typical deformities of hands and feet due to muscle weakness and imbalance. In countries where the disease is not endemic, diagnosis may have to await the results of skin smear examination, serological tests and skin or nerve biopsy.

Patterns of nerve involvement

Nerve trunks of the upper limbs are involved more often than those of the lower limbs. There is a pattern in the selection, site of involvement, risk of damage and chances of recovery (see Table 2.2). In the upper limb ulnar nerve paralysis is the most common and combined ulnar and median nerve paralysis is seen less frequently. Occasionally, triple nerve paralysis (paralysis of ulnar, median and radial nerves) may occur. Any other pattern is extremely rare.

Treatment

For purposes of treatment, patients are categorized as having *paucibacillary* (cases of indeterminate and tuberculoid leprosy) or *multibacillary* (cases of lepromatous and borderline leprosy) leprosy.

MULTIDRUG THERAPY

Combined chemotherapy with rifampicin as one of the drugs is the mainstay of treatment; however, the choice of drugs and duration of treatment depend on the type of disease. Following the recommendations of the World Health Organization, patients with *paucibacillary disease* are treated with rifampicin 600 mg once monthly and dapsone 100 mg once daily, for 6 months; patients with *multibacillary disease* are given rifampicin 600 mg and clofazi mine 300 mg once monthly and dapsone 100 mg and clofazimine 50 mg once daily, for 12 months. Reactions, especially acute neuritis, are treated with anti-inflammatory medication, of which prednisolone is the most important, and other supportive therapy.

NERVE DECOMPRESSION

Surgical decompression of a nerve trunk is sometimes required in order to improve perfusion of the nerve and allow the anti-leprosy and anti-inflammatory drugs to reach the affected segment and thus prevent or abort nerve damage. Surgical decompression is indicated: (a) in acute neuritis when, even while under treatment with corticosteroids, there is increasing neurological deficit; and (b) in cases of severe, unresponsive nerve pain, for relief of pain. Decompression involves tunnel release (often with excision of the medial epicondyle for the ulnar nerve) combined with incision of the epineurium over the entire sclerosed segment of the nerve. *Stripping the epineurium should not be done.*

TREATMENT OF NERVE ABSCESS

Cold abscesses associated with deteriorating neurological function and those that are likely to burst through the skin need to be excised or surgically evacuated. If there is no associated neural deficit, it is not necessary to intervene immediately, provided the patient can be reviewed periodically.

Table 2.2 Features of nerve trunk involvement in leprosy

Nerve affected	Preferred site	Involvement[a]	Motor paralysis	Recovery
Ulnar[c]	Above elbow/wrist	++++	++++	+
Median	Above wrist	++	++	++
Common peroneal	Back of knee	+++	+	++
Tibia	Behind ankle	+++	+++	[b]
Radial	Cutaneous division	+++	NA	NA
	Radial groove	++	(Forearm muscles only)	+++

[a] Thickening; [b] tenderness/pain; [c] most commonly involved nerve trunk; + uncommon; ++ common; +++ quite common; ++++ very common; NA, not applicable.

MANAGEMENT OF RESIDUAL PARALYSIS AND TROPHIC LESIONS

The long-term neuropathic complications of leprosy are dealt with in Chapter 11. The notorious deformities and disablement result from: (a) *local leprous granulomas* (as in the face); (b) *damage to nerves* of the hands and feet and consequent muscle paralysis; and (c) so-called '*trophic lesions*' (ulcers, shortening of digits and mutilations) arising from injuries to insensitive hands and feet. These conditions are prevented by early treatment of the disease, adequate treatment of neuritis and protection of anaesthetic hands and feet.

Paresis and established deformities can usually be corrected or at least improved by surgery. Although this is done mainly to improve function, restoration of normal appearance is also important for leprosy patients. Deformities such as claw fingers and drop foot stigmatize affected individuals as 'leprosy patients', with dire social consequences.

Individuals requiring surgery should have had antileprosy treatment and should not have had acute neuritis of any nerve trunk for at least 6 months prior to surgery. They must be well motivated and there should be proper pre-operative preparation with appropriate physiotherapy. Absence of facilities for pre- and postoperative therapy is an absolute contraindication for corrective surgery.

MYCOTIC INFECTIONS

Mycotic or fungal infection causes an indolent granulomatous reaction, often leading to abscess formation, tissue destruction and ulceration. When the musculoskeletal system is involved, it is usually by direct spread from the adjacent soft tissues. Occasionally, however, a bone or joint may be infected by haematogenous spread from a distant site.

These disorders are conveniently divided into 'superficial' and 'deep' infections.

Superficial mycoses These are primarily infections of the skin or mucous surfaces which spread into the adjacent soft tissues and bone. The more common examples are the *maduromycoses* (a group consisting of several species), *Sporothrix* and various species of *Candida*.

The *actinomycoses* are usually included with the superficial fungal infections. The causal organisms, of which *Actinomyces israelii* is the commonest in humans, are not really fungi but anaerobic bacilli with fungus-like appearance and behaviour.

Deep mycoses This group comprises infections by *Blastomyces*, *Histoplasma*, *Coccidioides*, *Cryptococcus*, *Aspergillus* and other rare fungi. The organisms, which occur in rotting vegetation and bird droppings, gain entry through the lungs and, in humans, may cause an influenza-like illness. Bone or joint infection is uncommon except in patients with compromised host defences.

MADUROMYCOSIS

This chronic fungal infection is seen mainly in northern Africa and the Indian subcontinent. The organisms usually enter through a cut in the foot; from there they spread through the subcutaneous tissues and along the tendon sheaths. The bones and joints are infected by direct invasion; local abscesses form and break through the skin as multiple sinuses. The patient may present at an early stage with a tender subcutaneous nodule (when the diagnosis is seldom entertained); more often he or she is seen when the foot is swollen and indurated, with discharging sinuses and ulcers (Figure 2.16). *X-rays* may show multiple bone cavities or progressive bone destruction. The organism can be identified in the sinus discharge or in tissue biopsies.

Treatment is unsatisfactory as there is no really effective chemotherapy. Intravenous amphotericin B is advocated, but it is fairly toxic and causes side effects such as headaches, vomiting and fever. Necrotic tissue should be widely excised. Even then it is sometimes difficult to stop further invasion, and amputation is sometimes necessary.

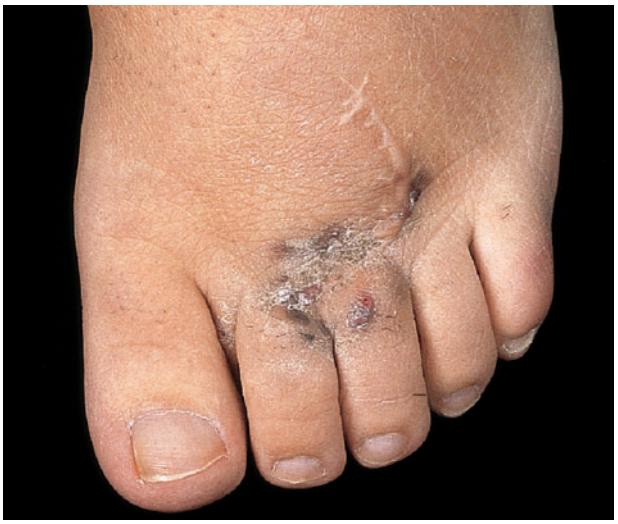


Figure 2.16 Maduromycosis This Mediterranean market-worker was perpetually troubled by tiny abscesses and weeping sinuses in her foot. X-rays showed that bone destruction had already spread to the tarsal bones, and after 2 years of futile treatment the foot had to be amputated.

Candidiasis

Candida albicans is a normal commensal in humans and it often causes superficial infection of the skin or mucous membranes. Deep and systemic infections are rare except under conditions of immunosuppression.

Candida osteomyelitis and arthritis may follow direct contamination during surgery or other invasive procedures such as joint aspiration or arthroscopy. The diagnosis is usually made only after tissue sampling and culture.

Treatment consists of thorough joint irrigation and curettage of discrete bone lesions, together with intravenous amphotericin B.

Actinomycosis

Infection is usually by *Actinomyces israelii*, an anaerobic Gram-positive bacillus. Although rare, it is important that it should be diagnosed because the organism is sensitive to antibiotics.

The most common site of infection is the mandible (from the mouth and pharynx), but bone lesions are also seen in the vertebrae (spreading from the lung or gut) and the pelvis (spreading from the caecum or colon). Peripheral lesions may occur by direct infection of the soft tissues and later extension to the bones. There may be a firm, tender swelling in the soft tissues, going on to form an abscess and one or more chronic discharging sinuses. *X-rays* may show cyst-like areas of bone destruction. The organism can be readily identified in the sinus discharge, but only on anaerobic culture.

Treatment, by large doses of benzylpenicillin G, tetracycline or erythromycin, has to be continued for several months.

The deep mycoses

Histoplasmosis, blastomycosis and coccidioidomycosis are rare causes of bone and joint infection, but they should always be considered in patients on immunosuppressive therapy who develop arthritis of one of the large joints or osteomyelitis in an unusual site. Diagnosis is usually delayed and often involves specialized microbiological investigations to identify the organism.

Treatment with intravenous amphotericin B is moderately effective. Operation may be necessary to drain an abscess or to remove necrotic tissue.

HYDATID DISEASE

Hydatid disease is caused by the tapeworm *Echinococcus*. Parasitic infestation is common among sheep farmers, but bone lesions are rare.

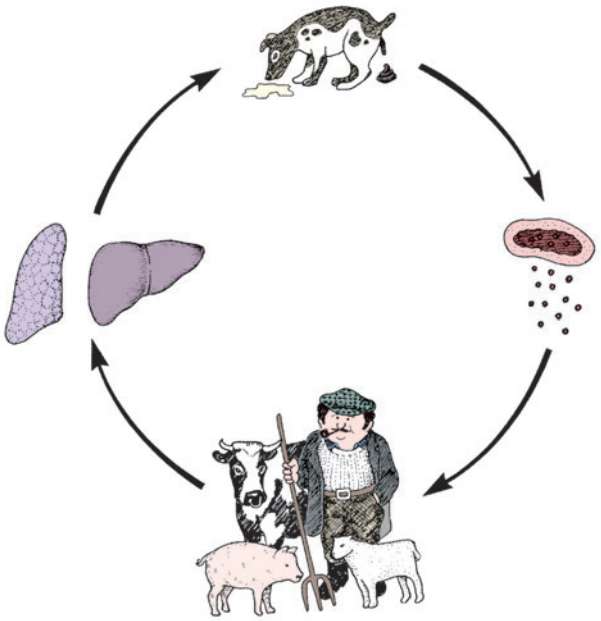


Figure 2.17 Hydatid disease The life cycle of the tapeworm which causes hydatid disease.

The organism, a cestode worm, has a complicated life cycle (Figure 2.17). The definitive host is the dog or some other carnivore that carries the tapeworm in its bowel. Segments of worm and ova pass out in the faeces and are later ingested by one of the intermediate hosts – usually sheep or cattle or man. Here the larvae are carried via the portal circulation to the liver, and occasionally beyond to other organs, where they produce cysts containing numerous scolices. Infested meat is then eaten by dogs (or humans), giving rise to a new generation of tapeworm.

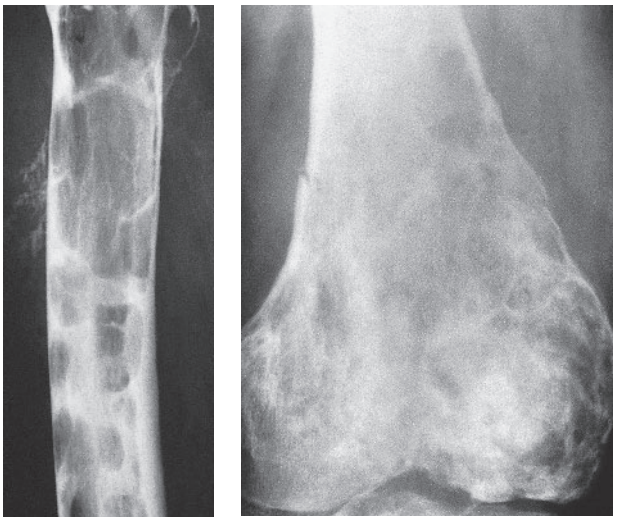
Scolices carried in the bloodstream occasionally settle in bone and produce hydatid cysts that slowly enlarge with little respect for cortical or epiphyseal boundaries. The bones most commonly affected are the vertebrae, pelvis, femur, scapula and ribs.

Clinical features

The patient may complain of pain and swelling, or may present for the first time with a pathological fracture or compression of the spinal cord. Infestation sometimes starts in childhood but the cysts take so long to enlarge that clinical symptoms and signs may not become apparent for many years. The diagnosis is more likely if the patient comes from a sheep-farming district.

Imaging

X-rays show solitary or multiloculated bone cysts, but only moderate expansion of the cortices (Figure 2.18). However, cortical thinning may lead to a pathological fracture. In the spine, hydatid disease may involve



(a) (b)

Figure 2.18 Hydatid disease of bone Two examples of hydatid involvement of bone: there is no expansion of the cortex in (a) and very little in (b).

adjacent vertebrae, with large cysts extending into the paravertebral soft tissues. These features are best seen on *CT* and *MRI*, which should always be performed if operative excision of the lesion is contemplated.

Investigations

Casoni's (complement fixation) test may be positive, especially in long-standing cases.

Diagnosis

Hydatid disease must be included in the differential diagnosis of benign and malignant bone cysts and cyst-like tumours. If the clinical and radiological features are not conclusive, needle biopsy should be considered, though there is a risk of spreading the disease.

Treatment

The antihelminthic drug albendazole is moderately effective in destroying the parasite. It has to be given in repeated courses: a recommended programme is oral administration of 10 mg per kg per day for 3 weeks, repeated at least 4 times with a 1-week 'rest' between courses. Liver, renal and bone marrow function should be monitored during treatment.

However, the bone cysts do not heal and recurrence is common. The indications for surgery are continuing enlargement or spread of the lesion, a risk of fracture, invasion of soft tissues and pressure on important structures. Curettage and bone grafting will lessen the risk of pathological fracture; at operation the cavity can be 'sterilized' with copious amounts of hypertonic saline, alcohol or formalin to lessen the risk of recurrence.

Radical resection, with the margin at least 2 cm beyond the cyst, is more certain, but also much more challenging. In a long bone, the space can sometimes be filled with a tumour-prosthesis, to include an arthroplasty if necessary. Large cysts of the vertebral column, or the pelvis and hip joint, are particularly difficult to manage in this way and in some cases surgical excision is simply impractical or impossible.

FINAL COMMENT

Infections are severe clinical entities that need to be considered in many clinical scenarios. When affecting bones or joints, and especially when implants are involved, microorganisms may attach and proliferate until the point of severely damaging the tissue and the general health of the patient and eventually proving fatal. Even if cured, an infection may seriously affect the appropriate function of the bone and joint, and cause long-term or even permanent disability.

Early clinical suspicion, adequate aetiological diagnosis, and properly staged treatment that usually includes surgery, antibiotics, and other actions, are crucial to control and eventually heal these complex diseases.

Future management of increasingly complex infections will require a deep knowledge of available diagnostic and therapeutic options and developments. From basic clinical reasoning to sophisticated laboratory tools, from appropriate surgical decisions to specific antibiotic regimes, a multidisciplinary approach is a major asset to successfully orient musculoskeletal infections.

FURTHER READING

Cierny G 3rd, Mader JT, Penninck JJ. A clinical staging system for adult osteomyelitis. *Clin Orthop Relat Res* 2003; **414**: 7–24.

Dartnell J, Ramachandran M, Katchburian M. Haematogenous acute and subacute paediatric osteomyelitis: a systematic review of the literature. *J Bone Joint Surg Br.* 2012; **94**(5): 584–95.

Donlan RM, Costerton JW. Biofilms: survival mechanisms of clinically relevant microorganisms. *Clin Microbiol Rev* 2002; **15**: 167–93.

Gristina AG. Biomaterial-centered infection: microbial adhesion versus tissue integration. *Science* 1987; **237**(4822): 1588–95.

Perez-Jorge C, Gómez-Barrena E, Horcajada JP, *et al.* Drug treatments for prosthetic joint infections in the era of multidrug resistance. *Expert Opin Pharmacother* 2016 Apr 7. [Epub ahead of print] PMID: 27054293. Available at: http://dx.doi.org/10.1080/14656566.2016.1176142 [Accessed 7 February 2017].

Tande AJ, Patel R. Prosthetic joint infection. *Clin Microbiol Rev* 2014; **27**(2): 302–45.

Thang SP, Tong AK, Lam WW, *et al.* SPECT/CT in musculoskeletal infections. *Semin Musculoskelet Radiol* 2014; **18**(2): 194–202.

Tsukayama DT, Estrada R, Gustilo RB. Infection after total hip arthroplasty: a study of the treatment of one hundred and six infections. *J Bone Joint Surg* 1996; **78A**: 512–23.

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Inflammatory rheumatic disorders

3

Christopher Edwards

The term 'inflammatory rheumatic disorders' covers a number of diseases that cause chronic pain, stiffness and swelling around joints and tendons. In addition, they are commonly associated with extra-articular features including skin rashes and inflammatory eye disease. Individuals with these diseases tend to die younger than their peers as a result of the effects of chronic inflammation. Many – perhaps all – are due to a faulty immune or inflammatory reaction resulting from a combination of environmental exposures against a background of genetic predisposition.

RHEUMATOID ARTHRITIS

Rheumatoid arthritis (RA) is the most common cause of chronic inflammatory joint disease. The most typical features are a symmetrical polyarthritis and tenosynovitis, morning stiffness, elevation of the erythrocyte sedimentation rate (ESR) and the appearance of autoantibodies (rheumatoid factor (RF) and anti-citrullinated peptide antibodies (ACPAs)) in the serum. Rheumatoid arthritis is a systemic disease and changes can be widespread in a number of tissues. Individuals with RA tend to die younger than their peers as a result of the effects of chronic inflammation on a number of organ systems. Chief among these is early ischaemic heart disease secondary to the effects of inflammation on the cardiovascular system.

The reported prevalence of RA in most populations is 1–2%, with a peak incidence in the fourth or fifth decades. Women are affected three times more commonly than men. Both the prevalence and the clinical expression vary between populations; the disease is more common (and generally more severe) in Caucasians living in the urban communities of Europe and North America than in the rural populations of Africa.

Cause

The cause of RA is still incompletely worked out. However, a great deal is now known about the circumstances in which RA develops, and hypotheses about its aetiology and pathogenesis have been suggested. Important factors in the evolution of RA are: (1) genetic susceptibility; (2) an immunological reaction, possibly involving a foreign antigen, preferentially focused on synovial tissue; (3) an inflammatory reaction in joints and tendon sheaths; (4) the appearance of rheumatoid factors (RF) and anti-citrullinated antibodies (anti-CCP or ACPA) in the blood and synovium; (5) perpetuation of the inflammatory process; and (6) articular cartilage destruction.

Genetic susceptibility A genetic association is suggested by the fact that RA is more common in first-degree relatives of patients than in the population at large; furthermore, twin studies have revealed a concordance rate of around 30% if one of the pair is affected. The human leucocyte antigen (HLA) DR4 occurs in about 70% of people with RA, compared to a frequency of less than 30% in normal controls. HLA-DR4 is encoded in the major histocompatibility complex (MHC) region on chromosome 6. There are strong associations between HLA-DR4 and RA. In particular, a key structural conformation within the HLA-DR4 binding groove called the 'shared epitope' seems important. This may suggest that a particular antigen that fits into this may be playing a part.

HLA Class II molecules appear as surface antigens on cells of the immune system (B lymphocytes, macrophages, dendritic cells), which can act as antigen-presenting cells (APCs). In some T-cell immune reactions, the process is initiated only when the antigenic peptide is presented in association with a specific HLA allele. It has been suggested that this is the case in people who develop RA; the idea is even more attractive if one proposes that the putative antigen has a special affinity for synovial tissue. So far no such antigen has been discovered.

The inflammatory reaction Once the APC/T-cell interaction is initiated, various local factors come into play and lead to a progressive enhancement of the immune response. There is a marked proliferation of

cells in the synovium, with the appearance of new blood vessel formation. Immune cells coordinate their action by the use of 'short-range hormones' (cytokines), which can activate inflammatory cells such as macrophages and B cells. Some cytokines, called chemokines, attract other inflammatory cells to the area.

Over recent years it has become clear that certain cytokines are important in RA. These include tumour necrosis factor (TNF), interleukin-1 (IL-1) and interleukin-6 (IL-6). The resulting synovitis, both in joints and in tendon sheath linings, is the hallmark of early RA.

Rheumatoid factor B-cell activation in RA leads to the production of anti-IgG autoantibodies, which are detected in the blood as 'rheumatoid factor' (RF). Low levels of RF can be found in many 'normal' individuals but, when the levels are high, an inflammatory disease is likely. Other autoimmune conditions such as systemic lupus erythematosus (SLE) and Sjögren's syndrome are also associated with the presence of RF.

In recent years other autoantibodies associated with RA have been identified. The most important are anti-cyclic citrullinated peptide antibodies (anti-CCP). The presence of anti-CCP is very specific for RA. Patients with a positive RF test tend to be more severely affected than those with a negative test.

Chronic synovitis and joint destruction Chronic rheumatoid synovitis is associated with the production of proteolytic enzymes, prostaglandins and the cytokines TNF and IL-1. Immune complexes are deposited in synovial joints, where they appear to augment the inflammatory process. This combination of factors leads to depletion of the cartilage matrix and, eventually, damage to cartilage and underlying bone. Vascular proliferation and osteoclastic activity, most marked at the edges of the articular surface, may contribute further to cartilage destruction and periarticular bone erosion.

Pathology

Rheumatoid arthritis is a systemic disease but the most characteristic lesions are seen in the synovium or within rheumatoid nodules. The synovium is engorged with new blood vessels and packed full of inflammatory cells.

JOINTS AND TENDONS

The pathological changes, if unchecked, proceed in four stages (Figure 3.1). Previously it was felt that having gone through these stages the disease activity could be 'burnt out'. This does not appear to be the case. In any one joint, features of different stages can be occurring simultaneously and even when joints are very badly destroyed the ongoing inflammation can continue to seriously damage systemic health by accelerating other disease processes such as ischaemic heart disease.

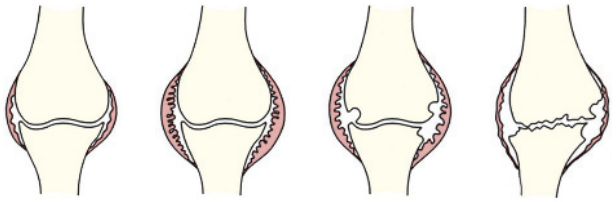


Figure 3.1 Rheumatoid arthritis – pathology
(a) Stage 1 – pre-clinical. (b) Stage 2 – synovitis and joint swelling. (c) Stage 3 – early joint destruction with peri-articular erosions. (d) Stage 4 – advanced joint destruction and deformity.

Stage 1: Pre-clinical Well before RA becomes clinically apparent, the immune pathology is already beginning. Raised ESR, C-reactive protein (CRP) and RF may be detectable years before the first diagnosis.

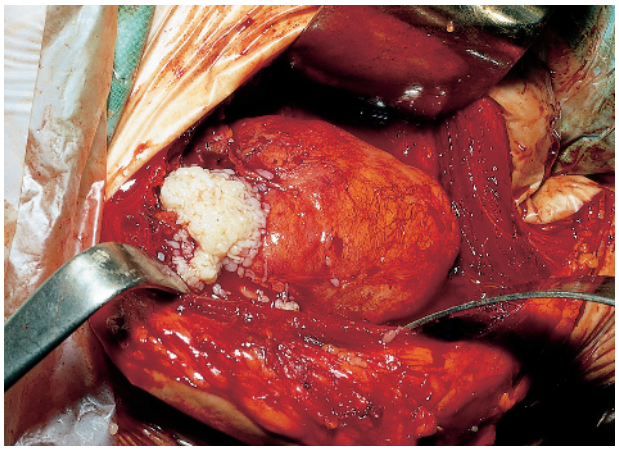
Stage 2: Synovitis Early changes are vascular congestion with new blood vessel formation, proliferation of synoviocytes and infiltration of the subsynovial layers by polymorphs, lymphocytes and plasma cells (Figure 3.2). There is thickening of the capsular structures, villous formation of the synovium and a cell-rich effusion into the joints and tendon sheaths. Although painful, swollen and tender, these structures are still intact and mobile, and the disorder is potentially reversible.

Stage 3: Destruction Persistent inflammation causes joint and tendon destruction. Articular cartilage is eroded, partly by proteolytic enzymes, partly by vascular tissue in the folds of the synovium, and partly due to direct invasion of the cartilage by a pannus of granulation tissue creeping over the articular surface. At the margins of the joint, bone is eroded by tissue invasion and osteoclastic resorption. Similar changes occur in tendon sheaths, causing tenosynovitis, invasion of the collagen bundles and, eventually, partial or complete rupture of tendons. A synovial effusion, often containing copious amounts of fibrinoid material, produces swelling of the joints, tendons and bursae.

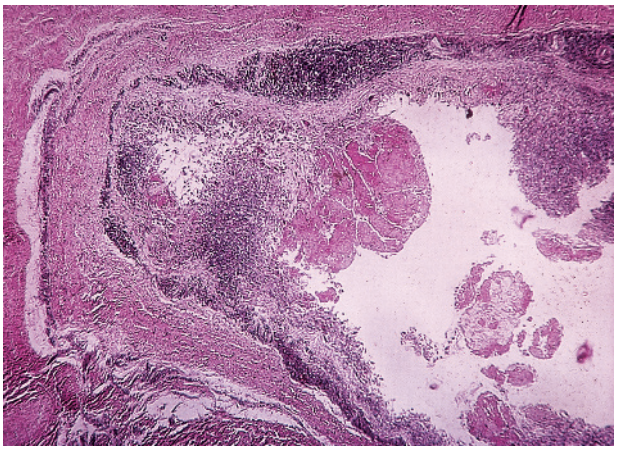
Stage 4: Deformity The combination of articular destruction, capsular stretching and tendon rupture leads to progressive instability and deformity of the joints. The inflammatory process usually continues but the mechanical and functional effects of joint and tendon disruption now become vital.

EXTRA-ARTICULAR TISSUES

Rheumatoid nodules The rheumatoid nodule is a small granulomatous lesion consisting of a central necrotic zone surrounded by a radially disposed



(a)



(b)

Figure 3.2 Rheumatoid synovitis (a) The macroscopic appearance of rheumatoid synovitis with fibrinoid material oozing through a rent in the capsule. (b) Histology shows proliferating synovium with round-cell infiltration and fibrinoid particles in the joint cavity (×120).

palisade of local histiocytes, and beyond that by inflammatory granulation tissue. Nodules occur under the skin (especially over bony prominences), in the synovium, on tendons, in the sclera and in many of the viscera.

Lymphadenopathy Not only the nodes draining inflamed joints, but also those at a distance such as the mediastinal nodes, can be affected. This, as well as a mild *splenomegaly*, is due to hyperactivity of the reticuloendothelial system. More severe splenomegaly can also be associated with neutropaenia as part of *Felty's syndrome*.

Vasculitis This can be a serious and life-threatening complication of RA. Involvement of the skin, including nailfold infarcts, is common but organ infarction can occur.

Muscle weakness Muscle weakness is common. It may be due to a generalized *myopathy* or *neuropathy*, but it is important to exclude spinal cord disease or cord compression due to vertebral displacement (atlantoaxial subluxation). Sensory changes may be part of a neuropathy, but localized sensory and motor symptoms can also result from *nerve compression* by thickened synovium (e.g. carpal tunnel syndrome).

Visceral disease The lungs, heart, kidneys, gastrointestinal tract and brain are sometimes affected. *Ischaemic heart disease* and *osteoporosis* are common complications.

Clinical features

The onset of RA is usually insidious, with symptoms emerging over a period of months. Occasionally the disease starts quite suddenly.

In the early stages the picture is mainly that of a polysynovitis, with soft-tissue swelling and stiffness (Figure 3.3). Typically, a woman of 30–40 years complains of pain, swelling and loss of mobility in the proximal joints of the fingers. There may be a previous history of 'muscle pain', tiredness, loss of weight and a general lack of well-being. As time passes, the symptoms 'spread' to other joints – the wrists, feet, knees and shoulders in order of frequency. Another classic feature is generalized stiffness after periods of inactivity, and especially after rising from bed in the early morning. This early morning stiffness typically lasts longer than 30 minutes.

Physical signs may be minimal, but usually there is symmetrically distributed swelling and tenderness of the metacarpophalangeal joints, the proximal interphalangeal joints and the wrists. Tenosynovitis is common in the extensor compartments of the wrist and the flexor sheaths of the fingers; it is diagnosed by feeling thickening, tenderness and crepitation over the back of the wrist or the palm while passively moving the fingers. If the larger joints are involved, local warmth, synovial hypertrophy and intra-articular effusion may be more obvious. Movements are often limited but the joints are still stable and deformity is unusual.

In the later stages joint deformity becomes increasingly apparent and the acute pain of synovitis is replaced by the more constant ache of progressive joint destruction. The combination of joint instability and tendon rupture produces the typical 'rheumatoid' deformities: ulnar deviation of the fingers, radial and volar displacement of the wrists, valgus knees, valgus feet and clawed toes. Joint movements are restricted and often very painful. About a third of all patients develop pain and stiffness in the cervical spine.

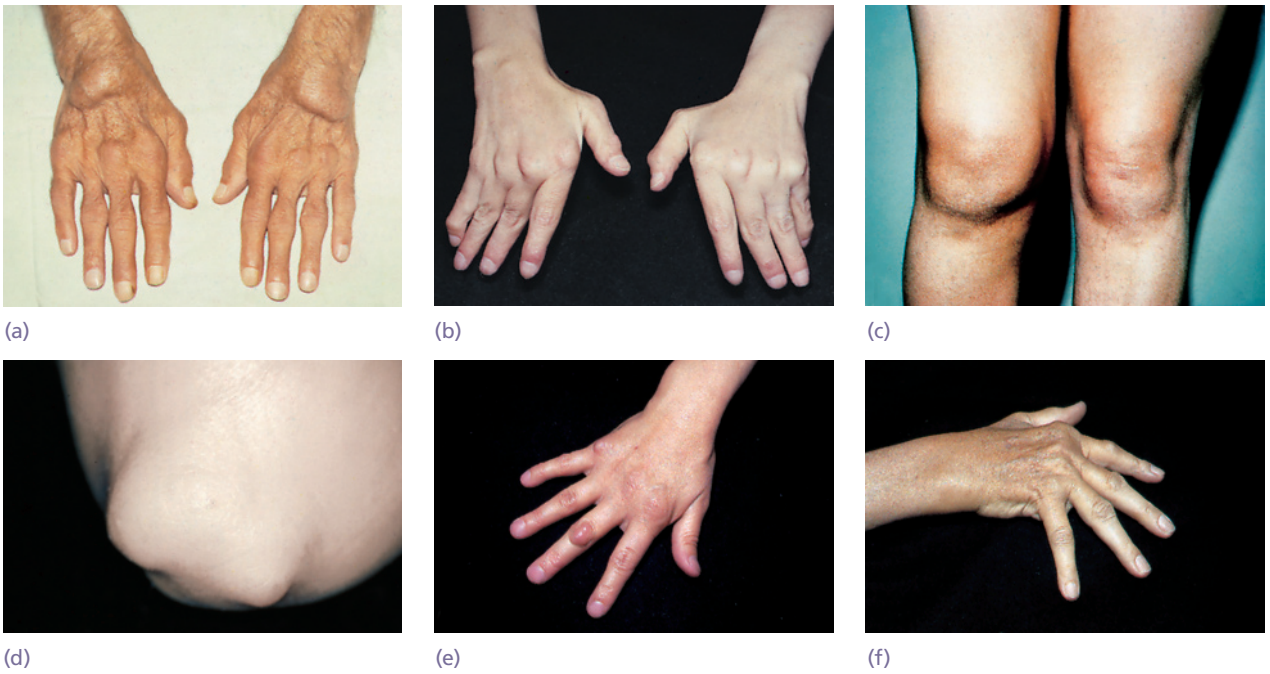


Figure 3.3 Rheumatoid arthritis – clinical features (a) Early features of swelling and stiffness of the proximal finger joints and the wrists. (b) The late hand deformities are so characteristic as to be almost pathognomonic. (c) Occasionally rheumatoid disease starts with synovitis of a single large joint (in this case the right knee). Extra-articular features include subcutaneous nodules (d,e) and tendon ruptures (f).

Function is increasingly disturbed and patients may need help with grooming, dressing and eating.

Extra-articular features These often appear in patients with severe disease. The most characteristic is the appearance of nodules. They are usually found as small subcutaneous lumps, rubbery in consistency, at the back of the elbows, but they also develop in tendons (where they may cause 'triggering' or rupture), in the viscera and the eye. They are pathognomonic of RA, but occur in only 25% of patients.

Less specific features include *muscle wasting*, *lymphadenopathy*, *scleritis*, *nerve entrapment syndromes*, *skin atrophy or ulceration*, *vasculitis* and *peripheral sensory neuropathy*. Marked visceral disease, such as *pulmonary fibrosis*, is rare.

Imaging

X-rays Early on, X-rays show only the features of synovitis: soft-tissue swelling and periarticular osteoporosis. The later stages are marked by the appearance of marginal bony erosions and narrowing of the articular space, especially in the proximal joints of the hands and feet (Figure 3.4). However, most individuals have evidence of erosions within 2 years. In advanced disease, articular destruction and joint deformity are obvious. Flexion and extension views of the cervical spine often show subluxation at the atlantoaxial or mid-cervical levels; surprisingly, this causes few symptoms in the majority of cases.

Ultrasound scanning and MRI The use of other imaging techniques to look at soft-tissue changes and early erosions within joints has become more common. Ultrasound can be particularly useful in defining the presence of synovitis and early erosions. Additional information on vascularity can be obtained if Doppler techniques are used.

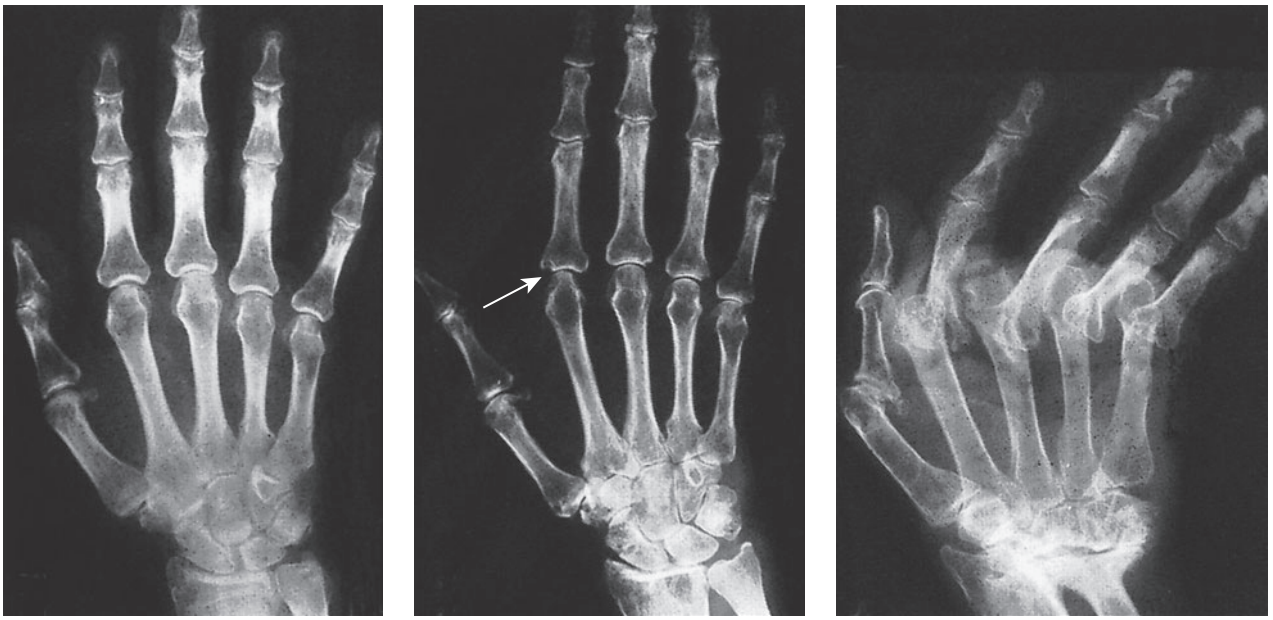
Blood investigations

Normocytic, hypochromic anaemia is common and is a reflection of abnormal erythropoiesis due to disease activity. It may be aggravated by chronic gastrointestinal blood loss caused by non-steroidal anti-inflammatory drugs. In active phases the ESR and CRP concentration are usually raised.

Serological tests for RF are positive in about 80% of patients and antinuclear factors are present in 30%. Neither of these tests is specific and neither is required for a diagnosis of rheumatoid arthritis. Newer tests such as those for anti-CCP antibodies have added much greater specificity but at the expense of sensitivity.

Synovial biopsy

Synovial tissue may be obtained by needle biopsy, via the arthroscope, or by open operation. Unfortunately, most of the histological features of rheumatoid arthritis are non-specific.



(a) (b) (c)

Figure 3.4 Rheumatoid arthritis – X-ray changes The progress of disease is well shown in this patient's X-rays. First, there was only soft-tissue swelling and periarticular osteoporosis; later juxta-articular erosions appeared (arrow); ultimately, the joints became unstable and deformed.

Diagnosis

The usual criteria for diagnosing rheumatoid arthritis are the presence of a bilateral, symmetrical polyarthritis involving the proximal joints of the hands or feet, and persisting for at least 6 weeks. If there are subcutaneous nodules or X-ray signs of periarticular erosions, the diagnosis is certain. *A positive test for RF in the absence of the above features is not sufficient evidence of rheumatoid arthritis, nor does a negative test exclude the diagnosis if the other features are all present.* The chief value of the RF tests is in the assessment of prognosis: persistently high titres herald more serious disease including extra-articular features.

Atypical forms of presentation are not uncommon. The early stages may be punctuated by spells of quiescence, during which the diagnosis is doubted, but sooner or later the more characteristic features appear. Occasionally, in older people, the onset is explosive, with the rapid appearance of severe joint pain and stiffness. Now and then (more so in young women) the disease starts with chronic pain and swelling of a single large joint and it may take months or years before other joints are involved. The presence of tenderness on squeezing across all metacarpophalangeal or metatarsophalangeal joints, early morning stiffness of at least 30 minutes and a raised ESR are highly suggestive of a diagnosis of rheumatoid arthritis. A rapid diagnosis is vital so that early treatment can be started with disease-modifying antirheumatic drugs.

In the differential diagnosis of polyarthritis several disorders must be considered.

Seronegative inflammatory polyarthritis Polyarthritis is a feature of a number of conditions including psoriatic arthritis, adult Still's disease, systemic lupus erythematosus and other connective-tissue diseases. These are considered in later sections.

Ankylosing spondylitis This is primarily an inflammatory disease of the sacroiliac and intervertebral joints, causing back pain and progressive stiffness; however, it may also involve the peripheral joints.

Reiter's disease/reactive arthritis The larger joints and the lumbosacral spine are the main targets. There is usually a history of urethritis or colitis and often also conjunctivitis.

Polyarticular gout Tophaceous gout affecting multiple joints can, at first sight, be mistaken for rheumatoid arthritis. On X-ray, the erosions are quite different from those of rheumatoid arthritis; the diagnosis is clinched by identifying typical birefringent urate crystals in the joint fluid or a nodular tophus.

It is a curious fact that, although both gout and RA are fairly common, the two conditions are rarely seen in the same patient. The reason for this is unknown.

Calcium pyrophosphate deposition disease This condition is usually seen in older people. Typically it affects large joints, but it may occur in the wrist and metacarpophalangeal joints as well. X-ray signs are fairly characteristic and crystals may be identified in synovial fluid or synovium.

Sarcoidosis Sarcoid disease sometimes presents with a symmetrical small-joint polyarthritis and no bone involvement; in other cases a large joint such as the knee or ankle may be involved. Erythema nodosum and hilar lymphadenopathy on chest X-ray are clues to the diagnosis.

Acute sarcoidosis usually subsides spontaneously within 6 months. *Chronic sarcoidosis* produces granulomatous infiltration of lungs, bone, synovium and other organs and is more common in Afro-Caribbean than Caucasian peoples. In addition to polyarthritis and tenosynovitis, there are usually X-ray features of punched-out 'cysts' and cortical erosions in the bones of the hands and feet. The ESR and serum angiotensin converting enzyme (SACE) may be raised. Biopsy of affected tissue shows typical noncaseating granulomas. Treatment with non-steroidal anti-inflammatory drugs (NSAIDs) may be adequate but in more intractable cases corticosteroids or other immunosuppressive therapies are necessary.

Lyme disease This tick-borne spirochaetal infection usually starts with a skin lesion and flu-like symptoms and then spreads to multiple organs. If the initial lesions are missed or left untreated, patients may present with an asymmetrical inflammatory polyarthritis affecting mainly the larger joints. It is most likely to be encountered in known endemic areas in North America, Europe and Asia. In late cases serological tests may be positive. Treatment with doxycycline or one of the newer cephalosporins is usually effective for the arthritic features.

Viral arthritis Viral infections are often associated with a transient polyarthralgia; flu-like illness and a rash will suggest the diagnosis. However, some infections – most typically parvovirus B19 – occasionally cause a symmetrical polysynovitis (including the finger joints) and early morning stiffness, symptoms which may last for several months or may recur over a few years. The absence of 'rheumatoid' X-ray features and subcutaneous nodules will raise suspicions about the diagnosis.

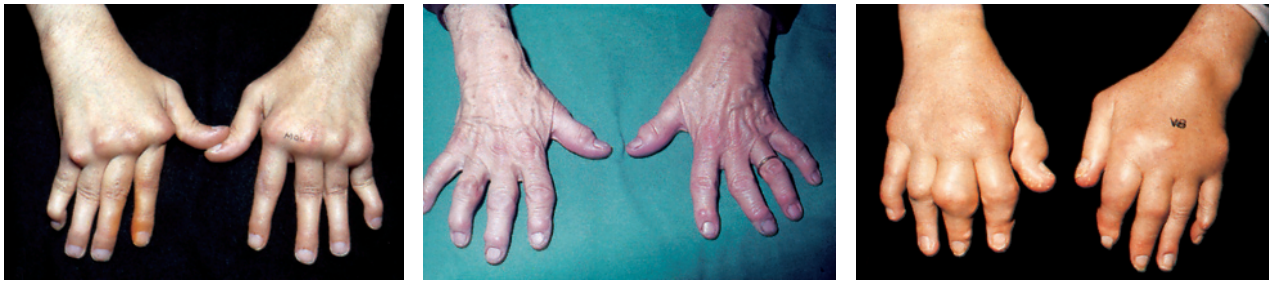
Polymyalgia rheumatica This condition, which is seen mainly in the middle-aged or elderly, is characterized by aching discomfort around the pectoral and pelvic girdles, post-inactivity stiffness and muscular weakness. The joints are not tender but the muscles may be. The ESR and CRP are almost always elevated. Corticosteroids (as little as 10 mg a day) provide rapid and dramatic relief of all symptoms, and this response is often used as a diagnostic test. The condition may be associated with, and certainly carries the risk of, giant cell arteritis which may result in blindness.

Osteoarthritis Polyarticular osteoarthritis (OA), which typically involves the finger joints, is often mistaken for RA. A moment's reflection will usually dispel any doubt: OA always involves the *distal* interphalangeal joints and causes a nodular arthritis with radiologically obvious osteophytes, whereas RA affects the *proximal* joints of the hand and causes predominantly erosive features (see Figure 3.5).

Some confusion may arise from the fact that RA, in its later stages, is associated with loss of articular cartilage and *secondary OA*. Enquiry into the early history will usually untangle the diagnosis. Sometimes, however, RA atypically affects only a few of the larger joints and it is then very difficult to distinguish from OA; X-ray features such as loss of articular cartilage throughout the entire joint and lack of hypertrophic bone changes (sclerosis and osteophytes) should suggest an inflammatory arthritis.

Treatment

There is no cure for rheumatoid arthritis. However, advances in therapy have revolutionized the treatment approach with associated major improvements in outcome. Medical treatment is guided by the principle that inflammation should be reduced rapidly and aggressively. A multidisciplinary approach is helpful from the beginning: ideally the therapeutic team should include a rheumatologist, orthopaedic surgeon, physiotherapist, occupational therapist,



(a) (b) (c)

Figure 3.5 Rheumatoid arthritis – differential diagnosis All three patients presented with painful swollen fingers. In (a) mainly the proximal joints were affected (rheumatoid arthritis); in (b) the distal joints were the worst (Heberden's osteoarthritis); in (c) there were asymmetrical nodular swellings around the joints (gouty tophi).

orthotist and social worker. Their deployment and priorities will vary according to the individual and stage of the disease.

At the onset of the disease both the patient and the doctor will be uncertain about the likely rate of progress. An attempt should be made to determine the likely prognosis. Poor prognosis is associated with female sex, multiple joint involvement, high ESR and CRP, positive RF and anti-CCP, younger age, high BMI, smoking and the presence of erosions at diagnosis.

PRINCIPLES OF MEDICAL MANAGEMENT

Treatment should be aimed at controlling inflammation as rapidly as possible. This is likely to require the use of corticosteroids for their rapid onset (initially oral doses of 30 mg of prednisolone or 120 mg i.m. methylprednisolone may be used). Steroids should be rapidly tapered to prevent significant side effects.

In addition, disease-modifying antirheumatic drugs (DMARDs) should be started at this time. The first choice is now methotrexate at doses of 10–25 mg/week. This may be used initially alone or in combination with sulphasalazine and hydroxychloroquine. Leflunomide can also be considered if methotrexate is not tolerated. Gold and penicillamine are associated with significant side effects and are now used very rarely.

Control of pain and stiffness with non-steroidal anti-inflammatory drugs (NSAIDs) may be needed, maintaining muscle tone and joint mobility by a balanced programme of exercise, and general advice on coping with the activities of daily living.

If there is no satisfactory response to DMARDs, it is wise to progress rapidly to biological therapies such as the TNF inhibitors infliximab, etanercept, golimumab, certolizumab and adalimumab. Other biological therapies include inhibitors of T-cell costimulation (abatacept), IL-6 (tocilizumab) and B-cell depleting therapies (rituximab).

Additional measures include the injection of corticosteroid preparations into inflamed joints and tendon sheaths. It is sometimes feared that such injections may themselves cause damage to articular cartilage or tendons. However, there is little evidence that they are harmful, provided they are used sparingly and with full precautions against infection.

Prolonged rest and immobility is likely to weaken muscles and lead to a worse prognosis. However, some splinting can be helpful at any stage of the disease.

PHYSIOTHERAPY AND OCCUPATIONAL THERAPY

Preventative splinting and orthotic devices may delay the march of events; however, it is important to encourage activity. If these fail to restore and maintain function, operative treatment is indicated.

SURGICAL MANAGEMENT

At first this consists mainly of soft-tissue procedures (synovectomy, tendon repair or replacement and joint stabilization); in some cases osteotomy may be more appropriate.

In late rheumatoid disease, severe joint destruction, fixed deformity and loss of function are clear indications for reconstructive surgery. Arthrodesis, osteotomy and arthroplasty all have their place and are considered in the appropriate chapters. However, it should be recognized that patients who are no longer suffering the pain of active synovitis and who are contented with a limited pattern of life may not want or need heroic surgery merely to improve their anatomy. Careful assessment for occupational therapy, the provision of mechanical aids and adjustments to their home environment may be much more useful. It appears safe to continue methotrexate during elective orthopaedic surgery. However, doses of corticosteroids should be as low as possible and biological therapies such as the TNF inhibitors should be stopped prior to surgery where possible.

BOX 3.1 KEY ELEMENTS IN MEDICAL TREATMENT

Identify patients with RA as early as possible.

Start disease-modifying antirheumatic drugs (DMARDs) immediately.

Consider combination therapy with multiple DMARDs.

If DMARDs fail, progress rapidly to biological therapies such as the TNF inhibitors.

Complications

Fixed deformities The perils of rheumatoid arthritis are often the commonplace ones resulting from ignorance and neglect. Early assessment and planning should prevent postural deformities, which will result in joint contractures.

Muscle weakness Even mild degrees of myopathy or neuropathy, when combined with prolonged inactivity, may lead to profound muscle wasting and weakness. This should be prevented by control of inflammation, physiotherapy and pain control, if possible; if not, the surgeon must be forewarned of the difficulty of postoperative rehabilitation.

Joint rupture Occasionally the joint capsule ruptures and synovial contents spill into the soft tissues. Treatment is directed at the underlying synovitis, i.e. splintage and injection of the joint, with synovectomy as a second resort.

Infection Patients with rheumatoid arthritis – and even more so those on corticosteroid therapy – are susceptible to infection. Sudden clinical deterioration, or increased pain in a single joint, should alert one to the possibility of septic arthritis and the need for joint aspiration.

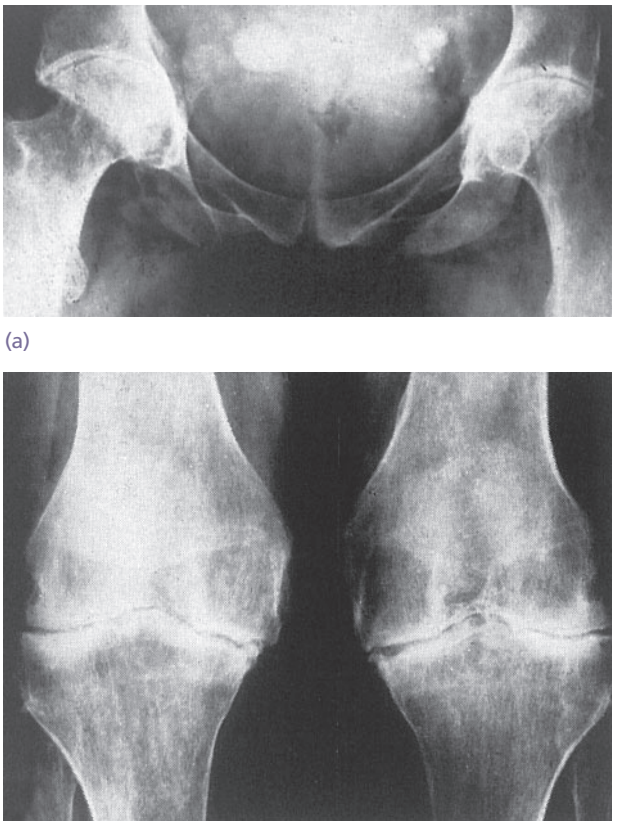
Spinal cord compression This is a rare complication of cervical spine (atlantoaxial) instability. The onset of weakness and upper motor neuron signs in the lower limbs is suspicious. If they occur, immobilization of the neck is essential and spinal fusion should be carried out as soon as possible.

Systemic vasculitis Vasculitis is a rare but potentially serious complication. Corticosteroids and immunosuppressives such as intravenous cyclophosphamide may be required.

Amyloidosis This is another rare but potentially lethal complication of long-standing rheumatoid arthritis. The patient presents with proteinuria and progressive renal failure. Finding amyloid in a rectal or renal biopsy makes the diagnosis. Aggressive control of inflammation has reduced this complication significantly.

Prognosis

Rheumatoid arthritis runs a variable course. When the patient is first seen, it is difficult to predict the outcome, but high titres of RF and anti-CCP, periarticular erosions, rheumatoid nodules, severe muscle wasting, joint contractures and evidence of vasculitis are bad prognostic signs. Women, on the whole, fare somewhat worse than men. Without effective treatment about 10% of patients improve steadily after the first attack of active synovitis; 60% have intermittent phases of disease activity and remission, but with a slow downhill course over many years; 20% have severe joint erosion, which is usually evident within the first 5 years (Figure 3.6); and 10% end up completely disabled. In addition, a reduction in life expectancy by 5–10 years is common and is often due to premature ischaemic heart disease. However, early aggressive medical treatment appears to reduce the morbidity and mortality.



(a)

(b)

Figure 3.6 Rheumatoid arthritis – aftermath After the acute inflammatory phase has passed, the patient may be left with features of secondary osteoarthritis, especially in the hips (a) and the knees (b).

AXIAL SPONDYLOARTHROPATHIES INCLUDING ANKYLOSING SPONDYLITIS

These are generalized chronic inflammatory diseases, but their effects are seen mainly in the spine and sacroiliac joints. Definitions have changed in the last decade to reflect the fact that ankylosing spondylitis (AS) is the end stage of a disease process best described by the term axial spondyloarthropathy (axial SPA). AS is characterized by pain and stiffness of the back, with variable involvement of the hips and shoulders and (more rarely) the peripheral joints. Its reported prevalence is 0.1–0.2% in Western Europe and North America, but it is much lower in Japanese and African peoples. Males are affected more frequently than females (estimates vary from 2:1 to 10:1) and the usual age at onset is between 15 and 25 years. There is a strong tendency to familial aggregation and association with the genetic markers HLA-B27 and ERAP1.

Cause

There is considerable evidence for regarding ankylosing spondylitis (AS) as a genetically determined immunopathological disorder. The disease is much more common in family members of patients than in the general population – HLA-B27 is present in over 95% of Caucasian patients and in half of their first-degree relatives (as compared with 8% of the general population); and racial groups with an unusually low prevalence of AS also show a very low prevalence of HLA-B27 (e.g. less than 1% in Japanese people).

There are various theories about the 'triggering factor' that initiates the abnormal immune response. It may be a bacterial antigen, which closely resembles HLA-B27 that induces an antibody response, which also targets the HLA-B27 positive cells; or, as in the case of RA, the HLA-B27 molecule may be involved in the presentation of a specific antigen to the T-cells, which then react with the antigen-presenting cells. Since classic ankylosing spondylitis is sometimes associated with genitourinary or bowel infection, and disorders such as Reiter's disease and ulcerative colitis cause vertebral and sacroiliac changes indistinguishable from those of ankylosing spondylitis, it has been suggested that the putative organism may be carried to the spine by local lymphatic drainage.

Pathology

There are two basic lesions: synovitis of diarthrodial joints and inflammation at the fibro-osseous junctions of syndesmotic joints and tendons. The preferential involvement of the insertion of tendons and ligaments (the entheses) has resulted in the term *enthesopathy*.

Synovitis of the sacroiliac and vertebral facet joints causes destruction of articular cartilage and periarticular bone. The costovertebral joints also are frequently involved, leading to diminished respiratory excursion. When peripheral joints are affected, the same changes occur.

Inflammation of the fibro-osseous junctions affects the intervertebral discs, sacroiliac ligaments, symphysis pubis, manubrium sterni and the bony insertions of large tendons. Pathological changes proceed in three stages: (1) an inflammatory reaction with cell infiltration, granulation tissue formation and erosion of adjacent bone; (2) replacement of the granulation tissue by fibrous tissue; and (3) ossification of the fibrous tissue, leading to ankylosis of the joint.

Ossification across the surface of the disc gives rise to small bony bridges or syndesmophytes linking adjacent vertebral bodies. If many vertebrae are involved the spine may become absolutely rigid.

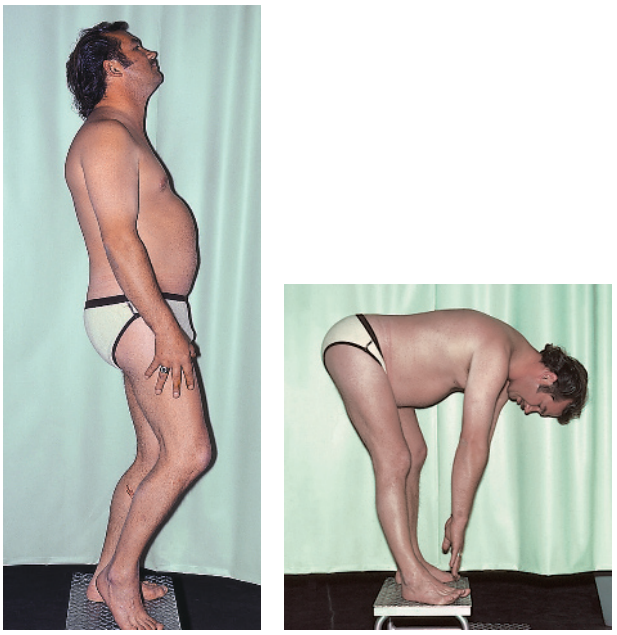
Clinical features

The disease starts insidiously: a teenager or young adult complains of backache and stiffness recurring at intervals over a number of years. This is often diagnosed as 'simple mechanical back pain', but the symptoms are worse in the early morning and after inactivity. Referred pain in the buttocks and thighs may appear as 'sciatica' and some patients are mistakenly treated for intervertebral disc prolapse. Gradually pain and stiffness become continuous and other symptoms begin to appear: general fatigue, pain and swelling of joints, tenderness at the insertion of the Achilles tendon, 'foot strain', or intercostal pain and tenderness.

Occasionally the disease starts with pain and slight swelling in a peripheral joint such as the ankle, or pain and stiffness of the hip. Sooner or later, though, backache will come to the fore. In women the axial skeletal disease may remain restricted to the sacroiliac joints making diagnosis challenging.

Early on there is little to see apart from slight flattening of the lower back and limitation of extension in the lumbar spine (Figure 3.7). There may be diffuse tenderness over the spine and sacroiliac joints, or (occasionally) swelling and tenderness of a single large joint.

In established cases the posture is typical: loss of the normal lumbar lordosis, increased thoracic kyphosis and a forward thrust of the neck; upright posture and balance are maintained by standing with the hips and knees slightly flexed, and in late cases these may become fixed deformities. Spinal movements are diminished in all directions, but loss of extension is always the earliest and the most severe disability. It is revealed dramatically by the 'wall test': the patient is asked to stand with his back to the wall; heels, buttocks, scapulae and occiput should all be able to touch the wall simultaneously. If extension is seriously diminished, the patient will find this impossible. In the most advanced stage the spine may be completely ankylosed from occiput to sacrum – sometimes in positions of grotesque deformity. Marked loss of cervical extension may restrict the line of vision to a few paces.



(a) (b)

Figure 3.7 Ankylosing spondylitis – early The cardinal clinical feature is marked stiffness of the spine. (a) This patient manages to stand upright by keeping his knees slightly flexed. (b) It looks as if he can bend down to touch his toes, but his back is rigid and all the movement takes place at his hips.

Chest expansion, which should be at least 7 cm in young men, is often markedly decreased. In old people, who may have pulmonary disease, this test is unreliable.

Peripheral joints (usually shoulders, hips and knees) are involved in over a third of the patients; they show the features of inflammatory arthritis – swelling, tenderness, effusion and loss of mobility. There may also be tenderness of the ligament and tendon insertions close to a large joint or under the heel.

Extraskeletal manifestations General fatigue and loss of weight are common. Acute anterior uveitis occurs in about 25% of patients; it usually responds well to treatment but, if neglected, may lead to permanent damage including glaucoma. Other extraskeletal disorders, such as aortic valve disease, carditis and pulmonary fibrosis (apical), are rare and occur very late in the disease.

Imaging

X-rays The cardinal sign – and often the earliest – is erosion and fuzziness of the sacroiliac joints. Later there may be periarticular sclerosis, especially on the iliac side of the joint and finally bony ankylosis.

The earliest vertebral change is flattening of the normal anterior concavity of the vertebral body ('squaring'). Later, ossification of the ligaments around the intervertebral discs produces delicate bridges (syndesmophytes) between adjacent vertebrae (Figure 3.8). Bridging at several levels gives the appearance of a 'bamboo spine'.

Osteoporosis is common in long-standing cases and there may be hyperkyphosis of the thoracic spine due to wedging of the vertebral bodies.

Peripheral joints may show erosive arthritis or progressive bony ankylosis.

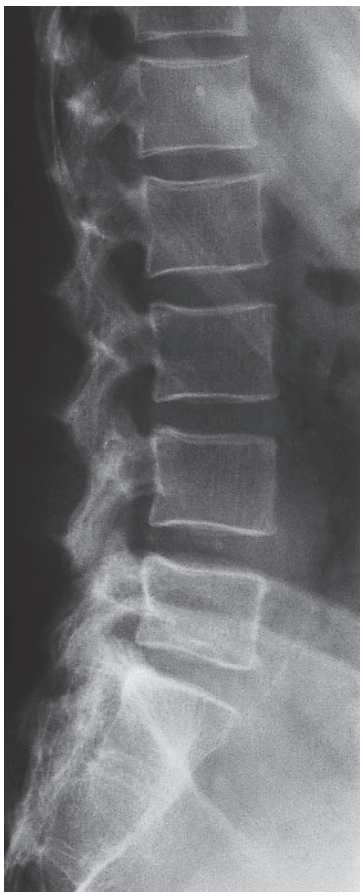
MRI MRI allows detailed investigation of sacroiliac joints and may show typical erosions and features of inflammation such as bone oedema. Various techniques including gadolinium contrast can be used to demonstrate inflammatory lesions in other areas of the spine.

Special investigations

The ESR and CRP are usually elevated during active phases of the disease. HLA-B27 is present in 95% of cases. Serological tests for rheumatoid factor are usually negative.

Diagnosis

Diagnosis is easy in patients with spinal rigidity and typical deformities, but it is often missed in those with early disease before radiographic changes are



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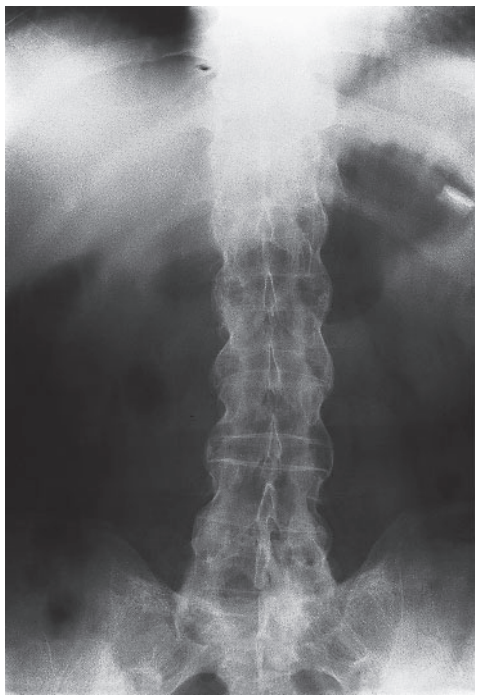

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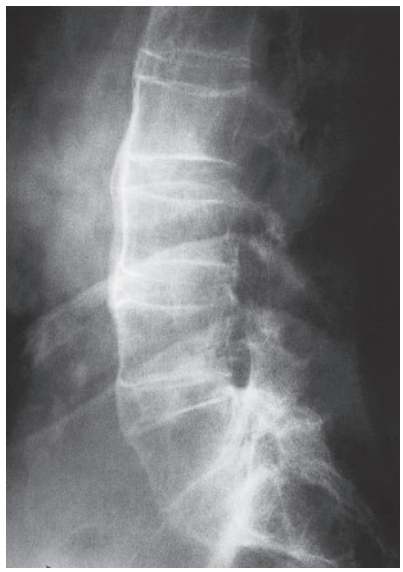


(c)

Figure 3.8 Ankylosing spondylitis – X-rays (a) An early sign is 'squaring' of the lumbar vertebrae. (b,c) Bony bridges (syndesmophytes) between the vertebral bodies convert the spine into a rigid column.

seen (non-radiographic axial SPA) or unusual forms of presentation. In over 10% of cases the disease starts with an asymmetrical inflammatory arthritis – usually of the hip, knee or ankle – and it may be several years before back pain appears. Atypical onset is more common in women, who may show less obvious changes in the sacroiliac joints. A history of AS in a close relative is strongly suggestive.

Mechanical disorders Low back pain in young adults is usually attributed to one of the more common disorders such as muscular strain, facet joint dysfunction or spondylolisthesis. These conditions differ from AS in several ways: the onset of pain is related to specific physical activities, stiffness is less pronounced and symptoms are eased rather than aggravated by inactivity. Tenderness is also more localized and the peripheral joints are normal.

Diffuse idiopathic hyperostosis (Forestier's disease) This is a fairly common disorder, predominantly of older men, characterized by widespread ossification of ligaments and tendon insertions. X-rays show pronounced but asymmetrical intervertebral spur formation and bridging throughout the dorsolumbar spine. Although it bears a superficial resemblance to AS, it is not an inflammatory disease, spinal pain and stiffness are seldom severe, the sacroiliac joints are not eroded and the ESR is normal.

Other seronegative spondyloarthropathies A number of disorders are associated with vertebral and sacroiliac lesions indistinguishable from those of ankylosing spondylitis. They are *Reiter's disease*, *psoriatic arthritis*, *ulcerative colitis*, *Crohn's disease*, *Whipple's disease* and *Behçet's syndrome*. In each there are certain characteristic features: the rash or nail changes of psoriasis, intestinal ulceration in inflammatory bowel disease, genitourinary and ocular inflammation in Reiter's disease, buccal and genital ulceration in Behçet's syndrome. Yet there is considerable overlap between them: all show some familial aggregation and many are associated with the histocompatibility antigen, HLAB27. Patients with one of these disorders (including AS) often have close relatives with another, or with a positive HLA-B27.

Treatment

The disease can be as damaging to a patient as rheumatoid arthritis but some continue to lead an active life. Treatment consists of: (1) general measures to maintain satisfactory posture and preserve movement; (2) anti-inflammatory drugs to counteract pain and stiffness; (3) the use of TNF inhibitors for severe disease (with inhibitors of other cytokines such as IL-17 and IL-12/23 in development); and (4) operations to correct deformity or restore mobility.

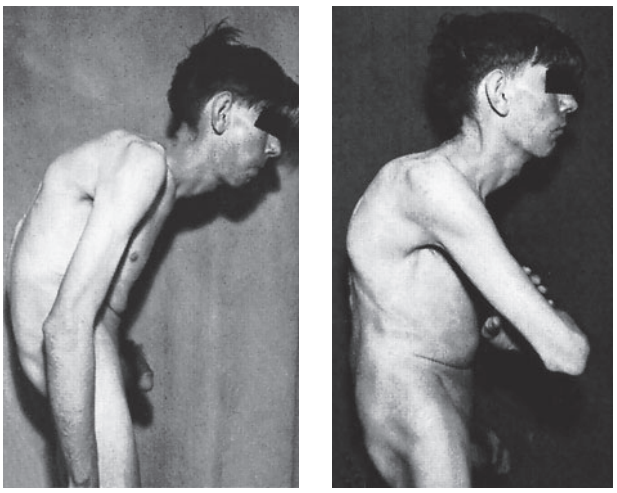
General measures Patients are encouraged to remain active and follow their normal pursuits. They should be taught how to maintain satisfactory posture and urged to perform spinal extension exercises every day. Swimming, dancing and gymnastics are ideal forms of recreation. Rest and immobilization are contraindicated because they tend to increase the general feeling of stiffness.

Non-steroidal anti-inflammatory drugs It is doubtful whether these drugs prevent or retard the progress to ankylosis, but they do control pain and counteract soft-tissue stiffness, thus making it possible to benefit from exercise and activity. They may have to be continued for many years.

TNF inhibitors With the introduction of the TNF inhibitors it has become possible to treat the underlying inflammatory processes active in AS. This can result in significant improvement in disease activity including remission. These therapies are generally reserved for individuals who have failed to be controlled with non-steroidal anti-inflammatory drugs.

Operation Significantly damaged hips can be treated by joint replacement, though this seldom provides more than moderate mobility. Moreover, the incidence of infection is higher than usual and patients may need prolonged rehabilitation.

Deformity of the spine may be severe enough to warrant lumbar or cervical osteotomy (Figure 3.9). These are difficult and potentially hazardous procedures; fortunately, with improved activity and exercise



(a) (b)

Figure 3.9 Ankylosing spondylitis – operative treatment Spinal osteotomy is occasionally performed to correct a severe, rigid deformity. (a) Before operation this man could see only a few paces ahead; (b) after osteotomy his back is still rigid but his posture, function and outlook are improved.

programmes, they are seldom needed. If spinal deformity is combined with hip stiffness, hip replacements (permitting full extension) often suffice.

Complications

Spinal fractures The spine is often both rigid and osteoporotic; fractures may be caused by comparatively mild injuries. The commonest site is C5–7, but it is prudent to X-ray the entire spine in accident victims who have AS. Treatment in these cases is directed at preventing further deformity.

Hyperkyphosis In long-standing cases the spine may become severely kyphotic, so much so that the patient has difficulty lifting his head to see in front of his feet.

Spinal cord compression This is uncommon, but it should be thought of in patients who develop long tract symptoms and signs. It may be caused by atlantoaxial subluxation or by ossification of the posterior longitudinal ligament.

Lumbosacral nerve root compression Patients may occasionally develop root symptoms, including lower limb weakness and paraesthesia, in addition to their 'usual' pelvic girdle symptoms.

PERIPHERAL SPONDYLOARTHROPATHIES

Reiter's syndrome and reactive arthritis

The syndrome described by Hans Reiter in 1916 (and 100 years before that by Benjamin Brodie) is a clinical triad of *urethritis*, *arthritis* and *conjunctivitis* occurring some weeks after either *dysentery* or *genitourinary infection* (Figure 3.10). It is now recognized that this is one of the classic forms of reactive arthritis, i.e. an aseptic inflammatory arthritis associated with non-specific infection (often urogenital or bowel).

Its prevalence is difficult to assess, but it is probably the commonest type of large-joint polyarthritis in young men. It is thought to occur in 1–3% of all people who develop either non-specific urogenital infection or *Shigella* dysentery, but its incidence may be as high as 25% in those who are HLA-B27 positive. Men are affected more often than women (the ratio is about 10:1), but this may simply reflect the difficulty of diagnosing the genitourinary infection in women. The usual age at onset is between 20 and 40 years, but children are affected too – perhaps after an episode of diarrhoea.

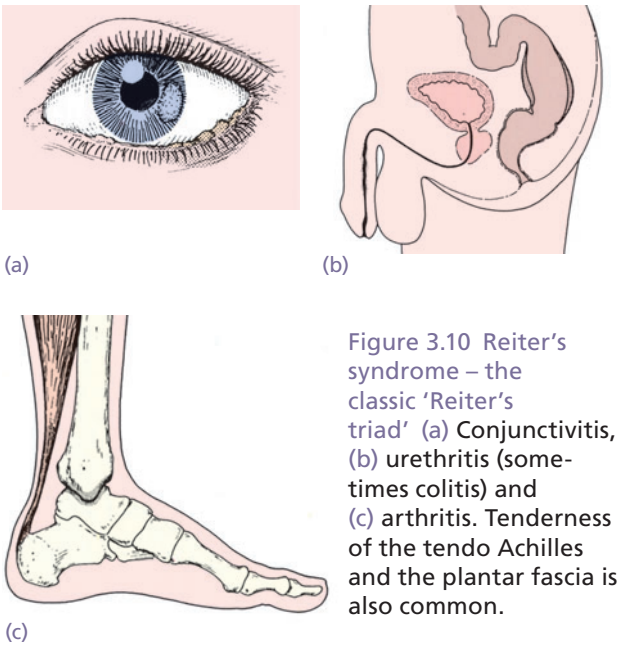


Figure 3.10 Reiter's syndrome – the classic 'Reiter's triad' (a) Conjunctivitis, (b) urethritis (sometimes colitis) and (c) arthritis. Tenderness of the tendo Achilles and the plantar fascia is also common.

Cause

Familial aggregation, overlap with other forms of seronegative spondyloarthropathy in first-degree relatives and a close association with HLA-B27 point to a genetic predisposition, the bowel or genitourinary infection acting as a trigger. Gut pathogens include *Shigella flexneri*, *Salmonella*, *Campylobacter* species and *Yersinia enterocolitica*. *Lymphogranuloma venereum* and *Chlamydia trachomatis* have been implicated as sexually transmitted infections. All these bacteria can survive in human cells; assuming that either the bacterium or a peptide bacterial fragment acts as the antigen, the pathogenesis could be the same as that suggested for ankylosing spondylitis.

Pathology

The pathological changes are essentially the same as those in ankylosing spondylitis, with the emphasis first on subacute large-joint synovitis and in some individuals with a chronic disease course tending towards sacroiliitis and spondylitis.

Clinical features

Acute phase The acute phase of the disease is marked by an asymmetrical inflammatory arthritis of the lower limb joints – usually the knee and ankle but often the tarsal and toe joints as well. The joint may be acutely painful, hot and swollen with a tense effusion, suggesting gout or infection. Tendo Achilles tenderness and plantar fasciitis (evidence of enthesopathy) are common, and the patient may complain of backache even in the early stage. Conjunctivitis, urethritis and bowel infections are often mild and easily

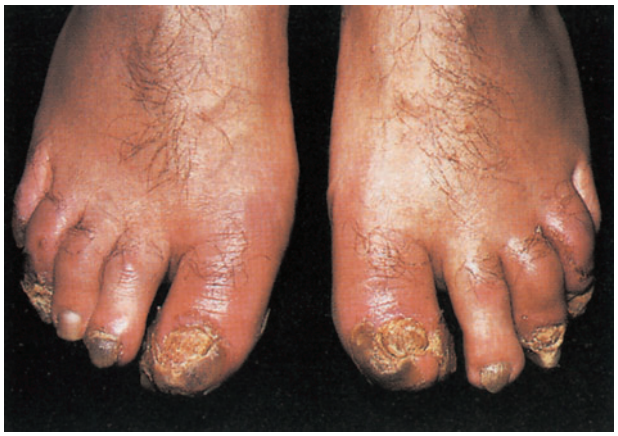


Figure 3.11 Reiter's disease – other features The characteristic pustular dermatitis of the feet – keratoderma blennorrhagicum.

missed; the patient should be carefully questioned about symptoms during the previous few weeks. Cystitis and cervicitis may occur in women.

Less frequent, but equally characteristic, features are a vesicular or pustular dermatitis of the feet (keratoderma blennorrhagica – Figure 3.11), balanitis and mild buccal ulceration.

The acute disorder usually lasts for a few weeks or months and then subsides, but most patients have either recurrent attacks of arthritis or other features of chronic disease.

Chronic phase The chronic phase is more characteristic of a spondyloarthropathy. Over half of the patients with Reiter's disease complain of mild, recurrent episodes of polyarthritis (including upper limb joints). About half of those again develop sacroiliitis and spondylitis with features resembling those of ankylosing spondylitis. Uveitis is also fairly common and may give rise to posterior synechiae and glaucoma.

X-rays

Sacroiliac and vertebral changes are similar to those of ankylosing spondylitis. If peripheral joints are involved, they may show features of erosive arthritis.

Special investigations

Tests for HLA-B27 are positive in 75% of patients with sacroiliitis. The ESR may be high in the active phase of the disease. The causative organism can sometimes be isolated from urethral fluids or faeces, and tests for antibodies may be positive.

Diagnosis

The diagnosis should be considered in any young adult who presents with an acute or subacute arthritis in the lower limbs. It is more likely to be missed in women, in children and in those with very mild (and often forgotten) episodes of genitourinary or bowel infection. Some patients never develop the full syndrome and one should be alert to the *formes fruste* with large-joint arthritis alone.

Gout and infective arthritis Reiter's disease, gout and infection should all be considered in the differential diagnosis of inflammation in a large peripheral joint. Examination of synovial fluid for organisms and crystals may provide important clues.

Gonococcal arthritis Gonococcal arthritis takes two forms: (1) bacterial infection of the joint; and (2) a reactive arthritis with sterile joint fluid. A history of genitourinary infection further complicates the distinction from Reiter's disease, and diagnosis may depend on identifying the organism or gonococcal antibodies.

Enteropathic arthritis Ulcerative colitis and Crohn's disease may be associated with subacute synovitis, causing pain and swelling of one or more of the peripheral joints. These subside when the intestinal disease is controlled.

Treatment

Initial treatment for Reiter's disease should be aimed at ensuring the infectious organism responsible has been cleared. This is particularly important for sexually transmitted infections such as *Chlamydia trachomatis*.

Even if the triggering infection is identified, treating it will have no effect on the reactive arthritis. However, there is some evidence that treatment of *Chlamydia* infection with tetracycline for periods of up to 3 months can reduce the risk of recurrent joint disease.

Symptomatic treatment could include the use of analgesia and non-steroidal anti-inflammatory drugs. If the inflammatory response is aggressive, local injection of corticosteroids or even intramuscular methylprednisolone may be useful. If symptoms and signs do not resolve, DMARDs used in the treatment of RA may be needed. Topical steroids may be used for uveitis.

Psoriatic arthritis

Psoriatic arthritis is characterized by seronegative polysynovitis, erosive (sometimes very destructive) arthritis, enthesitis and dactylitis and a significant incidence of sacroiliitis and spondylitis.

The prevalence of psoriasis is 1–2%, but only about 5% of those affected will develop psoriatic arthritis. The usual age at onset is 30–50 years (often later than the skin lesions).

Cause

As with the other spondyloarthropathies, there is a strong genetic component: patients often give a family history of psoriasis; there is a significantly increased incidence of other spondyloarthropathies in close relatives; and 60% of those with psoriatic spondylitis or sacroiliitis have HLA-B27.

Psoriatic skin lesions may well be a reactive phenomenon, and the joint lesions a form of 'reactive arthritis'. However, no specific trigger agent has thus far been identified.

Pathology

The joint changes are similar to those in rheumatoid arthritis – chronic synovitis with cell infiltration and exudate, going on to fibrosis. Cartilage and bone destruction may be unusually severe ('arthritis mutilans'). However, rheumatoid nodules are not seen. Sacroiliac and spine changes, which occur in about 30% of patients, are similar to those in ankylosing spondylitis.

Clinical features

The patient may present with one of several patterns of joint involvement. These include: arthritis of distal interphalangeal joints (Figure 3.12), 'arthritis mutilans', asymmetrical large joint oligoarthritis and patterns mimicking rheumatoid arthritis or ankylosing spondylitis. Psoriasis of the skin or nails usually precedes the arthritis, but hidden lesions (in the natal cleft or umbilicus) are easily overlooked.

The condition can progress slowly or very rapidly and may become quiescent. Sometimes (particularly in women) joint involvement is more symmetrical, and in these cases the condition may be indistinguishable from seronegative rheumatoid arthritis. Asymmetrical swelling of two or three fingers may be due to a combination of interphalangeal arthritis and tenosynovitis.

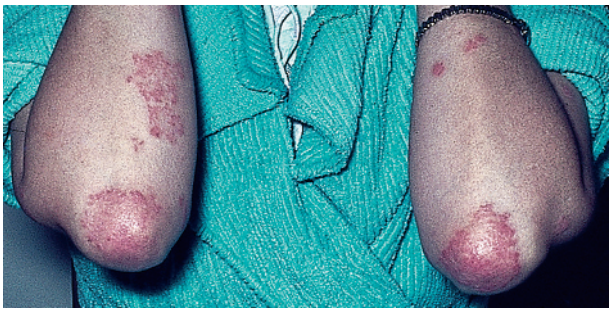
Sacroiliitis and spondylitis are seen in about one-third of patients, and occasionally this is the predominant change with a clinical picture resembling ankylosing spondylitis. As in the other spondyloarthropathies, heel pain (*enthesitis*) is not uncommon.

In the worst cases both the spine and the peripheral joints may be involved. Fingers and toes are severely deformed due to erosion and instability of the interphalangeal joints (*arthritis mutilans* – Figure 3.13).

Ocular inflammation occurs in about 30% of patients.

Imaging

X-ray examination may show severe destruction of the interphalangeal joints of the hands and feet; changes



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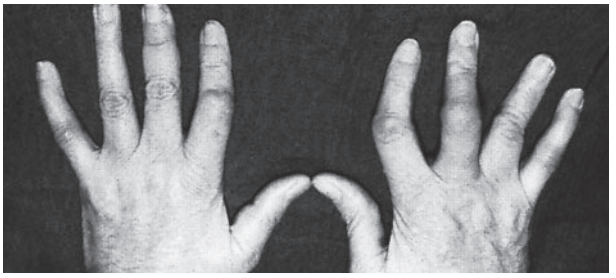

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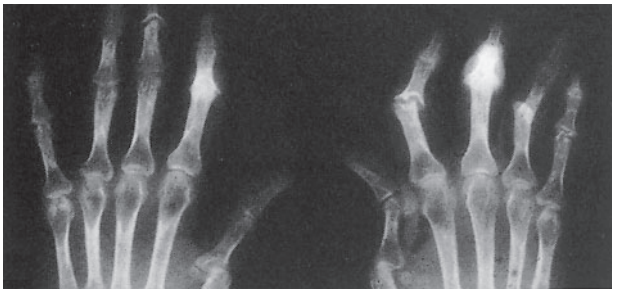
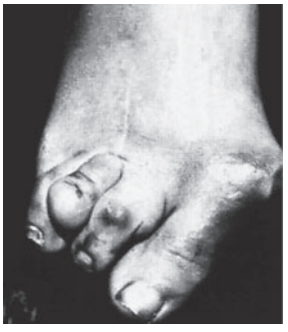


(c)

Figure 3.12 Psoriatic arthritis (1) (a) Psoriasis of the elbows and forearms; (b) typical finger deformities and (c) X-rays show distal joint involvement – clearly the disease is not simply rheumatoid arthritis in a patient with psoriasis.



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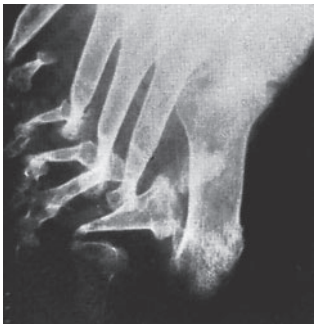


(b)

Figure 3.13 Psoriatic arthritis (2) (a and b) The feet and toes are often involved. In this case the patient developed a severely destructive form of the disease (arthritis mutilans).

in the large joints are similar to those of rheumatoid disease. Sacroiliac erosion is fairly common; if the spine is involved the appearances are identical to those of ankylosing spondylitis.

Ultrasound scanning and *MRI* may show greater definition of the extent and activity of synovitis.

Special investigations

Tests for rheumatoid factor are almost always negative. HLA-B27 occurs in 50–60%, especially in those with overt sacroiliitis.

Diagnosis

The main difficulty is to distinguish 'psoriatic arthritis' from 'psoriasis with seronegative RA'. The important distinguishing features of psoriatic arthritis are: (1) asymmetrical joint distribution; (2) involvement of distal finger joints; (3) the presence of sacroiliitis or spondylitis; and (4) the absence of rheumatoid nodules.

Treatment

In mild disease no more than topical preparations to control the skin disease and NSAIDs for the arthritis are needed. In resistant forms of arthritis, immunosuppressive agents (methotrexate) and TNF inhibitors have proved effective. Inhibitors of other cytokines such as IL-17 and IL-12/23 are in development and oral therapies that target intracellular signalling are also available (apremilast). Surgery may be needed for unstable joints. Arthrodesis of the distal interphalangeal joints may greatly improve function.

Enteropathic arthritis

Both Crohn's disease and ulcerative colitis may be associated with either peripheral arthritis or sacroiliitis and spondylitis.

Peripheral arthritis

Peripheral arthritis is fairly common, occurring in about 15% of patients with inflammatory bowel disease. Typically one or perhaps a few of the larger joints are involved. Pain and swelling may appear quite suddenly and last for 2–3 months before subsiding. Synovitis is usually the only feature but joint erosion can occur. Men and women are affected with equal frequency and there is no particular association with HLA-B27.

Treatment is directed at the underlying disorder: attacks of arthritis are often triggered by a flare-up of bowel disease and when the latter is brought under control the arthritis can disappear. Anti-inflammatory drugs should not generally be used as they may have a deleterious effect on the bowel disease. Other treatment options are local corticosteroid injection and disease-modifying treatments such as methotrexate. This may also improve the bowel disease. In severe cases TNF inhibitors may be needed.

Sacroiliitis and spondylitis

This pattern is seen in about 10% of patients with inflammatory bowel disease, and in half of these patients the clinical picture closely resembles that of ankylosing spondylitis. HLA-B27 is positive in 60% and there is an increased incidence of ankylosing spondylitis in close relatives. Unlike the peripheral arthritis, sacroiliitis shows no temporal relationship to gastrointestinal inflammation and its course is unaffected by treatment of the bowel disease. Management is the same as that of ankylosing spondylitis.

Complications

In addition to spondyloarthritis, there are several unusual but important complications of inflammatory bowel disease that may confuse the clinical picture.

Septic arthritis of the hip Infection may spread directly from the bowel. The patient presents with a fever and pain in the groin. Hip movements are limited and there may be swelling due to an abscess. Treatment is by antibiotics and operative drainage.

Psoas abscess In Crohn's disease a posterior fistula may track into the psoas sheath. The patient complains of back pain and may develop a typical psoas abscess with pain in the hip, limitation of movement and a tender mass in the groin. Treatment is by operative drainage of the abscess.

Osteopaenia Patients with chronic bowel disease often develop osteoporosis and osteomalacia – partly due to malabsorption and partly as a consequence of treatment with corticosteroids. Compression fractures of the spine may cause severe back pain.

Juvenile idiopathic arthritis

Juvenile idiopathic arthritis (JIA) is the preferred term for non-infective inflammatory joint disease of more than 3 months' duration in children under 16 years of age. It embraces a group of disorders in all of which pain, swelling and stiffness of the joints are common features. The prevalence is about 1 per 1000 children, and boys and girls are affected with equal frequency.

The cause is similar to that of rheumatoid arthritis: an abnormal immune response to some antigen in children with a particular genetic predisposition. However, rheumatoid factor is usually absent.

The pathology, too, may be like that of rheumatoid arthritis: primarily a synovial inflammation leading to fibrosis and ankylosis. Stiffening tends to occur in whatever position the joint is allowed to assume; thus flexion deformities are a common and characteristic feature. Chronic inflammation and alterations in the local blood supply may affect the epiphyseal growth plates, leading to both local bone deformities and an overall retardation of growth. However, cartilage erosion is less marked than in rheumatoid arthritis and severe joint instability is uncommon.

Clinical features

Children with JIA present in several characteristic ways. About 15% have a *systemic illness*, and arthritis only develops somewhat later; the majority (50–60%) have an *oligoarticular arthritis* affecting a few of the larger joints; about 10% present with *polyarticular arthritis*, sometimes closely resembling RA; the remaining 5–10% develop *enthesitis-related JIA*.

SYSTEMIC JIA

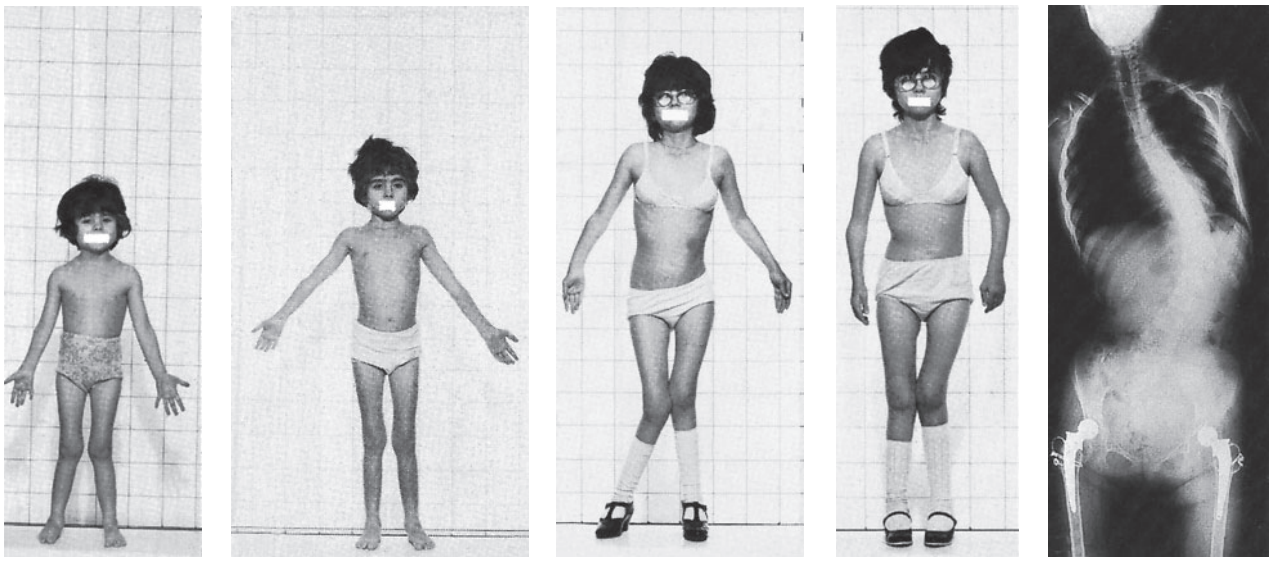
This, the classic *Still's disease*, is usually seen below the age of 3 years and affects boys and girls equally. It starts with intermittent fever, rashes and malaise; during these episodes, which occur almost daily, the child appears to be quite ill but after a few hours the clinical condition improves again. Less constant features are lymphadenopathy, splenomegaly and hepatomegaly. Joint swelling occurs some weeks or months after the onset; fortunately, it usually resolves when the systemic illness subsides, but it may go on to progressive seronegative polyarthritis, leading to permanent deformity of the larger joints and fusion of the cervical apophyseal joints. By puberty there may be stunting of growth, often abetted by the earlier use of corticosteroids.

OLIGOARTICULAR JIA

This is by far the commonest form of JIA. It usually occurs below the age of 6 years and is much more common in girls (Figure 3.14); occasionally older children are affected. Only a few joints are involved and there is no systemic illness. The child presents with pain and swelling of medium-sized joints (knees, ankles, elbows and wrists); sometimes only one joint is affected. Rheumatoid factor tests are negative but antinuclear antibodies (ANA) may be positive. A serious complication is chronic iridocyclitis, which occurs in about 50% of patients. The arthritis often goes into remission after a few years but by then the child is left with asymmetrical deformities and growth defects that may be permanent.

POLYARTICULAR JIA

Polyarticular arthritis, typically with involvement of the temporomandibular joints and the cervical spine, is usually seen in older children, mainly girls. The hands and wrists are often affected, but the classic deformities of rheumatoid arthritis are uncommon and rheumatoid factor is usually absent. In some cases, however, the condition is indistinguishable from adult rheumatoid arthritis, with a positive rheumatoid factor



(a) (b) (c) (d) (e)

3.14 Juvenile idiopathic arthritis (a–d) This young girl developed JIA when she was 5 years old. Here we see her at 6, 9 and 14 years of age. The arthritis has become inactive, leaving her with a knee deformity which was treated by osteotomy. Her eyes, too, were affected by iridocyclitis. (Courtesy of Mr Malcolm Swann and Dr Barbara Ansell). (e) X-ray of another young girl who required hip replacements at the age of 14 years and, later, surgical correction of her scoliosis.

test; these probably warrant the designation 'juvenile rheumatoid arthritis'.

ENTHESITIS-RELATED JIA

In older children – usually boys – the condition may take the form of sacroiliitis and spondylitis; hips and knees are sometimes involved as well. Tests for HLA-B27 are often positive and this should probably be regarded as 'juvenile ankylosing spondylitis'.

X-rays

In early disease non-specific changes such as soft tissue swelling may be seen, but X-ray is mainly useful to exclude other painful disorders. Later there may be signs of progressive joint erosion and deformity.

Investigations

The white cell count and ESR are markedly raised in systemic JIA, less so in the other forms. Rheumatoid factor tests are positive only in juvenile RA. Joint aspiration and synovial fluid examination may be essential to exclude infection or haemarthrosis.

Diagnosis

In the early stages, before chronic arthritis is fully established, diagnosis may be difficult. Systemic JIA may start with an illness resembling a viral infection. Oligoarticular JIA, especially if only one joint is involved, is indistinguishable from *Reiter's disease* or *septic arthritis* (if the signs are acute) or *tuberculous synovitis* (if they are more subdued).

Other conditions that need to be excluded are *rheumatic fever*, one of the *bleeding disorders* and *leukaemia*. In most cases the problem is resolved once the full pattern of joint involvement is established, but blood investigations, joint aspiration and synovial biopsy may be required to clinch the diagnosis.

Treatment

General treatment Systemic treatment is similar to that of rheumatoid arthritis, including the use of second-line drugs such as hydroxychloroquine, sulphasalazine or low-dose methotrexate for those with seropositive juvenile RA. Corticosteroids should be used only for severe systemic disease and for chronic iridocyclitis unresponsive to topical therapy. Severe inflammatory disease may need to be treated with cytokine inhibitors such as anti-TNF therapies.

Children and parents alike need sympathetic counselling to help them cope with the difficulties of social adjustments, education and training.

Local treatment The priorities are to prevent stiffness and deformity. Night splints may be useful for the wrists, hands, knees and ankles; prone lying for some period of each day may prevent flexion contracture of the hips. Between periods of splinting, active exercises are encouraged; these are started by the physiotherapist but the parents must be taught how to continue the programme.

Fixed deformities may need correction by serial plasters or by a spell in hospital on a continuous passive motion (CPM) machine; when progress is no longer being made, joint capsulotomy may help. For painful eroded joints, useful procedures include arthroplasty of the hip and knee (even in children), custom implants are occasionally required due to the small sizes required, and arthrodesis of the wrist or ankle.

Complications

Ankylosis While most patients recover good function, some loss of movement is common. Hips, knees and elbows may be unable to extend fully, and in the enthesitis-related form of JIA the spine, hips and knees may be almost rigid. Temporomandibular ankylosis and stiffness of the cervical spine can make general anaesthesia difficult and dangerous.

Growth defects There is a general retardation of growth, aggravated by prolonged corticosteroid therapy. In addition, epiphyseal disturbances lead to characteristic deformities: external torsion of the tibia, dysplasia of the distal ulna, underdevelopment of the mandible, shortness of the neck and scoliosis.

Fractures Children with chronic joint disease may suffer osteoporosis and they are prone to fractures.

Iridocyclitis This is most common in ANA-positive oligoarticular disease; untreated it may lead to blindness.

Amyloidosis In children with long-standing active disease there is a serious risk of amyloidosis, which may be fatal.

Prognosis

Fortunately, most children with JIA recover from the arthritis and are left with only moderate deformity and limitation of function. However, 5–10% are severely crippled and require treatment throughout life.

A significant number of children with JIA (about 3%) still die – usually as a result of renal failure due to amyloidosis, or following overwhelming infection.

CONNECTIVE TISSUE DISEASES

This term is applied to a group of closely related conditions that have features which overlap with those of rheumatoid arthritis. Like RA, these are 'autoimmune disorders', probably triggered

by environmental exposures, such as viral infections, in genetically predisposed individuals. They include systemic lupus erythematosus, scleroderma, Sjögren's syndrome, polymyositis, dermatomyositis and a number of overlap syndromes with features of more than one disease.

Systemic lupus erythematosus (SLE)

Systemic lupus occurs mainly in young females and may be difficult to differentiate from RA. Although joint pain is usual, it is often overshadowed by systemic symptoms such as malaise, anorexia, weight loss and fever. Characteristic clinical features are skin rashes (especially the 'butterfly rash' of the face), Raynaud's phenomenon, peripheral vasculitis, plenomegaly, and disorders of the kidney, heart, lung, eye and central nervous system. Anaemia, leucopaenia and elevation of the ESR are common. Tests for ANA are usually positive.

Treatment

Corticosteroids, hydroxychloroquine and immunosuppressants (mycophenolate mofetil or azathioprine), cytotoxics (cyclophosphamide) or biological therapies (rituximab) may be required for life. Progressive joint deformity is unusual and the arthritis can almost always be controlled.

Complications

A curious complication of SLE is avascular necrosis (usually of the femoral head). This may be due in part to the corticosteroid treatment, but the disease itself seems to predispose to bone ischaemia, possibly as a manifestation of the antiphospholipid (Hughes) syndrome which sometimes accompanies SLE.

FURTHER READING

Deighton CM, George E, Kiely PDW, *et al.* Updating the British Society for Rheumatology guidelines for anti-tumour necrosis factor therapy in adult rheumatoid arthritis (again). *Rheumatology* 2006; **45**: 649–52.

Kennedy T, McCabe C, Struthers G, *et al.* BSR guidelines on standards of care for persons with rheumatoid arthritis. *Rheumatology* 2005; **44**: 553–6.

Manadan AM, James N, Block JA. New therapeutic approaches for spondyloarthritis. *Curr Opin Rheumatol* 2007; **19**: 259–64.

Scott DL, Kingsley GH. Tumor necrosis factor inhibitors for rheumatoid arthritis. *N Engl J Med* 2006; **355**: 704–12.

Sidoropolous PL, Hatemi G, Song M, *et al.* Evidence-based recommendations for the management of ankylosing spondylitis: systematic literature search of the 3E Initiative in Rheumatology involving a broad panel of experts and practising rheumatologist. *Rheumatology* 2008; **47**: 355–61.

Crystal deposition disorders

4

Paul Creamer & Dimitris Kassimos

GOUT

Definition

Gout is a crystal deposition disease caused by deposition of monosodium urate crystals in joints and other tissues, secondary to hyperuricaemia.

History

No rheumatic disease has had more impact on the affairs of man than gout: throughout history there are reports of the great and the good suffering the agony of podagra (from the Greek for 'foot trap'). The term 'gout' derives from the Latin word *gutta*, meaning a drop: a common explanation for acute gout in early times being the concept of a poison dropping into the joint. Hippocrates (460–370 BC) clearly refers, in his *Aphorisms*, to the rarity of gout in premenopausal women and children. In 1683, the English physician Thomas Sydenham, himself a sufferer, gave a detailed clinical description of gout identifying a number of distinguishing features that we continue to recognize.

Epidemiology

Gout is often classified into primary and secondary forms. The great majority (95%) of cases occur in the absence of any obvious cause and are due to constitutional under-excretion of urate at the kidney. Secondary gout may be due to rare hereditary conditions or acquired disorders such myeloproliferative diseases, diuretic use or renal failure.

Gout is the most common inflammatory joint disease in men with peak onset in the fifth decade. Risk factors include family history (30-40%), alcohol (50%), renal impairment and obesity. Atherosclerosis, hyperlipidaemia and hypertension are important associated morbidities. A second group in whom gout is common are older females, invariably on diuretics. In both these groups under-excretion of urate results in hyperuricaemia.

Onset of gout in men prior to adulthood or in women before menopause is rare and should raise the possibility of an inherited disorder of urate metabolism resulting in overproduction. These include deficiency of hypoxanthine–guanine phosphoribosyltransferase (HGPRT): either complete (X-linked, Lesch–Nyhan syndrome) or, more commonly, partial (Kelley–Seegmiller syndrome).

Prevalence of gout varies widely but is increasing. It is estimated that 1–2.5% of adults in developed countries are affected.

Pathophysiology

Uric acid is predominantly produced in the liver as the end product of purine metabolism. Consumption of purine-rich foods may contribute to urate load but this effect is small compared to under-excretion. Hyperuricaemia is defined as serum uric acid (SUA) level over 420 μmol/L in men and 360 μmol/L in women. However, relative to most primates and all other mammals, all humans have high levels of serum urate, approaching saturation point (at which crystallization occurs). The reason for these high levels is the lack of uricase, the enzyme that breaks down urate to allantoin, which is more soluble and readily excreted. The human genome shows evidence of uricase but it has been selectively inactivated by a series of mutations, implying that maintenance of a high SUA has an evolutionary advantage. Urate is the most abundant natural antioxidant in the human body and may confer resistance to infection; recent studies have also examined a possible protective effect against neurodegeneration.

Renal excretion is complex: 90% of filtered urate is actively reabsorbed via urate transporter proteins (URAT1) in the proximal convoluted tubules. Further excretion and reabsorption occur in the distal tubules.

Hyperuricaemia is a prerequisite for formation of urate crystals but pH, temperature and the presence (or absence) of natural inhibitors are also important

– otherwise all hyperuricaemic patients would be constantly forming crystals. Stimulation of the NALP3 inflammasome and other humoral and cellular inflammatory mediators by monosodium urate crystals results in acute gouty arthritis with a neutrophilic synovitis. Chronic cumulative urate crystal formation in tissue fluids leads to deposition of monosodium urate crystals in the synovium, cartilage, tendons and soft tissues, resulting in tophi formation and chronic tophaceous gouty arthritis.

Clinical features

Asymptomatic hyperuricaemia is ten times more common than gout, and the majority of patients with hyperuricaemia do not develop gout. The annual incidence of gout is estimated to be about 5–10% in patients with SUA above 420 µmol/L.

Seventy per cent of acute gout affects the first metatarsophalangeal (MTP) joints (podagra). Other sites include the knee, ankle, midfoot, elbow and wrist. Attacks often begin at night and within a few hours the affected joint becomes red, hot, swollen and extremely painful. Gout is rarely just 'painful' or 'uncomfortable'; the patient may be unable to walk, or bear the touch of bedclothes. Systemic features such as fever and malaise may occur especially during polyarticular attacks. The natural history is for the attack to settle after 5–7 days, often with desquamation of the skin over the affected joint. Acute gout may also cause bursitis, tendinitis and, notable in the elderly, cellulitis of the lower limb.

Untreated, mass collections of urate (tophi) may be deposited in and around joints, notable at the elbows, over the small joints of the hands (Figure 4.1) and in the ear. In joints tophi result in erosive disease (chronic gouty arthropathy) (Figure 4.2).

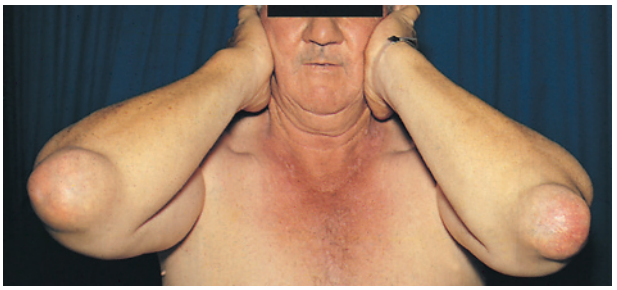
Comorbidities

Gout is associated with a number of comorbidities including *metabolic syndrome*, *cardiovascular disease*, *hypertension* and *diabetes*. Although a cause and effect relationship has not been clearly established, recent studies have suggested that hyperuricaemia may be an independent risk factor of *atherosclerotic disease*.

Investigation

Hyperuricaemia remains the cardinal feature of gout, but levels may be normal during acute attacks. If gout is suspected, repeated estimations of SUA over a period of a few weeks can be of great value. SUA is also useful when monitoring the effects of urate-lowering therapy.

Radiographs are unhelpful in the acute attack, either being normal or showing soft-tissue swelling.



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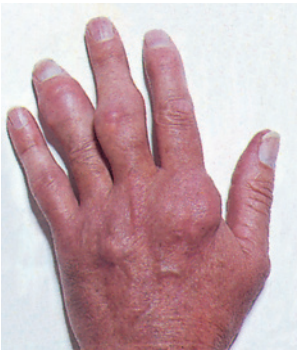

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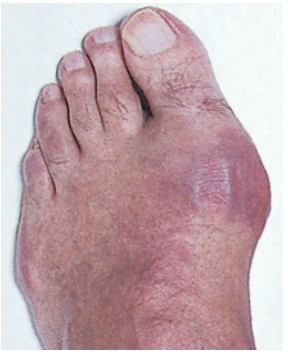


(c)

Figure 4.1 Gouty tophi (a) Large olecranon bursae with subcutaneous tophi over the elbows. (b,c) Tophaceous gout affecting the hands and feet; note the typical swelling of the first MTP joint.

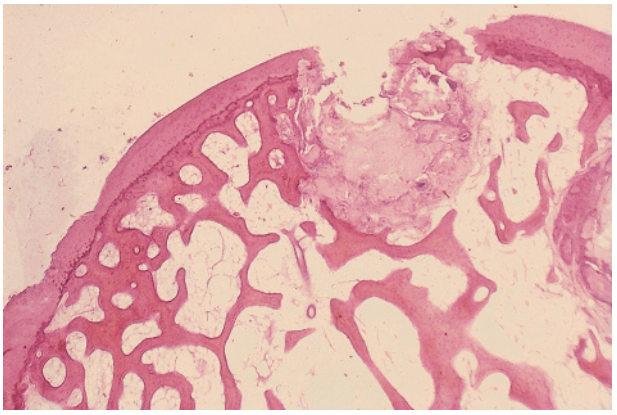


Figure 4.2 Gout pathology Histological section through a gouty MTP joint, showing the urate tophus eroding the articular surface.

In chronic gout, erosions may be seen: classically these are away from the joint margin, being 'punched-out' with a rounded or oval shape and overhanging edge (Figure 4.3).

Acute gout can only be diagnosed with certainty by identifying urate crystals in synovial fluid, bursa or aspirate of tophus. Crystals are needle-shaped and strongly negatively birefringent under polarized light (Figure 4.4). Absence of the crystals does not rule out

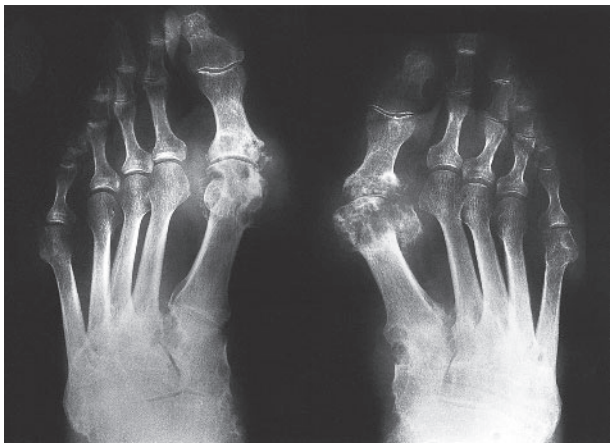
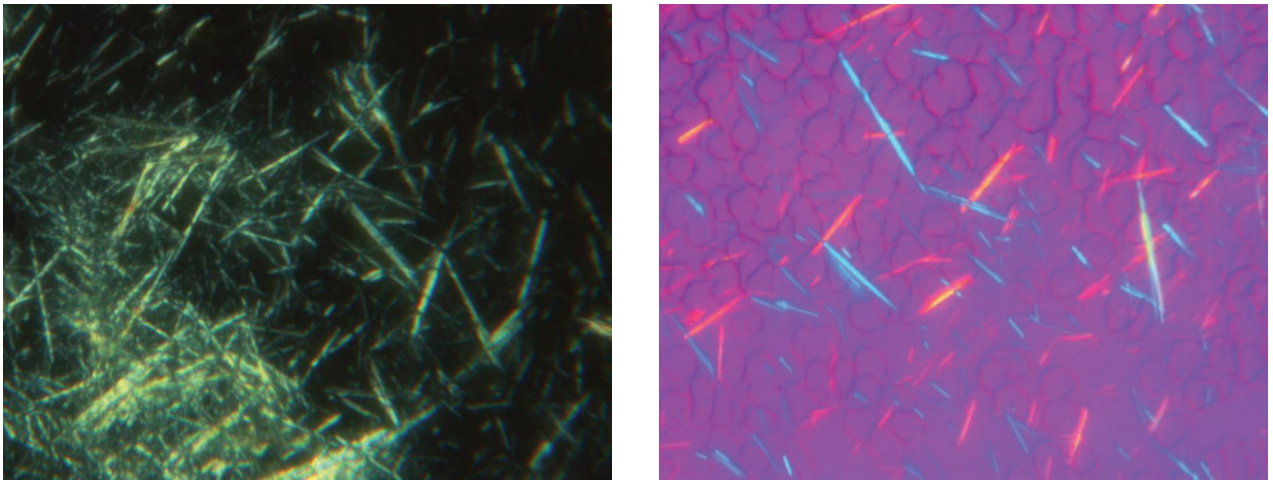


Figure 4.3 Chronic gout Note the large periarticular erosions – tophi consisting of uric acid deposits.

the diagnosis. In acute gout, synovial fluid is highly inflammatory, with white blood cell counts of ≥2,000 cells/mm^3. Even when uric acid crystals are seen, the joint fluid should also be examined for the simultaneous presence of other crystals, especially calcium pyrophosphate crystals, and should undergo Gram stain and culture to rule out co-infection.

Differential diagnosis

Septic arthritis is the most important differential diagnosis and for this reason, as well as attempting to identify urate crystals, urgent joint aspiration is generally indicated. The presence of acute polyarticular inflammation in an otherwise well individual may sway the differential diagnosis towards gout or another crystal disease and away from sepsis (see Table 4.1).



(a) (b)

Figure 4.4 Urate crystals Slender needles or rods, appearing bright white under normal light (a). In compensated polarized light they are strongly negatively birefringent (b). (Images courtesy of Andrew Bird, Senior Biomedical Scientist, Cytology, North Bristol NHS Trust.)

Table 4.1 Differential diagnosis of gout and septic arthritis

	Gout	Septic arthritis
Monoarticular	YES	YES
Polyarticular	++	+
Fever	++	+++
Onset of inflammation	Typically in hours, especially at night	Typically 1–2 days
Resolution of symptoms without treatment	Yes, 7–14 days	No resolution without i.v. antibiotics
Risk factors	Renal insufficiency, obesity hypertension, medication (diuretics, aspirin, cyclosporin)	Immunocompromise, intravenous drug use, prosthetic joint
Elevated systemic inflammatory markers (ESR, CRP)	YES	YES
WBC synovial fluid	2000–100 000, occasionally higher	50 000 typical, often 100 000
Synovial fluid microscopy	Negative birefringent, needle-shaped urate crystals	Pathogenic organism (occasionally negative)
Serum uric acid	High (occasionally during acute attack level can be normal)	Not informative

Polyarticular gout affecting the small joints of the hands may be mistaken for *rheumatoid arthritis*; a further confounder is the similar appearance of tophi and rheumatoid nodules, especially at the elbow. RA and gout seldom occur together. Other *inflammatory arthritis* (including psoriatic) are also differential diagnoses.

Pyrophosphate crystal deposition (see below) may cause an acute arthritis indistinguishable from gout though tending to affect large rather than small joints and being equally common in women and men. Articular calcification may show on X-ray. Demonstrating the CPP crystals in synovial fluid establishes the diagnosis.

Acute 'inflammatory' osteoarthritis may present as a single inflamed joint. Local intra-articular pathology such as pigmented villo-nodular synovitis should be considered though rarely results in the acute inflammation seen in gout. Finally, periarticular gout can cause inflammation that is primarily in the subcutaneous tissues and which simulates *cellulitis*: failure to respond to antibiotic should prompt consideration of gout as an alternative in a susceptible individual

Treatment

ACUTE GOUT

Anti-inflammatory agents (NSAIDs) are the drug of choice for the treatment of acute gout without comorbid diseases (e.g. indomethacin 50–75 mg bd, naproxen 500 mg bd, etorocoxib 120 mg). A 'decrescendo' regime is employed, using large doses at first, tapering over 5–7 days.

NSAIDs are unsuitable for many patients with gout due to renal impairment, congestive cardiac failure, peptic ulcer disease or use of anti-coagulant treatment. *Steroids* are probably the best alternative: perhaps intra-articular if only one joint is affected or orally or IM if joint access is difficult or gout is polyarticular.

Colchicine is also helpful, though limited by gastrointestinal side effects such as diarrhoea and nausea. Diarrhoea in the presence of acute podagra is an unfortunate combination and may put patients off ever taking colchicine again, which can deny them a potentially valuable medication. Treatment should be initiated early, perhaps at 0.5 mg four times per day, tapering over a week.

Non-drug treatment including rest, ice packs and splinting will help, as will powerful analgesia.

CHRONIC GOUT

Patient education is important in the management of chronic gout. Appropriate lifestyle modification, weight loss and dietary advice such as reduction of alcohol and fructose consumption can be powerful interventions.

In some patients with gout, a decision may be made to offer long-term *urate-lowering therapy (ULT)*. Relative indications for this may include: recurrent gout attacks (especially if acute attacks are difficult to treat, for example due to comorbidities), chronic gouty arthropathy, tophi, urate renal stones; however, the decision to initiate such lifelong treatment should always be made in conjunction with the patient. The aim of ULT is to maintain SUA below the saturation point for monosodium urate. ULT has the potential to 'cure' acute gout and also reverse crystal deposition. There is good evidence that treatment is more effective in both regards if the SUA is maintained at low levels, not simply within the 'normal range'. European League Against Rheumatism (EULAR) guidelines recommend a target SUA of less than 360 μmol/L. The British Society for Rheumatology (BSR) guidelines suggest an even stricter target of less than 300 μmol/L.

First-line ULT agents are those that reduce formation of urate by inhibiting xanthine oxidase. *Allopurinol* is the drug of choice. *As it is being introduced, it can trigger a paradoxical flare of gout, therefore it should not be started until at least 4 weeks after the last acute attack and prophylaxis should be offered (NSAID or low-dose colchicine) until the SUA is stable in the target range.* The starting dose is usually 100 mg/day, but 50 mg or even 50 mg on alternate days should be considered in the elderly or those with renal impairment. The dose is gradually titrated upwards until SUA is in the target range. Allopurinol is very safe though hypersensitivity may rarely occur.

Febuxostat is an alternative inhibitor of xanthine oxidase, metabolized and excreted by the liver, so no dose adjustment appears to be necessary in patients with mild-to-moderate renal impairment. Side-effect profile and the risk of gout flares are similar to allopurinol. It may be useful in patients with intolerance to allopurinol or who develop allopurinol hypersensitivity syndrome.

Other ULT agents are 'uricosuric', increasing excretion of urate at the kidney. In general they are less effective than allopurinol and are contraindicated in urolithiasis. Examples include probenecid and sulphinpyrazone, which act by inhibiting URAT1. Benzbromarone is an effective uricosuric but is not widely available.

Despite the availability of cheap, effective treatments, there is good evidence that gout is often poorly diagnosed and suboptimally managed. Compliance with ULT remains poor, with only 50% of patients still receiving allopurinol after the first year of therapy, emphasizing the role of education and explanation.

Surgery About 5% of patients with gout present to an orthopaedic surgeon, usually in one of three categories: infection of an affected area, poor hand

function (operation requested to improve function), or intolerable deformity (operation requested for cosmesis). Surgery on large tophaceous deposits may have a risk of delayed postoperative wound healing because of the poor circulation. However, severe infection may be less common as tophaceous material is generally regarded as bacteriostatic.

CALCIUM PYROPHOSPHATE CRYSTAL ASSOCIATED ARTHROPATHY (CPPD)

CPPD is usually the consequence of cartilage changes related to ageing, degeneration, enzymatic degradation or trauma which result in deposition of calcium pyrophosphate (CPP) crystals. It may also be associated with metabolic disorders that cause local changes in ionic calcium and pyrophosphate equilibrium and may, rarely, be familial. Recent EULAR guidelines (Table 4.2) have attempted to clarify the confusing nomenclature of CPPD.

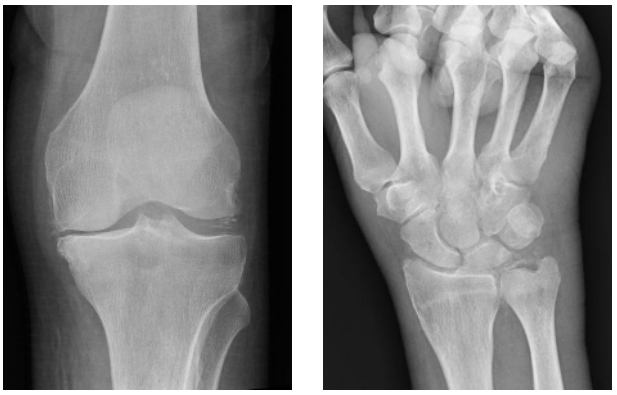
Pathology

Pyrophosphate is probably generated in abnormal cartilage by enzyme activity at chondrocyte surfaces, combining with calcium ions in the matrix where crystal nucleation occurs on collagen fibres. The crystals grow to form nests of amorphous material in the cartilage matrix.

CARTILAGE CALCIFICATION (CC)

Also known as *chondrocalcinosis*, this is a radiographic diagnosis, referring to the appearance of calcification, usually due to CPP deposition, in cartilage. This is generally seen in fibrocartilage, as cloudy, irregular opacities in the menisci of the knee (see Figure 4.5a), triangular fibrocartilage complex of the wrist (Figure 4.5b), pubic symphysis and intervertebral discs. It is often bilateral and symmetrical. CC may also occur in hyaline articular cartilage as a thin line parallel to the joint.

CC is strongly associated with age, prevalence rising from 10–15% in those aged 65–75 to 30–36% in those older than 85 years. A number of metabolic conditions have been linked to CC but some probably reflect a chance concurrence of common age-related conditions. Putative associations with diabetes, hypothyroidism and uraemia may fall into this category. The strongest evidence for association is with haemochromatosis, hypomagnesaemia, hypophosphatasia and, possibly, Wilson's disease, though small numbers limit the ability to link these conditions with certainty.



(a) (b)

Figure 4.5 Cartilage calcification CC (also known as chondrocalcinosis) at knee (a) and wrist (b).

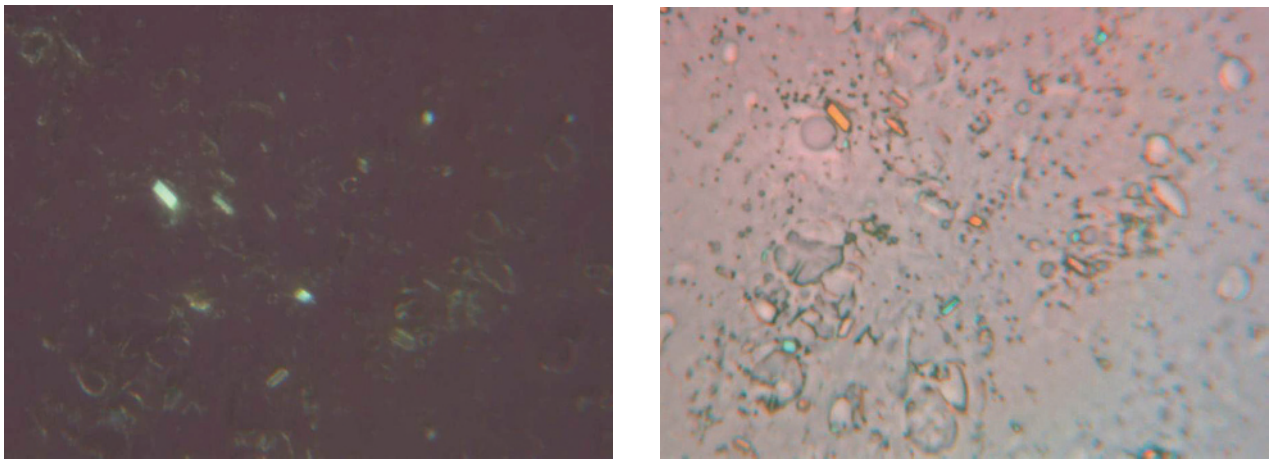
ACUTE CPP ARTHRITIS ('PSEUDOGOUT')

CPP crystals may trigger an acute inflammatory reaction within the joint. Direct trauma, intercurrent medical illness, surgery and blood transfusion are all potential triggers, though many attacks occur for no clear reason. This is the commonest cause of acute monoarthritis in the elderly, especially in hospital patients. Any joint may be involved but knee, wrist, shoulder and ankle are most common. Typical onset is with swelling and pain, which becomes severe over 6–24 hours. Fever is common and elderly patients in particular may appear systemically unwell.

Differentiation from gout can be very difficult, especially if the first MTP is affected ('*pseudopodagra*'). Acute CPP tends to have a slightly less acute onset, attacks last longer (7–10 days) and the pain may be marginally less severe than gout, but in practical terms crystal identification (positively birefringent

Table 4.2 European League Against Rheumatism (EULAR) terminology for CPP crystal deposition (Zhang et al., 2011)

CPP	Calcium pyrophosphate dehydrate crystals	
CPPD	Umbrella term for all instances of occurrence of CCP crystals	
CC	Cartilage calcification, as seen on X-ray or other imaging: usually (but not always) caused by CPP	Also known as 'chondrocalcinosis'
Clinical presentations associated with CPPD	Asymptomatic CPPD	No apparent clinical sequelae
	Acute CPP arthritis	Also known as 'pseudogout'
	OA with CPPD	CPPD in a joint that also shows OA
	Chronic CPP crystal inflammatory arthritis	Chronic inflammatory arthropathy associated with CPPD



(a) (b)

Figure 4.6 Pyrophosphate crystals CPPD crystals are seen as several different shapes (rods, barrels, rhomboid and ovoid). Only the larger ones are visible under normal light (a). In compensated polarized light they are weakly positively birefringent (b). (Images courtesy of Andrew Bird, Senior Biomedical Scientist, Cytology, North Bristol NHS Trust.)

rhomboid-shaped crystals) is required (Figure 4.6). CPP crystals are harder to identify than urate and failure to report does not exclude a diagnosis of acute CPP arthritis. X-rays may show CC.

The main differential diagnosis of acute CPP is gout. Other differentials are the same as those for gout (e.g. septic arthritis, reactive arthritis and other seronegative arthritidies). It follows that joint aspiration and examination of fluid for infection and crystals is mandatory. Acute CPP may be precipitated by trauma and a patient thought to have simple post-traumatic haemarthrosis may also have acute CPP.

Treatment options include rest, analgesia, aspiration and injection with steroid, NSAIDs. Colchicine may also be effective – a good response to this drug does not differentiate gout and acute CPP.

OSTEOARTHRITIS WITH CPPD

The presence of CPP crystals appears to modulate the development of osteoarthritis (OA). Characteristically, there is a hypertrophic reaction with marked osteophyte formation. There are the usual features of pain, stiffness, swelling, joint crepitus and loss of movement. Synovitis is more obvious than in OA without CPPD and there may be chronic symptoms with or without attacks of crystal-induced arthritis. The subgroup is commonest in older women, with polyarticular OA affecting knees in particular but also more unusual joints, such as the metacarpophalangeal (MCP), radiocarpal, midcarpal, glenohumeral, ankle and midfoot.

The characteristic X-ray features arise from a combination of intra-articular and periarticular calcification and OA. OA appearances are similar to those of non-CPPD disease apart from unusual distribution. In advanced cases, joint destruction may be marked, with the formation of loose bodies.

Haemochromatosis is an uncommon disorder of middle-aged people (usually men), resulting from chronic iron overload. The clinical features are those of cirrhosis and diabetes, with a typical bronze pigmentation of the skin. About half of the patients develop joint symptoms (particularly in the hands and fingers). As indicated above, it is associated with CC but may also result in a form of OA, though unusual in that MCP joints are affected. The plasma iron and iron-binding capacity are raised.

CHRONIC CPP CRYSTAL INFLAMMATORY ARTHRITIS

This presents as a chronic oligo- or polyarthritis with inflammatory symptoms and signs and often an acute phase response with elevated ESR or CRP. It thus forms part of the differential diagnosis of RA. There may be typical acute attacks of acute CPP arthritis. Commonest affected joints are knee, radiocarpal and glenohumeral.

BASIC CALCIUM PHOSPHATE CRYSTAL DEPOSITION DISEASE

Basic calcium phosphate (BCP) is a normal component of bone, as calcium hydroxyapatite crystals. BCP crystal deposition may occur in and around joints as a result of local tissue damage – strained or torn ligaments, tendon attrition and cartilage damage or degeneration. Such deposition is common, especially with increasing age. It is usually asymptomatic but may give rise to acute periarthritis or tendinitis. BCP crystals are also seen in some patients with an aggressive, atrophic form of OA; whether they cause the arthritis or simply reflect bone destruction from other causes remains uncertain.

Pathology

BCP crystals are deposited in relatively avascular or damaged parts of tendons and ligaments – most notably around the shoulder and knee – and also around chondrocytes in articular cartilage. The deposits grow by crystal accretion and eventually may be seen on X-ray. Macroscopically the BCP deposit has a chalky appearance. The deposit may be completely inert but, for reasons that are often unclear, may suddenly provoke an acute vascular reaction and inflammation. Crystal shedding into joints may give rise to synovitis.

Clinical features

Two clinical syndromes are associated with BCP crystal deposition: (1) an acute or subacute periarthritis; and (2) a chronic rapidly destructive arthritis.

ACUTE PERIARTHRITIS

This is by far the commonest form of BCP crystal deposition disorder affecting joints and it usually affects the rotator cuff. The patient, usually a female between 30 and 50 years of age, complains of pain around the shoulder. Symptoms may start suddenly, often at night and perhaps after minor trauma, and rise to a crescendo during which the tissues around the joint are swollen, warm and exquisitely tender. Although often difficult to differentiate, the tenderness is maximal around the tendon rather than the joint itself. Less commonly the knee may be affected. Surgical intervention may disclose a tense globule of creamy material oozing from between the frayed fibres of tendon or ligament.

Diagnosis is usually presumptive from the history. Radiographs may show calcification around the shoulder (Figure 4.7). Detection of BCP crystals is not practical in routine service: individual crystals are less than 0.1 µm long and cannot be seen on light microscopy. Alizarin red staining is very sensitive but may be non-specific and result in false positives. Transmission electron microscopy is the most accurate method of detection.

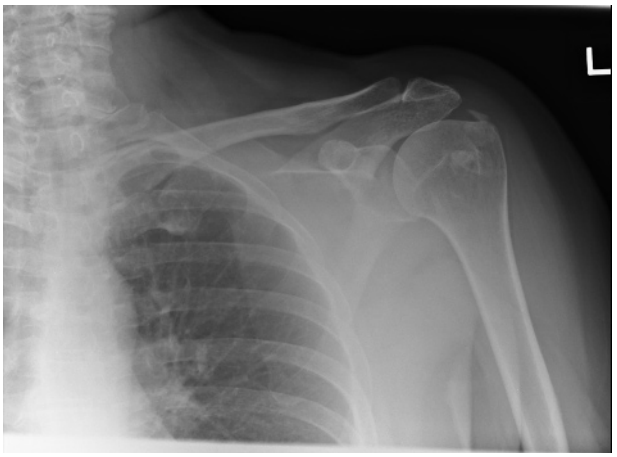
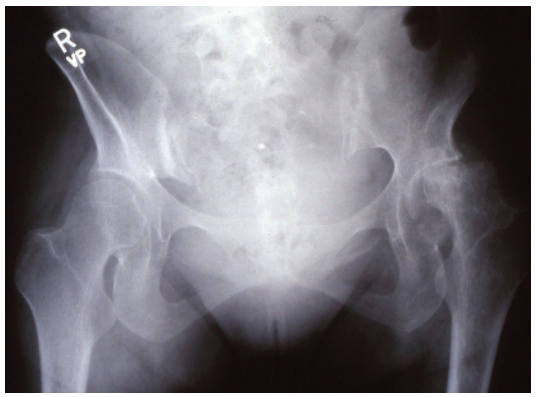


Figure 4.7 Calcification in supraspinatus

Treatment of acute periarthritis includes rest and non-steroidal anti-inflammatory drugs. Resistant cases may respond to local injection of corticosteroids during the acute attack; repeated injections for lesser pain may dampen the repair process in damaged tendons or ligaments and thus predispose to recurrence. Persistent pain and tenderness may call for operative removal of the calcific deposit or 'decompression' of the affected tendon or ligament.

CHRONIC DESTRUCTIVE ARTHRITIS

A destructive form of OA affecting the shoulder or hip (rarely, knee) has been described in elderly individuals, associated with rotator cuff defects and aggregates of BCP in the fluid. Patients are usually female and aged over 70, presenting with pain, swelling and loss of function of the affected shoulder. Night pain is often present. On examination a large effusion is typically seen: occasionally rupture of the capsule leads to extravasation of blood and synovial fluid into surrounding tissues. The glenohumeral joint has reduced movement in all planes. Aspiration of the joint is frequently blood-stained. X-ray (Figure 4.8) shows loss of the articular space, with little or no sclerosis or osteophyte formation. Rapid erosion and destruction



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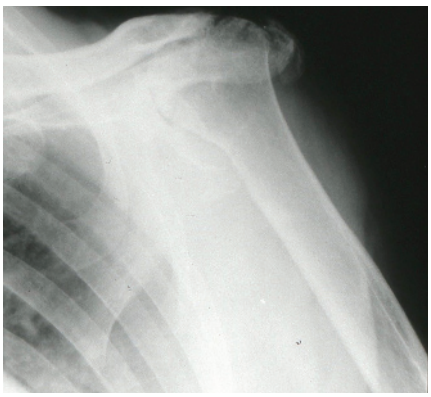


(b)

Figure 4.8 Apatite-associated destructive arthropathy (rapidly destructive OA) Radiographs of the hip (a) and shoulder (b). Common features are rapid progression to joint disruption, crumbling of the subarticular bone and periarticular ossification.

of subchondral bone may occur, resulting in instability and dislocation.

This syndrome was described in 1981 by McCarty and his colleagues from Milwaukee and acquired the sobriquet 'Milwaukee shoulder'. Large quantities of BCP can frequently be seen in the effusions: whether this represents the cause of the inflammation and joint destruction or is simply the effect of bone destruction due to an atrophic form of OA is unclear.

Management is often difficult: aspiration and steroid injection may be tried but effusions usually recur and the inevitable associated massive rotator cuff tears make surgery difficult.

SUMMARY

Crystal deposition diseases are common, although most are managed in primary care or by medical intervention. For the orthopaedic surgeon, their principal significance is as an important differential of acute monoarthritis: sepsis should always be excluded but an appreciation of acute crystal disease as an alternative cause may spare the patient prolonged antibiotics. Crystals may also be responsible for a destructive arthritis, requiring surgical intervention. Possible reasons to refer to rheumatology might include diagnostic uncertainty, severe recurrent attacks not responding to standard therapy, chronic tophaceous gout, or gout with renal disease or calculi. A number of drugs are now available to manage crystal arthropathy successfully in the long term and with careful treatment, outcomes can be excellent, especially for gout.

FURTHER READING

Gout

Diaz-Torné C, Perez-Herrero N, Perez-Ruiz F. New medications in development for the treatment of hyperuricemia of gout. *Curr Opin Rheumatol* 2015; **27**(2): 164–9.

Doherty M. New insights into the epidemiology of gout. *Rheumatology* 2009; **48**: ii2–ii8.

Khanna D, Fitzgerald JD, Khanna PP, *et al.* American College of Rheumatology guidelines for management of gout. Part 1: Systematic nonpharmacologic therapeutic approaches to hyperuricemia. *Arthritis Care Res (Hoboken)* 2012; **64**(10): 1431–46.

Lee JH, Park JY, Seo JW, *et al.* Surgical treatment of subcutaneous tophaceous gout. *J Plast Reconst Aesthet Sur* 2010; **63**: 1933–5.

Oda M, Satta Y, Takenaka O, *et al.* Loss of urate oxidase activity in humanoids and its evolutionary implications. *Mol Biol Evol* 2002; **19**: 640–53.

Richette P, Bardin T. Gout. *Lancet* 2010; **375**: 318–28.

Zhang W, Doherty M, Bardin T, *et al.* EULAR evidence based recommendations for gout. Part II: Management. Report of a task force of the EULAR standing committee for international clinical studies including therapeutics (ESCIST). *Ann Rheum Dis* 2006; **65**: 1312–24.

Zhang W, Doherty M, Pascual E, *et al.* EULAR evidence based recommendations for gout. Part I: Diagnosis. Report of a task force of the Standing Committee for International Clinical Studies including Therapeutics (ESCISIT). *Ann Rheum Dis* 2006; **65**:1301–11.

CPPD

Dieppe PA, Alexander GJM, Jones HE, *et al.* Pyrophosphate arthropathy: a clinical and radiological study of 105 cases. *Ann Rheum Dis* 1982; **41**: 371–6.

Ea HK, Liote F. Diagnosis and clinical manifestations of calcium pyrophosphate and basic calcium phosphate crystal deposition diseases. *Rheum Dis Clin N America* 2014; **40**: 207–29.

Zhang W, Doherty M, Bardin T, *et al.* European League Against Rheumatism recommendations for calcium pyrophosphate deposition. Part I: Terminology and diagnosis. *Ann Rheum Dis* 2011; **70**: 563–70.

BCP arthritis

McCarty DJ, Halverson PB, Carrera GF, *et al.* 'Milwaukee shoulder' – association of microspheroids containing hydroxyapatite crystals, active collagenase and neutral protease with rotator cuff defects. *Arthritis Rheum* 1981; **24**: 464–73.

Resnick CS, Resnick D. Crystal deposition disease. *Semin Arthritis Rheum* 1983; **12**: 390–403.

Schumacher HR, Miller JL, Ludivico C, *et al.* Erosive arthritis associated with apatite crystal deposition. *Arthritis Rheum* 1981; **24**: 31–7.

Osteoarthritis

5

Paul Dieppe & Ashley Blom

Osteoarthritis (OA) is by far the most common form of joint disease throughout the world. It is strongly associated with age, and extremely common in older people; some studies estimate that over 80% of people over 55 years of age have osteoarthritis of at least one joint. It mainly affects the hips, knees, spine, hands and feet. Hip and knee OA are the most important because of the high prevalence of pain and disability that they cause in older adults, and the massive healthcare resource input that results from this, particularly in terms of the provision of joint replacements.

The disease processes leading to OA result in a final common pathway of joint failure akin to heart or kidney failure. Clinically OA, or joint failure, is a very heterogeneous condition, but is the end result of biochemical and mechanical insult that exceeds the joint's ability to repair itself. Classification and subtyping are difficult. In the past people have tried to subset OA according to whether or not there is an obvious cause (primary versus secondary OA), whether symptoms are associated with joint damage or not (asymptomatic versus symptomatic OA), the extent and pattern of joint involvement (e.g. localized versus generalized OA), the type of bone reaction (hypertrophic versus atrophic OA), and whether the disease process is active, resulting in progression, or not (progressive versus inactive OA). Here we prefer to classify OA into two main categories – what has been called 'sporadic' or '*common-or-garden OA*' – by far the most frequent type, with its varying degrees of joint involvement (distribution), joint damage, and impact on peoples' lives, and '*atypical forms of OA*' that include joint failure that can result from a number of clear antecedent causes, and a variety of relatively unusual manifestations of joint failure.

We have therefore divided this chapter into three parts: first a description of joints and how diseases can affect them, then a discussion of 'common-or-garden' OA, and finally a brief description of some of the atypical variants of OA.

JOINTS

Joints occur where two or more discrete bones meet each other in the body. These joints can be fixed or can allow movement between the bones. There are three main types: fibrous joints, cartilaginous joints and synovial joints.

Fibrous joints join bone or cartilage by fibrous tissue and allow very little movement, such as in the sutures of the skull. Primary cartilaginous joints are between bone and hyaline cartilage. In secondary cartilaginous joints the two bone ends are covered in a thin layer of hyaline cartilage and these two ends are joined by interposed fibrous cartilage. An example of this is the symphysis pubis. The spine consists of a series of fibrocartilaginous joints as each vertebra is joined to its neighbour by an intervertebral disc consisting of fibrocartilage filled with gel. Even in cartilaginous joints that allow very little movement the joint can fail, particularly if subjected to heavy loads such as the acromioclavicular joint in weightlifters. Damage to the fibrocartilage can sometimes be accompanied by osteophyte formation. This can be thought of as a variant of OA. However, OA is usually defined as a condition of synovial joints.

Synovial joints (see Figure 5.1) have evolved in order to allow movement between bones. The adjoining bone ends are covered by extremely smooth hyaline cartilage (see Figure 5.2). This junction is enclosed within a joint capsule containing synovial fluid which bathes and lubricates the hyaline cartilage. The capsule is lined by a synovial membrane that contains cells called synoviocytes. These produce lubricant and hyaluronic acid which is responsible for viscosity of the synovial fluid. They also produce cytokines and growth factors and remove unwanted waste products, such as metabolites, from the synovial fluid. The capsule is reinforced by ligaments. These are arranged in such a way as to provide stability throughout the range of movement of the joint. Some synovial joints

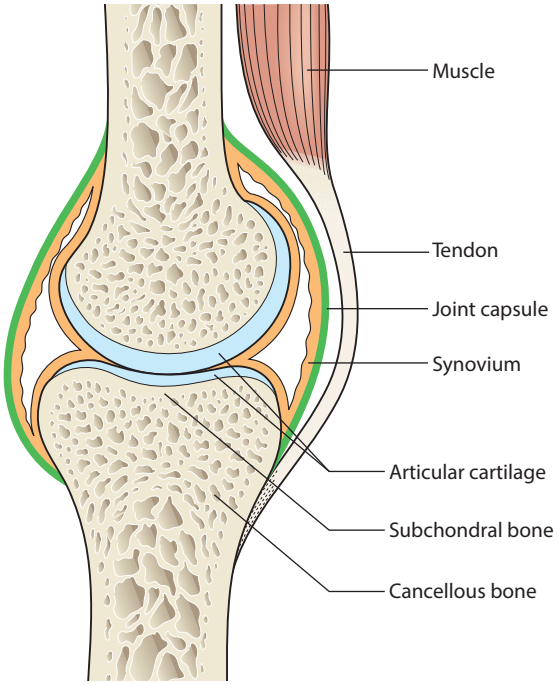


Figure 5.1 Diagram showing the components of a synovial joint

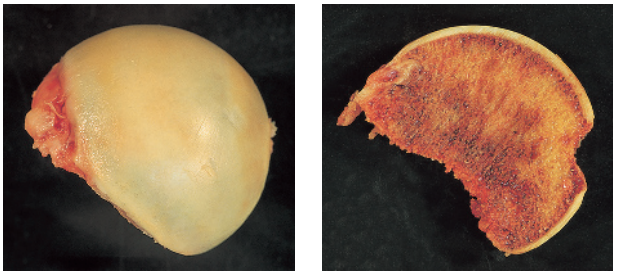


Figure 5.2 Normal articular cartilage Normal articular cartilage, smooth and glistening, is well preserved into old age. These specimens were taken from elderly patients with fractures of the femoral neck.

have intra-articular discs of fibrocartilage. The hip has a ridge of fibrocartilage called the labrum which deepens the articulation, whilst in the knee the same tissue has migrated further into the joint and has become the menisci. The menisci spread load through the knee and improve congruence, thereby improving stability. They thus protect the hyaline cartilage from both compressive and shear forces. Synovial joints move in a controlled way because of the muscle forces that act across them. Muscles are attached to bone by tendons. Ligaments attach to bones in a similar way. These areas of attachments by soft tissues to bone are called entheses and are subject to tensile forces.

All of the structures that compose synovial joints can fail and lead to the final common outcome of OA. Once one structure begins to fail, the surrounding structures are affected adversely and then also fail. Normally the insult, whether biological or mechanical, affects a number of structures simultaneously and the remaining structures secondarily. For example, a traumatic knee injury could lead to a tear of the meniscus, fracture of subchondral bone, disruption of the hyaline cartilage and stretching of entheses, all occurring simultaneously.

Subchondral bone supports the overlying hyaline cartilage. It is susceptible to fracture when subjected to great compressive force or to avascular necrosis when subjected to shear forces. Collapse of the subchondral bone leads to splitting of the overlying hyaline cartilage. Synovium can become inflamed due to chemical irritants such as crystals, or infection; additionally, the synovium is prone to inflammation resulting from systemic immune-related problems, as in rheumatoid arthritis. Synovitis of any cause results in the release of inflammatory mediators such as cytokines, and affects the production of hyaluronic acid, thereby altering the viscosity of the synovial fluid. The cytokines in the synovial fluid affect the catabolic and anabolic activities of the chondrocytes and osteocytes in the nearby cartilage and bone, as well as the capsule, leading to alterations in the normal integrity of these tissues, rendering them more susceptible to mechanical insults. The entheses are commonly stretched by injuries producing inflammation and oedema in the adjacent bone; they are also susceptible to inflammation in the seronegative spondarthropathies, such as ankylosing spondylitis. The menisci or the labrum are susceptible to tearing under excessive shear forces. Once their function is compromised, the hyaline cartilage can become exposed to abnormal load and can fail.

Failure of hyaline cartilage results in the subchondral bone being subjected to both increased load and direct pressure from synovial fluid. The final common pathway of all of these mechanisms is damage to hyaline cartilage, increased load on the underlying bone, cyst formation due to penetration of the subchondral bone by synovial fluid under pressure, and new bone formation on the joint margins (osteophytes), i.e. the development of osteoarthritis (OA). OA, therefore, can occur as a result of any form of joint disease.

‘COMMON-OR-GARDEN’ OA

Osteoarthritis (OA or joint failure) is relatively easy to define pathologically, being distinguished by focal areas of loss of articular cartilage within a synovial joint, accompanied by sclerosis of the underlying bone, and varying degrees of change in other joint tissues.

This pathology is reflected in characteristic changes in images of joints, particularly the plain radiograph, which is used to identify the pathology. This pathology is seen in most of the higher animal species, and it is the final common pathway of many forms of joint insult or injury; it is particularly common in some synovial joints in older humans. The pathology of OA is sometimes, but certainly not always, associated with the development of joint pain and other symptoms and signs, but it is difficult to describe or define clinical OA. This is the central dilemma of OA: *pathological changes of OA are very common in older people, but often asymptomatic; joint pain is very common in older people, and sometimes due to OA*. This means that clinicians need to think carefully about the cause of joint signs and symptoms in older people, before immediately labelling them as being due to OA on the basis of age or a radiographic appearance.

Prevalence and distribution

Most of the available data on the prevalence of OA comes from the developed Western world, and most of it is based on a radiographic definition of the condition, rather than a pathological or clinical one.

The joint sites most commonly involved are the knees, hips, hands, feet and spine. OA affects focal areas within joints: early in the disease only a localized area is affected, although later on it may spread to affect the whole joint. The sites most commonly affected in the knee are the anteromedial compartment of the tibiofemoral joint, and the lateral facet of the patellofemoral joint, in the hip it is the superolateral aspect that is most often damaged, and in the hands and feet it is the terminal (distal) interphalangeal joints, as well as the first MTP and thumb base that are most often involved.

Increasing age is a strong risk factor, and there are differences in prevalence and distribution in men and women. Figure 5.3 illustrates the overall prevalence in Western men and women of hip, knee and hand OA.

Some racial/ethnic differences exist in the prevalence and distribution of OA. For example, superolateral hip OA, which is very common in Caucasians, is relatively uncommon in people of Chinese origin. As explained later, this may be due to subtle differences in skeletal shape.

Aetiology and risk factors

OA has no single cause; rather, it is due to a variable combination of several risk factors affecting different individuals and different joint sites, which explains its heterogeneity. OA arises as a result of a mixture of both systemic predisposition and local biomechanical risk factors, as shown in Figure 5.4, and Box 5.1 shows the major risk factors currently known.

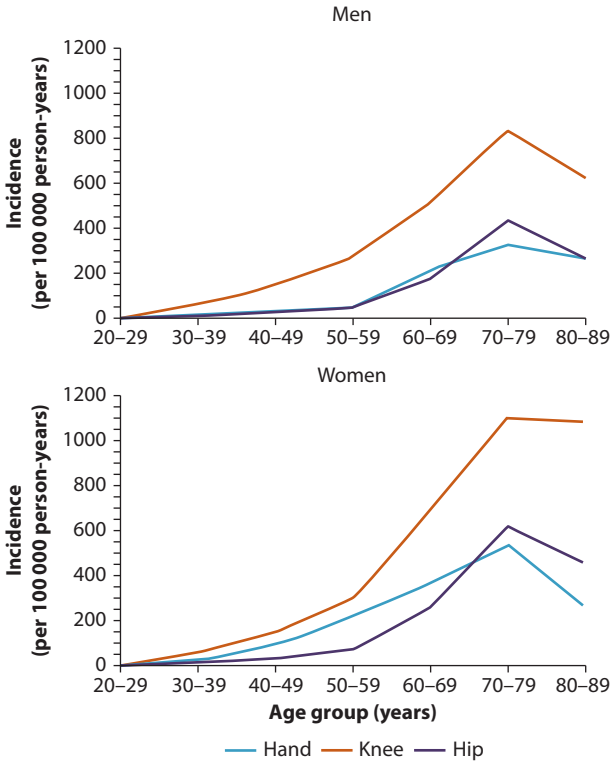


Figure 5.3 Incidence of symptomatic osteoarthritis (OA) of the hand, knee and hip as a function of age (Data from Fallon Community Health Plan (Oliveria et al., 1995) reproduced from Nelson & Jordan, 2015.)

GENETIC PREDISPOSITION

From twin studies and other data, it has been estimated that about 40% of the predisposition to OA may be genetic. However, there is no 'OA gene'; rather, several different sites within the genome each confer a small increased risk. Many of the sites associated with this increased risk relate to genes important for skeletal development, adding to other evidence that suggests that bone size and shape are important determinants of the likelihood of getting OA.

AGE

OA is strongly associated with increasing age. But this is not because age-related changes in joints are similar to those of OA – there are major differences in the 'pathology' of ageing joints from those of OA, and it has been suggested that we would need to live for over 200 years before the age-related changes in the joints alone (such as thinning of the cartilage) would cause OA. The association with age may have more to do with joint stability and muscles than joints. As we age, our cartilage gets thinner and our muscles get weaker, and the stability of major joints such as the knee may be affected in subtle but important ways by these changes. Some studies have suggested that muscle weakness precedes the development of knee OA.

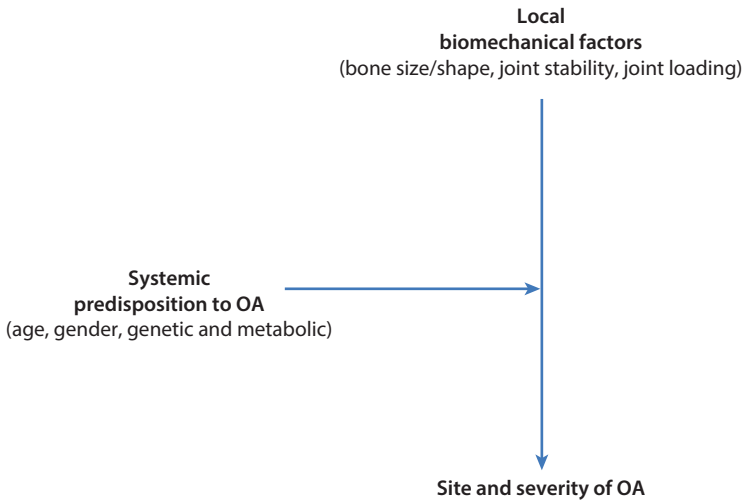


Figure 5.4 Systemic predisposition and local biomechanical risk factors in OA

BOX 5.1 MAJOR RISK FACTORS FOR OA

Systemic predisposition

- Genetics
- Age
- Gender
- Diet and obesity

Local biomechanical factors

- Abnormal joint shape and size
- Previous injury
- Neuromuscular problems
- Obesity
- Loading/occupational factors

Bone mineral density

(It is unclear whether this is a local or a systemic factor)

GENDER

As shown in Figure 5.3, there are differences in prevalence of OA between men and women. The reasons for this are not clear. Changes related to the female menopause appear to be particularly important, as knee OA prevalence in women rises sharply after the menopause, and inflammatory OA of the hands often starts during the menopause.

DIET AND OBESITY

Obesity is a strong risk factor, particularly for knee OA. It is also a risk factor for increased incidence of hand OA, suggesting that it may have some systemic influence, perhaps through changes in obesity-related biochemical factors such as leptin levels. In addition, there have been a number of studies to suggest that some vitamin deficiencies may be important in the development of OA, including vitamins C, D and K.

ABNORMAL JOINT SHAPE AND SIZE

Joint shape is an important risk factor, particularly for hip OA. Hip dysplasia predisposes you to hip OA in later life, and more subtle abnormalities of the size or shape of the head of the femur or acetabulum (such as the shape changes that cause femoroacetabular impingement – FAI), may be responsible for much of the common-or-garden hip OA seen. The differences in shape of hips in Chinese from that in Caucasians may explain the low prevalence of hip OA in Chinese people. It is possible that joint size and shape are also important in knee OA.

PREVIOUS INJURY

Injuries that affect the shape or stability of a joint predispose to OA. This is most apparent in joints which have a low prevalence of 'common-or-garden' OA such as the wrist or ankle – OA at these sites is usually due to a previous significant injury. At the knee joint, meniscal and ligament injuries, particularly ACL rupture, are important predisposing factors for OA.

NEUROMUSCULAR PROBLEMS

Severe neurological problems of specific types can lead to the important variant of OA called 'Charcot's joints'. Lesser forms of neurological change, including weak muscles, and loss of proprioception, may be important in 'common-or-garden' OA. In addition, joint laxity seems to predispose to OA. Conversely, spasticity results in very tight joints accompanied by abnormal joint loading leading to joint damage and secondary osteoarthritis. OA of the hip is particularly common in persons suffering from spastic cerebral palsy.

JOINT LOADING, OCCUPATION AND OBESITY

The extent to which normal or excessive joint use, including exercise, are risk factors for OA, or alternatively protective to the condition, is a contentious issue, and we do not yet understand exactly what aspects of joint loading matter most to joint health. However, certain specific occupations involving repetitive 'overuse' of joints can predispose to OA, resulting in special forms of the condition such as 'picca-thumpers thumb' (OA at the base of the thumb in people who spent their working days shifting printing blocks around with their thumbs). The fact that obesity is particularly important for knee OA indicates that a part of the risk is likely to be due to loading factors, as well as any systemic influence.

BONE MINERAL DENSITY

Long ago it was noted that people who came to hip replacement because of fractures caused by osteoporosis were unlikely to have hip OA. Subsequent studies have confirmed that, at both the knee and the hip, high bone mineral density is a risk factor for OA, and low bone mineral density is protective. It is not understood how these relationships operate, and it is unclear to what extent this is a systemic factor, or whether it is about local loading of cartilage and subchondral bone.

Pathology and pathogenesis

The OA process is mechanically driven but chemically mediated.

PATHOLOGY

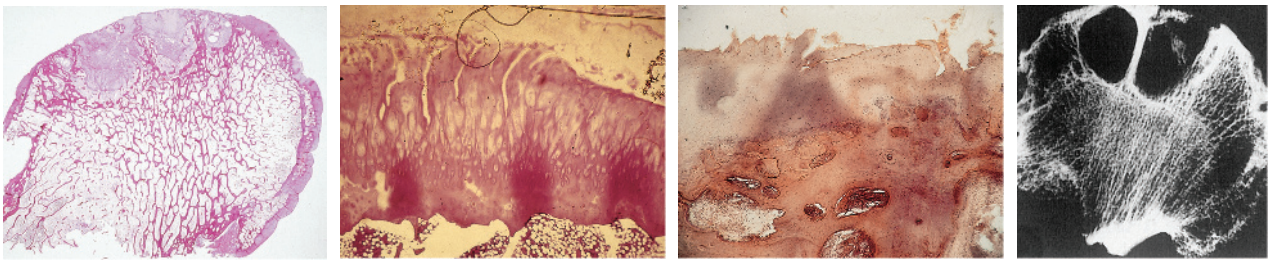
The key *pathological features* of OA are shown in Table 5.1, which also documents their radiographic correlates.

Historically, pathologists and academics interested in OA have concentrated on the changes seen in articular hyaline cartilage, more than the changes in other joint tissues. The cartilage changes include: early softening and swelling or articular cartilage, with an increase in its water content; intermediate fragmentation and fissuring of the cartilage surface; and late erosion down to the underlying bone (see Figure 5.5). This pathology is very well described, and several classification and scoring systems are available. The *Outerbridge classification* is most commonly used by orthopaedic surgeons. It originally described changes to the articular surface of the patella, but it is now used in all synovial joints, particularly as arthroscopy is now common and allows direct visualization of many joints.

Grade 1 Softening and swelling of the cartilage
Grade 2 Fragmentation and fissuring of the cartilage in an area less than ½ inch in diameter
Grade 3 Fragmentation and fissuring of the cartilage in an area more than ½ inch in diameter
Grade 4 Exposure of underlying bone.

Table 5.1 Key pathological features of OA

Pathology	Radiographic correlates
Focal areas of loss of articular cartilage	Joint space narrowing (if loss is extensive)
Bone growth at the joint margins	Osteophytes
Sclerosis of underlying bone	Sclerosis of subchondral bone
Cyst formation in underlying bone	Bone cysts
Loss of bone	Bone attrition
Varying degrees of synovial inflammation	Effusions may be apparent
Fibrosis and thickening of the joint capsule	Not visible on radiographs



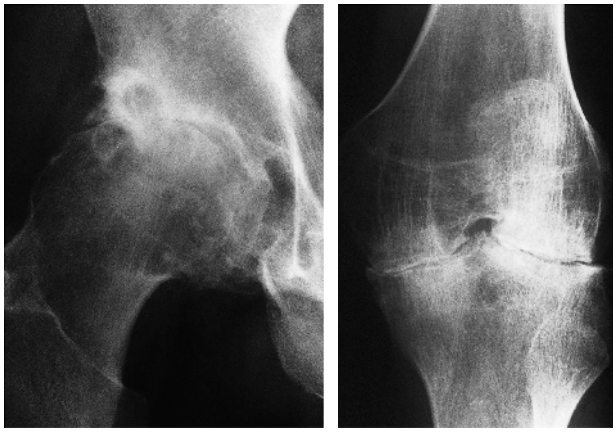
(a) (b) (c) (d)

Figure 5.5 Osteoarthritis – histology (a) Destructive changes (loss of articular cartilage and cyst formation) are most marked where stress is greatest; reparative changes are represented by sclerosis around the cysts and new bone formation (osteophytes) in less stressed areas. (b) In this high-power view, the articular cartilage shows loss of metachromasia and deep clefts in the surface (fibrillation). Attempts at repair results in (c) subarticular sclerosis and buds of fibrocartilage mushrooming where the articular surface is destroyed (d).

The cartilage changes are accompanied by extensive changes in the tidemark between bone and cartilage, with vascular invasion and extension of the calcified zone, as well as thickening of the subchondral bone. At the margins of the joint, periosteal cells proliferate and change their phenotype to form bone (osteophytes). In addition, there is usually some synovial inflammation, which may result in joint effusions, as well as thickening and fibrosis of the joint capsule, which may be extensive. In advanced cases the damage to the subchondral bone can lead to the formation of cysts, and loss of bone volume. All of these changes vary in extent in different individuals, and there are also some differences according to joint site, with, for example, unusual pathological features such as hyaluronan cyst formation being a feature of some hand OA.

The radiograph is a blunt instrument for revealing these pathological changes, but it is the only routinely available tool to detect them, and as long as the changes in the joint are severe enough, they result in characteristic joint space narrowing, osteophyte formation and subchondral bone sclerosis, which are pathognomonic of OA (see Figure 5.6). As with the pathological changes, there are a number of different scoring systems available to assess the severity of radiographic changes. The one used most commonly is the *Kellgren and Lawrence scoring system*, which divides OA X-ray changes into five categories:

0	*Normal*	No features of OA
1	*Doubtful*	Minimal osteophyte, doubtful significance
2	*Minor*	Definite osteophyte, no loss of joint space
3	*Moderate*	Some diminution of joint space
4	*Severe*	Advanced joint space loss and sclerosis of bone



(a) (b)

Figure 5.6 Osteoarthritis – X-rays The cardinal features of osteoarthritis are remarkably constant whether in (a) the hip or (b) the knee.

PATHOGENESIS

As already explained, the OA process is initiated by a mixture of systemic predisposing factors interacting with local mechanical influences that affect the site and severity of the OA changes, but the changes themselves are chemically mediated.

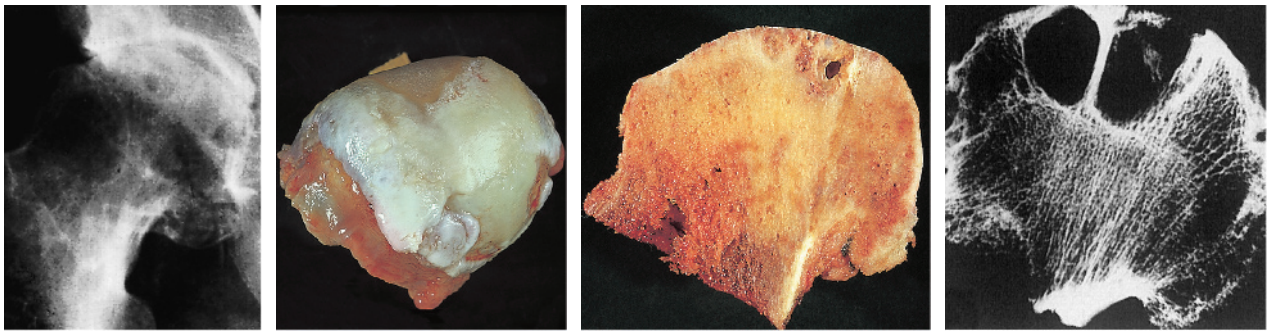
There are many different hypotheses about how the process is mediated. As with OA pathology, research on OA pathogenesis has been dominated by work on articular cartilage rather than other tissues, although it is now seen as a disorder of the whole synovial joint organ.

A lot of current work revolves around the generation of local cytokines and proteolytic enzymes within the joint, and cell signalling pathways that link chondrocyte activity to changes in the subchondral bone, synovium and capsule. The main emphasis of much OA research remains on trying to understand how sparsely distributed cells in cartilage (chondrocytes) maintain the integrity of the articular cartilage in normal joints, and the anabolic and catabolic processes that result in OA. The early changes in cartilage appear to result from collagenase enzymes disrupting the integrity of the type II collagen matrix which encloses the hydrophilic proteoglycans, leading to swelling and softening; subsequently, more proteolysis and damage to proteoglycans, as well as collagen, results in the fissuring and loss of volume. It is less clear how the changes at the tidemark and in subchondral bone are mediated, where increased calcification and angiogenesis are switched on, and trabecular thickening is seen, perhaps in part as a result of subchondral micro-fractures of trabeculae (see Figure 5.7).

One of the problems encountered by those studying OA is to detect it in its earliest stages, before the pathology is severe enough to become apparent on an X-ray or be clinically relevant. Animal models of the condition help here, and there has been interest in what comes first – changes in the cartilage, bone, synovium or other parts of the joint? The evidence, not surprisingly, is that they occur together and are linked and, contrary to earlier ideas, it is apparent that changes in the bone and soft tissues can occur in the earliest stages of the process in humans. However, the OA process and the homeostasis of the normal synovial joint are yet to be well understood.

NATURAL HISTORY AND OUTCOMES: INCIDENT-VS-PROGRESSIVE OA

In the past OA has been talked about as a degenerative condition, a progressive one, and something that cannot get better. None of these ideas is correct. It is not a degenerative process; rather, the changes that are going on in cartilage and other



(a) (b) (c) (d)

Figure 5.7 Osteoarthritis – pathology (a) The X-ray shows loss of articular cartilage at the superior pole and cysts in the underlying gone; the specimen (b) shows that the top of the femoral head was completely denuded of cartilage and there are large osteophytes around the periphery. In the coronal section (c) the subarticular cysts are clearly revealed. (d) A fine-detail X-ray shows the extent of the subarticular bone destruction.

tissues are very active ones. The term 'degenerative joint disease' should be avoided as it conveys old age, inevitability and negativity to both patients and healthcare professionals alike. Similarly, OA is not necessarily progressive. Natural history and physiological imaging studies (such as bone scintigraphy) suggest that it goes through periods of activity and quiescence. Thus a joint may be mechanically compromised in a susceptible person, and respond by activation of the OA process, leading to some changes to cartilage and bone, and the characteristic radiographic changes. This may result in the process then becoming quiescent for long periods of time, although physical examination and radiographs of the joint will still reveal the changes of OA. There is some evidence that the risk factors for progression are a little different from those for initiation of the process – for example, malalignment of a joint is more important for progression of OA than for initiation.

Clinically, it is also clear that patients can have periods of more severe symptoms, followed by quiescent periods, and in a significant proportion of cases (probably about 30%) time results in clinical improvement rather than deterioration.

Repair of the pathological changes is less common but can occur: spontaneous improvement in hip OA is well described, as is joint repair in response to major mechanical treatment interventions, such as osteotomy at the knee, or mechanical unloading by use of Ilizarov frames for ankle OA. However, the repaired OA joint is not normal, as the hyaline articular cartilage is replaced by fibrocartilage.

Symptoms and signs

Pain is the main clinical problem. But, as already noted and shown in Figure 5.8, there is a poor correlation between the radiographic evidence of OA in joints, and the prevalence of the clinical symptoms such as pain.

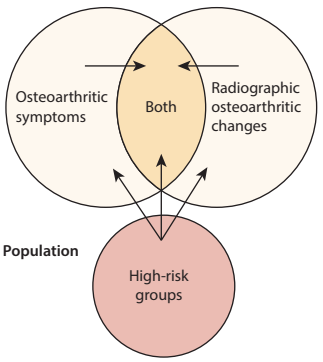


Figure 5.8 The relationship between osteoarthritic symptoms and radiological features of osteoarthritis (From Dieppe & Lohmander, 2005.)

The most common symptoms and clinical signs reported by/seen in people with clinical OA are documented in Box 5.2.

PAIN

Most people with clinical OA report discomfort or pain in or around the joints affected, but their pain experiences vary hugely, both over time, between individuals, and, as outlined further below, according to the joints affected. It is very common for patients to have trouble describing their pain. They often refer to the sensation as a deep-seated discomfort similar to a toothache emanating from within the joint. Reported experiences vary from: a dull ache after exercise; through to the more common moderate, activity-related pain; to excruciating, continuous pain and pain at night. Severe pain that wakes the patient nightly is a particularly debilitating symptom of severe arthritis and leads to sleep deprivation. Many people report that no two days are the same, the pain experience being variable and seemingly inexplicable.

BOX 5.2 MAJOR SYMPTOMS AND SIGNS OF CLINICAL OA

Symptoms

- Pain (the nature and severity of which is very variable)
- Joint stiffness (particularly short-lasting stiffness after a period of inactivity)
- Fatigue
- Sleep disturbance
- Depression
- Reduced functional ability and activities

Signs

- Tenderness of the joint
- Bony swelling
- Reduced range of movement with pain at the end of the range
- Crepitus on movement of the joint
- Weakness and wasting of muscles acting on the joint
- Signs of inflammation (usually fairly mild)
- Deformity and instability in severe cases

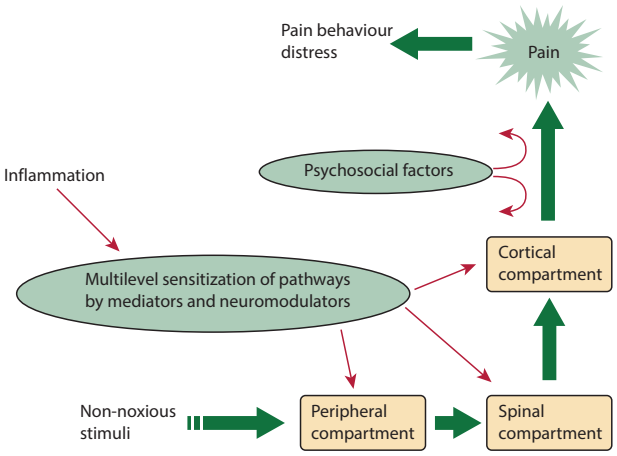


Figure 5.9 The pathogenesis of pain in osteoarthritis (From Dieppe & Lohmander, 2005.)

We do not know why OA sometimes causes pain. There are no nocioceptive receptors in the cartilage, which is the major tissue affected pathologically, but nocioceptive pathways do occur in subchondral bone, in periosteum and in the synovium and capsule of the joint. It would appear likely that some of the variation in the pain experience is related to differences in where the nocioceptive drive is coming from in different people/joints; there is good evidence for bone being a major source of pain in advanced knee OA, but in other sites/severity, it may be different (see Figure 5.9).

In addition to there being a nocioceptive drive to cause pain, pain amplification systems, both locally in the joint and within the central nervous system, can contribute to the pain experience in chronic OA. Once pain has become chronic, amplification pathways can be activated locally and at both spinal and cortical levels, leading to chronic and more widespread pain, which is difficult to treat, particularly if accompanied by mood changes. Anxiety and depression both affect the perception of pain and its response to interventions.

OTHER SYMPTOMS

Stiffness, or gelling of the joint after inactivity, is a classical feature of OA, resulting in people having difficulty getting moving after resting. This is particularly common in the early morning after first awakening. The cause is not known. Less well-appreciated symptoms of OA include fatigue, sleep disturbance caused by pain and anxiety/depression. Each of these is very common, and can have a big impact on individuals, and, as discussed below, may need management separately from any attempt to deal with joint damage or pain.

SIGNS

On examination the joints affected may appear swollen, there may be evidence of wasting and weakness of surrounding muscles (as in quadriceps muscle wasting at the knee, or the Trendelenburg sign at the hip which signifies weakness of hip abductors) and, in advanced OA, joint deformity. Palpation may reveal bony swelling at the margin of the joint, signs of mild inflammation, such as heat over the joint line, and an effusion. On movement there is usually a reduced range, with pain at the end of the range, and crepitus may be felt as the joint is moved; in advanced disease instability may be detected.

As emphasized in this chapter, the challenge for the clinician is to ascertain whether the symptoms and signs are the result of OA or of some other articular or periarticular problem.

SYMPTOMS AND SIGNS AT DIFFERENT JOINT SITES

Hips Pain is usually felt in the groin, laterally over the hip and radiates down the anterolateral aspect of the thigh to the knee. Occasionally the pain can radiate beyond the knee. Referred pain felt only in the knee is not uncommon, and clinicians should always consider hip OA as a cause of isolated knee pain. Pain is worse on exercise and walking distance is reduced. Pain at rest and night pain can be particularly troublesome. Stiffness is usually experienced first thing in the morning and after having sat still for a while, but it quickly resolves on movement to be replaced by pain. Complex movements, such as getting in and out of a motorcar or putting on socks, which involve deep

flexion combined with rotation, are often difficult or impossible to perform. Patients struggle with stairs and in the absence of a banister may only manage stairs on all fours.

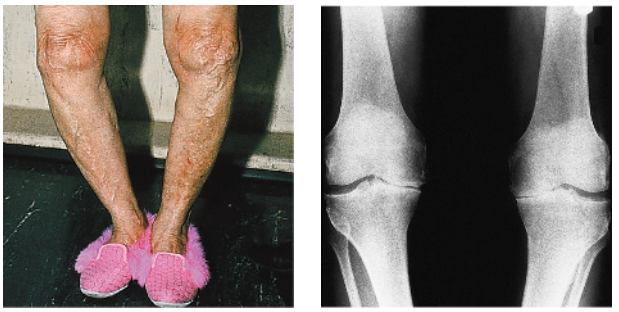
Examination reveals an antalgic gait, characterized by an uneven cadence, in which less time is spent in the stance phase of the painful limb. There is a globally reduced range of movement with internal rotation often restricted early in the disease progression. Joint movement is limited by pain at the extremes of movement.

Knees Knee osteoarthritis occurs most commonly in the medial tibiofemoral joint but can occur in all three compartments and is often tricompartmental. Isolated patellofemoral OA is probably due to altered biomechanics of the extensor mechanism. Pain is felt globally over the knee and the proximal tibia. In isolated patellofemoral OA the pain is felt anteriorly over the knee and is often worst when ascending or descending stairs as the patella is compressed against the femur. As in the hip, the pain is a deep-seated aching sensation related to exercise. Rest pain and night pain develop in the later stages of the disease. Patients sometimes report audible crepitus (crackling or grating sounds) coming from the knee as well as symptoms of instability (a feeling that the knee is going to give way). They may notice gradual deformity of the knee, in particular varus deformity (see Figure 5.10), but less commonly valgus deformity. Fixed flexion deformity means that the knees cannot lock in full extension and thus patients cannot stand comfortably for prolonged periods due to muscle fatigue. Loss of flexion beyond 90 degrees makes standing from a sitting position difficult as patients cannot move their centre of gravity anterior to their mid-coronal plane. Swelling and stiffness are common features.

Examination reveals an antalgic gait, wasting of quadriceps muscles, joint effusion, joint deformity, and crepitus palpable and sometimes audible on movement. The joint deformity may be passively correctable. Deformity is towards the compartment most severely affected, usually varus deformity with predominantly medial compartment OA. There is sometimes tenderness along the joint line and palpable osteophytes that can be tender.

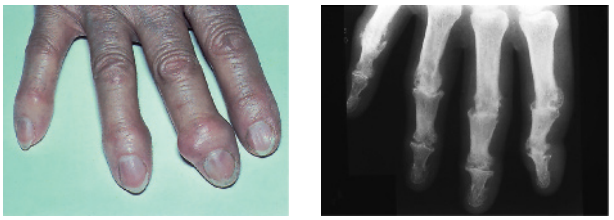
Hands The joint sites commonly affected in the hand are the distal interphalangeal joints (DIPs) and the thumb base (both the radiocarpal and scaphotrapeziod joints); less commonly, proximal interphalangeal joints and metacarpophalangeal joints are also involved (see Figure 5.11).

OA of the hand is strongly associated with OA at other joint sites, especially the knee, and with genetic predisposition to OA, suggesting that it is a feature of generalized 'common-or-garden' OA, but it has several unique pathological and clinical features, not often seen at other sites. It is far more common in women



(a) (b)

Figure 5.10 Polyarticular (generalized) osteoarthritis of the knees



(a) (b)

Figure 5.11 Polyarticular (generalized) osteoarthritis of the hands An almost invariable feature of the polyarticular OA is involvement of the terminal finger joints – Heberden's nodes.

than in men and often starts relatively abruptly around the time of the menopause (sometimes called 'menopausal OA') with painful inflammation in the distal interphalangeal joints: over time (years) the inflammation settles and the joint is left with the typical pathological features of OA. Erosions can occur ('erosive OA'), and cysts containing hyaluronan that protrude at the margins of the joints are not uncommon. Distal interphalangeal joint OA is not generally a major problem in terms of function, but thumb base OA can be, as it leads to instability and difficulty with pinch grip.

Other joints Almost any joint can be affected by OA, particularly if it is damaged by severe trauma. However, there are peculiarities to the phenotype of the condition at different sites. For example, elbow OA is almost always asymptomatic (just causing loss of full extension of the elbow), while shoulder OA is more likely to result in severe bone destruction (a condition sometimes called 'Milwaukee shoulder') than is OA at other joint sites.

Differential diagnosis, investigation and assessment

The major problem is the common assumption that any joint symptoms in an older person are due to OA, compounded by the fact that they are very likely to

have radiographic changes of OA, even if that is not the cause of their symptoms.

Joint pain may:

- be referred from above (e.g. hip OA causing knee pain)
- be due to a periarticular problems
- come from the joint itself
- be due to central nervous system pain sensitization (as in 'fibromyalgia')
- result from a complex mixture of the above.

In people with OA we think that there is often a complex mixture of several mechanisms underlying their symptoms, with nocioceptive drives coming from both the joint and the periarticular tissues, and a degree of pain sensitization complicating the picture.

Clinical examination may help to differentiate between these different types of pain problem: areas of localized tenderness *may* reflect articular or periarticular pathology (but beware, referred tenderness can occur), and the presence of widespread pain or allodynia (severe skin sensitivity) may indicate pain sensitization. If movement of the joint reproduces the patient's common symptoms, this suggests that joint pathology is the problem, but it is not easy to find out why a joint is painful, or whether OA pathology is the cause.

Routine investigations do not help. All blood tests are generally normal in OA (although there might be a small rise in CRP levels), but tests for evidence of systemic inflammation (such as the ESR or CRP) may be useful in differentiating OA from inflammatory forms of joint disease. As explained, the plain radiograph is simply an historical record of preceding joint changes, and it is not possible to use other imaging techniques such as MRI or scintigraphy to detect pathophysiology in routine care.

ASSESSMENT

Pain, the main symptom of OA, is a subjective experience which cannot easily be measured or assessed. Similarly, it is not easy to ascertain the severity of the functional problems that any individual patient may be experiencing. So the assessment of severity and of the likelihood of a good response to interventions such as surgery is difficult in routine clinical practice. One is largely reliant on what the patient says, supplemented by the observation of gait, any difficulties the patient has undressing, dressing or getting onto or off the examination couch and clinical examination.

For research purposes a number of instruments are available to assess the severity and impact of OA. Patient self-assessment questionnaires such as the 'WOMAC', Oxford hip, knee and shoulder scores, Hip Dysfunction and Osteoarthritis Outcome Score 'HOOS' (for hips), Knee Dysfunction and Osteoarthritis Outcome Score 'KOOS' (for knees) and the Australian–Canadian Hand Osteoarthritis Index 'AUSCAN' (for hands) are often used. In addition, semi-objective tests of function, such as walking speed, can be measured to assess disability, and special imaging techniques such as MRI may be used to assess the degree of joint damage. However, plain radiographs are so characteristic as to make other imaging studies unnecessary. The four cardinal signs are *osteophyte formation*, *joint space narrowing*, *sclerosis* of the underlying bone, and *subchondral bone cysts.*

Again for research purposes it is important to try to understand whether the disease process is active or not, and what tissues are undergoing change. Academics have invested heavily in the development of biochemical markers of the OA process, such as blood or urinary levels of degradation products of cartilage, as well as functional imaging techniques (fMRI) in order to try to understand more about the disease, its responsiveness to different therapies, and its activity, but no clear consensus on what measures are most useful has yet emerged.

Management

Many different interventions are available for the treatment of people with OA, and there are a plethora of guidelines available.

Interventions are generally divided into symptomatic therapies and disease-modifying therapies. As yet, there are no drugs with proven ability to modify the disease process, although claims have been made for many different agents. Disease modification can occur in response to mechanical interventions, such as joint distraction and osteotomy. All other interventions are symptomatic.

The main symptoms of OA (pain, stiffness, fatigue, and anxiety/depression) are very susceptible to so-called 'placebo' and 'nocebo' effects. Placebo is generally thought of as a sham or dummy intervention, and we know that sham surgery can work well in OA, but what placebo research teaches us is that symptoms of conditions such as OA are highly responsive to the whole context in which any therapy is administered. If patients and clinicians feel safe and trusting of each other, and if the clinician is able to validate the patient's experiences (i.e. the patient feels fully understood), then outcomes will be good, whatever the intervention. And the opposite is also true.

SYMPTOMATIC THERAPY

Figure 5.12 illustrates the basic principles of the symptomatic management of OA.

As noted, there are several evidence-based, published guidelines on the management of OA. This discussion is based on the UK National Institute for Health and Care Excellence (NICE) 2014 guideline

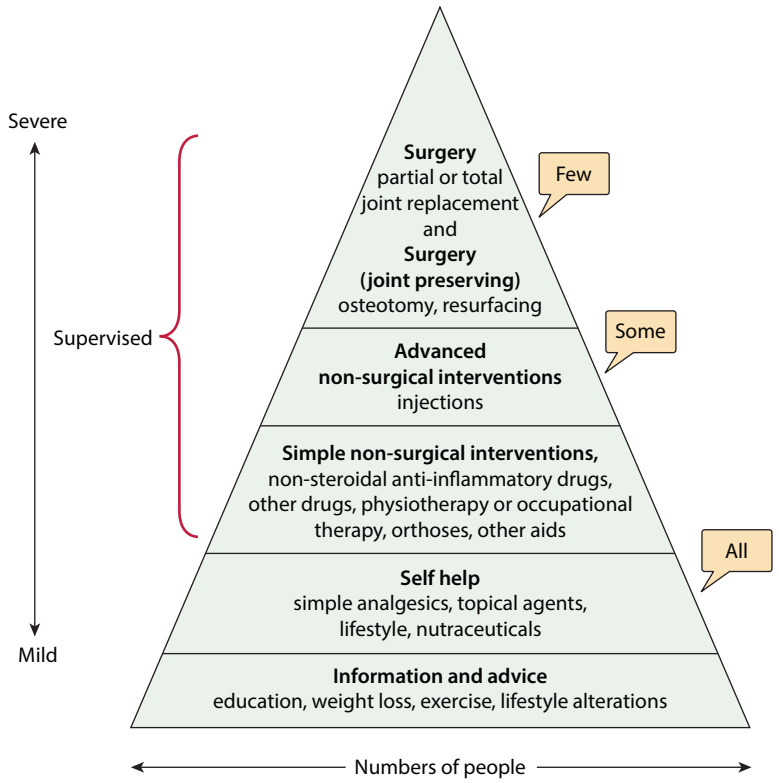


Figure 5.12 The pyramid of treatment for symptomatic osteoarthritis (From Dieppe & Lohmander, 2005.)

CG177: *Osteoarthritis: Care and Management*, which recommends that clinicians follow the steps described below.

Step 1 *Take a holistic approach and encourage self-management.* This means assessing the impact of the OA on the individual's quality of life, function, mood, relationships and activities, thinking about their context and any comorbidities (including depression) and thinking about the potential risks and benefits of any intervention. Self-management strategies include considering alterations of diet (particularly to lose weight), alteration in activities, changing footwear, taking a more positive approach to the condition and exercising more.

Step 2 *Introduce the 'core treatments' appropriate for most people with OA.* These include the provision of information about the condition and its management, helping people to increase their exercise level and to do specific exercises to strengthen muscles around affected joints, footwear advice, and help with weight loss for those who are obese.

Step 3 *Introduce specific non-surgical interventions.* These may be pharmacological or non-pharmacological.

The *non-pharmacological* interventions of proven value include supervised courses of physical therapy, the use of aids and devices to reduce instability of to help with functional problems, walking aids such as sticks or crutches, some electrotherapy techniques, such as TENS machines for pain control, and some manipulations, particularly in the case of hip OA with a reduced range of motion of the hip.

Pharmacological options include the use of regular or on-demand paracetamol, and topical NSAIDs, which are considered to be the first choices for pain. More recent evidence has suggested that paracetamol is no better than placebo, but both have been demonstrated to reduce pain. If they are insufficient, NSAIDs, COX-2 inhibitors and opioid analgesics can be considered. Intra-articular local anaesthetic and corticosteroid injections can be considered as an adjunct to other treatments, but hyaluronan injections are not recommended because of lack of evidence.

The very widely used 'nutriceuticals' such as glucosamine and chondroitin are not recommended because of lack of the lack of robust evidence to support their use.

Step 4 *Consider surgical options.* There are many different surgical options available, as discussed in more detail in other chapters in this book. Surgery is broadly divided into joint realignment, joint fusion, joint excision and joint replacement (arthroplasty), which may be total or partial (such as unicompartmental knee replacement). Joint debridement such as arthroscopic knee debridement has largely been discredited by randomized controlled trials of sham surgery versus

debridement, which showed no treatment benefit. Surgery is usually confined to end-stage disease once pain has become refractory to other treatment options. Hip and knee replacements are particularly successful treatments for advanced OA, often resulting in complete resolution of pain and a dramatic improvement in function and quality of life. However, a small but important minority of patients do not benefit from joint replacement (between 5 and 15%, lack of response being more common for knee replacement than hip replacement) and have persistent severe pain in the long-term post surgery despite no evidence of technical issues with the surgery performed.

PRINCIPLES OF MANAGEMENT

The natural history, presentation, impact and prognosis are different for each of the major joint sites affected, and management needs to be based on prognosis and impact in particular, so here we provide a brief outline of the principles of management of hip, knee and hand OA.

Hip OA This responds relatively well to physical therapy and walking aids. It is important to assess leg length and think about corrective footwear, and shock-absorbing shoes can help. Among the most useful interventions are the use of a stick in the contralateral hand to reduce loading while walking, and physiotherapy aimed at increasing the range of motion and improving muscle strength and pelvic stability.

OA of the hip often progresses relatively rapidly, so patients may have a story of a relatively long period of mild problems, such as exercise-related aching in the groin, followed by the development of severe pain over a period of a few weeks or months. If pain becomes very severe, interfering with activities and sleep, then hip replacement is likely to be the best option.

Spontaneous recovery from severe hip OA occasionally occurs, particularly in those who stay active, but this cannot be predicted or relied upon. The mainstays of treatment are weight loss, walking aids, physiotherapy and simple analgesics combined with NSAIDs. When these treatment modalities no longer control symptoms, total hip replacement is usually very effective.

Knee OA This often responds well to simple non-surgical interventions, and it often remains relatively stable and mild for many years, during which people can adjust to it, and find ways of making sure it interferes with life minimally. Knee OA is strongly related to obesity, and relatively recent research has shown that even modest amounts of weight loss can result in marked reduction in symptoms, so this should be prioritized. Keeping the quadriceps muscles strong is important, as they are key to knee stability.

Topical NSAIDs are useful, as are shoes or appliances that reduce impact loading (shock absorbing) and/or adjustments to unload the most affected compartments (usually the medial tibiofemoral compartment). Patella strapping is useful for patellofemoral OA. Corticosteroid injections can result in good pain relief for relatively short periods of time (a few weeks to months) and can, therefore, be of great value as an adjunct to a course of physical therapy, or to help a patient manage a planned, important life event such as a wedding. There are many surgical options if the condition becomes severe. These include osteotomies of various types, and unicompartmental or total joint replacements. Arthroscopic joint lavage is not recommended because of insufficient evidence for efficacy over and above its very big placebo effect.

Hand OA Osteoarthritis in the hand often has a relatively good long-term prognosis, unless there is severe thumb base disease. Management therefore usually involves strategies that reduce pain without putting the patient at risk. Topical NSAIDs and capsaicin are of proven value for interphalangeal joint disease. There are good surgical options for advanced thumb base OA.

It is important to re-emphasize the fact that most people with OA do not progress to a severe enough state to warrant surgical intervention, and that the prognosis is generally relatively good. Therefore, interventions should be kept as simple and as safe as possible. Most people with OA are older adults, many of whom have comorbidities that can make pharmacological and surgical interventions more hazardous than usual. In addition, as noted above, symptomatic OA responds well to context (placebo) effects, meaning that it is crucially important to make sure that whatever is being offered in terms of advice or intervention is offered in a positive manner, and in a safe environment for the patient, in which he or she feels 'heard'. As the song says, *'It ain't what you do it's the way that you do it.'*

OA VARIANTS

In the previous part of the chapter we described the commonest form of 'sporadic' or 'common-or-garden' OA. But the pathology and pathogenesis of OA can be seen as the final common pathway of many different types of joint insult – synovial joints can only respond to injury or abnormality in a restricted manner, as they have limited repair capacity.

Here we briefly describe some of the conditions that can predispose to OA, as well as some interesting anatomical variants that differ in phenotype from 'common-or-garden' OA.

Anatomical and pathological variants

As mentioned above, OA involves varying degrees of inflammatory reaction in the synovium, and bone change, in addition to cartilage damage. In some instances the inflammatory or bony components are so great as to dominate the clinical and pathological features of the condition. Examples include the following:

- *Atrophic destructive OA including rapidly progressive hip OA and 'Milwaukee shoulder'*. In these conditions there is a rapid period in which the joint damage progresses with extensive bone loss, such that the whole head of the femur of humerus may 'disappear' (see Figure 5.13). There is very little osteophyte formation; the synovial fluid is usually bloody, and contains large numbers of apatite crystals, which may contribute to the damage. The cause is unknown.
- *Hypertrophic OA* is at the other end of the spectrum of joint pathology. It is particularly common at the hip and knee, and characterized by massive osteophyte formation, with relatively little damage to subchondral bone or cartilage (see Figure 5.14). It may be associated with calcium pyrophosphate dihydrate (CPPD) crystal deposition (chondrocalcinosis), and/or with diffuse idiopathic skeletal hyperostosis (DISH), each of which seem to be associated with extensive osteophyte formation (as well as enthesis calcification in DISH).
- *Erosive inflammatory OA of the interphalangeal joints*. As mentioned above, OA of the terminal interphalangeal joints of the hand can be associated with a lot of inflammation and the development of joint erosions of the sort normally associated with rheumatoid arthritis.

OA resulting from biochemical abnormalities within the cartilage

If the articular hyaline cartilage is compromised by some genetic or metabolic abnormality, it becomes more susceptible to damage, initiating the whole OA process. Examples include the following:

- *Genetic abnormalities in type II collagen* (for example, Kniest Syndrome) *or other key components*. Rare forms of familial OA exist in which the

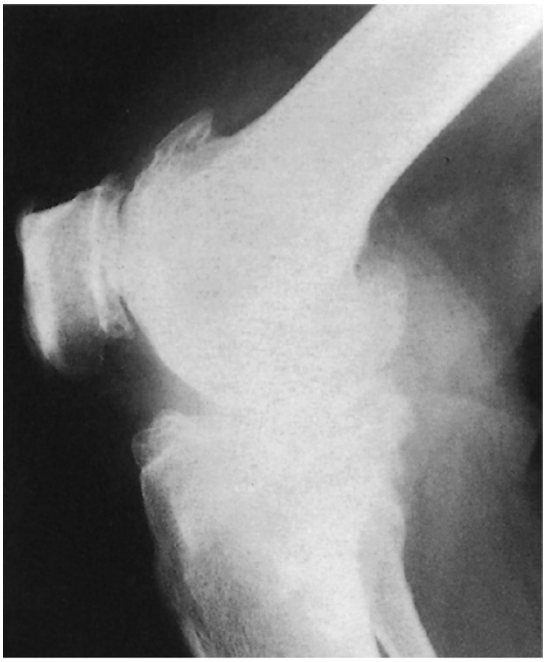
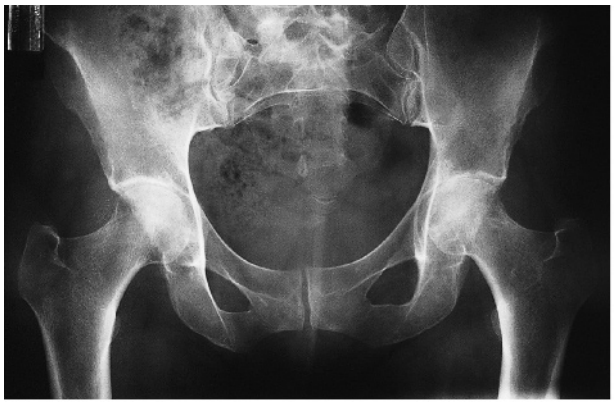


Figure 5.14 Diffuse idiopathic skeletal hyperostosis – DISH The large bony outgrowths around the knee suggest something more than the usual OA.



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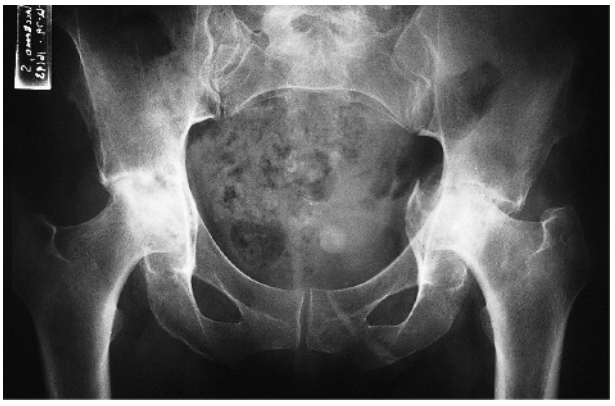


(b)

Figure 5.13 Rapidly destructive osteoarthritis (a) X-ray obtained when the patient was first seen, complaining of pain in the left hip. This shows the typical features of an atrophic form of osteoarthritis on the painful side. (b) Eleven months later there is marked destruction of the left hip, with crumbling of both the femoral head and acetabular floor, and similar features are beginning to appear on the right side.

condition develops in early adulthood, and affects large numbers of joints. They are sometimes due to a definable genetic abnormality in a component of joint tissues, such as type II collagen, which is the main form of collagen found in articular hyaline cartilage. The phenotype of this condition is subtly different from that of 'common-or-garden OA', indicating that such simple genetic defects cannot explain most OA.

- *Alkaptonuria, haemochromatosis and other metabolic conditions.* Similarly, inherited conditions that alter the biochemistry of cartilage or other joint components can cause premature OA, which may have a phenotype similar to that of the genetic varieties alluded to above, although the expression is different in the various metabolic conditions known to cause OA.
- *Kashin Beck disease*, an extremely rare polyarticular form of OA affecting 1 in 6 million people in the Northern China and Eastern Siberia. Those affected have symptoms of joint pain, polyarticular swelling and deformity from childhood. Adults have short stature and their radiographs reveal distorted epiphyses and tubular long bones.

OA resulting from major abnormalities of joint shape or stability

As outlined above, minor abnormalities in the anatomy of a joint, perhaps congenital, can predispose it to OA. In addition, some injury or disorder that leads to acquired changes in joint stability or shape can result in premature OA. These can be generalized or focal. Examples of generalized conditions that affect bone shape include:

- skeletal dysplasias (see Chapter 8)
- Mseleni joint disease
- mucopolysaccharidoses (see Chapter 8).

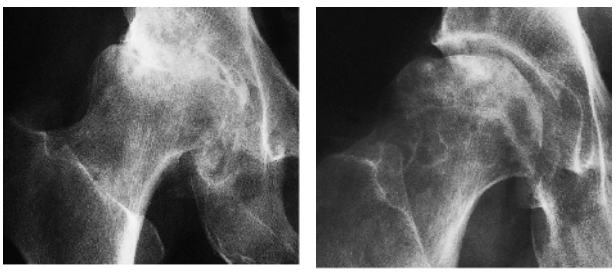
Mseleni joint disease is an endemic form of osteoarthritis that affects the Tsonga people who live in Northern Zululand in South Africa. There appears to be a strong genetic component, with a prevalence of 5% of the population, but with women more often affected than men. There appear to be two phenotypes and may in fact be two related conditions. The first phenotype has radiographic features with elements of multiple epiphyseal dysplasia and affects both genders from an earlier age; the second phenotype is a form of protrusio acetabulae occurring almost exclusively in women.

Focal causes include intra-articular fractures and the childhood conditions that cause abnormal development of the hip such as developmental dysplasia of the hip (DDH), Perthe's disease and slipped capital femoral epiphyses (SCFE). These are explained in detail in Chapter 19.

OA resulting from neuromuscular or vascular problems

Muscle weakness may be a major aetiological factor in the development of OA in older people. In addition, muscle disease or loss of the normal innervation of the joint can lead to atypical forms of OA; furthermore, if the blood supply to subchondral bone is damaged, the OA process can be initiated. The best examples are the following:

- *Neuropathic arthritis*, resulting from syphilis, diabetes, syringomyelia and other forms of neurological abnormality. Neuropathic arthritis is a progressive, destructive form of joint disease associated with loss of sensation. The sensory loss leads to a relative lack of pain, but the joint damage is severe, and similar to that seen in atrophic arthritis (see above). Severe deformity and instability are common. Unusually, the midfoot, and ankle are often involved.
- *Spastic cerebral palsy* which causes abnormal joint loading resulting in premature OA.
- *OA resulting from avascular necrosis of the femoral head* or femoral component of the knee (see Figure 5.15), as explained in detail in Chapter 6.



(a) (b)

Figure 5.15 Differential diagnosis – osteoarthritis and osteonecrosis (a) Osteoarthritis with marked subarticular bone collapse is sometimes mistaken for osteonecrosis. The clue to the diagnosis is that in OA the articular 'space' (cartilage) is progressively reduced before bone collapse occurs, whereas in primary osteonecrosis (b) articular cartilage is preserved even while the underlying bone crumbles.

Chondrocalcinosis, pseudogout and pyrophosphate arthropathy

Calcification of articular hyaline cartilage, or of the fibrocartilage pads within synovial joints, such as the menisci of the knee joint ('*chondrocalcinosis*'), is common. It is strongly age-related (like OA), and rare before the age of 50. It is occasionally due to a familial predisposition, or the existence of some metabolic disorder, such as hyperparathyroidism or hypophosphatasia, but it is usually sporadic and idiopathic. It is often a chance radiographic finding and completely asymptomatic.

The material that is most commonly found in calcified menisci is made of aggregates of tiny calcium pyrophosphate dihydrate (CPPD) crystals. If these crystals get dislodged from the menisci, entering the synovial cavity, they can trigger an intense, self-limiting synovitis, just like gout – hence the name '*pseudogout*'. The knee is to pseudogout what the first metatarsophalangeal joint is to gout. Attacks result in painful, hot swollen joints, and can be relieved by aspiration of the joint and injection of steroids, if not contraindicated, and the use of non-steroidal anti-inflammatory drugs.

A variant of osteoarthritis called '*pyrophosphate arthropathy*' has been described in association with widespread chondrocalcinosis, and is characterized by an unusual distribution for common-or-garden OA, affecting sites such as the wrist, where chondrocalcinosis is common. However, to what extent this is a distinct disease entity is still disputed, and there is no known treatment for the chondrocalcinosis.

FURTHER READING

Beswick AD, Wylde V, Gooberman-Hill R, *et al*. What proportion of patients report long-term pain after total hip or knee replacement for osteoarthritis? A systematic review of prospective studies in unselected patients. *BMJ Open* 2012 Feb 22; **2**(1): e000435.

Dieppe PA, Lohmander LS. Pathogenesis and management of pain in osteoarthritis. *Lancet* 2005 Mar 12–18; **365**(9463): 965–73.

Glyn-Jones S, Palmer AJ, Agricola R, *et al*. Osteoarthritis. *Lancet* 2015 Jul 25; **386**(9991): 376–87.

Hochberg MC, Silman AJ, Smolen JS, *et al*. (eds.). *Rheumatology*, 6th edn. Section 13: Osteoarthritis and related disorders. Philadelphia: Elsevier, 2015.

MacKichan F, Wylde V, Dieppe P. The assessment of musculoskeletal pain in the clinical setting. *Rheum Dis Clin North Am* 2008 May; **34**(2): 311–30.

National Institute for Health and Care Excellence (NICE). Clinical guideline CG177: *Osteoarthritis: Care and Management*, 2014. Available at: https://www.nice.org.uk/guidance/cg177 [Accessed 22 February 2017]

Nelson AE, Jordan JM. Osteoarthritis: Epidemiology and classification. In: Hochberg MC, *et al*. (eds.), *Rheumatology,* 6th edn. Philadelphia: Elsevier, 2015; pp 1433–40.

Oliveria SA, Felson DT, Reed JL, *et al*. Incidence of symptomatic hand, hip and knee osteoarthritis among patients in a health maintenance organization. *Arthritis Rheum* 1995; **38**: 1134–41.

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Osteonecrosis and osteochondritis

6

Jason Mansell & Michael Whitehouse

Avascular necrosis (AVN) has long been recognized as a complication of femoral neck fractures, the usual explanation being traumatic disruption of the blood supply to the femoral head. Osteonecrosis also appears as a distinctive feature in a number of non-traumatic disorders: joint infection, Perthes' disease, caisson disease, Gaucher's disease, systemic lupus erythematosus (SLE), high-dosage corticosteroid administration and alcohol abuse, to mention only the more common ones (see Box 6.1). Whatever the cause, the condition, once established, may come to dominate the clinical picture, demanding attention in its own right.

Aetiology and pathogenesis

Certain sites are peculiarly susceptible to ischaemic necrosis including the femoral head, the femoral condyles, the head of the humerus, the capitulum and the proximal parts of the scaphoid and talus. These subarticular regions lie at the most distant parts of the bone's vascular territory, and they are largely enclosed by cartilage, giving restricted access to local blood vessels. The subchondral trabeculae are further compromised in that they are sustained largely by a system of endarterioles with limited collateral connections.

Another factor which needs to be taken into account is that the vascular sinusoids which nourish the marrow and bone cells, unlike arterial capillaries, have no adventitial layer and their patency is determined by the volume and pressure of the surrounding marrow tissue, which itself is encased in unyielding bone. The system functions essentially as a closed compartment within which one element can expand only at the expense of the others. Local changes such as decreased blood flow, haemorrhage or marrow swelling can, therefore, rapidly spiral to a vicious cycle of ischaemia, reactive oedema or inflammation, marrow swelling, increased intraosseous pressure and further ischaemia (see Figure 6.1).

BOX 6.1 MAIN CONDITIONS ASSOCIATED WITH NON-TRAUMATIC OSTEONECROSIS

Infections
- Osteomyelitis
- Septic arthritis

Haemoglobinopathy
- Sickle-cell disease

Storage disorders
- Gaucher's disease

Caisson disease
- Dysbaric osteonecrosis

Coagulation disorders
- Familial thrombophilia
- Hypofibrinolysis
- Hypolipoproteinaemia
- Thrombocytopenic purpura

Other
- Perthes' disease
- Cortisone administration
- Alcohol abuse
- Systemic lupus erythematosus (SLE) (? increase in antiphospholipid antibodies)
- Pregnancy (? decreased fibrinolysis; ? fatty liver)
- Anaphylactic shock
- Ionizing radiation

The process described above can be initiated in at least four different ways:

1 disruption of the local blood supply
2 venous stasis and retrograde arteriolar stoppage
3 intravascular thrombosis
4 compression of capillaries and sinusoids by marrow swelling.

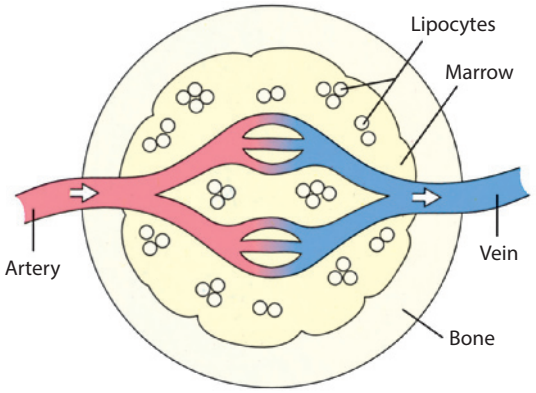


Figure 6.1 Avascular necrosis – pathogenesis
The medullary cavity of bone is virtually a closed compartment containing myeloid tissue, marrow fat and capillary blood vessels. Any increase in fat cell volume will reduce capillary circulation and may result in bone ischaemia.

Ischaemia, in the majority of cases, is due to a combination of several of these factors.

TRAUMATIC OSTEONECROSIS

In traumatic osteonecrosis the vascular anatomy is particularly important. In fractures and dislocations of the hip, the retinacular vessels supplying the femoral head are easily torn. If, in addition, there is damage to or thrombosis of the ligamentum teres, osteonecrosis is inevitable. Little wonder that displaced fractures of the femoral neck are complicated by osteonecrosis in over 20% of cases. Undisplaced fractures also sometimes result in subchondral necrosis; this may be due to thrombosis of intraosseous capillaries or sinusoidal occlusion due to marrow oedema.

Other injuries which are prone to osteonecrosis are fractures of the scaphoid and talus. Significantly, in these cases it is always the proximal fragment which suffers. This is because the principal vessels enter the bones near their distal ends and take an intraosseous course from distal to proximal. Impact injuries and osteoarticular fractures at any of the convex articular surfaces behave in the same way and often develop localized ischaemic changes. These small lesions are usually referred to as 'osteochondroses' and many of them have acquired eponyms which are firmly embedded in orthopaedic history.

NON-TRAUMATIC OSTEONECROSIS

The mechanisms here are more complex and may involve several pathways to intravascular stasis or thrombosis, as well as extravascular swelling and capillary compression.

Intravascular thrombosis Various mechanisms leading to capillary thrombosis have been demonstrated in patients with non-traumatic osteonecrosis. Over 80% of cases are associated with high-dosage corticosteroid medication and/or alcohol abuse. These conditions give rise to hyperlipidaemia and fatty degeneration of the liver. Research has indicated that fat embolism plays a part, giving rise to capillary endothelial damage, platelet aggregation and thrombosis. Other studies have suggested that thrombophilia and hypofibrinolysis are important aetiological factors in both adult osteonecrosis and Perthes' disease. Other coagulopathies have been implicated, including antiphospholipid deficiency in SLE and enhanced coagulability in sickle-cell disease. It seems likely that coagulation abnormalities of one sort or another play at least a contributory role in some of the disorders associated with non-traumatic osteonecrosis.

Extravascular marrow swelling High-dosage corticosteroid administration and alcohol overuse cause fat cell swelling in the marrow, a feature which is very obvious in bone specimens obtained during joint replacement. There is a rise in intraosseous pressure and contrast venography shows slowing of venous blood flow from the bone. This increase in marrow fat volume in the femoral head is thought to cause sinusoidal compression, venous stasis and retrograde ischaemia leading to trabecular bone death; in other words, the establishment of a compartment syndrome.

Whichever of these mechanisms offers the primary pathway to non-traumatic bone ischaemia, it is almost certain that both intravascular and extravascular factors come into play at a fairly early stage and each enhances the effect of the other (see Figures 6.2 and 6.3).

Pathology and natural history

Bone cells die after 12–48 hours of anoxia, yet for days or even weeks the gross appearance of the affected segment remains unaltered. During this time the most striking histological changes are seen in the marrow: loss of fat cell outlines, inflammatory cell infiltration, marrow oedema, the appearance of tissue histiocytes, and eventual replacement of necrotic marrow by undifferentiated mesenchymal tissue.

A characteristic feature of ischaemic segmental necrosis is the tendency to bone repair, and within a few weeks one may see new blood vessels and osteoblastic proliferation at the interface between ischaemic and live bone. As the necrotic sector becomes demarcated, vascular granulation tissue advances from the surviving trabeculae and new bone is laid down upon the dead; it is this increase in mineral mass that later produces the radiographic appearance of increased density or 'sclerosis'.

Reparative new bone formation proceeds slowly and probably does not advance for more than 8–10 mm into the necrotic zone. With time, structural failure begins to occur in the most heavily loaded part of the

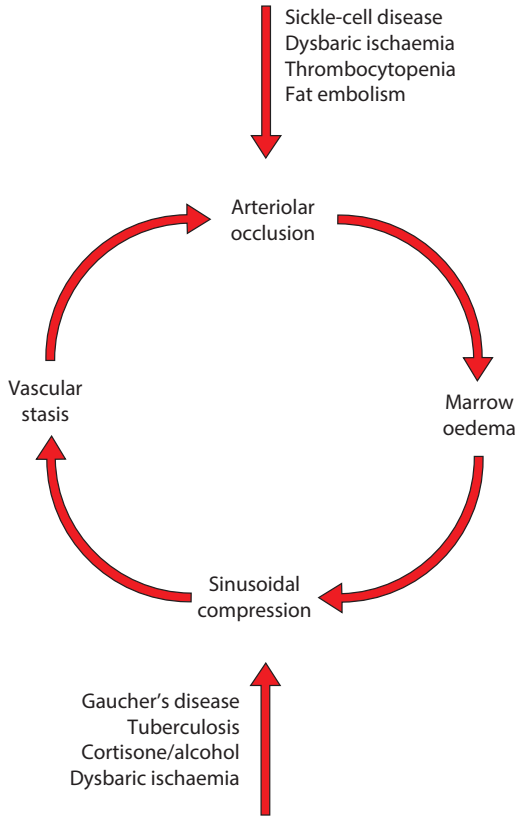


Figure 6.2 Avascular necrosis Algorithm showing how various disorders may enter the vicious cycle of capillary stasis and marrow engorgement.

necrotic segment. Usually this takes the form of a linear tangential fracture close to the articular surface, possibly due to shearing stress. The crack may break through the articular cartilage and at operation it may be possible to lift the 'lid' off the necrotic segment like the cracked shell of a hard-boiled egg.

However, until very late the articular cartilage retains its thickness and viability. In the final stages, fragmentation of the necrotic bone leads to progressive deformity and destruction of the joint surface (see Figure 6.4). The size of the necrotic segment, as defined by the hypo-intense band in the T1 weighted MRI, is usually established at the time of the initiating ischaemic event, and from then on it rarely increases; indeed, there is evidence that non-traumatic lesions sometimes diminish in size and occasionally even disappear. In persistent lesions, the rate of bone collapse depends largely on the site and extent of the necrotic segment: lesions which lie outside the normal stress trajectories may remain structurally intact while those that involve large segments of the load-bearing surface usually collapse within 3 years (see 'Staging the lesion' below).

Clinical features

The earliest stage of bone death is asymptomatic; by the time the patient presents, the lesion is usually advanced. Pain is a common complaint. It is felt in or near a joint, and perhaps only with certain movements. Some patients complain of a 'click' in the joint,

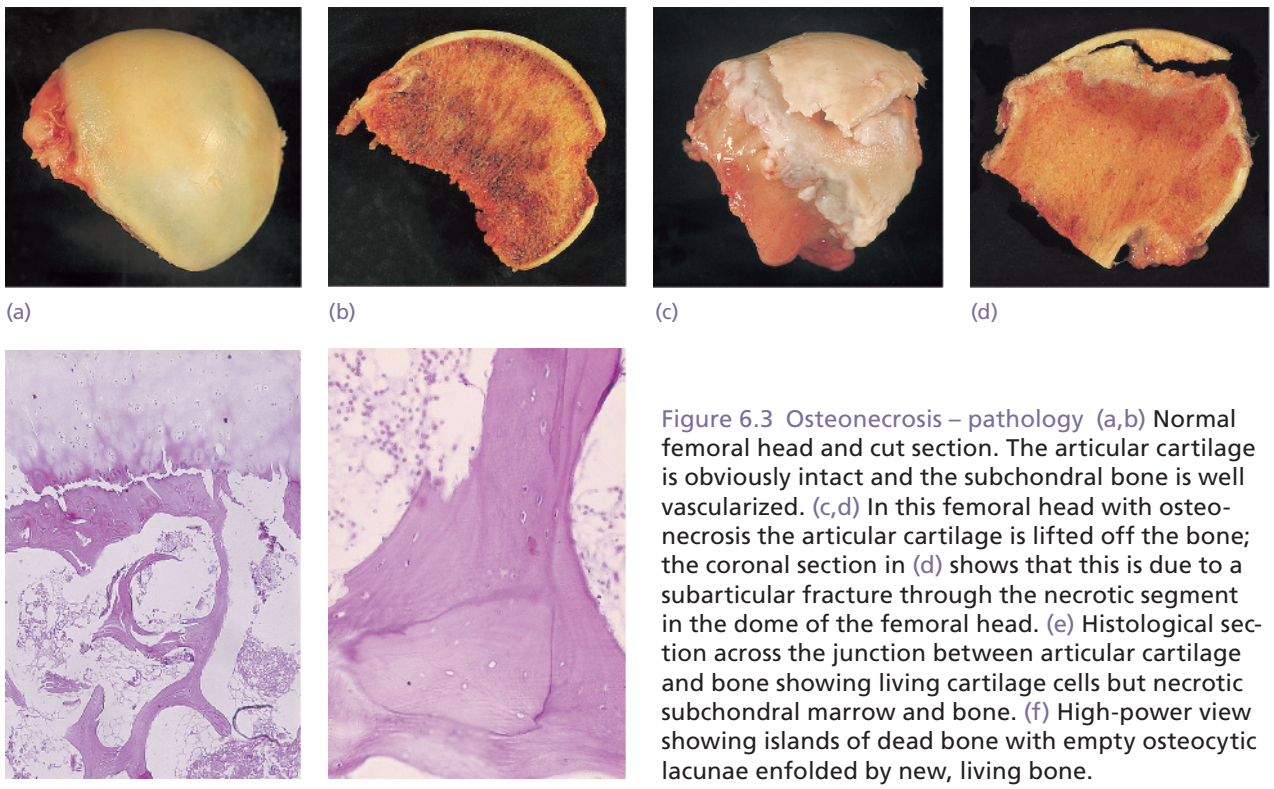
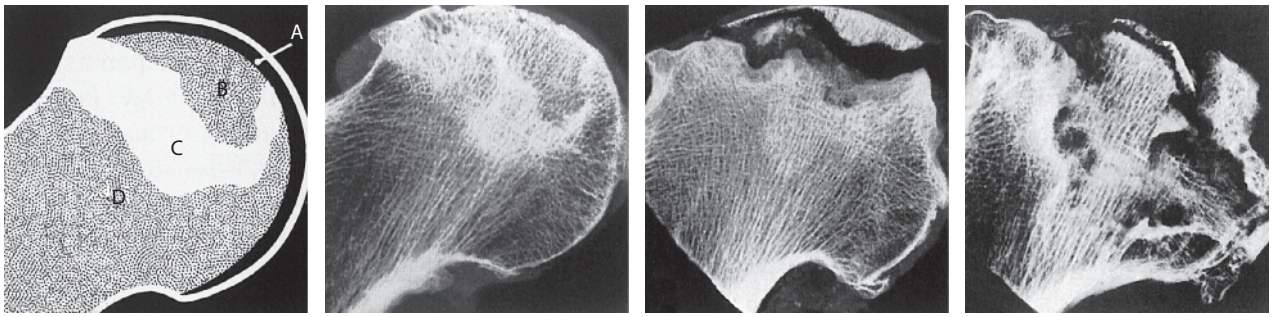


Figure 6.3 Osteonecrosis – pathology (a,b) Normal femoral head and cut section. The articular cartilage is obviously intact and the subchondral bone is well vascularized. (c,d) In this femoral head with osteonecrosis the articular cartilage is lifted off the bone; the coronal section in (d) shows that this is due to a subarticular fracture through the necrotic segment in the dome of the femoral head. (e) Histological section across the junction between articular cartilage and bone showing living cartilage cells but necrotic subchondral marrow and bone. (f) High-power view showing islands of dead bone with empty osteocytic lacunae enfolded by new, living bone.



(a) (b) (c) (d)

Figure 6.4 Avascular necrosis of bone – pathology (a) This is a diagrammatic guide to the fine-detail X-rays of necrotic femoral heads (b–d) which show the progress of osteonecrosis. The articular cartilage (A) remains intact for a long time. The necrotic segment (B) has a texture similar to that of normal bone, but it may develop fine cracks. New bone surrounds the dead trabeculae and causes marked sclerosis (C). Beyond this the bone remains unchanged (D). In the later stages the necrotic bone breaks up and finally the joint surface is destroyed.

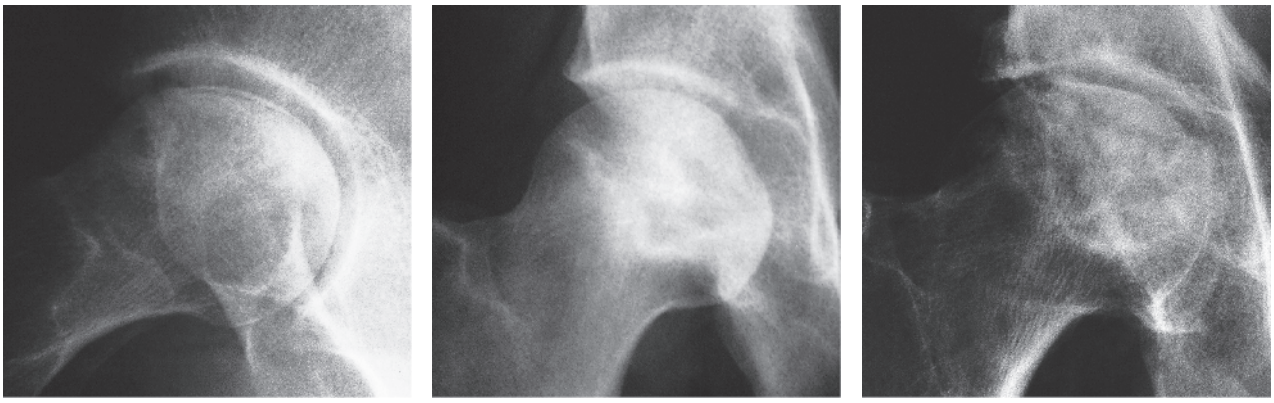
probably due to snapping or catching of a loose articular fragment. In the later stages the joint becomes stiff and deformed. Local tenderness may be present and, if a superficial bone is affected, there may be some swelling. Movements may be restricted; in advanced cases there may be fixed deformities.

Imaging

X-ray The early signs of ischaemia are confined to the bone marrow and cannot be detected by plain X-ray examination. X-ray changes, when they appear (seldom before 3 months after the onset of ischaemia), are due to (1) reactive new bone formation at the boundary of the ischaemic area, and (2) trabecular failure in the necrotic segment. An area of increased radiographic density appears in the subchondral bone; soon afterwards, suitable views may show a thin tangential fracture line just below the articular surface – the '*crescent sign*'. In the late stages there is distortion of the articular surface and more intense 'sclerosis', now partly due to bone compression in a collapsed segment.

Occasionally the necrotic portion separates from the parent bone as a discrete fragment. However, it is now recognized that in the case of the femoral head and the medial femoral condyle such necrotic fragments may have resulted from small osteoarticular fractures which only later failed to unite and lost their blood supply.

With all the changes described here (and this is the cardinal feature distinguishing primary avascular necrosis from the sclerotic and destructive forms of osteoarthritis) the 'joint space' retains its normal width because the articular cartilage is not destroyed until very late (see Figure 6.5).

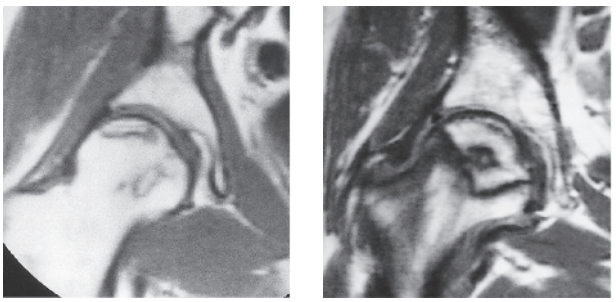


(a) (b) (c)

Figure 6.5 Avascular necrosis – X-rays (a) The earliest X-ray sign is a thin radiolucent crescent just below the convex articular surface where load bearing is at its greatest. This represents an undisplaced subarticular fracture in the early necrotic segment. (b) At a later stage the avascular segment is defined by a band of increased density due to vital new bone formation. At this stage the femoral head may still be spherical and (unlike osteoarthritis) the articular space is still well-defined. (c) In late cases there is obvious collapse and distortion of the articular surface.

Radioscintigraphy Radionuclide scanning with ^{99m}Tc sulphur colloid, which is taken up in myeloid tissue, may reveal an avascular segment. This is most likely in traumatic avascular necrosis, where a large segment of bone is involved, or in sickle-cell disease where a 'cold' area contrasts significantly with the generally high nuclide uptake due to increased erythroblastic activity. ^{99m}Tc-HDP scans (in the bone phase) may also show a 'cold' area, particularly if a large segment of bone is avascular (e.g. after fracture of the femoral neck). More often, however, the picture is dominated by *increased* activity, reflecting hyperaemia and new bone formation in the area around the infarct.

Magnetic resonance imaging MRI is the most reliable way of diagnosing marrow changes and bone ischaemia at a comparatively early stage (Figure 6.6). The first sign is a band-like low-intensity signal on the T1 weighted spin echo (SE) image (and a similar but high-intensity signal on the short-tau inversion recovery (STIR) image), corresponding to the interface between ischaemic and normal bone. The site (Figure 6.7) and size of the demarcated necrotic zone have been used to predict the progress of the lesions (see Chapter 19).



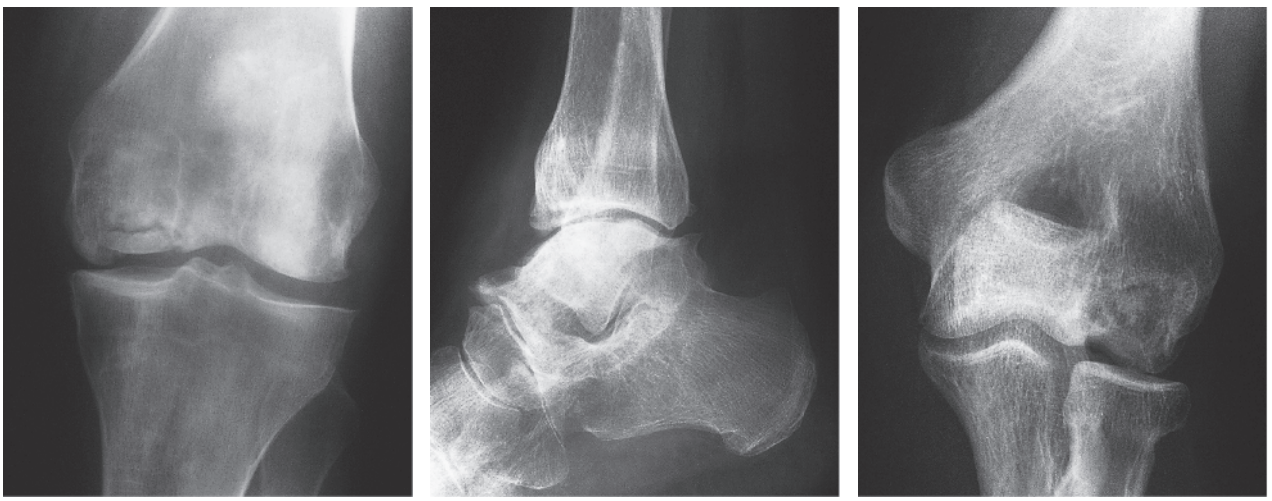
(a) (b)

Figure 6.6 Osteonecrosis – MRI (a) Before any change is discernible on the plain X-ray, MRI will show a typical hypointense band in the T1 weighted image, outlining the ischaemic segment beneath the articular surface. (b) In this case the size of the ischaemic segment is much larger – and the likelihood of bone crumbling much greater.

Computed tomography CT involves considerable radiation exposure and it is not very useful for diagnosing osteonecrosis. However, it does show the area of bone destruction very clearly and it may be useful in planning surgery.

Tests for haemodynamic function

During the early stage of ischaemic necrosis the intramedullary pressure is often markedly raised. This phenomenon is most easily demonstrated in the femoral head. A cannula introduced into the metaphysis enables measurements to be taken (1) at rest and (2) after rapid injection of saline. The normal resting pressure is 10–20 mmHg, rising by about 15 mm after saline injection; in early osteonecrosis both the intramedullary pressure and the response to saline injection may be increased three- or four-fold. Venous stasis can also be demonstrated by venography after injection of radio-opaque medium into the bone. Similar findings have been recorded in osteoarthritis, but the change is not nearly as marked as in osteonecrosis.



(a) (b) (c)

Figure 6.7 Osteonecrosis – distribution The most common sites for osteonecrosis are the head of the femur, the head of the humerus and, as shown here, the medial condyle of the femur, the talus and the capitulum. All these areas are located beneath convex articular surfaces; osteonecrosis is seldom seen beneath a concave articular surface.

Staging the lesion

In 1980 Ficat and Arlet introduced the concept of *radiographic staging* for osteonecrosis of the hip. The system has since been updated to account for imaging modalities such as MRI.

Stage 0 is preclinical and preradiological and AVN can be suggested only if it is has been diagnosed in, for example, the opposite hip.
Stage 1 is the early resorptive phase. The plain X-rays are normal but changes become visible on MRI or bone scintigraphy.
Stage 2 is the reparative stage where the femoral head contour is still normal but there are early signs of reactive change in the subchondral area.
Stage 3 is early collapse of the femoral head. There are clear-cut X-ray signs of osteonecrosis with evidence of structural damage and distortion of the bone outline.
Stage 4 shows collapse of the articular surface and signs of secondary OA.

MRI has proven to be more reliable than X-rays as a predictor of outcome.

The location and size of the necrotic segment in Ficat stages 1–3 are defined by the hypo-intense band on the T1 weighted MRI. Two general observations can be made: (1) the size of the ischaemic segment is determined at a very early stage and it rarely increases after that; (2) small lesions which do not involve the maximally loaded zone of the articular surface tend not to collapse, whereas large lesions extending under the maximally loaded articular surface break down in over 60% of cases. Shimizu's classification is particularly useful in planning treatment; this is discussed in Chapter 19.

The most widely used system, which permits comparison between series from different participating centres, is the one promoted by the International Association of Bone Circulation and Bone Necrosis (*Association Research Circulation Osseous – ARCO*) which applies mainly to femoral head necrosis (Table 6.1).

Table 6.1 ARCO staging of osteonecrosis

Stage	Observations
Stage 0	Patient asymptomatic and all clinical investigations 'normal' Biopsy shows osteonecrosis
Stage 1	X-rays normal MRI or radionuclide scan shows osteonecrosis
Stage 2	X-rays and/or MRI show early signs of osteonecrosis but no distortion of bone shape or subchondral 'crescent sign' Sub-classification by area of articular surface involved: A = less than 15% B = 15–30% C = more than 30%
Stage 3	X-ray shows 'crescent sign' but femoral head still spherical Sub-classification by length of 'crescent'/articular surface: A = less than 15% B = 15–30% C = more than 30%
Stage 4	Signs of flattening or collapse of femoral head: A = less than 15% of articular surface B = 15–30% of articular surface C = more than 30% of articular surface
Stage 5	Changes as above plus loss of 'joint space' (secondary OA)
Stage 6	Changes as above plus marked destruction of articular surfaces

Diagnosis of the underlying disorder

In many cases of osteonecrosis an underlying disorder will be obvious from the history: a known episode of trauma, an occupation such as deep-sea diving, a family background of Gaucher's disease or sickle-cell disease. There may be a record of high-dosage corticosteroid administration; for example, after renal transplantation. However, smaller doses (e.g. as short-term treatment for asthma or as an adjunct in neurosurgical emergencies) and even topical steroid preparations can also be dangerous in patients with other risk factors. Combinations of drugs (e.g. corticosteroids and azathioprine, or corticosteroids after a period of alcohol abuse) can be potent causes of osteonecrosis; occasionally corticosteroids have been given without the patient's knowledge.

Alcohol abuse is often difficult to determine because patients tend to hide the information. There is no biochemical marker that is specific for high alcohol intake but elevation of three or four of the following is suggestive: aspartate transaminase, gamma glutamyl transferase (GGT), serum urate, serum triglyceride and mean red cell volume.

Ideally patients with very early non-traumatic osteonecrosis, and children with early Perthes' disease, should undergo laboratory tests for coagulopathies; this is justified by reports of cases in which the condition has been halted or reversed by treatment with antithrombotic preparations. Unfortunately, the tests are expensive and there is understandable resistance to adopting this approach in routine management.

In cases of suspected SLE, antiphospholipid antibodies may be measured.

Prevention

Where risk factors for osteonecrosis are recognized, preventive steps can be taken especially in the management of corticosteroid medication and alcohol abuse.

Corticosteroids should be used only when essential and in minimal effective dosage. *It is important also to be aware of the cumulative effect of even moderate doses of corticosteroids in patients with a history of alcohol abuse*. Anoxia must be prevented in patients with haemoglobinopathies. Decompression procedures for divers and compressed-air workers should be rigorously applied.

Treatment

In planning treatment, all the factors that influence the natural course of the condition must be taken into account: the general medical background, the type of ischaemic necrosis, the site and extent of the necrotic segment, its stage of development, the patient's age and capacity for bone repair, the persistence or otherwise of the aetiological agent and its effect on bone turnover.

Only general principles will be discussed here; the treatment of osteonecrosis in specific sites is dealt with in the appropriate chapters on regional orthopaedics.

EARLY OSTEONECROSIS

While the bone contour is intact there remains the hope that structural failure can be prevented. Some lesions heal spontaneously and with minimal deformity; this is seen especially in areas which are not heavily loaded: the non-weight-bearing joints, the superomedial part of the femoral head and the non-weight-bearing surfaces of the femoral condyles and talus. Here one can afford to pursue a watchful waiting policy.

There is low-level evidence that drug therapy (such as bisphosphonates, statins and low molecular weight heparin) may have some efficacy in early stages of the disease process. Lesions in heavily loaded joints have a poor prognosis and will probably end in structural failure if left untreated. Simple measures to reduce loading of weight-bearing joints may help, though their value has not been proven. If the bone contour is still intact, an 'unloading' osteotomy will help to preserve the anatomy while remodelling proceeds. This approach is applicable especially to the hip and knee.

Medullary decompression and bone grafting may have a place in ARCO stage 1 and 2 osteonecrosis of the femoral head (Chapter 19) and there is some recent evidence that rates of femoral head collapse can be reduced through augmentation with mesenchymal stem cells.

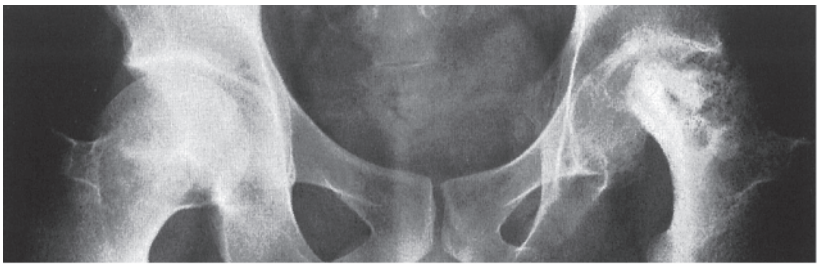
INTERMEDIATE STAGE OSTEONECROSIS

Once there is structural damage and distortion of the articular surface, conservative operations are inappropriate. However, the joint may still be salvageable and in this situation realignment osteotomy – either alone or combined with curettage and bone grafting of the necrotic segment – has a role.

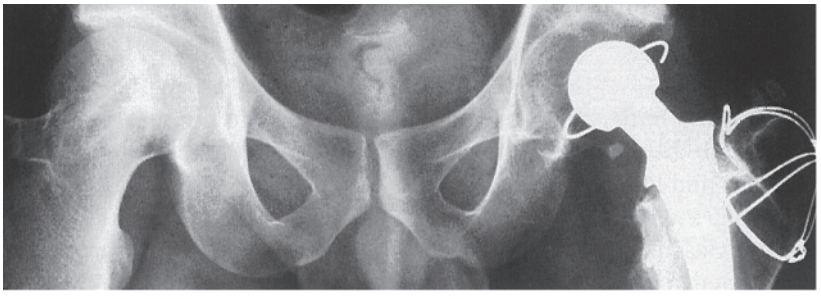
If mobility can be sacrificed without severe loss of function (e.g. in the ankle or wrist), arthrodesis will relieve pain and restore stability.

LATE STAGE OSTEONECROSIS

Destruction of the articular surface may give rise to pain and severe loss of function. Three options are available: (1) non-operative management, concentrating on pain control, modification of daily activities and, where appropriate, splintage of the joint; (2) arthrodesis of the joint (e.g. the ankle or wrist); or (3) partial or total joint replacement, the preferred option for the shoulder, hip and knee (see Figure 6.8).



(a)



(b)

Figure 6.8 Osteonecrosis – treatment (a) Alcohol abuse has led to bilateral femoral head necrosis, advanced on the left but detectable only by MRI on the right. (b) The left hip had to be replaced; at the same time the right side was treated by drilling of the femoral neck (medullary decompression). This X-ray was taken 8 years later.

SYSTEMIC DISORDERS ASSOCIATED WITH OSTEONECROSIS

Drug-induced necrosis

Alcohol, corticosteroids, immunosuppressives and cytotoxic drugs, either singly or in combination, are the commonest causes of non-traumatic osteonecrosis. 'At risk' doses for these drugs have not been established; the threshold depends not only on the total intake but also on the time over which the intake is spread and the presence or absence of other risk factors for the development of osteonecrosis. A cumulative dose of 2000 mg of prednisone equivalent administered over several years (e.g. in the treatment of inflammatory arthritis) is less likely to cause osteonecrosis than the same dose given over a period of a few months (e.g. after organ transplantation). It is important to bear in mind that multiple causative agents have an additive effect; thus, osteonecrosis has been encountered after comparatively short courses and low doses of corticosteroids (totals of 800 mg or less), but in these cases an additive factor can almost always be identified.

The threshold dose for alcohol is equally vague. However, based on the known dose relationship of alcohol-induced fatty degeneration of the liver, it is probably around 150 mg of ethanol per day (for men) – the equivalent of 300 mL of spirits, 1.2 litres of table wine or 3 litres of beer – continuing for over 2 years. The dose for women is considerably less. Asking patients 'How much do you drink?' is unlikely to elicit an accurate response. However, the presence of raised serum triglyceride and GGT levels, together with an increased mean corpuscular volume (MCV), is suggestive of excessive alcohol intake.

Sickle-cell disease

Sickle-cell disease is a genetic disorder in which the red cells contain abnormal haemoglobin (HbS). Instead of having an X in the Hb beta chain, the replacement of this residue with Y results in Hb precipitation at low oxygen tensions. In deoxygenated blood there is increased aggregation of the haemoglobin molecules and distortion of the red cells, which become somewhat sickle-shaped. At first this is reversible and the cells reacquire their normal shape when the blood is oxygenated. Eventually, however, the red cell membrane becomes damaged and the cells are permanently deformed.

The sickle-cell trait, which originated in West and Central Africa centuries ago, is an example of natural selection for survival in areas where malaria was endemic. From there, the gene was carried to countries along the Mediterranean, the Persian Gulf, parts of India and across the Atlantic where it appears in people of Afro-American descent. In recent years it has spread more widely in Europe but it is rarely encountered south of the equator.

Sickle-cell disease is most likely in homozygous offspring (those with HbS genes from both mother and father), but it may also occur in heterozygous children with HbS/C haemoglobinopathy and HbS/thalassaemia.

Inheritance of one HbS gene and one normal β-globin gene confers the (heterozygous) *sickle-cell trait*; HbS concentration is low and sickling occurs only under conditions of hypoxia (e.g. under inefficient anaesthesia, in extreme cold, at very high altitudes and when flying in unpressurized aircraft). In the established disorder, the main clinical features are due to a combination of chronic haemolytic anaemia and a tendency to clumping of the sickle-shaped cells which results in diminished capillary flow and recurrent episodes of intracapillary thrombosis.

Secondary changes such as trabecular coarsening, infarctions of the marrow, periostitis and osteonecrosis are common. Complications include hyperuricaemia (due to increased red cell turnover) and an increased susceptibility to bacterial infection.

Clinical features

Children during the first two years of life may present with swelling of the hands and feet. X-rays at first seem normal, but later there may be suggestive features such as marrow densities and periosteal new bone formation ('dactylitis'). These changes are usually transient, but treatment is required for pain. In older children a typical feature is recurrent episodes of severe pain, sometimes associated with fever. These 'crises', which may affect almost any part of the body, are thought to be due to infarcts.

Osteonecrosis of the femoral head is common, both in children (when it is sometimes mistaken for Perthes' disease) and in young adults, in whom other causes of non-traumatic osteonecrosis have to be excluded. Males and females are affected with almost equal frequency. The child develops a painful limp and movements are restricted.

X-rays may show no more than a diffuse increase in density of the epiphysis; however, in most cases the changes are very similar to those of Perthes' disease, usually going on to flattening of the epiphysis. In young adults there are both destructive lesions and diffuse sclerosis of the femoral head (see Figure 6.9). The head of the humerus and the femoral condyles may be similarly affected.

Other bone changes are due to a combination of marrow hyperplasia and medullary infarctions.

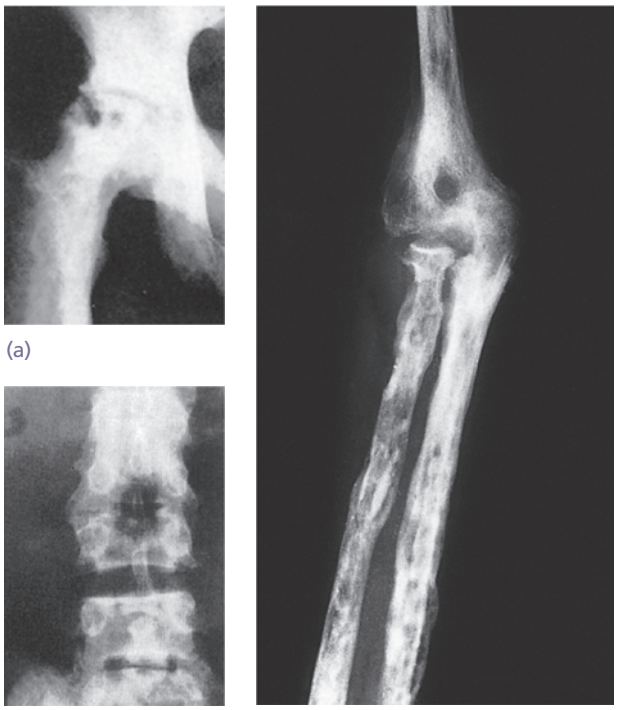


Figure 6.9 Sickle-cell disease (a) Typical features of osteonecrosis are seen in the femoral head, often accompanied by patchy areas of bone destruction and endosteal sclerosis in the femoral shaft. (b) The spine also may be involved, producing appearances similar to those of bone infection. (c) In severe cases infarctions of tubular bones may resemble osteomyelitis, with sequestra and a marked periosteal reaction.

Trabecular coarsening and thickening of the cortices may be mistaken for signs of infection.

Bacterial osteomyelitis and *septic arthritis*, sometimes involving multiple sites, are serious complications, particularly in children. In over 50% of cases the organism is *Salmonella*.

Treatment

A follow-up study of untreated children with femoral head necrosis due to sickle-cell disease showed that 80% of them had permanently damaged hips with severe loss of function. This may be due to recurrent infarction and inflammatory changes in the joint. Hypoxic conditions favouring the occurrence of crises should be avoided. If episodes of bone pain are frequent, transfusions may be necessary to reduce the concentration of HbS. During a crisis, the patient should be given adequate analgesia and should be kept fully oxygenated. Infections should be guarded against, or treated promptly with the appropriate antibiotics.

Femoral head necrosis in children should be treated in the same way as Perthes' disease (see 'Perthes' disease' in Chapter 19, The hip). Adults are treated along the lines described in the section 'Osteonecrosis' in Chapter 19. The emphasis in all cases should be on conservatism. Anaesthesia carries definite risks; failure to maintain adequate oxygenation may recipitate vascular occlusion in the central nervous system, lungs or kidneys. Prophylactic antibiotics are advisable as the risk of postoperative infection is high.

Caisson disease and dysbaric osteonecrosis

Decompression sickness (caisson disease) and osteonecrosis are important causes of disability in deep-sea divers and compressed-air workers building tunnels or underwater structures. Under increased air pressure the blood and other tissues (especially fat) become supersaturated with nitrogen; if decompression is too rapid, the gas is released as bubbles, which cause local tissue damage, generalized embolic phenomena and intracapillary coagulation. Prolonged compression may also cause swelling of marrow fat cells and decreased intramedullary blood flow, possibly due to oxygen toxicity.

The symptoms of decompression sickness, which may develop within minutes, are pain near the joints ('the bends'), breathing difficulty and vertigo ('the staggers'). In the most acute cases there can be circulatory and respiratory collapse, severe neurological changes, coma and death. Only 10% of patients with bone necrosis give a history of decompression sickness.

Radiological bone lesions have been found in 17% of compressed-air workers in the UK; almost half the lesions are juxta-articular – mainly in the humeral head and femoral head – but microscopic bone death is much more widespread than X-rays suggest.

Clinical and X-ray features The necrosis may cause pain and loss of joint movement, but many lesions remain 'silent' and are found only on routine X-ray examination. Medullary infarcts cause mottled calcification or areas of dense sclerosis. Juxta-articular changes are similar to those in other forms of osteonecrosis.

Management The aim is prevention; the incidence of osteonecrosis is proportional to the working pressure, the length of exposure, the rate of decompression and the number of exposures. Strict enforcement of suitable working schedules has reduced the risks considerably. The treatment of established lesions follows the principles already outlined.

Gaucher's disease

Gaucher's disease (see also Chapter 8) is a familial metabolic disorder caused by inherited deficiency of

lysosomal enzyme glucocerebrosidase and characterized by accumulation of glucosylceramide in the lysosomes of reticuloendothelial cells to produce Gaucher's cells. The effects are seen chiefly in the liver, spleen and bone marrow. Accumulation of Gaucher's cells in the bone marrow triggers a series of events that lead to skeletal pathology, of which osteonecrosis is the worst. The hip is most frequently affected, but lesions also appear in the distal femur, the talus and the head of the humerus. Bone ischaemia is usually attributed to the increase in medullary cell volume and sinusoidal compression, but it is likely that other effects (abnormal cell emboli and increased blood viscosity) are equally important.

Clinical features

Bone necrosis may occur at any age but is of particular concern in children due to the risk of disturbing normal growth and inhibiting the child's ability to achieve optimal bone mass. It usually causes pain around one of the larger joints (usually the hip). In long-standing cases movements are restricted. There is a tendency for the Gaucher deposits to become infected and the patient may present with septicaemia. Blood tests reveal anaemia, leucopenia and thrombocytopenia. A diagnostic, though inconstant, finding is a raised serum acid phosphatase level.

X-ray

The appearances resemble those in other types of osteonecrosis, and 'silent' lesions may be found in a number of bones. A special feature (due to replacement of myeloid tissue by Gaucher cells) is expansion of the tubular bones, especially the distal femur, producing the Erlenmeyer flask appearance (see Figure 6.10). Cortical thinning and osteoporosis may lead to pathological fracture. Dual-energy X-ray absorptiometry is used to evaluate bone mineral density in patients with Gaucher's disease and is capable of providing a quantitative assessment of bone involvement and monitoring changes in bone density in response to therapy.

Treatment

The condition can now be treated by replacement of the missing enzyme and there is evidence that this will reduce the incidence of bone complications. The management of established osteonecrosis follows the principles outlined earlier. However, there is a greater risk of infection following operation and suitable precautions should be taken. For adults, total joint replacement is probably preferable to other procedures.

Radiation necrosis

Ionizing radiation, if sufficiently intense or prolonged, may cause bone death. This is due to the combined effects of damage to small blood vessels, marrow cells and bone cells. Such changes, which are dose-related, often occurred in the past when low-energy radiation was in use. Nowadays, with megavoltage apparatus and more sophisticated planning techniques, long-term bone damage is much less likely; patients who present with osteonecrosis are usually those who were treated some years ago. Areas affected are mainly the shoulder and ribs (after external irradiation for breast cancer), the sacrum, pelvis and hip (after irradiation of pelvic lesions) and the jaws (after treatment of tumours around the head and neck).

Pathology

Unlike the common forms of ischaemic necrosis, which always involve subchondral bone, radiation necrosis is more diffuse and the effects more variable. Marrow and bone cells die, but for months or even years there may be no structural change in the bone. Gradually, however, stress fractures appear and

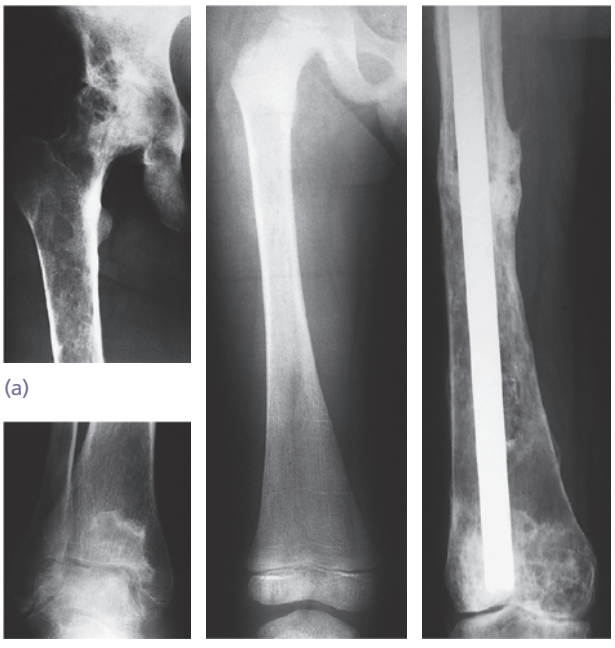


Figure 6.10 Gaucher's disease (a) Gaucher deposits are seen throughout the femur. The cortices are thin and there is osteonecrosis of the femoral head. (b) Bone infarction is seen in the distal end of the tibia and the talus. (c) The typical Erlenmeyer flask appearance is seen in the X-ray of this teenager. (d) Ten years later the bone changes are much more marked, the cortices are extremely thin and the patient has obviously suffered a pathological fracture.

may result in widespread bone destruction. A striking feature is the absence of repair and remodelling. The surrounding bone is usually osteoporotic; in the jaw, infection may follow tooth extraction.

Clinical features

The patient usually presents with pain around the shoulder, the hip, the sacrum or the pubic symphysis. There will always be a history of previous treatment by ionizing radiation, though this may not come to light unless appropriate questions are asked. There may be local signs of irradiation, such as skin pigmentation, and the area is usually tender. Movements in the nearby joint are restricted. General examination may reveal scars or other evidence of the original lesion.

X-rays show areas of bone destruction and patchy sclerosis (see Figure 6.11); in the hip there may be an unsuspected fracture of the acetabulum or femoral neck, or collapse of the femoral head.

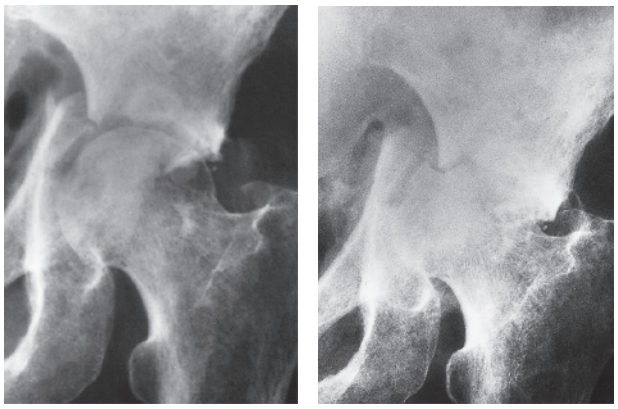
Treatment

Treatment depends on the site of osteonecrosis, the quality of the surrounding bone and the life expectancy of the patient. If a large joint is involved (e.g. the hip), joint replacement may be considered; however, bone quality is often poor and there is a high risk of early implant loosening. Nevertheless, if pain cannot be adequately controlled, and if the patient has a reasonable life expectancy, joint replacement is justified.

Osteochondrosis (osteochondritis)

The terms 'osteochondrosis' or 'osteochondritis' have for many years been applied to a group of conditions in which there is demarcation, and sometimes separation and necrosis, of a small segment of articular cartilage and bone. The affected area shows many of the features of ischaemic necrosis, including death of bone cells in the osteoarticular fragment and reactive vascularity and osteogenesis in the surrounding bone. The disorder occurs mainly in adolescents and young adults, often during phases of increased physical activity, and it may be initiated by trauma or repetitive stress.

The pathogenesis of these lesions is still not completely understood. Impact injuries can cause oedema or bleeding in the subarticular bone, resulting in capillary compression or thrombosis and localized ischaemia. The critical event may well be a small osteochondral fracture, too faint to show up on plain X-ray examination but often visible on MRI. If the crack fails to unite, the isolated fragment may lose its blood supply and become necrotic. Traction injuries may similarly damage the blood supply to an apophysis. However, it is thought that there must be other predisposing factors, for the condition is sometimes multifocal and sometimes runs in families.



(a) (b)

Figure 6.11 Radiation necrosis – X-rays This patient received radiation therapy for carcinoma of the bladder. One year later he developed pain in the left hip and X-ray showed (a) a fracture of the acetabulum. Diagnosis of radiation necrosis was confirmed when (b) the fracture failed to heal and the joint crumbled.

Clinical presentation

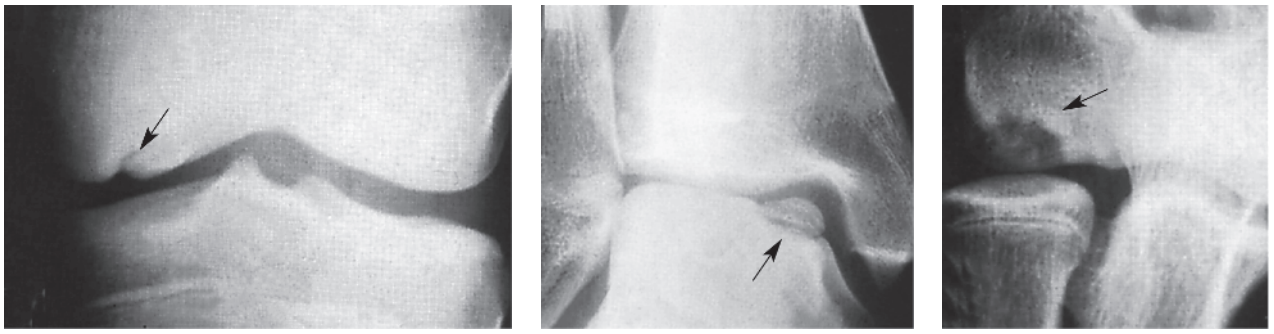
The classic example of this disorder is the condition known as *osteochondritis dissecans*. This occurs typically in young adults, usually men, and affects particular sites: the inner (medial) surface of the medial femoral condyle in the knee, the anteromedial corner of the talus in the ankle, the superomedial part of the femoral head, the humeral capitulum and the head of the second metatarsal bone. (Note that these are all slightly bulbous areas with convex articular surfaces.) The patient usually complains of intermittent pain; sometimes there is swelling and a small effusion in the joint. If the necrotic fragment becomes completely detached, it may cause locking of the joint or unexpected episodes of 'giving way' in the knee or ankle.

Imaging

X-rays must be taken with the joint in the appropriate position to show the affected part of the articular surface in tangential projection. The dissecting fragment is defined by a radiolucent line of demarcation (Figure 6.12). When it separates, the resulting loose body or donor site may be obvious.

The early changes (i.e. before demarcation of the dissecting fragment) are better shown by *MRI*: there is decreased signal intensity in the area around the affected osteochondral segment.

Radionuclide scanning with ^{99m}Tc-HDP shows markedly increased activity in the same area.



(a) (b) (c)

Figure 6.12 Osteochondritis dissecans The osteochondral fragment usually remains in place at the articular surface. The most common sites are (a) the medial femoral condyle, (b) the talus and (c) the capitulum.

Treatment

Treatment in the early stage consists of load reduction and restriction of activity. In young people complete healing may occur, though it can take up to 2 years. For a large joint such as the knee, it is generally recommended that partially detached fragments be pinned back in position after roughening of the base, while completely detached fragments should be pinned back only if they are fairly large and completely preserved. These procedures may be carried out by arthroscopy. If the fragment becomes detached and causes symptoms, it should be fixed back in position or else completely removed.

Treatment of osteochondrosis at the elbow, wrist and metatarsal head is discussed in the relevant chapters.

'Spontaneous' osteonecrosis of the knee ('SONK')

This condition is similar to osteochondritis dissecans of the medial femoral condyle, but it is distinguished by three important features: it appears in *elderly people* (usually women) who are *osteoporotic* and the lesion invariably appears on the *highest part* of the medial femoral condyle. A detailed description is given in Chapter 20.

BONE MARROW OEDEMA SYNDROME

In 1959 an uncommon clinical syndrome characterized by pain and *transient osteoporosis* of one or both hips affecting women in the last trimester of pregnancy was described. It is now recognized that the condition can occur in patients of either sex and at all ages from late adolescence onwards. Although quite distressing at its onset, the condition typically lasts for only 6–12 months, after which the symptoms subside and radiographic bone density is restored. Sometimes successive joints are affected ('regional migratory osteoporosis'), with similar symptoms occurring at each site.

The aetiology of this condition is obscure. The intense activity shown on radionuclide scanning suggests a neurovascular abnormality akin to that of reflex sympathetic dystrophy (RSD). However, there are no trophic changes in the soft tissues and no long-term effects, such as one sees in RSD. The demonstration of diffuse changes on MRI – low signal intensity on T1 weighted images (Figure 6.13) and matching high signal intensity on T2 weighted images – corresponding to the areas of increased scintigraphic activity are characteristic of *bone marrow oedema*, and this is now thought to be an important aspect of transient osteoporosis. What causes it is still unknown.

Similar 'marrow oedema changes' are sometimes seen in areas around typical lesions of osteonecrosis and it has been suggested that transient osteoporosis is due to a sub-lethal, reversible episode of ischaemia associated with reactive hyperaemia in the surrounding bone. Many would disagree with this hypothesis;

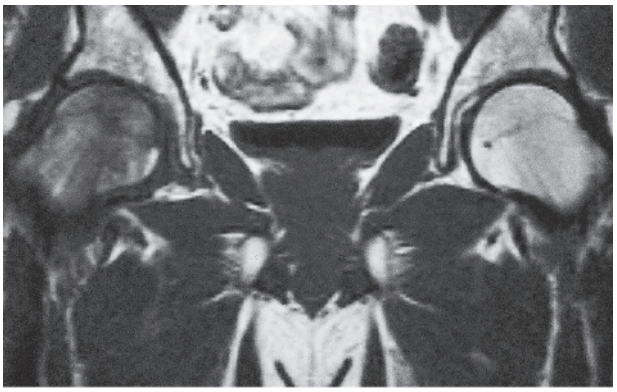


Figure 6.13 Bone marrow oedema MRI showing the typical diffuse area of low signal intensity in the right femoral head in the T1 weighted image.

Table 6.2 Differences between transient bone marrow oedema and osteonecrosis

	Bone marrow oedema	Osteonecrosis
Sex distribution (M:F)	1:3	1:1
Predisposing factors	Pregnancy	Systemic disorders Corticosteroids
Onset	Acute	Gradual
Clinical progress	Self-limiting	Progressive
X-ray	Osteopenia	Sclerosis
Scintigraphy	Increased activity	Reduced activity
MRI	Diffuse changes	Focal changes
Histology	Marrow oedema Minimal bone death	Marrow necrosis Bone necrosis

the most significant differences between the two conditions are listed in Table 6.2. The issue is important because transient osteoporosis has until now been regarded as a reversible disorder which requires only symptomatic treatment while osteonecrosis often calls for operative intervention.

FURTHER READING

Glueck CJ, Freiberg R, Tracy T, *et al.* Thrombophilia and hypofibrinolysis: pathophysiologies of osteonecrosis. *Clin Orthop* 1997b; **334**: 43–56.

Guerra JJ, Steinberg ME. Distinguishing transient osteoporosis from avascular necrosis of the hip. *J Bone Joint Surg* 1995; 77**A**: 616–24.

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7 Metabolic and endocrine bone disorders

Emma Clark & Jon Tobias

Metabolic bone disorders are associated with critical alterations in the regulation of bone formation, bone resorption and distribution of minerals in bone. Clinical features arise from both systemic responses to changes in mineral exchange and local effects of abnormal bone structure and composition. Orthopaedic surgeons deal mainly with the bone abnormalities (e.g. rickety deformities in growing bones or insufficiency fractures in the elderly) but it is important also to be aware of the systemic disorders that may lie behind apparently straightforward 'orthopaedic' defects and to understand the unseen metabolic changes that influence the outcome of many of our surgical interventions.

BONE AND BONES

Understanding of disorders of the musculoskeletal system begins with a basic knowledge of the anatomical structure and physiology of the bones and joints – the framework that supports the body, protects the soft tissues, transmits load and power from one part of the body to another and mediates movement and locomotion.

Figure 7.1 summarizes the different stages of bone development. Embryonic development of the limbs begins with the appearance of the arm buds at about 4 weeks from ovulation and the leg buds shortly afterwards. These at first have the appearance of miniature paddles but by around 5 weeks the finger and toe rays become differentiated. By then primitive skeletal elements and pre-muscle masses have begun to differentiate in the limbs. From about 6 weeks after ovulation the primitive cartilaginous bone-models start to become vascularized and primary ossification centres appear in the chondroid anlage. By now spinal nerves are growing into the limbs. At 7 or 8 weeks cavitation occurs where the joints will appear and during the next few weeks the cartilaginous epiphyseal precursors become vacularized. Between 8 and 12 weeks the primitive joints and synovium become defined.

From then onwards further development goes hand in hand with growth. Bone formation in the cartilaginous model progresses along the diaphysis but the epiphyseal ends remain unossified until after birth. The entire sequence has been aptly summarized as *condensation* → *chondrification* → *ossification*.

Soon after birth secondary ossification centres begin to appear in the still cartilaginous ends of the tubular bones, a process that will occur during childhood in all the *endochondral bones* (bones formed in cartilage). By then each bone end is defined as an *epiphysis*, the growth plate between the epiphysis and the rest of the bone as the *physis*, the adjacent end of the long bone the metaphysis, and the shaft as the *diaphysis*.

Longitudinal growth continues up until late adolescence, by a process of *endochondral bone formation*, whereby cartilage formed beneath the growth plate becomes calcified to produce the primary spongiosa, which is then replaced by the secondary spongiosa following vascular invasion. Cessation of longitudinal growth is heralded by the growth plate becoming ossified, resulting in fusion of the epiphysis to the metaphysis.

An increase in bone circumference occurs by a different process, namely *periosteal bone formation*. The latter involves small generative cuboidal cells in the deepest layer of the periosteum. In contrast to longitudinal growth, periosteal bone formation does not involve the intermediary formation of cartilage. Whereas longitudinal growth ceases once growth plates have fused, periosteal bone formation may continue life-long, depending on the anatomical site.

Where bones connect with each other, i.e. at the joints, the contact surfaces remain cartilaginous. *Diarthrodial joints* (freely movable, synovial joints) comprise *hyaline cartilage*, which is ideally suited to permit low-friction movement and to accommodate both compressive and tensile forces. In *synarthroses*,

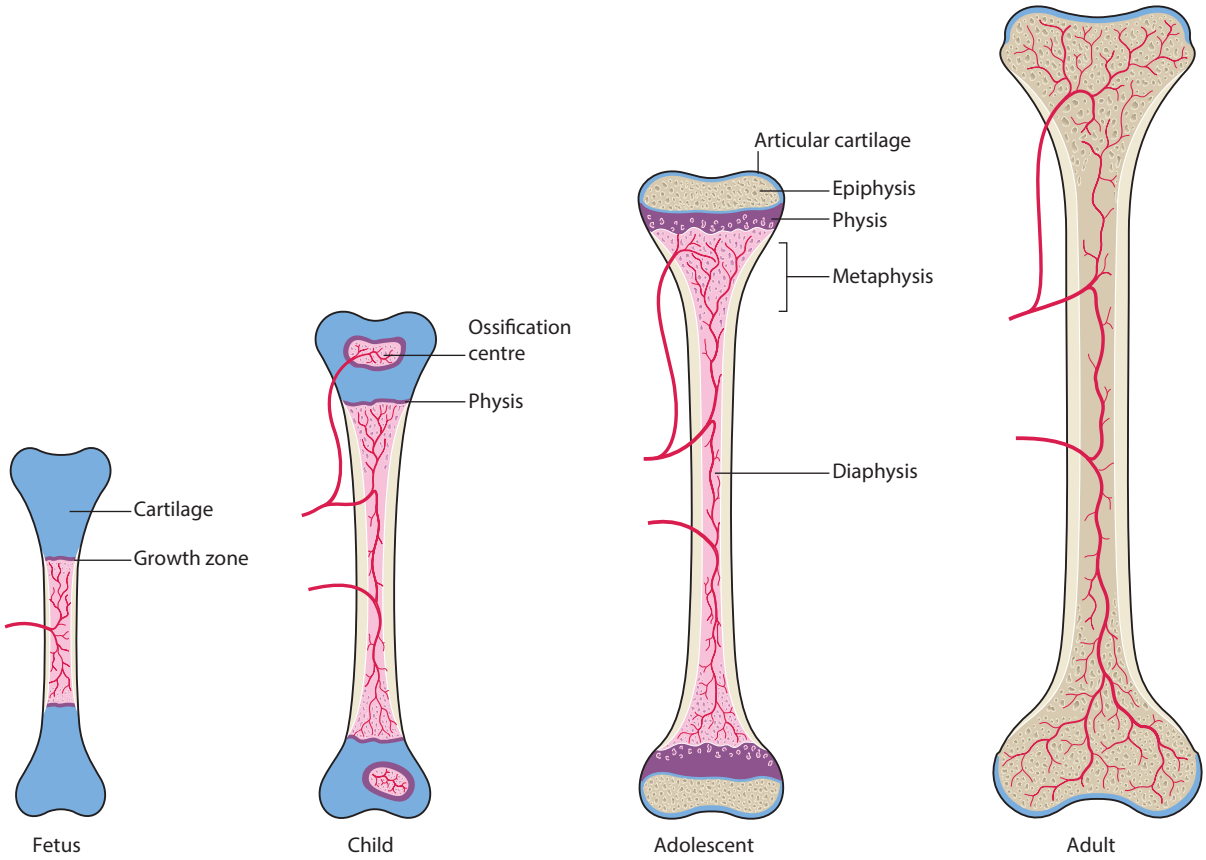


Figure 7.1 Stages in bone development Schematic representation of the stages in the development of a tubular bone showing the progress from diaphyseal ossification, through endochondral growth at the physis and increase in width of the diaphysis by sub-periosteal appositional bone formation.

where greater resistance to shearing forces is needed, the interface usually consists of tough *fibrocartilage* (e.g. the pubic symphysis).

BONE COMPOSITION AND STRUCTURE

Bones as structural organs have three main functions: *support*, *protection* and *leverage*. They support every part of the body in a wide variety of positions and load-bearing; they protect important soft tissues such as the brain, the spinal cord, the heart and the lungs; they provide space and structural support for cells involved in haematopoiesis; and they act as jointed levers that facilitate a range of movements.

Bone as tissue has an equally important role as a mineral reservoir which helps to regulate the composition – and in particular the calcium ion concentration – of the extracellular fluid. For all its solidity, it is in a continuous state of flux, its internal shape and structure changing from moment to moment in concert with the normal variations in mechanical function and mineral exchange.

All modulations in bone structure and composition are brought about by cellular activity, which is regulated by hormones and local factors; these agents, in turn, are controlled by alterations in mineral ion concentrations. Disruption of this complex interactive system results in systemic changes in mineral metabolism and generalized skeletal abnormalities.

BONE COMPOSITION

Bone consists of a largely collagenous matrix which is impregnated with mineral salts and populated by cells (*osteoblasts* and *osteoclasts*).

The matrix

Type I collagen fibres, derived from tropocollagen molecules produced by osteoblasts, make up over 80% of the unmineralized matrix. They consist of collagen fibrils comprising a triple helix, with the overall structure stabilised by cross-linking between adjacent fibrils. Collagen is responsible for the skeleton's tensile strength. It also serves as a scaffold on which the mineral component – crystalline hydroxyapatite – is deposited.

Other non-collagenous proteins exist in small amounts in the mineralized matrix – mainly *sialoproteins* (*osteopontin*), *osteonectin*, *osteocalcin* (*bone Gla protein*) and *alkaline phosphatases*. Their functions have not been fully elucidated but they appear to be involved in the regulation of bone cells and matrix mineralization. Osteocalcin is produced only by osteoblasts and its concentration in the blood is, to some extent, a measure of osteoblastic activity. Bone matrix also has large concentrations of local regulatory factors including TFG-beta, which stimulate osteoblast differentiation, and may act as coupling factors whereby degradation of bone matrix during resorption leads to release of factors that recruit osteoblasts for the subsequent formation phase.

Bone mineral

Almost half the bone volume is mineral matter – mainly *calcium* and *phosphate* in the form of *crystalline hydroxyapatite* which is laid down in osteoid at the calcification front. The interface between bone and osteoid can be labelled by administering tetracycline, which is taken up avidly in newly mineralized bone and shows as a fluorescent band on ultraviolet light microscopy. In mature bone the proportions of calcium and phosphate are constant and the molecule is firmly bound to collagen. It is important to appreciate that in life *'demineralization' of bone occurs only by resorption of the entire matrix.*

While the collagenous component lends tensile strength to bone, the crystalline mineral enhances its ability to resist compression.

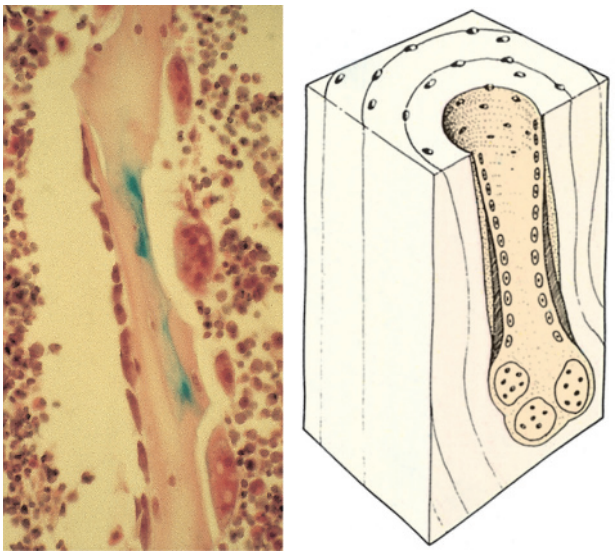
Unmineralized matrix is known as *osteoid*; in normal life it is seen only as a thin layer on surfaces where active new bone formation is taking place, but the proportion of osteoid to mineralized bone increases significantly in rickets and osteomalacia.

Bone cells

There are of three types of bone cell: osteoblasts, osteocytes and osteoclasts.

Osteoblasts Osteoblasts are concerned with bone formation and osteoclast activation. They are derived from mesenchymal precursors in the bone marrow and the deep layer of the periosteum. Differentiation is controlled by a number of interacting growth factors, including bone morphogenetic proteins (BMPs).

Mature osteoblasts form rows of small (20 μm) mononuclear cells along the free surfaces of trabeculae and haversian systems where *osteoid* is laid down prior to calcification (see Figure 7.2). They are rich in alkaline phosphatase (ALP) and are responsible for the production of type I collagen as well as the non-collagenous bone proteins and for the mineralization of bone matrix (Peck and Woods, 1988). Stimulated



(a) (b)

Figure 7.2 Bone cells (a) Histological section showing a trabecula lined on one surface by excavating osteoclasts and on the other surface by a string of much smaller osteoblasts. These two types of cell, working in concert, continuously remodel the internal bone structure. (b) In compact bone the osteoclasts burrow deeply into the existing bone, with the osteoblasts following close behind to re-line the cavity with new bone.

by PTH, they play a critical role in the initiation and control of osteoclastic activity.

At the end of each bone remodelling cycle the osteoblasts either remain on the newly formed bone surface as quiescent lining cells or they become embedded in the matrix as 'resting' osteocytes. During advanced ageing their numbers decrease.

Osteocytes Osteocytes are osteoblasts that become entombed in their own matrix. Lying in their bony lacunae, they communicate with each other and with the surface lining cells by slender cytoplasmic processes. They are sensitive to mechanical stimuli and communicate information and changes in stress and strain to the active osteoblasts (Skerry et al., 1989) which can then modify their osteogenic activity accordingly. One of the principle ways they achieve this is thought to be through secretion of sclerostin (see 'Bone formation' below). Osteocytes also play an important role in mineral homeostasis, particularly phosphate homeostasis by virtue of the fact that they are the primary source of the phosphaturic factor, fibroblast growth factor (FGF) 23 (see FGF23 in 'Mineral exchange' below).

Osteoclasts These large multinucleated cells are the principal mediators of bone resorption. They develop from mononuclear precursors in the haemapoietic marrow (the same lineage as macrophages) under the influence of local osteoblastic stromal cells

that generate an essential osteoclast differentiating factor – *receptor activator of nuclear factor-κβ ligand (RANKL)* – which binds with a specific receptor site (RANK) on the osteoclast precursors.

Mature osteoclasts have a foamy appearance, due to the presence of numerous vesicles in the cytoplasm. In response to appropriate stimuli the osteoclast forms a sealed attachment to a bone surface, where the cell membrane develops a ruffled border within which bone resorption takes place. This process, and the important interactions between RANKL and RANK, are discussed further in 'Bone resorption' below.

Following resorption of the bone matrix, the osteoclasts are left in shallow excavations – Howship's lacunae – along free bone surfaces (see Figure 7.2). By identifying these excavations one can distinguish 'resorption surfaces' from the smooth 'formation surfaces' or 'resting surfaces' in histological sections.

Bone structure

When viewed under polarized light microscopy, bone tissue can be seen to have been laid down in a haphazard manner known as *woven bone*, or in parallel sheets known as *lamellar bone*. Woven bone occurs where bone tissue is formed in its immature state, as in the early stages of fracture healing, where it acts as a temporary weld before being replaced by mature bone, or when formed in pathological states such as infection or Paget's disease.

In lamellar bone, collagen fibres are arranged parallel to each other to form multiple layers (or laminae) with the osteocytes lying between the lamellae. Unlike woven bone, which is laid down in fibrous tissue, lamellar bone forms only on existing bone surfaces.

Lamellar bone exists in two structurally different forms, *compact (cortical) bone* and *cancellous (trabecular) bone.*

Compact bone

Compact (cortical) bone is dense to the naked eye. It is found where support matters most: the outer walls of all bones but especially the shafts of tubular bones, and the subchondral plates supporting articular cartilage. It is made up of compact units – haversian systems or osteons – each of which consists of a central canal (the haversian canal) containing blood vessels, lymphatics and nerves and enclosed by closely packed, more or less concentric lamellae of bone. Between the lamellae lie osteocytes, bedded in lacunae which appear to be discrete but which are in fact connected by a network of fine canaliculae. The haversian canal offers a free surface lined by bone cells; its size varies, depending on whether the osteon is in a phase of resorption or formation. During resorption osteoclasts eat into the surrounding lamellae and the canal widens out; during formation osteoblasts lay down new lamellae on the inner surface and the canal closes down again.

Cancellous bone

Cancellous (trabecular) bone has a honeycomb appearance; it makes up the interior meshwork of all bones and is particularly well developed in the ends of the tubular bones and the vertebral bodies. The structural units of trabecular bone are flattened sheets or spars that can be thought of as unfolded osteons. Three-dimensionally the trabecular sheets are interconnected (like a honeycomb) and arranged according to the mechanical needs of the structure, the thickest and strongest along trajectories of compressive stress and the thinnest in the planes of tensile stress. The interconnectedness of this meshwork lends added strength to cancellous bone beyond the simple effect of tissue mass. The spaces between trabeculae – the 'opened-out' vascular spaces – contain the marrow and fine sinusoidal vessels that course through the tissue, nourishing both marrow and bone.

Trabecular bone is obviously more porous than cortical bone. Though it makes up only one-quarter of the total skeletal mass, it provides two-thirds of the total bone surface. Add to this the fact that it is covered with marrow and it is easy to understand why trabecular bone acts as the principle reservoir for calcium in the body, and why the effects of metabolic disorders are usually seen first here.

Haversian system

Bones vary greatly in size and shape. At the most basic level, however, they are similar: compact on the outside and spongy on the inside. Their outer surfaces (except at the articular ends) are covered by a tough *periosteal membrane*, the deepest layer of which consists of potentially bone-forming cells. The inner, endosteal, surfaces are irregular and lined by a fine *endosteal membrane* in close contact with the marrow spaces.

The osteonal pattern in the cortex is usually depicted from two-dimensional histological sections. A three-dimensional reconstruction would show that the *haversian canals* are long, branching channels running in the longitudinal axis of the bone (see Figure 7.3). These connect extensively with each other and with the endosteal and periosteal surfaces by smaller channels (*Volkmann canals*). In this way the vessels in the haversian canals form a rich anastomotic network between the medullary and periosteal blood

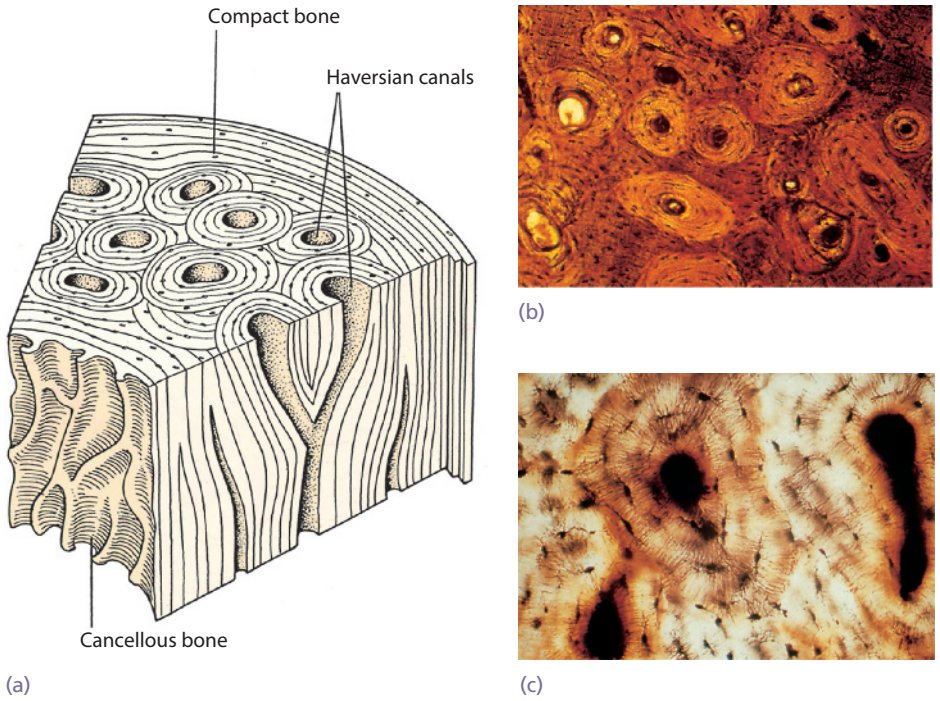


Figure 7.3 The haversian systems (a) A schematic diagram representing a wedge taken from the cortex of a long bone. It shows the basic elements of compact bone: densely packed osteons, each made up of concentric layers of bone and osteocytes around a central haversian canal which contains the blood vessels; outer laminae of sub-periosteal bone; and similar laminae on the interior surface (endosteum) merging into a lattice of cancellous bone. (b,c) Low- and high-power views showing the osteons in various stages of formation and resorption.

supply (see Figure 7.4). Blood flow in this capillary network is normally centrifugal – from the medullary cavity, which is fed by a nutrient artery, outwards. The outermost layers of the cortex are normally also supplied by periosteal vessels; if the medullary vessels are blocked or destroyed, the periosteal circulation can take over entirely and the direction of blood flow is reversed.

BONE DEVELOPMENT AND GROWTH

Bones develop in two different ways: by ossification of a prior cartilage model or framework (*endochondral ossification*) and by direct *intramembranous ossification*.

Endochondral ossification

This is the usual manner in which tubular bones develop, and it is also the process involved in longitudinal growth of bones. At birth the cartilage model is complete and ossification has already begun at the centre of the diaphysis. After secondary ossification of the epiphyseal ends has begun, further growth in length takes place in the still-cartilaginous zone between the extending area of diaphyseal bone and the epiphysis. In this way the still-cartilaginous zone between the ossifying diaphysis and the epiphysis gradually narrows down but does not disappear until late adolescence. This actively growing cartilage disc is called the *physis* (growth plate), situated between the epiphysis and the diaphysis.

The growth plate consists of four distinct zones (see Figure 7.5). Co-extensive with the epiphysis is a *zone of resting chondrocytes* in haphazard array. This merges into a *proliferative zone* in which the chondrocytes are lined up longitudinally; being capable of interstitial growth, they add progressively to the overall length of the bone. The older cells in this zone (those 'left behind' nearest the advancing new bone of the diaphysis) gradually enlarge and constitute a *hypertrophic zone*. Close to the interface between cartilage and bone the cartilage becomes calcified (probably with the involvement of alkaline phosphatase produced by the hypertrophic cells); this *zone of calcified cartilage* finally undergoes osteoclastic resorption and, with the ingrowth of new blood vessels from the metaphysis, ossification. Woven bone is laid down on the calcified scaffolding and this in turn is replaced by lamellar bone which forms the newest part of the bone shaft, now called the *metaphysis*.

It should be noted that a similar process takes place in the late stage of fracture repair.

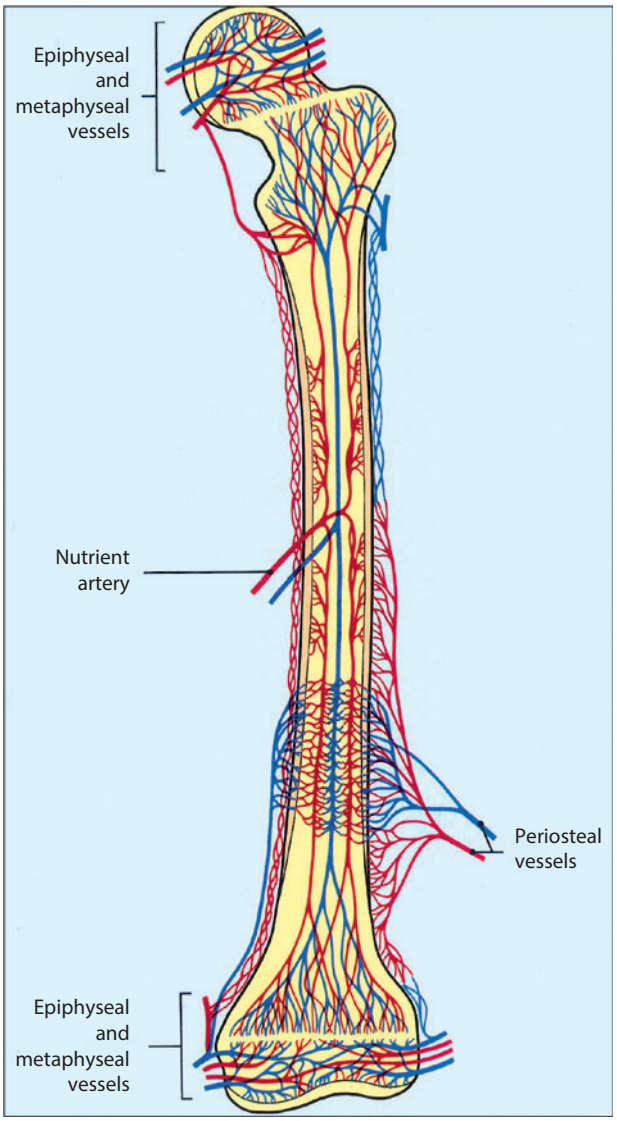


Figure 7.4 Blood supply to bone Schematic presentation of blood supply in tubular bones. (Reproduced from Bullough (1985). By kind permission of Dr Peter G Bullough and Elsevier.)

Intramembranous ossification

With the growth in length, the bone also has to increase in circumference and, since a tubular bone is an open cylinder, this inevitably demands that the medullary cavity increase in size proportionately. New bone is added to the outside by direct ossification at the deepest layer of the periosteum where mesenchymal cells differentiate into osteoblasts (*intramembranous, or 'appositional', bone formation*) and old bone is removed from the inside of the cylinder by osteoclastic *endosteal resorption*.

Intramembranous periosteal new bone formation also occurs as a response to periosteal stripping due to trauma, infection or tumour growth, and its appearance is a useful radiographic pointer.

Bone modelling

Bone modelling describes the process by which changes in the overall size and shape of bone are accomplished. Though primarily occurring during longitudinal growth, outward growth via intramembranous bone formation may continue long after the former has ceased through closure of growth plates. Whereas the outer dimension of long bones continues to expand during the third decade as part of the process of peak bone mass achievement, at certain sites such as the femoral neck, expansion may continue lifelong (Duan et al., 2003). In contrast to bone modelling which is driven by bone formation, bone resorption occurs as a secondary process intended to maintain overall shape, as in the maintenance of the flared ends of bone during longitudinal growth.

Peak bone mass

Dual energy X-ray absorptiometry (DXA) scans are used to measure the amount of skeletal tissue by evaluation of calcium content, termed bone mass (see 'Measurement of bone mass' below). Bone mass increases during childhood, in keeping with the increase in bone size which is characterized by acceleration around puberty, followed by a less rapid gain until the cessation of linear growth. Once longitudinal growth has ceased, further skeletal consolidation continues, with peak bone mass reached in the late twenties (Figure 7.6). Peak bone mass remains stable into later adulthood, until processes contributing to bone loss take effect, particularly oestrogen deficiency after the menopause in women (see below). Skeletal consolidation involves a number of factors including periosteal expansion and modelling, secondary mineralization of newly formed bone, and in-filling of haversian canals due to reduced bone remodelling after longitudinal growth has ceased.

By the end of bone growth, mean bone mass is about 5–10% greater in young men than in young women, due mainly to increased appositional bone formation when androgen levels rise after puberty (Seeman, 2003). The level of peak bone mass attained is thought to be important in terms of risk of fractures and osteoporosis in later life. Of the many genetic factors that have been identified as influencing bone mineral density (BMD) and fracture risk (Estrada et al., 2012), several of these are likely to act by influencing peak bone mass. Environmental factors are probably also important, such as physical activity and diet. Disorders affecting peak bone mass acquisition have important effects in terms of subsequent fracture risk, including anorexia nervosa, primary amenorrhea, and inflammatory paediatric disorders such as inflammatory bowel disease.

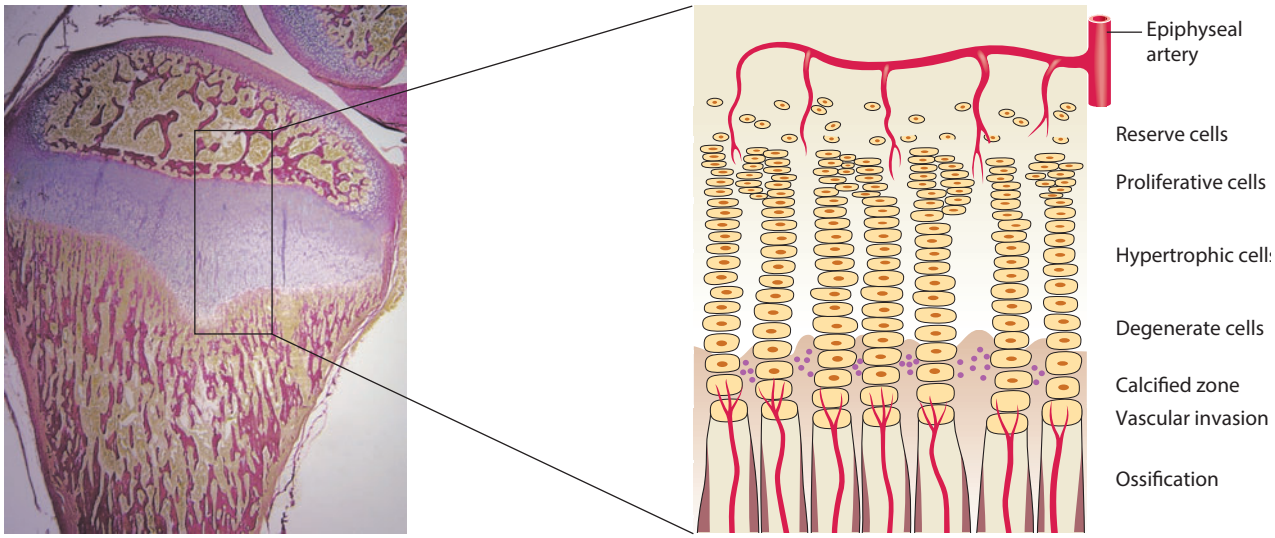


Figure 7.5 Endochondral ossification Histological section of a growing endochondral bone with a schematic figure showing the layers of the growth disc (physis). (Reproduced from Bullough (1985). Second figure by kind permission of Dr Peter G Bullough and Elsevier.)

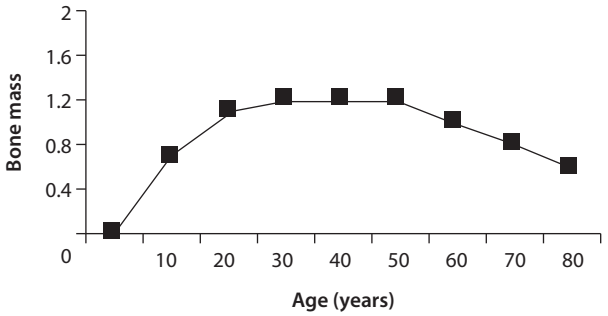


Figure 7.6 Bone mineral density over the life course Graph showing change in bone mass in women from birth to mid 80s.

BONE MAINTENANCE

As well as contributing to bone growth and modelling, bone formation and resorption continue as a lifelong process. These two processes are carefully coordinated as part of the remodelling cycle which serves to maintain bone integrity. They are also activated during repair following injury, for example after a fracture.

BONE RESORPTION

Bone resorption is carried out by the *osteoclasts* under the influence of stromal cells (including *osteoblasts*) and both local and systemic activators. Though it has long been known that parathyroid hormone (PTH) promotes bone resorption, osteoclasts have no receptor for PTH. The hormone acts indirectly through its effect on 1,25- $(OH)_2D_3$ and osteoblasts.

Proliferation of osteoclastic progenitor cells requires the presence of an osteoclast differentiating factor produced by the stromal osteoblasts after stimulation by, for example, PTH, glucocorticoids or pro-inflammatory cytokines. It is now known that this 'osteoclast differentiating factor' is the receptor activator of nuclear factor-ќβ ligand (RANKL for short), and that it has to bind with a RANK receptor on the osteoclast precursor in the presence of a macrophage colony-stimulating factor (M-CSF) before full maturation and osteoclastic resorption can begin. Osteoprotegerin (OPG), which is expressed by osteoblasts, is able to inhibit the differentiation of osteoclast precursors by preferentially binding with RANKL (acting as a 'decoy' receptor) and so reducing bone resorption by preventing RANKL from binding with its receptor on the osteoclast precursor. Pharmacological inhibition with denosumab which is a monoclonal RANKL antibody, leads to marked suppression of bone resorption and has been found to be a useful therapeutic agent for conditions like osteoporosis associated with generalized increases in resorption (see below). Use of denosumab to inhibit the RANK/RANKL/OPG system is also helpful in treating disorders associated with localized increases in bone resorption, including bone erosions associated with rheumatoid arthritis, Paget's disease of bone, skeletal metastases particularly from breast and prostate cancer, and multiple myeloma.

Before mature osteoclasts start to resorb bone, osteoblasts are thought to first 'prepare' the resorption site by removing osteoid from the bone surface while

other matrix constituents act as osteoclast attractors. During resorption, each osteoclast forms a sealed attachment to the bone surface where the cell membrane folds into a characteristic ruffled border within which hydrochloric acid and proteolytic enzymes are secreted. At this low pH, minerals in the matrix are dissolved and the organic components are destroyed by lysosomal enzymes. Cathepsin K (organic matrix degradation) and carbonic anhydrase II, the CLC-7 chloride channel and the ATPase proton pump (hydrochloric acid secretion), are all essential for this process. Calcium and phosphate ions are absorbed into the osteoclast vesicles from where they pass into the extracellular fluid and, ultimately, the bloodstream. Defects in osteoclast function following mutations affecting the function of these proteins lead to excess accumulation of bone, skeletal fragility and bone deformity, as occurs in osteopetrosis and, in the case of cathepsin K deficiency, pycnodysostosis (Figure 7.7). The cathepsin K inhibitor odanacatib is currently in development as a possible new anti-resorptive drug to treat osteoporosis (Brixen et al., 2013).

In cancellous bone excess osteoclast activity results in thinning and sometimes actual perforation of existing trabeculae. In cortical bone the cells either enlarge an existing haversian canal or burrow into the compact bone to create a *cutting cone*. During bone resorption, products from bone breakdown are released into the circulation. Release of calcium and phosphate contributes to overall exchange of these minerals.

Bone formation

Bone formation is carried out by teams of osteoblasts, which are recruited to a bone surface or haversian system and proceed to secrete osteoid, composed of type I collagen fibrils, which becomes deposited on the adjacent bone surface. Bone mineral, in the form of hydroxyapatite crystals, subsequently become deposited in spaces between collagen fibrils. Though essentially a passive process, this is dependent on an adequate calcium × phosphate product in extracellular fluid.

Whereas osteoid is rapidly mineralized following its synthesis, a process of secondary mineralization takes place after bone formation is complete. The latter process is time-dependent, and its extent depends on skeletal maturity. Skeletal maturity also influences the degree of collagen cross-link formation, a process whereby adjacent type I collagen fibrils undergo further protein binding to enhance tensile strength. The amino acid sequence where these cross-links occur, termed N-terminal telopeptide, are largely specific to bone, and their level in plasma following release into the circulation following bone resorption forms the basis of clinical measurement of bone turnover.

Like bone resorption, bone formation is regulated by a combination of systemic and locally produced factors acting to promote osteoblast differentiation. Arguably the most important local factors in regulating osteoblast differentiation are the bone morphogenetic proteins (BMPs) and the Wnt signalling system, both of which comprise complex systems of multiple ligands, cell surface receptors, intracellular signalling pathways and endogenous inhibitors. Recognition of the role of these systems in bone formation has helped in understanding rare bone diseases characterized by excessive bone formation. For example, activating mutations of the BMP receptor Activin A receptor type I (ACVR1) causes the bone disease *fibrous ossificans progressiva*, which is associated with progressive transformation of muscle into bone. BMPs are produced commercially to enhance local osteogenesis, for example to treat non-union of fractures or fuse bones (Rihn et al., 2008).

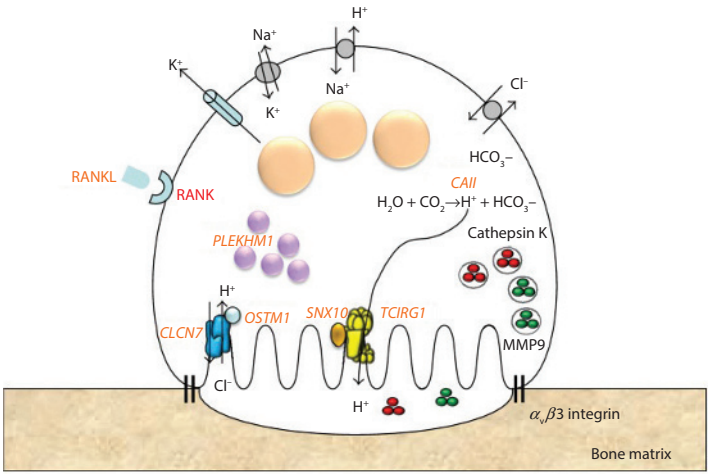


Figure 7.7 Mechanisms involved in osteoclastic bone resorption Cellular localization and protein involved in osteoclast differentiation and function. The genes mutated in human osteopetrosis are red in bold. Figure from review by Coudert et al. (2015).

In terms of the Wnt pathway, activating mutations of the Wnt receptor LRP5 lead to marked elevations in bone mass (Whyte et al., 2004). The Wnt ligand sclerostin, the production of which by osteocytes is suppressed by mechanical strain, is thought to play an important role in regulating bone cell activity in response to skeletal loading. Loss of function mutations in sclerostin lead to sclerosteosis, a rare familial disorder characterized by marked elevations in bone mass, and complications such as cranial nerve palsies arising from bone overgrowth. The sclerostin antibody romosozumab is currently in development as a new anabolic treatment for osteoporosis (McClung et al., 2014).

Bone remodelling

Osteoblast and osteoclast activity are coordinated during bone remodelling, which describes a process by which previously formed bone is removed and then replaced in a specific sequence comprising the bone remodelling cycle. This process, which determines the internal architecture of bone, occurs not only during growth but throughout life. Bone remodelling serves several crucial purposes: 'old bone' is continually replaced by 'new bone' and in this way the skeleton is protected from the excess accumulation of fatigue damage and the risk of stress failure; bone turnover is sensitive to the demands of function, and trabeculae are fashioned (or refashioned) in accordance with the stresses imposed upon the bone, the thicker and stronger trabeculae following the trajectories of compressive stress and the finer trabeculae lying in the planes of tensile stress; besides, the maintenance of calcium homeostasis requires a constant turnover of the mineral deposits which would otherwise stay locked in bone.

At each *remodelling site* work proceeds in an orderly sequence (see Figure 7.8). Prompted by the osteoblasts, osteoclasts gather on a free bone surface and proceed to excavate a cavity. After 2–4 weeks resorption ceases; the osteoclasts undergo apoptosis and are phagocytosed. There is a short quiescent period, then the excavated surface is covered with osteoblasts and for the next 3 months osteoid is laid down and mineralized to leave a new 'packet' of bone (or *osteon*). The entire *remodelling cycle* takes from 4 to 6 months and at the end the boundary between 'old' and 'new' bone is marked by a histologically identifiable 'cement line'.

The osteoblasts and osteoclasts participating in each cycle of bone turnover work in concert, together acting as a *bone remodelling unit* (of which there are more than a million at work in the adult skeleton at any time). Resorption and formation are *coupled*, the one ineluctably following the other. Systemic hormones and local growth factors are involved in coordinating this process; indeed, it is likely that PTH and

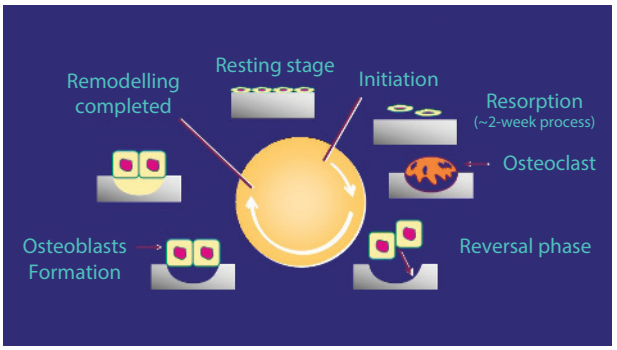


Figure 7.8 The remodelling cycle On activation of bone resorption the surface layer of un-mineralized osteoid is removed and bone resorption by osteoclasts begins, the membrane of the osteoclasts taking on a ruffled appearance at the site of active bone resorption. When bone resorption is complete, the cement line is laid down during the reversal phase. The unmineralized osteoid synthesized by osteoblasts fills the resorption cavity. The osteoid is then mineralized, the bone surface finally being covered by lining cells and a thin layer of unmineralized osteoid. (Adapted from Bone et al. (2000).)

$1,25\text{-}(OH)_2D$ are involved in initiating both formation and resorption. This ensures that (at least over the short term) a balance is maintained, though at any moment and at any particular site one or other phase may predominate. Many other factors influence bone remodelling, including other systemic hormones such as sex steroids, dietary factors and mechanical stresses. Together these impinge on local regulatory factors described above that regulate osteoblast and osteoclast activity.

In the long term, change does occur. The annual rate of bone turnover in healthy adults has been estimated as 4% for cortical bone and 25% for trabecular bone (Parfitt, 1988). The rate may be increased or decreased either by alterations in the number of remodelling units at work or by changes in the remodelling time. During the first half of life formation slightly exceeds resorption and bone mass increases; in later years resorption exceeds formation and bone mass steadily diminishes. Connecting trabeculae may be perforated or lost, further diminishing bone strength and increasing the likelihood of fragility fractures. Rapid bone loss is usually due to excessive resorption rather than diminished formation.

Mineral exchange

Calcium and phosphorus have an essential role in a wide range of physiological processes. Over 98% of the body's calcium and 85% of its phosphorus are tightly packed as hydroxyapatite crystals in bone and capable of only very slow exchange. A small amount

exists in a rapidly exchangeable form, either in partially formed crystals or in the extracellular fluid and blood where their concentration is maintained within very narrow limits by an efficient homeostatic mechanism involving intestinal absorption, renal excretion and mineral exchange in bone.

A number of essential metabolic processes require extracellular calcium levels to be maintained within a very narrow range, which is achieved through the action of parathyroid hormone (PTH). Transient alterations in blood levels are rapidly compensated for by changes in renal tubular absorption. A more persistent fall in extracellular calcium concentration can be accommodated by increasing bone resorption. Exchange of extracellular with skeletal calcium is also strongly dependent on phosphate levels. For example, any tendency for the extracellular solubility calcium × phosphate product to increase due to a rise in phosphate stimulates deposition of both these ions in bone as hydroxyapatite crystals, leading to a reciprocal decline in calcium. Phosphate levels are regulated by a separate hormone, FGF23.

Calcium

Calcium is essential for normal cell function and physiological processes such as blood coagulation, nerve conduction and muscle contraction. An uncompensated fall in extracellular calcium concentration (hypocalcaemia) may cause tetany; an excessive rise (hypercalcaemia) can lead to depressed neuromuscular transmission.

The main sources of calcium are dairy products, green vegetables and soya (or fortified foods). The recommended daily intake for adults is 800–1000 mg (20–25 mmol), and ideally this should be increased to 1200 mg during pregnancy and lactation. Children need less, about 200–400 mg per day.

About 50% of the dietary calcium is absorbed (mainly in the upper gut) but much of that is secreted back into the bowel and only about 200 mg (5 mmol) enters the circulation. The normal concentration in plasma and extracellular fluid is 2.2–2.6 mmol/L (8.8–10.4 mg/dL). Much of this is bound to albumin as well as other proteins; about half (1.1 mmol) is ionized and effective in cell metabolism and the regulation of calcium homoeostasis.

Calcium absorption in the intestine is promoted by vitamin D metabolites, particularly 1,25-$(OH)_2$ vitamin D, and requires a suitable calcium/phosphate ratio. Absorption is inhibited by excessive intake of phosphates (common in soft drinks), oxalates (found in tea and coffee), phytates (chapati flour) and fats, by the administration of certain drugs (including corticosteroids) and in malabsorption disorders of the bowel.

Urinary excretion varies between 2.5 and 5 mmol (100–200 mg) per 24 hours. If the plasma ionized calcium concentration falls, PTH is released and causes (a) increased renal tubular reabsorption of calcium and reduced renal tubular reabsorption of phosphate and (b) a switch to increased 1,25-$(OH)_2$ vitamin D production and enhanced intestinal calcium absorption. If the calcium concentration remains low, calcium is drawn from the skeleton by increased bone resorption through the influence of PTH.

Phosphorus

Apart from its role (with calcium) in the composition of hydroxyapatite crystals in bone, phosphorus is needed for many important metabolic processes, including energy transport and intracellular cell signalling. It is abundantly available in the diet and is absorbed in the small intestine, more or less in proportion to the amount ingested; however, absorption is reduced in the presence of antacids such as aluminium hydroxide, which binds phosphorus in the gut. Phosphate excretion is extremely efficient, but 90% is reabsorbed in the proximal tubules. Plasma concentration – almost entirely in the form of ionized inorganic phosphate (Pi) – is normally maintained at 0.9–1.3 mmol/L (2.8–4.0 mg/dL).

The solubility product of calcium and phosphate is held at a fairly constant level; any increase in the one will cause the other to fall. Pi is regulated by the phosphaturic hormone FGF23. The other main regulators of plasma phosphate concentration are PTH and 1,25-$(OH)_2$D. If Pi rises abnormally, a reciprocal fall in calcium concentration will stimulate PTH secretion which in turn will suppress urinary tubular reabsorption of Pi, resulting in increased Pi excretion and a fall in plasma Pi. High Pi levels also result in diminished 1,25-$(OH)_2$D production, causing reduced intestinal absorption of phosphorus.

Magnesium

Magnesium plays a small but important part in mineral homeostasis. The cations are distributed in the cellular and extracellular compartments of the body and appear in high concentration in bone. Magnesium is necessary for the efficient secretion and peripheral action of parathyroid hormone. Thus, if hypocalcaemia is accompanied by hypomagnesaemia, it cannot be fully corrected until normal magnesium concentration is restored.

Vitamin D

Vitamin D, through its active metabolites, is principally concerned with calcium absorption and transport and (acting together with PTH) bone remodelling. Target organs are the small intestine and bone. There are two main forms: vitamin D2 (ergocalciferol), which

is synthetic, and vitamin D3 (cholecalciferol), which is naturally occurring. Whether these two forms are equipotent clinically is a matter of ongoing debate.

Vitamin D2 is exclusively obtained from supplements, whereas vitamin D3 may be obtained from supplements, from the diet, or indirectly by the action of ultraviolet light on the precursor 7-dehydrocholesterol in the skin. For people who do not receive adequate exposure to bright sunlight, the recommended daily requirement for adults is 400–800 IU (10–20 μg) per day – the higher dose for those over 70 years of age. In most countries this is obtained mainly from exposure to sunlight; those who lack such exposure are likely to suffer from vitamin D deficiency unless they take dietary supplements.

Vitamin D itself is inactive. Conversion to active metabolites takes place first in the liver by 25-hydroxylation to form 25-hydroxycholecalciferol [25-OHD], and then in the kidneys by further hydroxylation to 1,25-dihydroxycholecalciferol [1,25-$(OH)_2$D] (calcitriol) (see Figure 7.9).

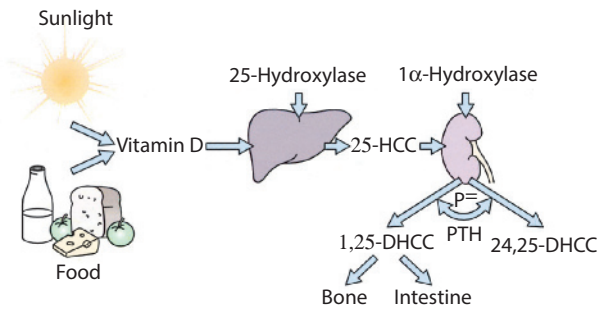


Figure 7.9 Vitamin D metabolism The active vitamin D metabolites are derived either from the diet or by conversion of precursors when the skin is exposed to sunlight. The inactive 'vitamin' is hydroxylated, first in the liver and then in the kidney, to form the active metabolites 25-HCC and 1,25-DHCC.

1,25-$(OH)_2$D is the active hormone, acting to increase serum calcium levels in response to calcium stress (see Table 7.1). The one-alpha hydroxylase enzyme responsible for the latter conversion is activated mainly by parathyroid hormone (PTH), but also by other hormones (including oestrogen and prolactin) and a low concentration of phosphate. If the PTH concentration falls and phosphate remains high, 25-OHD is converted alternatively to 24,25-$(OH)_2$D which is inactive. During negative calcium balance, production switches to 1,25-$(OH)_2$D in response to PTH secretion; the increased 1,25-$(OH)_2$D then helps to restore the serum calcium concentration.

1,25-$(OH)_2$D acts on the *lining cells of the small intestine* to increase the absorption of calcium and phosphate. *In bone* it promotes osteoclastic resorption; it also enhances calcium transport across the osteoblast cell membrane and indirectly assists with osteoid mineralization.

Some anti-epileptic drugs interfere with the vitamin D metabolic pathway and may cause vitamin D deficiency.

Although the concentration of active metabolites can be measured in serum samples, the best indicator of vitamin D status is 25-OHD concentration (serum 1,25-$(OH)_2$D has a half-life of only 15 hours and is therefore not as good an indicator). Generally, 25-OHD levels of >50 nmol/L are considered sufficient, though some advocate higher levels (e.g. >75 nmol/L). Levels below 50 nmol/L are frequently observed, particularly during winter months at more northern latitudes, and in high risk populations such as older and housebound individuals.

Parathyroid hormone

Parathyroid hormone (PTH) is the major regulator of extracellular calcium concentration, acting on the

Table 7.1 Regulation of mineral metabolism by PTH and 1,25-$(OH)_2$ vitamin D

	Source	Secretion increased by…	Secretion decreased by…	Effects on intestine	Effects on kidney	Effects on bone	Effect on serum Ca and Pi
PTH	Parathyroid gland	Fall in serum Ca	Rise in serum Ca Increase in 1,25-$(OH)_2$D	No direct effect but Ca absorption increased through 1,25-$(OH)_2$D	Increase in 1,25$(OH)_2$D Increased resorption of Ca Increased excretion of phosphate	No direct effect but resorption increased via action on 1,25-$(OH)_2$D	Rise in serum Ca Fall in serum Pi
1,25$(OH)_2$ vit D	Kidney tubule	Fall in serum Ca Fall in serum Pi Rise in serum PTH	Rise in serum Ca Rise in serum Pi Fall in PTH	Increased absorption of Ca Increased absorption of phosphate		Osteoclasto-genesis and increased bone resorption	Rise in serum Ca and Pi

renal tubules, the renal parenchyma, the intestine and bone (see Table 7.1).

PTH, which can readily be measured in blood samples, is produced by the parathyroid glands. Secretion is regulated by plasma ionized calcium, with increases and decreases in calcium respectively suppressing and stimulating PTH release. This regulation is mediated by the calcium-sensing receptor (CaSR), which is activated by decreased ionized calcium, leading to suppression of PTH secretion. Rare CaSR inactivating mutations result in failure of PTH suppression leading to hypercalcaemia (familial hypocalciuric hypercalcaemia). In contrast, rare CaSR activating mutations cause hypoparathyroidism and hypocalcaemia.

Acting on the *renal tubules*, PTH increases phosphate excretion by restricting its reabsorption, and conserves calcium by increasing its reabsorption. These responses rapidly compensate for any change in plasma ionized calcium.

Acting on the *kidney parenchyma*, PTH controls hydroxylation of the vitamin D metabolite 25-OHD: a rise in PTH concentration stimulates conversion to the active metabolite 1,25-$(OH)_2$D and a fall in PTH causes a switch towards the inactive metabolite 24,25-$(OH)_2$D.

In the *intestine* PTH has the indirect effect of stimulating calcium absorption by promoting the conversion of 25-OHD to 1,25-$(OH)_2$D in the kidney.

In *bone* PTH acts to promote osteoclastic resorption and the release of calcium and phosphate into the blood. This it does not by direct action on osteoclasts but by stimulating osteoblastic activity, increased expression of RANKL and diminished production of osteoprotegerin (OPG). Furthermore, the PTH induced rise in 1,25-$(OH)_2$D has the effect of stimulating osteoclastogenesis. The net effect of these complex interactions is a prolonged rise in plasma calcium.

FGF23

FGF23 is produced by osteocytes, and acts to lower serum Pi by promoting renal Pi excretion through suppression of renal tubular Pi reabsorption. FGF23 levels are influenced by dietary Pi intake, and by calcitriol levels; the latter reduces serum Pi by increasing Pi excretion via stimulation of FGF23 release, leading to a reciprocal increase in serum calcium.

Altered circulating levels of FGF23, which can be readily measured, are associated with a variety of genetic and acquired disorders. In many of these cases, the clinical manifestations reflect reduced skeletal mineralization as a result of a reduction in the calcium × phosphate product due to excess FGF23 activity, giving a clinical picture of hypophosphatemia and vitamin D-resistant osteomalacia (see "hypophosphataeic rickets and osteomalacia" below. For example, in autosomal dominant hypophosphataemic rickets (ADHR), FGF23 levels are elevated due to a gain of function mutation causing resistance to enzymatic cleavage. Similarly, sex-linked hypophosphataemic rickets (XLH), autosomal recessive hypophosphataemic rickets type I (ARHR1) and autosomal recessive hypophosphataemic rickets type 2 (ARHR2) are associated with overproduction of FGF23 in osteocytes due to loss of function mutations in other genes, namely *PHEX*, *DMP1* and *ENPP1* respectively (Ruppe and Jan de Beur, 2013). Conversely, a number of mutations have been identified leading to decreased FGF23 levels, resulting in familial tumoural calcinosis in which hyperphosphataemia occurs, leading to soft-tissue calcification.

OTHER INFLUENCES ON BONE METABOLISM

As well as regulating calcium and phosphate homeostasis, hormones such as PTH have profound effects on bone turnover as a consequence of their effects on osteoclastic bone resorption. A number of other hormones and other factors exist which have no specific role in mineral exchange, but nonetheless influence bone remodelling and turnover, and contribute to the development of osteoporosis in which bone remodelling is increased (see the section on 'Osteoporosis' below).

Gonadal hormones

Sex steroids have a major role in the attainment of peak bone mass as a consequence of their role in puberty, which is associated with not only rapid growth but also a rapid gain in bone mass. Sex steroids are also important for maintaining bone mass subsequently, with loss of sex steroids in later life leading to significant bone loss in both males and females.

Oestrogen itself acts to suppress longitudinal growth and periosteal expansion, and its rise around puberty contributes to growth plate closure. The major impact of oestrogen deficiency on the skeleton in cases of primary amenorrhoea is a reduction in bone mineral density, particularly at trabecular-rich sites such as the spine. Oestrogen also acts to preserve bone mass, with its loss at the menopause leading to an increase in bone turnover, a decline in bone mineral density, a deterioration in bone architecture, and a consequential increase in fracture risk. This action involves suppression of bone resorption, as a consequence of increased production of OPG, thereby interfering with osteoclast differentiation and bone resorption.

Androgens stimulate bone growth, and act to maintain bone mass, through a combination of suppression of bone resorption and stimulation of bone formation. Although androgens mainly influence the skeleton in males and oestrogen in females, lower levels of oestrogens and androgens in males and females respectively are also thought to exert protective effects on the skeleton, particularly where the primary sex steroid is deficient.

Other hormones

Glucocorticoid excess in Cushing's disease is associated with adverse effects on bone metabolism characterized by suppressed bone formation, leading to a decrease in BMD and increased risk of osteoporosis. Glucocorticoid therapy has similar effects when given at higher doses to suppress inflammation, but there is little evidence that glucocorticoid inhalers for asthma, or glucocorticoid replacement in Addison's disease, adversely influence bone metabolism.

Thyroxine excess in thyrotoxicosis, or over-replacement with thyroxine in hypothyroidism, is associated with increased bone turnover and is a recognized risk factor for low BMD and osteoporosis.

As well as playing an important role in skeletal growth, *growth hormone (GH)* is thought to influence bone remodelling. GH deficiency is associated with reduced BMD as well as short stature, which may be an indication for continued GH replacement after the cessation of longitudinal growth.

Calcitonin is produced by C cells of the thyroid, acts to reduce osteoclast activity, and in the past was used as an anti-resorptive drug treatment for osteoporosis. No physiological role is recognized in humans and, although previously used to treat osteoporosis, this has been superseded due to limited efficacy.

Mechanical stress

One of the primary roles of the skeleton is to provide an endoskeleton, transmitting forces applied by muscles acting as external levers for the purposes of locomotion and other activities. The skeleton is designed to withstand different types of forces such as compression, tension, shear and torsion. However, at any one site a specific type of force tends to predominate; for example, compression is the predominant force acting on lumbar vertebrae while tensile forces predominate at the superior surface of the femoral neck. The direction and thickness of trabeculae in cancellous bone are related to regional stress trajectories. This is recognized in Wolff's law (1896), which states that the architecture and mass of the skeleton are adjusted to withstand the prevailing forces imposed by functional need or deformity (see Figure 7.10). This has led to the concept of the mechanostat, whereby bone remodelling and bone mass are regulated to ensure that bone strain (defined as deformation in response to an externally applied load per unit length) is kept within a target range (Frost, 1987).

Bone loading provides a more effective osteogenic stimulus when applied intermittently at high intensity, as opposed to continuously at lower intensity (Rubin and Lanyon, 1984). Osteocytes play a major role in sensing strains caused by mechanical deformation, transmitting information via the extensive network of canaliculi adjoining adjacent cells within the cortex. These signalling pathways are thought to involve sclerostin/Wnt signalling, with strain causing suppression of osteocyte secretion leading to stimulation of osteoblast activity. As well as stimulating bone formation, mechanical strain also enhances bone mass by suppression of bone resorption. Reduced mechanical stimulation causes bone loss and leads to osteoporosis in paraplegia and other conditions associated with absent or reduced weight-bearing activity such as space travel.

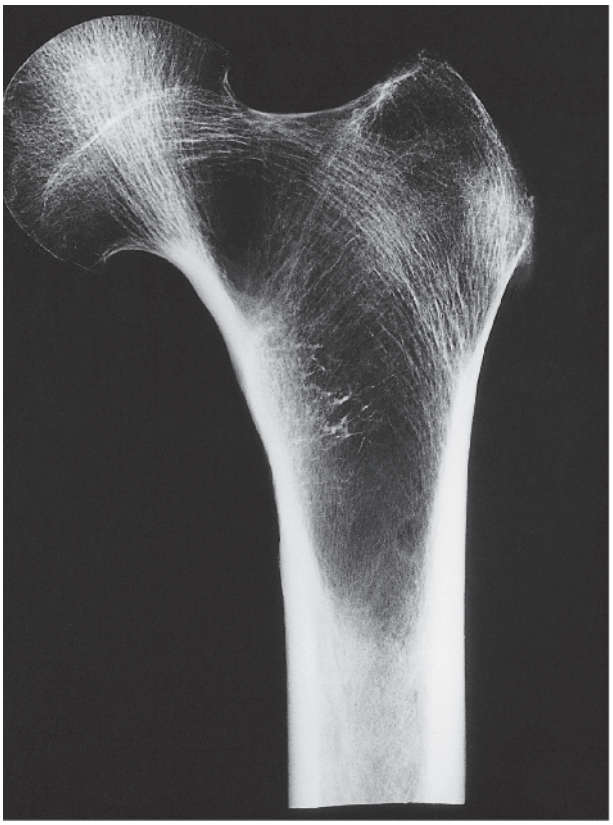


Figure 7.10 Wolff's Law Wolff's Law is beautifully demonstrated in the trabecular pattern at the upper end of the femur. The thickest trabeculae are arranged along the trajectories of greatest stress.

Dietary factors

Conditions associated with low protein and calorie intake such as anorexia nervosa are recognized to influence bone metabolism and turnover, leading to a decrease in BMD and increased risk of osteoporosis, particularly where onset is at a relatively young age such that peak bone mass is affected. Although other mechanisms such as amenorrhoea contribute to reduced bone mass in anorexia nervosa, low protein and calorie intake are also major factors. Low protein and calorie intake also contribute to reductions in BMD caused by other conditions including inflammatory bowel disease. The occurrence of low protein and calorie intake is often associated with reduced intake of specific dietary constituents such as calcium and vitamin D, which also contributes to reductions in BMD in this context.

Several other dietary factors have been suggested to influence bone metabolism. Low vitamin K intake has been suggested as a risk factor for low BMD and osteoporosis, possibly by virtue of its involvement in osteocalcin metabolism. High protein and other diets associated with relatively high acid production (as quantified by renal net acid excretion) may adversely influence bone metabolism, consistent with the fact that acidosis stimulates bone resorption.

METABOLIC BONE DISORDERS

ASSESSMENT

Patients with metabolic bone disorders usually appear to the orthopaedic surgeon in one of the following guises:

- a child with bone deformities (*rickets*)
- an elderly person with a fracture of the femoral neck or a vertebral body following comparatively minor trauma (*postmenopausal osteoporosis*)
- an elderly patient with bone pain and multiple compression fractures of the spine (*osteomalacia*)
- a middle-aged person with hypercalcaemia and pseudogout (*hyperparathyroidism*)
- someone with multiple fractures and a history of prolonged *corticosteroid treatment.*

X-rays may show *stress fractures, vertebral fractures, cortical thinning, loss of trabecular structure* or merely an ill-defined loss of radiographic density – *radiographic osteopenia* – which can signify either osteomalacia or osteoporosis.

History

Children are likely to be brought for examination because of failure to thrive, below-normal growth or deformity of the lower limbs. Adults may complain of back pain, the sudden onset of bone pain near one of the large joints or symptoms suggesting a full-blown fracture following some comparatively modest injury. Generalized muscle weakness is common in osteomalacia.

Details such as the patient's sex, age, race, onset of menopause, nutritional background, level of physical activity, previous illnesses, medication and operations are important. The onset and duration of symptoms and their relationship to previous disease or trauma should be carefully considered, especially in older people who may have suffered insufficiency fractures. Other causal associations are retarded growth, malnutrition, dietary fads, intestinal malabsorption, alcohol abuse and cigarette smoking.

A careful family history may yield clues to heritable disorders associated with osteoporosis and vulnerability to fracture.

Examination

The patient's appearance may be suggestive of an endocrine or metabolic disorder: the moon face and Cushingoid build of hypercortisonism; the smooth, hairless skin of testicular atrophy; physical underdevelopment and bone deformities in rickets. Thoracic kyphosis is a non-specific feature of vertebral osteoporosis.

X-rays

Decreased skeletal radiodensity is a late and unreliable sign of bone loss; it becomes apparent only after a 30% reduction in mineral or skeletal mass, and even then one cannot tell whether this is due to *osteoporosis* (a decrease in bone mass) or *osteomalacia* (insufficient mineralization of bone) or a combination of both. Sometimes the term *radiological osteopenia* is used to describe a mild or moderate loss of radiodensity in bone X-rays without implying whether this is pathological.

A more reliable sign of osteoporosis is the presence of obvious fractures – new and old – especially in the spine, ribs, pubic rami or corticocancellous junctions of the long bones. Small stress fractures are more difficult to detect: they may be found in the femoral neck, the proximal part of the femur or the upper end of the tibia.

In addition to these general signs of reduced bone mass or defective mineralization, there may be specific features of bone disorders such as rickets, hyperparathyroidism, metastatic bone disease or myelomatosis.

Measurement of bone mass

The gold standard investigation for measurement of bone mass is the dual-energy X-ray absorptiometry

(DXA) scan. Additional techniques are available, but are reserved for research purposes. These research tools include peripheral quantitative computer tomography (pQCT) and calcaneal ultrasound (US).

Dual-energy X-ray absorptiometry (DXA) This is now the method of choice. Precision and accuracy are excellent, radiation exposure is not excessive and it is low-cost (Blake and Fogelman, 2009). Measurements are usually taken from the lumbar spine and hip, but they can also be taken from the distal forearm and total body (Figure 7.11). Additional investigations carried out by the DXA machine and software that can help improve fracture prediction include Vertebral Fracture Assessment (VFA) and Trabecular Bone Score (TBS). Bone mass values from DXA scans are two-dimensional estimates of bone density presented as the number of standard deviations below the young adult mean (T score) and the number of standard deviations below the age-matched mean (Z score). The World Health Organization (WHO) has defined osteoporosis as a T score below –2.5, osteopenia as a T score between –1.0 and –2.5, and normal bone mass as above –1.0 (WHO Technical Support Series). VFA can be useful as it provides a low-resolution image of the entire lateral spine that can be assessed for the presence of vertebral fractures. TBS provides an additional measurement of bone microarchitecture based on bone texture of the lumbar spine.

Quantitative computed tomography (QCT) QCT permits measurement of mineral content per unit volume of bone, which is a true three-dimensional measurement of bone density. It also provides separate values for cortical and cancellous bone.

Calcaneal ultrasound (US) Measurements made of the speed and attenuation of sound through bone can be related to bone density and architecture.

Indications for bone densitometry

The main indications for using bone densitometry are:

- in adults over the age of 50 who have experienced a low trauma fracture, as part of Fracture Liaison Services (FLS) to allow assessment of future fracture risk to enable decisions on whether preventative treatment should be initiated
- in all women over the age of 65 and all men over the age of 75 to assess risk of future fracture
- to assess the degree and progress of bone loss in patients with clinically diagnosed metabolic bone disease or conditions such as hyperparathyroidism, corticosteroid-induced osteoporosis, gonadal deficiency or other endocrine disorders
- to monitor the effect of treatment for osteoporosis
- in patients on medications known to adversely affect bone health such as corticosteroids or aromatase inhibitors.

Biochemical tests

Serum calcium and phosphate concentrations should be measured in the fasting state, and it is the ionized calcium fraction that is important.

Serum bone alkaline phosphatase (ALP) concentration is an index of osteoblastic activity; it is raised in osteomalacia and in disorders associated with high bone turnover (hyperparathyroidism, Paget's disease, bone metastases).

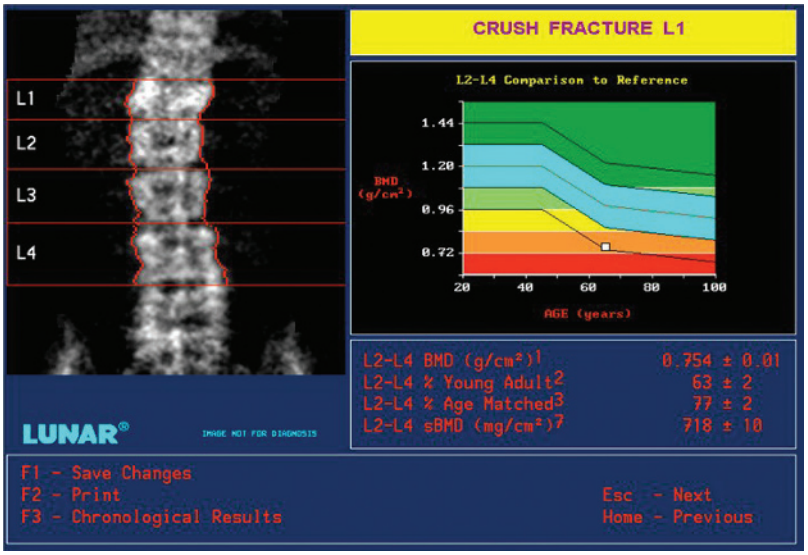


Figure 7.11 DXA output The output from a DXA scan showing the lumbar spine image on the left, and the measured BMD compared to the reference range on the right.

Parathyroid hormone (PTH) activity can be estimated from serum assays of the COOH terminal fragment. However, in renal failure the test is unreliable because there is reduced clearance of the COOH fragment.

Vitamin D activity is assessed by measuring the serum 25-OHD concentration. Serum 1,25-$(OH)_2$D levels do not necessarily reflect vitamin uptake but are reduced in advanced renal disease.

Urinary calcium and phosphate excretion can be measured. Significant alterations are found in malabsorption disorders, hyperparathyroidism and other conditions associated with hypercalcaemia.

Bone turnover markers can be measured if identification of high or low bone turnover states will be useful. The most common markers are the bone formation marker, serum type I collagen extension propeptide (P1NP), and the bone resorption marker, serum C-terminal cross-linking telopeptide of type I collagen (CTX).

NB: Laboratory reports should always state the normal range for each test, which may be different for infants, children and adults.

Bone biopsy

Standardized bone samples are obtained from the iliac crest and can be examined (without prior decalcification) for histological bone volume, osteoid formation and the relative distribution of formation and resorption surfaces (Figure 7.12). The rate of bone remodelling can also be gauged by labelling the bone with tetracycline on two occasions (2 weeks apart) before obtaining the biopsy. Tetracycline is taken up in new bone and produces a fluorescent strip on ultraviolet light microscopy. By measuring the distance between the two labels, the rate of new bone formation can be calculated. Characteristically in osteomalacia there is a decrease in the rate of bone turnover and an increase in the amount of uncalcified osteoid.

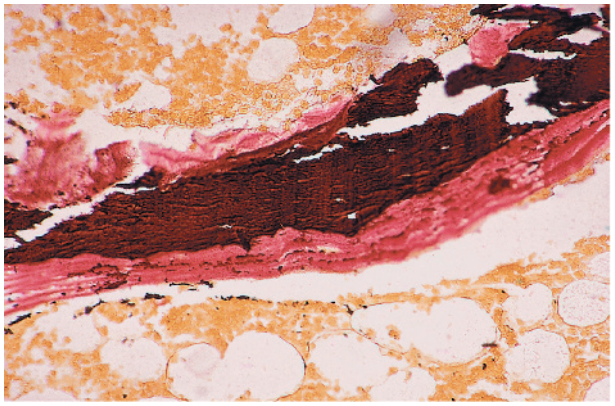


Figure 7.12 Bone biopsy von Kossa stain showing the unusually wide osteoid layer (in red) in a patient with osteomalacia.

OSTEOPOROSIS

Osteoporosis as a clinical disorder is characterized by an abnormally low bone mass and defects in bone structure, a combination which renders the bone unusually fragile and at greater than normal risk of fracture in a person of that age, sex and race. Although the cancellous regions are more porous and the cortices thinner than normal, the existing bone is fully mineralized (Figure 7.13).

Bone depletion may be brought about by predominant bone resorption, decreased bone formation or a combination of the two. It seems self-evident that the main reason for the loss of bone strength is the reduction in bone mass; however, in the remaining trabecular bone there may also be a loss of structural connectivity between bone plates, and this so alters the mechanical properties that the loss of strength is out of proportion to the diminution in bone mass. As a consequence, the bone – particularly around the diaphyseo-metaphyseal junctions in tubular bones and in the mainly cancellous vertebral bodies – eventually

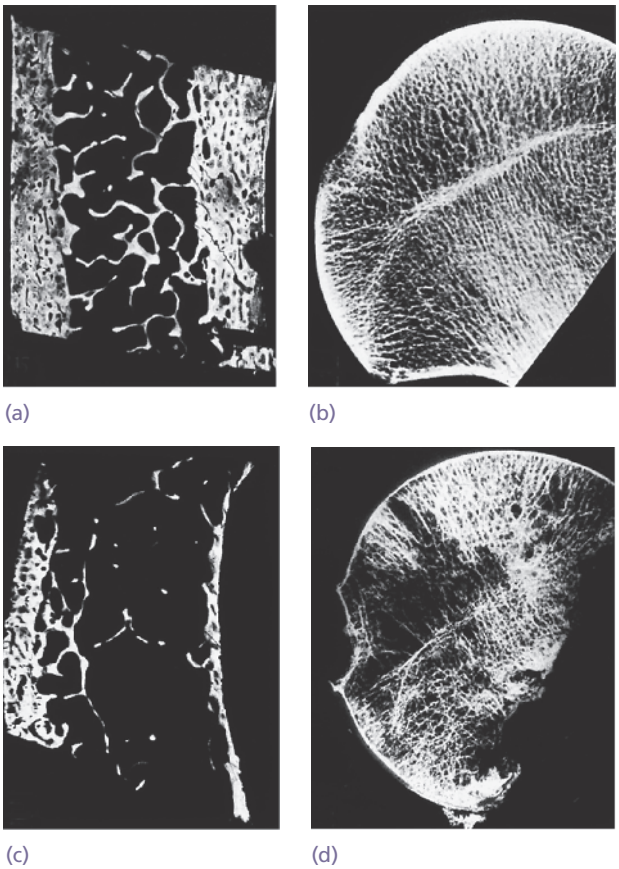


Figure 7.13 Age-related changes in bone These fine-detail X-rays of iliac crest biopsies and femoral head slices show the marked contrast between trabecular density in a healthy 40-year-old woman (a,b) and one of 75 years (c,d).

reaches a state in which a comparatively modest stress or strain causes a fracture.

This section deals with *generalized osteoporosis*, but it should not be forgotten that osteoporosis is sometimes confined to a particular bone or group of bones – *regional osteoporosis* (for example due to disuse, immobilization, inflammation or pregnancy) – which is usually reversible once the cause is addressed.

From the onset of the menopause and for the next 10 years the rate of bone loss in women accelerates to about 3% per year, occurring predominantly in trabecular bone. This steady depletion is due mainly to excessive resorption – osteoclastic activity seeming to be released from the restraining influence of gonadal hormone. (Similar changes are seen in younger women about 5 years after oophorectomy.) About 30% of white women will lose bone to the extent of developing postmenopausal osteoporosis.

From the age of 65 or 70 years the rate of bone loss in women gradually tails off and by the age of 75 years it is about 0.5% per year. This later phase of depletion is due mainly to diminishing osteoblastic activity (Parfitt, 1988).

Men are affected in a similar manner, but the phase of rapid bone loss occurs 15–20 years later than in women, at the climacteric.

Bone mass and bone strength

It is important to recognize that throughout life, and regardless of whether *bone mass* increases or decreases, the degree of *mineralization* in normal people varies very little from age to age or from one person to another.

With advancing years the loss of bone mass is accompanied by a *disproportionate loss of bone strength*, which is explained in a number of ways.

1 The absolute diminution in bone mass is the most important, but not the only, factor.
2 With increased postmenopausal bone resorption, perforations and gaps appear in the plates and cross-spars of trabecular bone; not all these defects are repaired and the loss of structural connectivity further reduces the overall strength of the bone.
3 In old age the decrease in bone cell activity makes for a slower remodelling rate; old bone takes longer to be replaced and microtrauma takes longer to be repaired, thus increasing the likelihood of stress failure.

This tendency to increased bone fragility with age is counteracted to some extent in tubular bones by the fact that they actually increase in diameter as their cortices become thinner. During each remodelling cycle resorption exceeds formation on the endosteal surface while formation slightly exceeds resorption on the periosteal surface. Simple mechanics can show that, of two cylinders with equal mass, the one with a greater diameter and thin walls is stronger than one with thicker walls but lesser diameter. Nonetheless, the age-related decrease in bone mass leads to a clear increase in fracture risk with increasing age (see Figure 7.14).

The boundary between 'normal' age-related bone loss and a clinical disorder (*osteoporosis*) is poorly defined. Factors that have an adverse influence on bone mass are shown in Box 7.1. Ageing individuals also often have some degree of *osteomalacia* due to lack of dietary vitamin D and poor exposure to

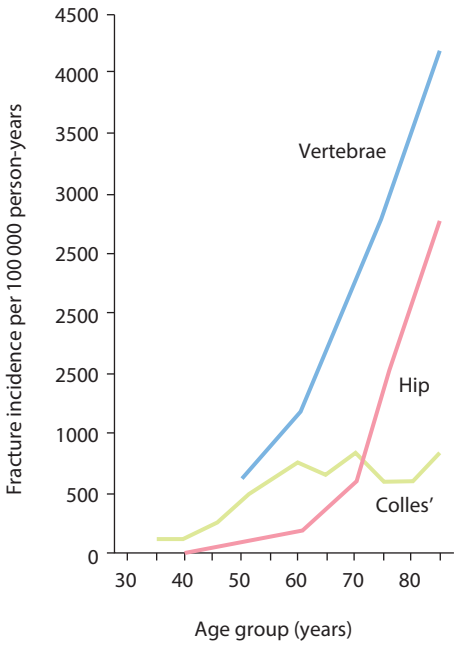


Figure 7.14 Fracture incidence The incidence of fractures of the vertebrae, hip and wrist rises progressively after the menopause.

BOX 7.1 RISK FACTORS FOR OSTEOPOROSIS

Age

Female

Previous fragility fracture

Current use or frequent recent use of oral or systemic glucocorticoids

Family history of hip fracture

Low body mass index (BMI) (less than 18.5 kg/m^2)

Smoking

Alcohol intake of more than 14 units per week for women and more than 21 units per week for men

sunlight, and this added to the normal age-related bone depletion makes them more vulnerable than usual to insufficiency fractures.

X-rays

Osteoporosis cannot be diagnosed from plain radiographs. 'Radiological osteopenia' is a term sometimes used to describe bone which appears to be less 'dense' than normal on X-ray. Typical signs of radiological osteopenia are loss of trabecular definition, thinning of the cortices and insufficiency fractures. Compression fractures of the vertebral bodies, sometimes described as wedging or compression of the vertebral end plates, are typical of severe postmenopausal osteoporosis (Figure 7.15). Osteoporotic vertebral fractures can be identified by experienced radiologists, or via systematic approaches such as quantitative morphometry whereby percentage height reductions are used to grade fractures as mild (20–25% reduction in height), moderate (25–40% reduction in height) or severe (>40% height reduction). Moderate and severe vertebral fractures predict future hip fractures, but mild do not, probably because the majority of mild anterior height loss is due to degenerative change rather than osteoporotic fracture.

Assessment of fracture risk

Increasingly, physicians are moving away from simple identification and treatment of low bone density from DXA scans to individualized assessment of fracture risk. This is because bone density is only one risk factor for fracture. There are now many tools available to calculate a patient's probability of fracture over the next few years based on a combination of clinical risk factors plus bone density. Examples of these tools include FRAX (www.shef.ac.uk/frax/) and QFracture (www.Qfracture.org). Treatment decisions can then be made on the basis of whether the patient's individual risk of fracture is high enough to warrant medication. There is no standardized definition of 'high' fracture risk, and choice of treatment thresholds differ according to country, often based on willingness to pay.

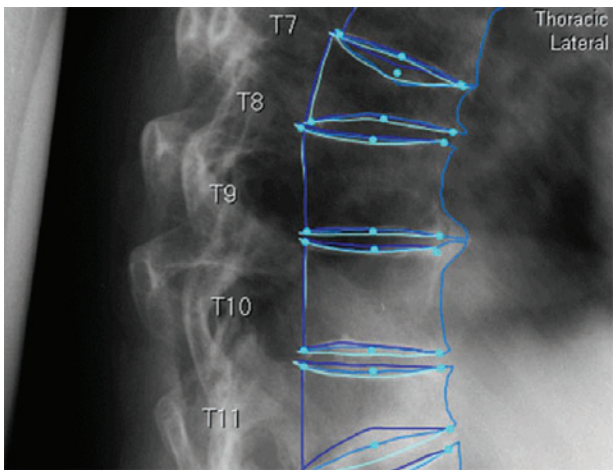


Figure 7.15 Vertebral fractures and their grading
A lateral spinal image showing the quantitative morphometry method of grading of osteoporotic vertebral fractures by utilising six points on each vertebral body. There is a severe fracture at T8 and a moderate fracture at T11.

Postmenopausal osteoporosis

Postmenopausal osteoporosis is an exaggerated form of the physiological bone depletion that normally accompanies ageing and loss of gonadal activity (see 'Gonadal hormones' above). Around the menopause, and for the next 10 years, bone loss normally accelerates to about 3% per year compared with 0.3% during the preceding two decades. This is due mainly to increased bone resorption, the withdrawal of oestrogen having removed one of the normal restraints on osteoclastic activity. Genetic influences play an important part in determining when and how this process becomes exaggerated, but a number of other risk factors have been identified (see Box 7.1).

Clinical features and investigations

Osteoporosis is asymptomatic unless fractures occur. The fractures are classically low trauma defined as a fall from standing height or less. Fracture of the distal radius (Colles' fracture) is usually the first fracture to occur, followed by vertebrae and hip unless treatment is initiated. Osteoporotic vertebral fractures are particularly difficult to diagnose as they may be clinically silent. Less than a third of vertebral fractures are diagnosed, and this is an important healthcare gap because older women with vertebral fractures are at one of the highest risks of future fracture. In severe cases, significant height loss (often exceeding 4 cm) and thoracic kyphosis can occur due to multiple vertebral fractures (see Figure 7.16). However, milder height loss and smaller kyphoses are most commonly due to degenerative change.

As well as assessment of fracture risk, the rate of bone turnover is either normal or slightly increased; measurement of excreted collagen cross-link products and telopeptides may suggest a high-turnover type of bone loss.

Once any fractures have been managed, if Z scores on the DXA scan are low, tests should be performed to rule out other causes of osteoporosis such as hyperparathyroidism, malignant disease or hypercortisonism.

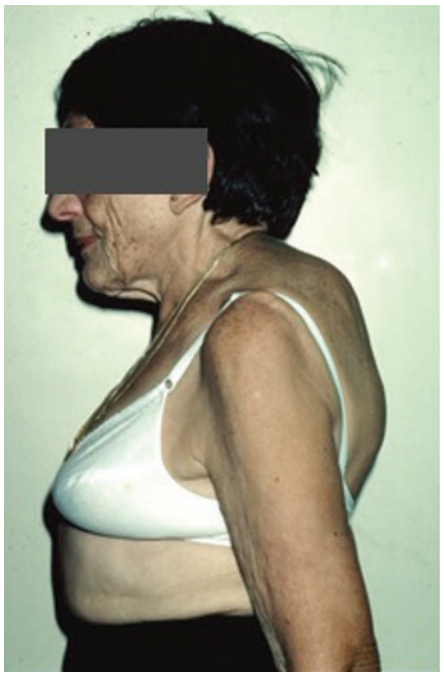


Figure 7.16 Dowager's hump Kyphotic deformity of the thoracic spine in a patient with osteoporosis.

Prevention

Adults over the age of 50 who have a low-energy fracture should have their fracture risk assessed by one of the clinical tools such as FRAX as well as bone density assessment by DXA. Those above the local treatment thresholds should be offered medications to reduce their fracture risk. This screening of people with fractures is now commonplace, and often through well-developed Fracture Liaison Services. Primary screening for people who have not sustained fractures is not recommended unless there is a concern about secondary osteoporosis. All women should be advised on lifestyle choices to maintain healthy bones. These include diets rich in calcium and vitamin D, weight-bearing physical activity and to avoid smoking and excessive consumption of alcohol. If necessary, the recommended daily requirements should be met by taking calcium and vitamin D supplements; these measures have been shown to reduce the risk of low-energy fractures in elderly women (Chapuy et al., 1994).

Treatment

The current goal of treatment is to reduce risk of future fracture. There is also increasing interest in a 'treat to target' approach, but this is not yet widespread. Medications are now recommended for a period of time, typically 3–5 years, at which point the need for ongoing treatment should be reconsidered by further assessment of fracture risk.

Calcium and vitamin D supplements are not treatments to reduce fracture risk. However, all the randomized controlled trials of medications to reduce fracture risk ensured all participants were taking these supplements. It is sensible to consider them if there is any suggestion that diet or sunlight exposure is likely to be low. Hormone replacement therapy (HRT) and calcitonin are no longer recommended as treatments to reduce fracture risk.

Bisphosphonates Bisphosphonates taken orally are now regarded as the first-line medication for reduction of fracture risk in postmenopausal women, although intravenous formulations are also available. For example, zoledronate can be given once per year intravenously. They act by reducing osteoclastic bone resorption and the general rate of bone turnover. They have been shown to prevent bone loss and to reduce the risk of vertebral and hip fractures (Boonen et al, 2005). Gastrointestinal side effects are the most common adverse event with oral preparations. Rarer, more serious adverse events mainly associated with the intravenous preparations include atypical femoral fractures and osteonecrosis of the jaw (ONJ).

Denosumab This is an antibody to RANKL, essential for promoting osteoclastogenesis. It is a subcutaneous injection given every 6 months and it has been shown to reduce the risk of both hip and vertebral fractures (Cummings et al., 2005). Uniquely, it has quite a rapid 'off-set' on stopping the medication, with rapid reversal of bone turnover. Potential adverse events also include atypical femoral fractures and ONJ.

Parathyroid hormone Preotact and Teriparatide (recombinant human parathyroid hormone 1–34) are anabolic agents, given intermittently at low doses that stimulate bone formation to a greater and earlier extent than bone resorption (Pleiner-Duxneuner et al., 2009). They prevent fractures, prevent corticosteroid-induced osteoporosis and are occasionally used in unlicensed situations such as healing of non-union or atypical femoral fractures.

Selective oestrogen receptor modulators (SERMs) Raloxifene (Delmas et al., 2002) is licensed for fracture risk reduction in women and has been shown to reduce the risk of vertebral fractures. The main side effect is hot flushes, but use can also increase the risk of venous thromboembolism.

Strontium ranelate A strontium salt of ranelic acid, strontium ranelate is thought to increase bone formation and reduce bone resorption *in vitro*. It is given as a sachet of granules to be dissolved in water and drunk once per day. Clinical trials show a reduction in the risk of vertebral fractures (Meunier et al., 2004) and non-vertebral fractures. The use of strontium is

limited by the contraindications: current or previous venous thromboembolism, ischaemic heart disease, peripheral arterial disease and/or cerebrovascular disease.

Recent advances in drug treatment Cathepsin K is a lysosomal cysteine proteinase with high collagenase activity. Cathepsin K inhibition (Odanacatib) is a promising new treatment to reduce fracture risk (Chapurlat, 2015), with a phase III trial, unpublished but reported at scientific meetings, showing a reduction in vertebral, hip and non-vertebral fractures. Sclerostin is a glycoprotein inhibitor of osteoblast Wnt signalling produced by osteocytes that has been recognized as a new target for therapeutic intervention in patients with osteoporosis. Romosozumab is a humanized anti-sclerostin monoclonal antibody that has been demonstrated to increase bone formation. A large phase III controlled study is currently underway. Abaloparatide is a new synthetic peptide analog of human parathyroid hormone-related peptide (PTHrP), and a phase III trial for this is also awaited.

Management of fractures Femoral neck and other long-bone fractures may need operative treatment. Methods are described in the relevant chapters in Section 3.

Vertebral fractures can be painful and patients will need analgesic treatment. Physiotherapy should initially be aimed at maintaining muscle tone; if pain is adequately controlled, patients should be encouraged to walk and, when symptoms allow, they can be introduced to postural training. Spinal orthoses may be needed for support and pain relief, but they cannot be expected to correct any structural deformity. Vertebral augmentation such as kyphoplasty or vertebroplasty are occasionally called for to treat extremely painful vertebral fractures that have caused symptoms for more than 6–8 weeks.

Osteoporosis in men

With the gradual depletion in androgenic hormones, men eventually suffer the same bone changes as postmenopausal women, only this occurs about 15 years later unless there is some specific cause for testicular failure. *Osteoporotic fractures in men under 60 years of age should arouse the suspicion of some underlying disorder – notably hypogonadism, metastatic bone disease, multiple myeloma, liver disease, renal hypercalciuria, alcohol abuse, malabsorption disorder, malnutrition, glucocorticoid medication or anti-gonadal hormone treatment for prostate cancer.* Other causes of secondary osteoporosis are shown in Box 7.2.

Treatment is much the same as for postmenopausal osteoporosis. Vitamin D and calcium supplementation is important; bisphosphonates are the anti-resorptive drug of choice. If testosterone levels are unusually low, hormone treatment should be considered, although bisphosphonates reduce fracture risk in the setting of low testosterone.

BOX 7.2 SECONDARY CAUSES OF OSTEOPOROSIS

Endocrine
- Hypogonadism in either sex including untreated premature menopause, treatment with aromatase inhibitors or androgen deprivation therapy
- Hyperthyroidism
- Hyperparathyroidism
- Hyperprolactinaemia
- Cushing's disease
- Diabetes

Respiratory
- Cystic fibrosis
- Smoking-related lung disease

Metabolic
- Homocystinuria

Chronic renal disease

Gastrointestinal
- Coeliac disease
- Inflammatory bowel disease
- Chronic liver disease
- Chronic pancreatitis
- Other causes of malabsorption

Rheumatological
- Rheumatoid arthritis
- Other inflammatory arthropathies

Haematological
- Multiple myeloma
- Haemoglobinopathies
- Systemic mastocytosis

Immobility
- Neurological injury
- Neurological disease

(Based on NICE (2012) CG146: *Osteoporosis: Assessing the risk of fragility fracture*)

Secondary osteoporosis

Among the numerous causes of secondary osteoporosis, hypercortisonism, gonadal hormone deficiency, hyperthyroidism, multiple myeloma, chronic

alcoholism and immobilization will be considered further. In addition, the impact of obesity and diabetes on bone health will be discussed.

Glucocorticoid-induced osteoporosis

Glucocorticoid overload occurs in endogenous Cushing's disease or after prolonged treatment with corticosteroids. This often results in severe osteoporosis, especially if the condition for which the drug is administered is itself associated with bone loss – for example, rheumatoid arthritis.

Glucocorticoids have a complex mode of action. The deleterious effect on bone is mainly by suppression of osteoblast function, but it also causes reduced calcium absorption, increased calcium excretion and stimulation of PTH secretion (Hahn, 1980).

Treatment Oral corticosteroid dosage should be kept to a minimum, but there is no evidence that steroid-based inhalers for asthma or glucocorticoid replacement for adrenal insufficiency adversely affect bone. All patients on corticosteroids should have their fracture risk assessed using the standard clinical tools such as FRAX, although it must be remembered that FRAX is likely to underestimate fracture risk in those on high doses of corticosteroids. If people on glucocorticoids are identified as being at high risk of future fracture, oral and intravenous bisphosphonates are licensed for preventing corticosteroid-induced osteoporosis and can also reduce fracture risk in the presence of corticosteroids. Fractures are treated as and when they occur.

Gonadal hormone insufficiency

Oestrogen lack is an important factor in postmenopausal osteoporosis. It also accounts for osteoporosis in younger women who have undergone oophorectomy, and in pubertal girls with ovarian agenesis and primary amenorrhoea (Turner's syndrome). Treatment is the same as for postmenopausal osteoporosis. Amenorrhoeic female athletes, and adolescents with anorexia nervosa, may become osteoporotic and have a high fracture risk.

A decline in testicular function probably contributes to the continuing bone loss and rising fracture rate in men over 70 years of age. A more obvious relationship is found in young men with overt hypogonadism; this may require long-term treatment with testosterone. Alternatively, bisphosphonates are effective in reducing fracture risk in the context of hypogonadism.

Hyperthyroidism

Thyroxine speeds up the rate of bone turnover, but resorption exceeds formation. Osteoporosis is quite common in untreated hyperthyroidism, but fractures usually occur only in older people who suffer the cumulative effects of the menopause and thyroid overload. In the worst cases osteoporosis may be severe with spontaneous fractures, a marked rise in serum alkaline phosphatase, hypercalcaemia and hypercalciuria. Treatment is needed for both the osteoporosis and the thyrotoxicosis.

Multiple myeloma and carcinomatosis

Generalized osteoporosis, anaemia and a high ESR are characteristic features of myelomatosis and metastatic bone disease. Bone loss is due to overproduction of local osteoclast-activating factors. Treatment with bisphosphonates may reduce the risk of fracture.

Alcohol excess

This is a common and often neglected cause of osteoporosis at all ages, with the added factor of an increased tendency to falls and other injuries. Bone changes are due to a combination of decreased calcium absorption, liver failure and a toxic effect on osteoblast function. Alcohol also has a mild glucocorticoid effect.

Immobilization

The worst effects of stress reduction are seen in states of weightlessness; bone resorption, unbalanced by formation, leads to hypercalcaemia, hypercalciuria and severe osteoporosis. Lesser degrees of osteoporosis are seen in bedridden patients, and regional osteoporosis is common after immobilization of a limb. The effects can be mitigated by encouraging mobility, exercise and weight-bearing.

Obesity

Obesity is traditionally viewed as being beneficial to bone health because of the well-established positive effect of mechanical loading. However, it is increasingly recognized that obesity is associated with an increased risk of particular types of fractures, such as ankle fractures. This may be via increased mechanical force due to heavier body weight during the injury event, via hormones such as leptin and adiponectin secreted by adipose tissue, or because of the association between obesity and type 2 diabetes (see below), metabolic syndrome, impaired glucose tolerance and inflammation which may be associated with poor bone health.

Diabetes

People with type 1 diabetes have a reduced bone mass and increased risk of fragility fractures compared to

people without diabetes. However, despite having normal or above-normal bone density, people with type 2 diabetes are susceptible to low-trauma fractures, even after adjusting for age, physical activity and body weight. 'Diabetic bone disease' is a term being used to describe the bone health of people with type 2 diabetes, and it is thought to related to poor bone quality, perhaps due to the influence of hyperglycaemia, diabetic complications or other lifestyle factors (Adami, 2009).

Other conditions

There are many other causes of secondary osteoporosis, including hyperparathyroidism (which is considered below), rheumatoid arthritis (see Chapter 3), ankylosing spondylitis (see Chapter 3) and subclinical forms of osteogenesis imperfecta (see Chapter 8). The associated clinical features usually point to the diagnosis.

HYPOCALCAEMIA

Regulatory pathways described above, particularly PTH, are generally very effective at maintaining normocalcaemia, which is required for a number of essential cellular processes. Symptomatic hypocalcaemia is therefore rare, but when it is present it needs to be treated promptly. Causes of hypocalcaemia are listed in Box 7.3. The most important are hypoparathyroidism, severe vitamin D deficiency, and chronic kidney disease.

BOX 7.3 CAUSES OF HYPOCALCAEMIA

Vitamin D deficiency
- ***Acquired:*** Dietary deficiency, malabsorption
- ***Congenital:*** 1 alpha-hydroxylase mutation, vitamin D receptor mutation

Chronic renal failure

Hypoparathyroidism
- ***Congenital:*** Di George syndrome, calcium-sensing receptor (activating) mutations, pseudohypoparathyroidism
- ***Acquired:*** Autoimmune, post-parathyroidectomy

Hypomagnesaemia

Pancreatitis

Hyperphosphataemia

Drug-induced (denosumab, zoledronate, cinacalcet)

Hypoparathyroidism This is an important cause of hypocalcaemia immediately following parathyroidectomy, during which calcium levels need to be carefully monitored. Rarely, hypoparathyroidism can develop as part of an autoimmune disease, due to iron overload resulting from haemochromatosis, or due to magnesium deficiency which interferes with PTH release. Congenital hypoparathyroidism may result from failure to develop parathyroid glands (DiGeorge syndrome), and from rare genetic disorders associated with activating mutations of the calcium-sensing receptor.

Diagnosis of hypoparathyroidism is supported by finding of a low serum calcium and elevated serum phosphate and low or absent PTH. In cases of activating mutations of the calcium-sensing receptor, PTH levels may be close to the normal range, but they are nonetheless inappropriately low given the low calcium level.

Pseudohypoparathyroidism is a rare genetic cause of hypocalcaemia in which peripheral sensitivity to PTH is impaired due to a loss of function mutation in the Gs alpha subunit of G proteins involved in mediating peripheral responses to PTH. In contrast to type II, types 1a and 1b are both characterized by a defective urinary cyclic AMP response to PTH, which can be useful in confirming the diagnosis. Type Ia is associated with specific morphological features of Albright's hereditary osteodystrophy such as short stature, short fourth and fifth metacarpals and sometimes mild mental retardation. Pseudopseudohypoparathyroidism refers to a condition characterized by similar phenotypic features of Albright's hereditary osteodystrophy, but with normal calcium metabolism.

Clinical features

Mild hypocalcaemia itself is often asymptomatic. Occasionally, severe hypocalcaemia develops acutely and can potentially be fatal. Typical features include tetany, numbness/paraesthesias and muscle spasms. More severe features include convulsions, cardiac arrhythmias, and laryngeal spasm. Typical signs are the Trousseau sign (tetany caused by inflating a blood-pressure cuff), Chvostek's sign (facial spasms caused by tapping over the facial nerve), and prolongation of the QT interval on ECG.

Investigations

Radiographs may show a variety of appearances depending on the underlying cause. Certain conditions associated with chronic hypocalcaemia, such as pseudohypoparathyroidism, can paradoxically be associated with soft-tissue calcification, for example of

the basal ganglia and subcutaneous tissues. Finding of a low serum calcium should be followed up by further investigations as indicated, such as analysis of calcium-sensing receptor mutations if there is a suggestion of congenital hypoparathyroidism on the basis of a family history, low phosphate and low PTH; urinary cyclic AMP excretion in response to PTH should be analysed if serum PTH is elevated suggesting peripheral insensitivity. Magnesium levels should also be checked.

Treatment

Acute hypocalcaemia is a medical emergency and needs to be promptly treated with intravenous calcium, usually in the form of calcium gluconate. Chronic hypocalcaemia is often managed with vitamin D metabolites such as calcitriol, however caution needs to be used as calcitriol treatment can lead to significant hypercalciuria, especially in hypoparathyroidism. This increases the risk of renal complications such as nephrocalcinosis and nephrolithiasis. In patients with chronic hypoparathyroidism, replacement treatment with teriparatide may be helpful. In cases of activating mutations of the calcium-sensing receptor, newly developed inhibitors may play a role in future.

RICKETS AND OSTEOMALACIA

Rickets and osteomalacia are distinct from osteoporosis (see Table 7.2). The two are different expressions of the same disease: inadequate mineralization of bone in children is called rickets; in adults it is known as osteomalacia. Osteoid throughout the skeleton is incompletely calcified, and the bone is therefore 'softened' (*osteomalacia*). This leads to an increase in bone fragility and fracture risk. In children, the growth plate is also disrupted and, in combination with softening of bone, leads to characteristic deformities (*rickets*).

Table 7.2 Osteomalacia and osteoporosis compared

	Osteomalacia	Osteoporosis
Similarities	Common in ageing women Prone to pathological fracture Decreased bone density	
Individual	Unwell	Well
Pain	Generalized chronic ache	Pain only after fracture
Muscles	Weak	Normal
Looser's zones	YES	NO
Alkaline phosphatase	Increased	Normal
Serum phosphorus	Decreased	Normal
Ca × P	<2.4 mmol/L	>2.4 mmol/L

Reduced serum 1,25-OHD is by far the most common cause of rickets/osteomalacia, usually due to a lack of 25-OHD substrate, resulting from a combination of nutritional deficiency and lack of sunlight exposure. Several risk factors are recognized including childhood and pregnancy when vitamin D requirements are higher, strict veganism, dress codes which limit sun exposure, and nursing-home residence where poor diet is compounded by reduced sunlight exposure. Other causes include intestinal malabsorption of which coeliac disease is the most common, and defective vitamin D activation; decreased 25-hydroxylation is seen in liver disease and treatment with anticonvulsants; reduced 1α-hydroxylation is present in renal disease, nephrectomy and genetic 1α-hydroxylase deficiency.

In contrast, *vitamin D resistant rickets/osteomalacia* is caused by phosphate deficiency and has distinct clinical features (see below).

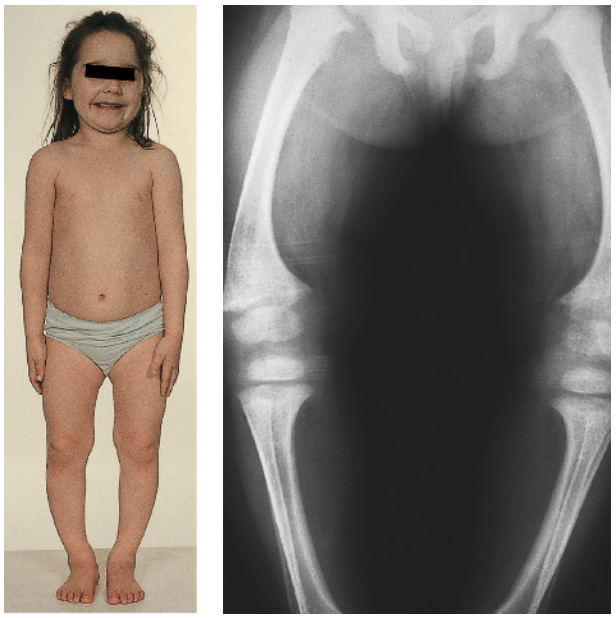
Pathology

Rickets is associated with the inability to calcify the intercellular matrix in the growth plate, causing chondrocytes to pile up irregularly, increase in the width of the growth plate, poor mineralization of the zone of calcification and sparse bone formation in the zone of ossification. The new trabeculae are thin and weak, and the metaphysis becomes broad and cup-shaped. *Osteomalacia* is characterized by widened osteoid seams and thinner cortices.

Clinical features

Children The infant with *rickets* may present with tetany or convulsions due to hypocalcaemia. Later the parents may notice that there is a failure to thrive, listlessness and muscular flaccidity. Early bone changes are deformity of the skull (craniotabes) and thickening of the knees, ankles and wrists from growth plate overgrowth. Enlargement of the costochondral junctions ('rickety rosary') and lateral indentation of the chest (Harrison's sulcus) may also be present. Lower limb deformities such as coxa vara and bowing of the femora and tibiae may develop after weight-bearing, while overall growth may be stunted (see Figure 7.17).

Adult *Osteomalacia* may have an insidious course and patients may complain of relatively non-specific symptoms such as widespread bone pain and muscle weakness. Unexplained pain in the hip or one of the long bones may presage a stress fracture. *Osteomalacia* increases the risk of fractures throughout the skeleton. When present, muscle weakness is of a proximal distribution, causing a 'waddling' gait.



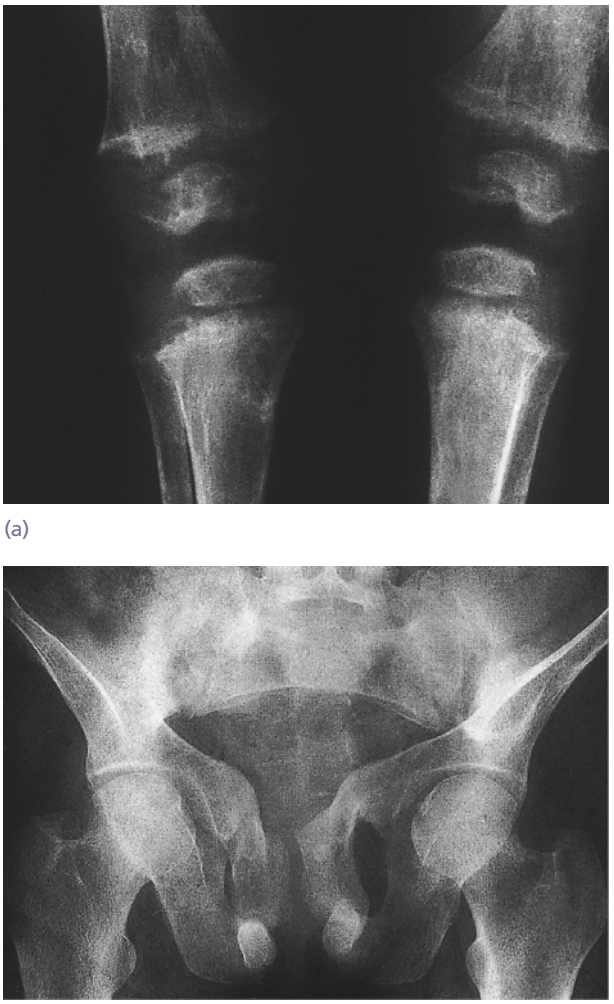
(a) (b)

Figure 7.17 Rickets In countries with advanced health systems nutritional rickets is nowadays uncommon. This 5-year-old girl, after investigation, was found to have familial hypophosphataemic rickets. In addition to the obvious varus deformities on her legs, (a) her lower limbs are disproportionately short compared to her upper body. (b) X-ray of another child with classical nutritional rickets, showing the well-marked physes, the flared metaphyses and the bowing deformities of the lower limb bones.

X-rays

Children In active *rickets* there is thickening and widening of the growth plate, cupping of the metaphysis and, sometimes, bowing of the diaphysis (see Figure 7.18). The metaphysis may remain abnormally wide even after healing has occurred. If the serum calcium remains persistently low, there may be signs of *secondary hyperparathyroidism*: subperiosteal erosions are at the sites of maximal remodelling such as the radial aspects of the proximal and middle phalanges of the middle and index fingers, medial borders of the proximal humerus, femoral neck, distal femur and proximal tibia.

Adults The classical lesion of *osteomalacia* is the Looser zone, a thin transverse band of rarefaction in an otherwise normal-looking bone (see Figure 7.19). These zones, seen especially in the pubic rami, medial proximal femur and axillary edge of the scapula, are due to incomplete stress fractures which heal with callus lacking in calcium. Vertebral fractures may show either a characteristic biconcave appearance or wedge-shaped deformities indistinguishable from osteoporotic fractures. There may also be signs of *secondary hyperparathyroidism*.



(a)

(b)

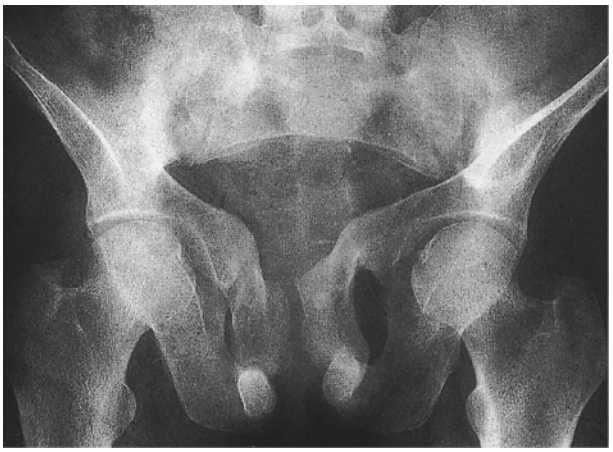
Figure 7.18 Rickets – X-rays X-rays obtained at two points during growth in a child with nutritional rickets. The typical features such as widening of the physis and flaring of the metaphysis are well marked (a). After treatment the bones have begun to heal but the bone deformities are still noticeable (b).

Biochemistry

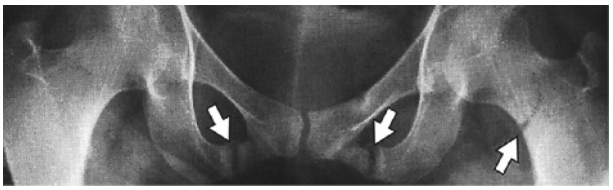
Overt hypocalcaemia is relatively uncommon in adults presenting with osteomalacia. More commonly, serum calcium is maintained at the lower part of the normal range as a result of secondary hyperparathyroidism, the latter causing raised levels of PTH and ALP (ALP may increase further following a fracture, or during bone healing after initiation of vitamin D replacement), and reduced serum phosphate. Low vitamin D status is indicated by a very low 25-OHD level (typically <10 nmol/L).

Bone biopsy

With clear-cut clinical and X-ray features the diagnosis is usually clear cut. In less typical cases a bone biopsy



(a)



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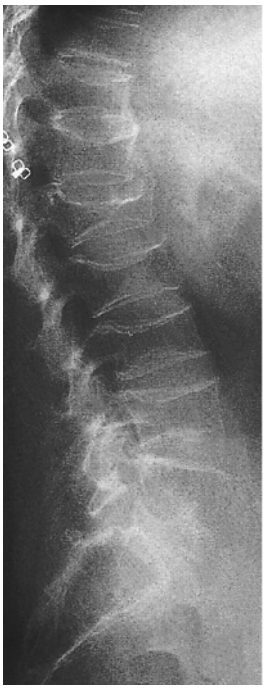


(c)

Figure 7.19 Osteomalacia
Characteristic features of osteomalacia: (a) indentation of the acetabula producing the trefoil or champagne glass pelvis; (b) Looser's zones in the pubic rami and left femoral neck; (c) biconcave vertebrae.

will provide the answer. Osteoid seams are both wider and more extensive, and tetracycline labelling shows that mineralization is defective (see Figure 7.12).

Treatment

In high-risk populations, osteomalacia is generally preventable by dietary modification or use of vitamin D supplements. For example, 400 IU per day is recommended in high-risk groups such as pregnant women (Gov.uk, 2012). However, larger doses may be necessary in patients with malabsorption, in whom 25-OHD levels should be checked to confirm adequate replacement. Most vitamin D preparations are in the form of combined supplements with calcium, which may be helpful since calcium deficiency may aggravate the effects of vitamin D deficiency. That said, significant calcium deficiency requiring replacement in its own right is rare, although patients with malabsorption and inflammatory bowel disease may be at particular risk. Difficulty in tolerating combined calcium and vitamin D supplements due to upper GI side effects is generally caused by the calcium component and can be overcome by use of vitamin D only supplements.

Whereas long-term vitamin D supplementation is generally required following the diagnosis of osteomalacia, initial replenishment requires a loading dose of 300–600 000 IU. Although the risk of hypercalcaemia is low, vitamin D loading doses tend to be administered either as two depo injections of a drug such as ergocalciferol spaced 4 weeks apart, or over an equivalent period using high-strength oral vitamin D (available at 10 000 and 50 000 IU doses). More recently, the bioavailability of vitamin D following IM depo injections has been called into question, and use of high-strength oral vitamin D as a loading dose is currently favoured.

Hypophosphataemic rickets and osteomalacia

Rarely, osteomalacia occurs secondary to renal phosphate wasting as a consequence of impaired renal tubular reabsorption of phosphate. This may be as part of a more generalized renal tubular disorder, which can produce a variety of biochemical abnormalities, including chronic phosphate depletion and osteomalacia. Isolated phosphate wasting is a consequence of excess FGF23 production. This may be acquired as a result of an FGF-secreting tumour.

Several rare genetic diseases are also associated with chronic hypophosphataemia. Although these have different modes of inheritance, in most cases they are characterized by excess serum levels of FGF23. As well as affecting the skeleton due to a reduction in mineralization secondary to lowering of the calcium × phosphate product, these disorders may be characterized by renal pathology due to an excessive calcium × phosphate product in renal tubules, leading to renal tubular defects, nephrocalcinosis and nephrolithiasis.

Familial hypophosphataemic rickets This is the commonest heritable phosphate wasting genetic disorder. It is an X-linked genetic disorder with dominant

inheritance, caused by a mutation in the *PHEX* gene, which leads to inappropriately elevated FGF23 levels. The condition starts in infancy or soon after and causes bony deformity of the lower limbs if it is not recognized and treated. During infancy the children look normal but deformities of the lower limbs such as genu valgum or varum develop when they begin to walk and growth is below normal. There is no myopathy. During adulthood there is a tendency to develop heterotopic bone formation around some of the larger joints and in the longitudinal ligaments of the spinal canal that may give rise to enthesopathies and neurological symptoms. As in other forms of osteomalacia these patients are at increased risk of fractures including stress fractures, but in contrast to other forms of osteomalacia bones can appear sclerotic. Biochemically these patients have low levels of phosphate, but serum calcium and PTH levels are usually normal.

Treatment requires the use of phosphate (up to 3 g per day, to replace that which is lost in the urine) and large doses of vitamin D (to prevent secondary hyperparathyroidism due to phosphate administration). *If calcitriol is given instead, plasma calcium concentration should be monitored in order to forestall the development of hypercalciuria and nephrocalcinosis.* The development of FGF23-blocking antibodies holds out the option of more effective and better tolerated treatment.

Bony deformities may require bracing or osteotomy. If the child needs to be immobilized, vitamin D must be stopped temporarily to prevent hypercalcaemia from the combined effects of treatment and disuse bone resorption.

Oncogenic osteomalacia This is caused by FGF-23-secreting tumours, particularly vascular tumours such as haemangiopericytomas, and also fibrohistiocytic lesions such as giant cell tumours and pigmented villonodular synovitis. Often the tumour is clinically silent and patients present with symptoms such as bone pain related to osteomalacia. Diagnosis is confirmed by finding of an elevated serum FGF23. Although resection of the primary leads to prompt resolution, identifying the site of the primary can be challenging and require extensive imaging.

HYPOPHOSPHATASIA

Hypophosphatasia is a rare heritable cause of rickets and osteomalacia resulting from deficiency of alkaline phosphatase bone isoenzyme. It has a range of severity and can present in the perinatal period, infancy, childhood and adulthood. Clinical features include weakness and limb deformities, and radiographs may reveal characteristic 'tongues' of hypomineralization extending from the growth plate. In adults, the condition can present with recurrent metatarsal stress fractures, and thigh or hip pain due to pseudofractures (Whyte, 2013). The main serum abnormality is a low or low normal level of alkaline phosphatase. Enzyme replacement therapy has been found to be effective at treating severely affected infants and children (Whyte et al., 2012), leading to interest in identifying and treating milder causes.

CHRONIC KIDNEY DISEASE MINERAL BONE DISORDER

Chronic kidney disease is associated with skeletal and soft-tissue abnormalities termed chronic kidney disease mineral bone disorder (CKD-MBD); the specific skeletal manifestations are termed renal osteodystrophy. Although CKD-MBD may coexist with osteoporosis, it should always be considered as a separate diagnosis in patients with CKD stage 4/5 (i.e. eGFR <30 mL/min) presenting with a low trauma fracture. Rarely, it may occur in patients with more mild degrees of renal impairment.

Pathology

In CKD-MBD, osteoblast function is impaired due to systemic consequences of CKD including acidosis and increased sclerostin levels. Despite defective osteoblast function at a cellular level, bone turnover is generally increased in part due to higher FGF23 levels. The latter, which are observed early in CKD, represent a physiological response to phosphate retention (a direct consequence of the reduced glomerular filtration rate), and until CKD stage 5 is reached (i.e. eGFR <15 mL/min), is generally successful in preventing phosphate retention by inhibiting phosphate tubular reabsorption. High bone turnover is also a consequence of raised PTH levels known as secondary hyperparathyroidism. The latter is a common feature of CKD, representing a physiological response to reduced levels of calcitriol as a consequence of impaired renal 1 alpha hydroxylase activity.

Three distinct abnormalities are seen pathologically:

- *High turnover disease* is observed most commonly. As well as evidence of increased bone turnover in the form of increased numbers of osteoblasts and osteoclasts, there may be typical features of hyperparathyroidism including osteitis fibrosa and woven bone.
- *Adynamic bone disease* is observed in a small proportion, with evidence of reduced bone turnover including reduced levels of bone resorption and particularly bone formation.
- *Mineralization* defects may also be present, as indicated by widened osteoid seams and defective mineralization on tetracycline labelling.

Clinical features

CKD-MBD is associated with an increased fracture risk, even at relatively mild levels of renal impairment. This may be exacerbated by concurrent risk factors for osteoporosis such as hypogonadism and glucocorticoid therapy. In more advanced CKD-MBD, when features of renal osteodystrophy are present, bone pain may occur, combined with fractures. In children, growth retardation occurs combined with deformities seen in rickets. Slipped epiphyses may also occur, particularly at the hips when it can cause limping (see Figure 7.18). A major clinical manifestation of CKD-MBD is vascular calcification, secondary to hyperphosphataemia and raised FGF23 levels, resulting in hypertension and increased cardiovascular mortality. In heterotopic calcification, a wide range of soft tissues may be affected, including periarticular tissues (tumoral calcinosis) (Hruska and Seifert, 2013).

X-rays

There are few specific skeletal manifestations of CKD-MBD radiologically. A rare manifestation is the 'rugger jersey spine' caused by osteosclerosis of the superior and inferior vertebral end plates. There may be evidence of increased soft-tissue calcification, particularly vascular calcification. DXA scans may not be helpful as fracture risk may be elevated in the presence of a relatively normal BMD.

Biochemistry

Raised PTH is a cardinal feature of CKD stage 5, often increasing to many times the upper normal limit; indeed, PTH levels less than two times the upper normal limit may be indicative of adynamic bone disease. Although calcium levels tend to fall as a consequence of reduced calcitriol, they are generally maintained within the normal range by secondary hyperparathyroidism. Extremely high PTH levels and hypercalcaemia may develop due to tertiary hyperparathyroidism as a result of autonomous unregulated PTH secretion, following prolonged secondary hyperparathyroidism.

Treatment

Hyperphosphataemia and secondary hyperparathyroidism can be treated by restricting the intake of phosphorus and taking phosphate binders, and by administering a vitamin D analogue, most commonly alfacalcidol. Calcium-sensing receptor agonists such as cinacalcet may also be used to inhibit PTH secretion. However, the biochemical changes are complex and treatment should always be managed by a renal specialist.

Drugs for treating osteoporosis can also be used to treat bone fragility in CKD patients, but in CKD stages 4 and 5 this is somewhat controversial. Oral bisphosphonates are probably safe but may need to be administered at lower dose or greater dosage interval. IV zoledronate should be avoided as it may precipitate acute renal failure. Denosumab runs the risk of acute hypocalcaemia, hence calcium levels should be checked 1–2 weeks after administration. Aside from general safety and renal toxicity, the efficacy of these agents in reducing fracture risk in CKD stages 4 and 5 is unclear. Extrapolation of their benefits in osteoporosis may justify their use, particularly in high bone turnover states given their anti-resorptive action. Theoretically, these agents may be less effective or even harmful in adynamic bone disease where bone turnover is already suppressed. Some advocate performing a bone biopsy to exclude adynamic bone disease before using these agents, but this is often not practical, and there is currently no clinical data to suggest that the efficacy of anti-osteoporotic drugs in CKD is predicted from bone biopsy.

HYPERCALCAEMIA

As previously mentioned, regulatory pathways described above, particularly PTH, are generally very effective at maintaining normocalcaemia, which is required for a number of essential cellular processes. Underlying causes of hypercalcaemia need to be identified and managed promptly. Causes of hypercalcaemia are listed in Box 7.4. The most important are primary hyperparathyroidism and malignancy.

Clinical features

Clinical features vary with the degree of hypercalcaemia: a mild elevation of serum calcium concentration may cause no more than general lassitude, polyuria and polydipsia. With plasma levels between 3 and 3.5 mmol/L, patients may complain of anorexia, nausea, muscle weakness and fatigue. Those with severe hypercalcaemia (<3.5 mmol/L) have a plethora of symptoms including abdominal pain, nausea, vomiting, severe fatigue and depression. In long-standing cases patients may develop kidney stones or nephrocalcinosis due to chronic hypercalciuria; some complain of joint symptoms, due to chondrocalcinosis. The clinical picture is summarized in the adage 'moans, groans, bones and stones'.

There may also be symptoms and signs of the underlying cause, which should always be sought.

Hyperparathyroidism is an important cause of hypercalcaemia. *Primary hyperparathyroidism* is usually due to an adenoma or hyperplasia. *Tertiary hyperparathyroidism* is when secondary hyperplasia leads to

BOX 7.4 CAUSES OF HYPERCALCAEMIA

Increased calcium/vitamin D resorption

Increased bone resorption

- Primary and tertiary hyperparathyroidism
- Malignancy
- Hyperthyroidism

Granulomatous disorders

- Sarcoidosis
- Tuberculosis
- Histoplasmosis

Drugs

- Lithium
- Thiazides
- Theophylline toxicity
- Vitamin A toxicity
- Vitamin D excess

Familial

- MEN I and II
- Familial hypocalciuric hypercalcaemia

Other

- Addison's disease
- Phaeochromocytoma
- Solid tumours
- Prolonged immobilization

autonomous overactivity. Both of these cause hypercalcaemia. Primary is more common and the diagnosis of this is supported by finding hypercalcaemia, hypophosphataemia and a raised serum PTH.

Malignancy is another important cause of hypercalcaemia and may be due to locally increased bone resorption resulting from secondary bony metastases or multiple myeloma, or more generalized increased bone resorption mediated by parathyroid hormone-related peptide (PTHrP) secreted by solid tumours. Multiple endocrine neoplasia (MEN) syndromes can also cause hypercalcaemia.

Familial hypocalciuric hypercalcaemia (FHH), caused by inactivating mutations in the gene for the calcium-sensing receptor, has a clinical spectrum of hypercalcaemia ranging from life-threatening disorders in the case of neonatal FHH to an asymptomatic biochemical abnormality found on a routine blood testing.

Investigations

Radiographs may show a variety of appearances depending on the underlying cause, including evidence of a primary lung tumour on chest radiography, or evidence of granulomatous diseases such as tuberculosis or sarcoidosis. Finding of a high serum calcium should be followed up by measurement of parathyroid hormone (PTH). If this is normal or high, 24-hour urinary calcium excretion should be measured. The combination of normal/high PTH plus normal or high urinary calcium excretion confirms primary or tertiary hyperparathyroidism. The combination of normal/high PTH plus low urinary calcium excretion suggests familial hypocalciuric hypercalcaemia. Suppressed levels of PTH in the context of hypercalcaemia require symptom-guided investigations for malignancy and tests for other endocrinopathies such as hyperthyroidism (measure thyroid-stimulating hormone, TSH) or adrenal insufficiency (measure cortisol).

Treatment

Acute severe hypercalcaemia is a medical emergency and needs to be promptly treated with intravenous fluids. Intravenous bisphosphonates and glucocorticoids can also be used as second line. Long-term management needs to be guided towards the underlying cause. Primary hyperparathyroidism is often treated with surgery.

HYPERPARATHYROIDISM

Excessive secretion of PTH may be *primary* (usually due to an adenoma or hyperplasia), *secondary* (due to persistent hypocalcaemia) or *tertiary* (when secondary hyperplasia leads to autonomous overactivity).

Pathology

Overproduction of PTH enhances calcium conservation by stimulating tubular absorption, intestinal absorption and bone resorption. The resulting hypercalcaemia increases glomerular filtration of calcium to such an extent that there is hypercalciuria despite the augmented tubular reabsorption. Urinary phosphate is also increased, due to suppressed tubular reabsorption. The main effects of these changes are seen in the kidney: calcinosis, stone formation, recurrent infection and impaired function. There may also be calcification of soft tissues.

There is a general loss of bone substance. In severe cases, osteoclastic hyperactivity produces subperiosteal erosions, endosteal cavitation and replacement of the marrow spaces by vascular granulations and fibrous tissue (osteitis fibrosa cystica). Haemorrhage and giant-cell reaction within the fibrous stroma may give rise to brownish, tumour-like masses, whose liquefaction leads to fluid-filled cysts.

Primary hyperparathyroidism

Primary hyperparathyroidism is usually caused by a solitary adenoma in one of the small glands. Patients are middle-aged (40–65 years) and women are affected twice as often as men. Many remain asymptomatic and are diagnosed only because routine biochemistry tests unexpectedly reveal a raised serum calcium level.

Clinical features

Symptoms and signs are mainly due to *hypercalcaemia*: anorexia, nausea, abdominal pain, depression, fatigue and muscle weakness. Patients may develop polyuria, kidney stones or nephrocalcinosis due to chronic hypercalciuria. Some complain of joint symptoms, due to chondrocalcinosis. Only a minority (probably less than 10%) present with bone disease; this is usually generalized osteoporosis rather than the classic features of osteitis fibrosa, bone cysts and pathological fractures.

X-rays

Typical radiographic features are osteoporosis (sometimes including vertebral collapse) and areas of cortical erosion (Figure 7.20). Hyperparathyroid 'brown tumours' should be considered in the differential diagnosis of atypical cyst-like lesions of long bones. The classical – and almost pathognomonic – feature, which should always be sought, is subperiosteal cortical resorption of the middle phalanges. Non-specific features of hypercalcaemia are renal calculi, nephrocalcinosis and chondrocalcinosis.

Biochemical tests

There may be hypercalcaemia, hypophosphataemia and a raised serum PTH concentration. Serum alkaline phosphatase is raised with osteitis fibrosa.

Diagnosis

It is necessary to exclude other causes of hypercalcaemia (*multiple myeloma*, *metastatic disease*, *sarcoidosis*) in which PTH levels are usually depressed. Hyperparathyroidism also comes into the differential diagnosis of all types of *osteoporosis* and *osteomalacia*.

Treatment

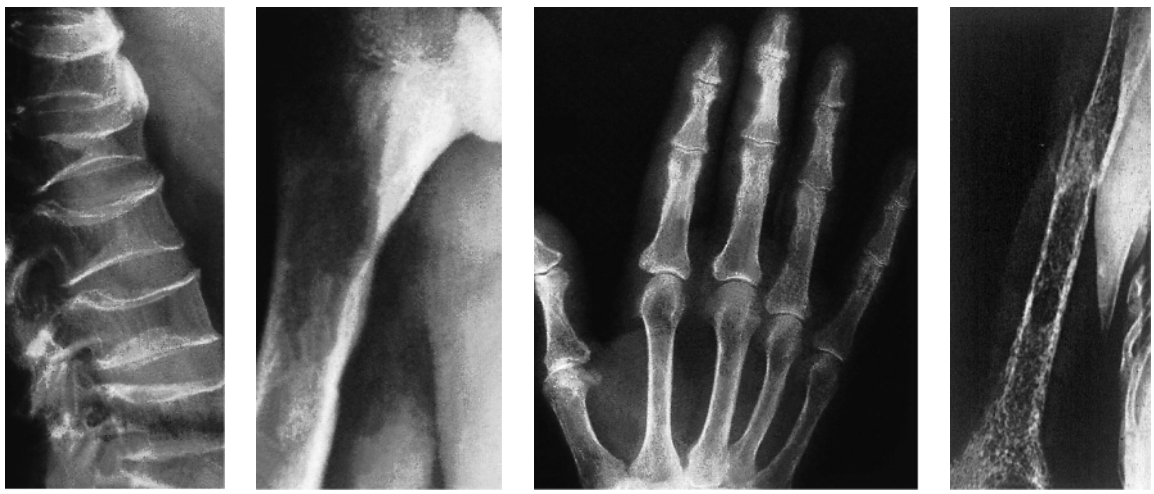
Treatment is usually conservative and includes adequate hydration and decreased calcium intake. The indications for parathyroidectomy are marked and unremitting hypercalcaemia, recurrent renal calculi, progressive nephrocalcinosis and severe osteoporosis.

Postoperatively there is a danger of severe hypocalcaemia due to brisk formation of new bone (the 'hungry bone syndrome'). This must be treated promptly, with one of the fast-acting vitamin D metabolites.

Hypercalcaemia of malignancy

Pathology

Hypercalcaemia of malignancy is due to either local or generalized increase in bone resorption. Secondary bony metastases (usually from prostate, breast, lung, kidney or thyroid primaries) or multiple myeloma cause local bone resorption due to cytokine induced



(a) (b) (c) (d)

Figure 7.20 Hyperparathyroidism (a) This hyperparathyroid patient with spinal osteoporosis later developed pain in the right arm; (b) an X-ray showed cortical erosion of the humerus; (c) he also showed typical erosions of the phalanges. (d) Another case, showing 'brown tumours' of the humerus and a pathological fracture.

bone lysis. More generalized increase in bone resorption can be mediated by parathyroid hormone-related peptide (PTHrP) secreted by solid tumours such as head and neck squamous cancers. Hodgkin's lymphoma causes hypercalcaemia through increased production of calcitriol. Multiple endocrine neoplasia (MEN) syndromes (benign or malignant tumours of endocrine tissues inherited in an autosomal dominant pattern) can also cause hypercalcaemia. In MEN type 1, hyperfunction of the parathyroid glands cause hypercalcaemia. In MEN type 2, the hypercalcaemia is associated with medullary thyroid carcinoma.

Clinical features

Symptoms and signs are mainly due to the malignancy plus the hypercalcaemia: localized pain due to bony metastases is a common presentation along with anorexia, nausea, abdominal pain, depression, fatigue and muscle weakness. Pathological fractures also occur, but often due to the malignant deposit in bone.

Investigations

Investigations for hypercalcaemia of malignancy need to be directed towards finding the primary tumour and should be based on symptoms, signs and basic blood tests. A history of smoking, cough, haemoptysis or shortness of breath should direct investigations towards the lungs, but there should be a low threshold for imaging (usually CT scanning) of the entire chest, abdomen and pelvis. A skeletal survey or nuclear medicine bone scan can be useful to identify all bone metastases (Figure 7.21). A myeloma screen (serum and urine electrophoresis) should be performed.

Treatment

Treatment of acute severe hypercalcaemia of malignancy should be with intravenous fluids. Intravenous bisphosphonates and glucocorticoids can also be used as second line. Management of the underlying malignancy can resolve the hypercalcaemia. Local radiotherapy to bone metastases can provide temporary

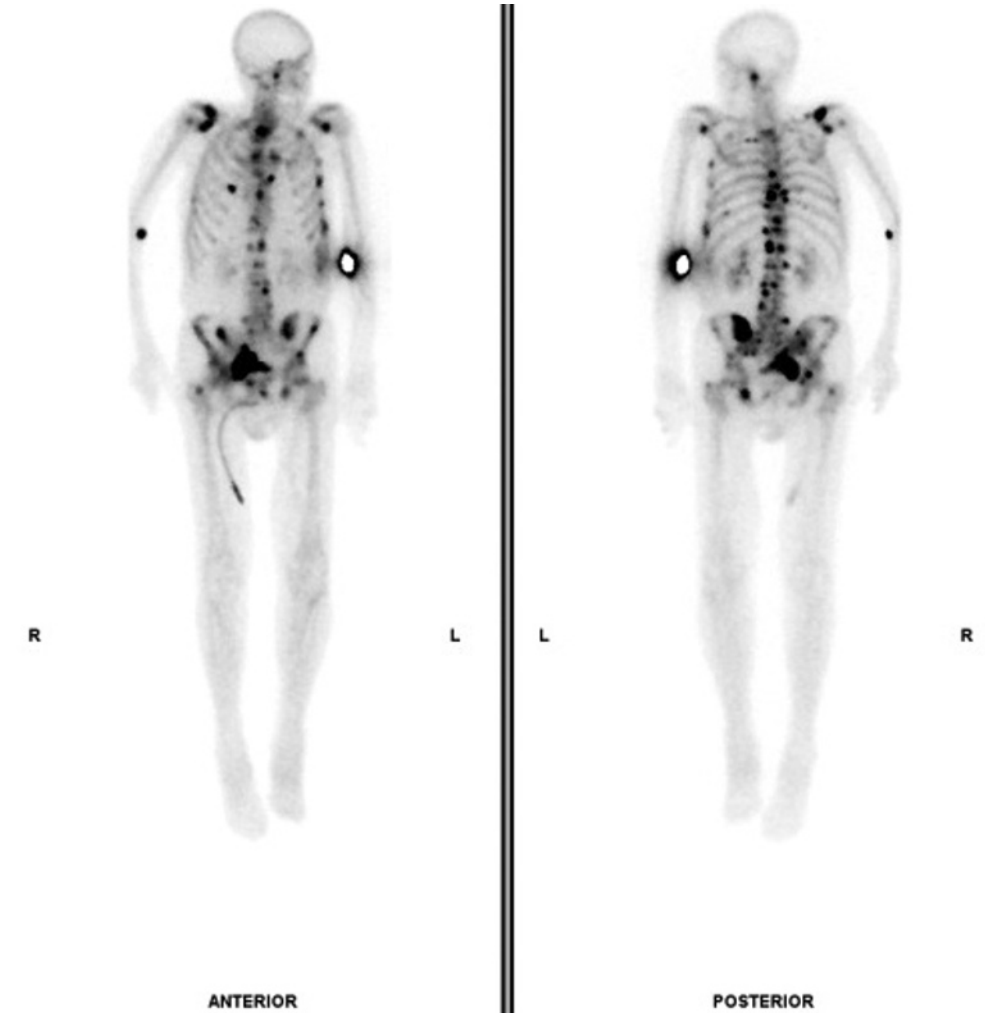


Figure 7.21 Nuclear medicine bone scan Multiple bony metastases can be seen clearly. (Image provided with kind permission from Dr Paul McCoubrie, Consultant Radiologist, North Bristol NHS Trust.)

relief of pain and hypercalcaemia caused by local bone resorption.

PAGET'S DISEASE OF BONE (PDB)

Paget's disease of bone (PDB) is characterized by localized sites of increased bone turnover followed by enlargement and thickening of the bone, but the internal architecture is abnormal and the bone is unusually brittle. It is largely restricted to people of Anglo-Saxon descent, and to North America, Britain, Western Europe and Australia. Recent declines in prevalence attest to the role of environmental factors though these remain elusive. Genetic factors also play an important role: 15–30% of cases are familial, of which around 30% are caused by *SQTM1* mutations (Siris and Roodman, 2013). Rare forms of severe, early-onset PDB are recognized, caused by activating mutations of the *RANKL* gene.

Pathology

PDB may appear in one or several sites; in the long bones it starts at the metaphysis and progresses slowly towards the diaphysis, leaving altered architecture behind. The characteristic cellular change is a marked increase in osteoclastic and osteoblastic activity. Bone turnover is accelerated, plasma alkaline phosphatase is raised (a sign of osteoblastic activity) and there is an increased serum level of collagen crosslinks as reflected by CTX (due to osteoclastic activity).

In the osteolytic (or 'vascular') stage there is avid resorption of existing bone by large osteoclasts, the excavations being filled with vascular fibrous tissue. In adjacent areas osteoblastic activity produces new woven and lamellar bone, which in turn is removed by osteoclasts. This alternating activity extends to both endosteal and periosteal surfaces, so the bone increases in thickness but is structurally weak and prone to deformation. Gradually, osteoclastic activity abates and the eroded areas fill with new lamellar bone, leaving an irregular pattern of cement lines that mark the limits of the old resorption cavities; these 'tidemarks' produce a marbled or mosaic appearance on microscopy (see Figure 7.22). In the late, osteoblastic, stage the thickened bone becomes increasingly sclerotic and brittle.

Clinical features

PDB affects men and women equally. Only occasionally does it present in patients under 50, but from that age onwards it becomes increasingly common. The disease may for many years remain localized to part or the whole of one bone – the pelvis and tibia are the commonest sites, and the femur, skull, spine and clavicle the next commonest.

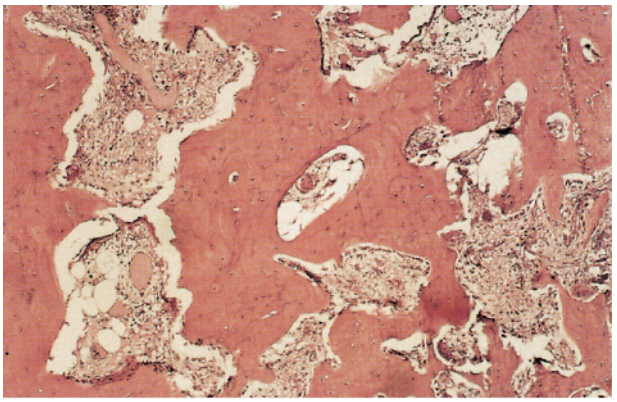


Figure 7.22 Paget's disease – histology Section from pagetic bone, showing the mosaic pattern due to overactive bone resorption and bone formation. The trabeculae are thick and patterned by cement lines. Some surfaces are excavated by osteoclastic activity while others are lined by rows of osteoblasts. The marrow spaces contain fibrovascular tissue.

Most people with PDB are asymptomatic, the disorder being diagnosed when an X-ray is taken for some unrelated condition or after the incidental discovery of a raised serum alkaline phosphatase level. When patients do present, it is usually because of pain or deformity, or some complication of the disease.

The pain is a dull constant ache, worse in bed when the patient warms up, but rarely severe unless a fracture occurs or sarcoma supervenes.

Deformities are seen mainly in the lower limbs. Long bones bend across the trajectories of mechanical stress; thus the tibia bows anteriorly and the femur anterolaterally (see Figure 7.23a). The limb looks bent and feels thick, and the skin is unduly warm - hence the term 'osteitis deformans'. If the skull is affected, it enlarges; the patient may complain that old hats no longer fit. The skull base may become flattened (platybasia), giving the appearance of a short neck.

Cranial nerve compression may lead to impaired vision, facial palsy, trigeminal neuralgia or deafness. Another cause of deafness is otosclerosis. Vertebral thickening may cause spinal cord or nerve root compression.

Steal syndromes, in which blood is diverted from internal organs to the surrounding skeletal circulation, may cause cerebral impairment and spinal cord ischaemia. If there is also spinal stenosis the patient develops typical symptoms of 'spinal claudication' and lower limb weakness.

X-rays

The appearances are characteristic. During the resorptive phase there may be localized areas of osteolysis;

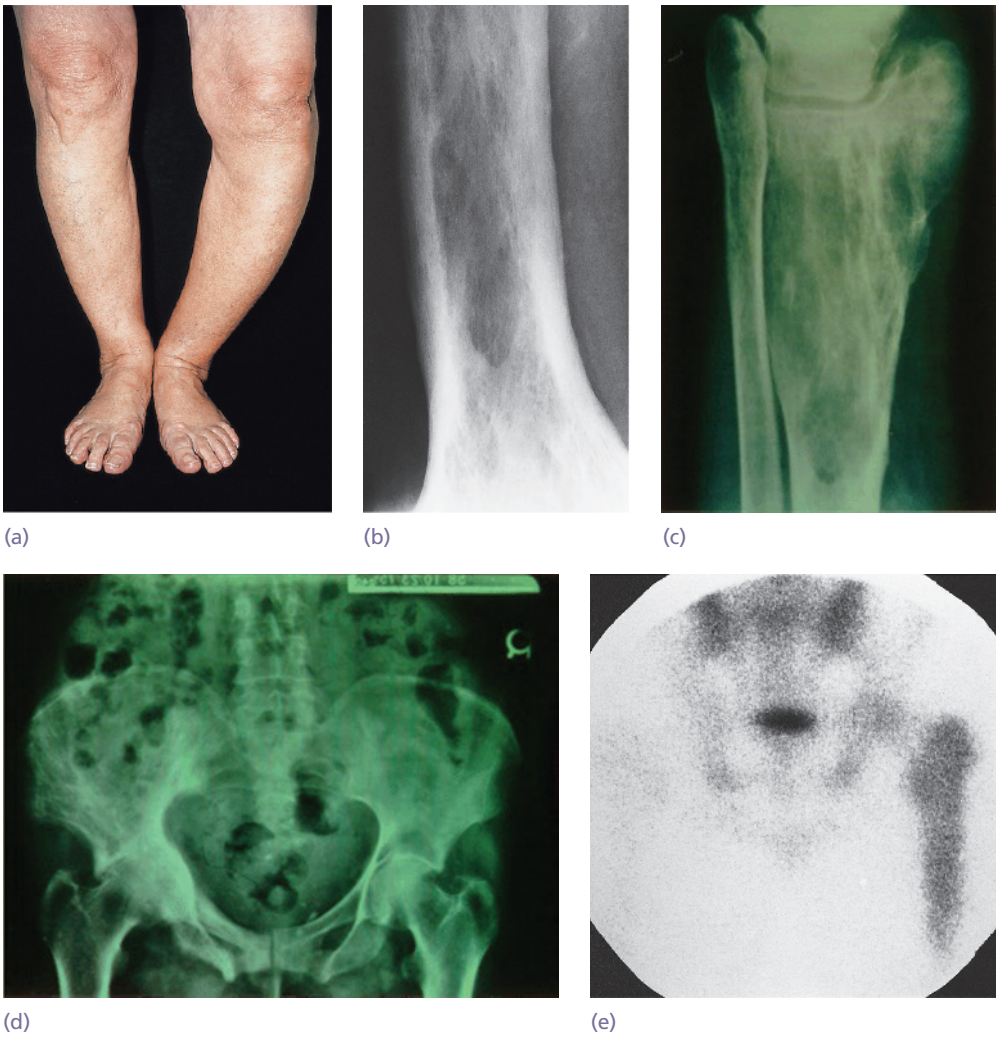


Figure 7.23 Paget's disease (a) Deformity of the tibia due to Paget's disease. (b) Flame-shaped area of osteopenia. (c,d) AP radiographs showing coarse trabecular patterning of the upper tibia and fibula (c), and right hemi-pelvis (d). (e) Radionucleotide scan showing increased activity in the left femur.

most typical is the flame-shaped lesion extending along the shaft of the bone (Figure 7.23b), or a circumscribed patch of osteoporosis in the skull (osteoporosis circumscripta). Later the bone becomes thick and sclerotic, with coarse trabeculation (Figure 7.23c,d). The femur or tibia sometimes develops fine cracks on the convex surface – stress fractures that heal with increasing deformity of the bone. Occasionally the diagnosis is made only when the patient presents with a pathological fracture. Radionucleotide scans can be helpful in showing the distribution of active lesions; the distribution of increased uptake within an affected bone is relatively extensive, which can be helpful in distinguishing from other causes such as malignant secondary deposits (Figure 7.23e).

Biochemical investigations

Serum calcium and phosphate levels are usually normal, though patients who are immobilized may develop hypercalcaemia. The most useful routine test is measurement of the serum ALP concentration (which reflects osteoblastic activity and extent of the disease), particularly the bone isoenzyme. ALP level can be useful in evaluating overall activity and response to treatment. It may remain within the normal range in monostotic PDB confined to a single site.

Complications

Fractures These are common, especially in the weight-bearing long bones. In the femoral neck they are often vertical; elsewhere the fracture line is usually partly transverse and partly oblique. In the femur there is a high rate of non-union; for femoral neck fractures prosthetic replacement and for shaft fractures early internal fixation are recommended. Small stress fractures may be very painful; they resemble Looser's zones on X-ray, except that they occur on convex surfaces.

Osteoarthritis OA of the hip or knee is not merely a consequence of abnormal loading due to bone deformity; in the hip it seldom occurs unless the innominate bone is involved. The X-ray appearances suggest an atrophic arthritis with sparse remodelling, and at operation joint vascularity is increased.

Nerve compression and spinal stenosis Occasionally the first abnormalities to be detected, these may call for definitive surgical treatment. Local bone hypertrophy may cause hearing loss.

Bone sarcoma Osteosarcoma arising in an elderly patient is almost always due to malignant transformation in Paget's disease. The frequency of malignant change is probably around 1%. It should always be suspected if a previously diseased bone becomes more painful, swollen and tender. Occasionally it presents as the first evidence of PDB. The prognosis is extremely grave.

High-output cardiac failure This is a rare but important general complication. It is due to prolonged, increased bone blood flow.

Hypercalcaemia If the patient is immobilized for prolonged periods, hypercalcaemia may occur.

Intra-operative bleeding Bleeding from cut bony surfaces is common.

Treatment

IV zoledronate given as a single infusion (dose 4–5 mg) is very effective at treating active PDB; as well as inducing remission in the great majority of patients, patients often remain in remission for several years thereafter. Whereas remission as reflected by a reduction in bone pain and ALP (which is often normalized) generally occurs, whether long-term sequelae, such as deafness and deformity, are prevented is currently unclear. Other than renal impairment (eGFR <30 mL/min), IV zoledronate has no major contraindications or cautions, and it is well tolerated apart from a risk of flu-like symptoms for 24 hours after the first infusion. IV zoledronate should be considered in patients with symptoms related to PDB, even if there is some uncertainty as in patients with pelvic involvement and hip pain in whom some of the symptoms may be attributable to hip OA. Treatment should also be considered in patients who are considered at imminent risk of fracture, for example the presence of flame-shaped lytic lesions of the femur or tibia on radiographs.

Surgery The main indication for operation is a pathological fracture, which (in a long bone) usually requires internal fixation. When the fracture is treated, the opportunity should be taken to straighten the bone. Other indications for surgery are painful osteoarthritis (total joint replacement), nerve entrapment (decompression) and severe spinal stenosis (decompression). Some sources advocate pretreatment with IV zoledronate to limit blood loss, which may be excessive in these cases.

FURTHER READING

Adami S. Bone health in diabetes: considerations for clinical management. *Curr Med Res Opin* 2009; **25**(5): 1057–72.

Bickerstaff DR, Douglas DL, Burke PH, *et al.* Improvement in the deformity of the face in Paget's disease treated with diphosphonates. *J Bone Joint Surg* 1990; **72B**: 132–6.

Black DM, Cummings SR, Karpf DB, *et al.* Randomised trial of effect of alendronate on risk of fracture in women with existing vertebral fractures. *Lancet* 1996; **348**: 1535–41.

Black DM, Bilezikian JP, Ensrud KE, *et al.* One year of alendronate after one year of parathyroid hormone. *N Engl J Med* 2005; **353**: 555–65.

Blake GM, Fogelman I. The clinical role of dual energy X-ray absorptiometry. *Eur J Radiol* 2009; **71**(3): 406–14.

Bone HG, Adami S, Rizzoli R, *et al.* Weekly administration of alendronate: rationale and plan for clinical assessment. *Clin Ther* 2000; **22**: 15–28.

Boonen S, Laan R, Barton I, *et al.* Effect of osteoporosis treatments on risk of non-vertebral fractures: Review and meta-analysis of intention to treat studies. *Osteoporos Int* 2005; **16**(10): 1291–8.

Brighton CT, McCluskey WP. Cellular response and mechanisms of action of electrically induced osteogenisis. In: Peck WA (ed.), *Bone and Mineral Research/4.* Amsterdam: Elsevier, 1986.

Brixen, K, Chapurlat R, Cheung AM, *et al.* Bone density, turnover, and estimated strength in postmenopausal women treated with odanacatib: a randomized trial. *J Clin Endocrinol Metab* 2013; **98**(2): 571–80.

Bullough PG. *Atlas of Orthopaedic Pathology: With Clinical and Radiological Correlations,* 2nd edn. Baltimore: University Park Press, 1985.

Chapurlat RD. Odanacatib: a review of its potential in the management of osteoporosis in postmenopausal women. *Ther Adv Musculoskel Dis* 2015; 7(3): 103–9.

Chapuy MC, Arlot ME, Delmas PD, *et al.* Effect of calcium and chlecalciferol treatment for three years on hip fractures in elderly women. *BMJ* 1994; **308**: 1081–2.

Coudert AE, de Vernejoul MC, Muraca M, *et al.* Osteopetrosis and its relevance for the discovery of new functions associated with the skeleton. *Int J Endocrinol* 2015; DOI 10.1155/2015/372156.

Cummings SR, Black DM, Nevitt MC, *et al.* Bone density at various sites for prediction of hip fractures. *Lancet* 1993; **341**: 72–5.

Cummings SR, San Martin J, McClung MR, *et al.* Denosumab for prevention of fractures in postmenopausal women with osteoporosis. *N Eng J Med* 2005; **361**: 756–65.

Delmas P, Ensrud K, Adachi J, *et al.* Efficacy of raloxifene on vertebral fracture risk reduction in postmenopausal women with osteoporosis: four-year results from a randomized clinical trial. *J Clin Endocrinol Metab* 2002; **87**: 3609–17.

Duan Y, Beck TJ, Wang XF, *et al.* Structural and biomechanical basis of sexual dimorphism in femoral neck fragility has its origins in growth and aging. *J Bone Miner Res* 2003; **18**: 1766–74.

El Hajj Fuleihan G, Testa MA, Angell JE, *et al.* Reproducibility of DXA absorptiometry: a model for bone loss estimates. *J Bone Miner Res* 1995; **10**: 1004–14.

Estrada K, Styrkarsdottir U, Evangelou E. Genome-wide meta-analysis identifies 56 bone mineral density loci and reveals 14 loci associated with risk of fracture. *Nature Genet* 2012; **44**(5): 491–501.

Frost HM. Bone 'mass' and the 'mechanostat': a proposal. *Anatomical Record* 1987; **219**, 1–9.

Genant HK, Engelke K, Fuerst T, *et al.* Noninvasive assessment of bone mineral and structure: state of the art. *J Bone Miner Res* 1996; **11**: 707–30.

Gov.uk. Vitamin D – advice on supplements for at risk groups. Letter from UK Chief Medical Officers, 2012. Available at: https://www.gov.uk/government/publications/vitamin-d-advice-on-supplements-for-at-risk-groups [Accessed 23 February 2017]

Hahn TJ. Drug-induced disorders of vitamin D and mineral metabolism. *Clin Endocrinol Metab* 1980; **9**: 107–29.

Horowitz MC. Cytokines and estrogen in bone: anti-osteoporotic effects. *Science* 1993; **260**: 626–7.

Hruska KA, Seifert M. Pathophysiology of CKD-MBD. In: Rosen CJ (ed.), *Primer on the Metabolic Bone Diseases and Disorders of Mineral Metabolism*. Oxford: Wiley-Blackwell, 2013, pp. 632–9.

HYP Consortium. A gene (PEX) with homologies to endopeptidases is mutated in patients with X-linked hypophosphatemic rickets. *Nature Genet* 1995; **11**: 130–6.

Liberman UA, Downs RW Jr, Dequeker J, *et al.* Effect of oral alendronate on bone mineral density and the incidence of fractures in postmenopausal osteoporosis. *N Engl J Med* 1995; **333**: 1437–43.

Lindsay R, Hart DM, Clark DM. The minimum effective dose of estrogen for prevention of postmenopausal bone loss. *Obstet Gynecol* 1984; 63: 759–63.

Masud T, Mootoosamy I, McCloskey EV, *et al.* Assessment of osteopenia from spinal radiographs using two different methods: the Chingford study. *Br J Radiol* 1996; **69**: 451–6.

McClung MR, Grauer A, Boonen S, *et al.* Romosozumab in postmenopausal women with low bone mineral density. *N Engl J Med* 2014; **370**(5): 412–20.

McClung MR, Lewiecki ME, Cohen S, *et al.* Denosumab in postmenopausal women with low bone mineral density. *N Engl J Med* 2006; **354**: 821–31.

Meunier P, Roux C, Seeman E, *et al.* The effects of strontium ranelate on the risk of vertebral fracture in women with postmenopausal osteoporosis. *N Engl J Med* 2004; **350**: 459–68.

National Institute for Health and Care Excellence (NICE). Clinical guideline CG146: *Osteoporosis: Assessing the risk of fragility fracture*, 2012. Available at: https://www.nice.org.uk/guidance/cg146 [Accessed 22 February 2017]

Nesbitt T, Drezner MK. Hepatocyte production of phosphatonin in HYP mice. *J Bone Miner Res* 1996; **11**: S136.

Pak CYC, Sakhall K, Adams-Huet B, *et al.* Treatment of postmenopausal osteoporosis with slow-release sodium fluoride. *Ann Intern Med* 1995; **123**: 401–8.

Parfitt AM. Bone remodelling: relationship to the amount and structure of bone, and the pathogenesis and prevention of fractures. In: Riggs BL, Melton LJ III (eds.), *Osteoporosis*. New York: Raven Press, 1988, pp. 45–93.

Peck WA, Woods WL. The cells of bone. In: Riggs BL, Melton LJ III (eds.), *Osteoporosis*. New York: Raven Press, 1988, pp. 1–44.

Pleiner-Duxneuner J, Zwettler E, Paschalis E, *et al.* Treatment of osteoporosis with parathyroid hormone and teriparatide. *Calcif Tissue Int* 2009; **84**(3): 159–70.

Rebel A, Basle M, Poulard A, *et al.* Towards a viral aetiology for Paget's disease of bone. *Metab Bone Dis Relat Res* 1981; 3: 235–8.

Rihn JA, Gates C, Glassman SD, *et al.* The use of bone morphogenetic protein in lumbar spine surgery. *J Bone Joint Surg Am* 2008; **90**(9): 2014–25.

Rosen HN, Dresner-Pollak R, Moses AC, *et al.* Specificity of urinary excretion of cross-linked N-telopeptides of type I collagen as a marker of bone turnover. *Calcif Tissue Int* 1994; **54**: 26–9.

Rubin LT, Lanyon CE. Regulation of bone formation by applied dynamic loads. *J Bone and Joint Surg* 1984; **66A**: 397–40.

Ruppe DR, Jan de Beur SM. Disorders of phosphate homeostasis. In: Rosen CJ (ed.), *Primer on the Metabolic Bone Diseases and Disorders of Mineral Metabolism*. Oxford: Wiley-Blackwell, 2013, pp. 601–12.

Seeman E. Periosteal bone formation: a neglected determinant of bone strength. *New Engl J Med* 2003; **349**: 320–3.

Siris ES, Roodman GD. Paget's disease of bone. In: Rosen CJ (ed.), *Primer on the Metabolic Bone Diseases and Disorders of Mineral Metabolism*. Oxford: Wiley-Blackwell, 2013, pp. 659–68.

Skerry TM, Bitensky L, Chayen J, *et al.* Early strain-related changes in enzyme activity in osteocytes following bone loading in vivo. *J Bone Miner Res* 1989; **4**: 783–8.

Wolf J. *The Law of bone remodelling*. Berlin Heiderlberg New York: Springer, 1986 (translation of the German 1892 edition).

World Health Organization. Assessment of fracture risk and its application to screening for postmenopausal osteoporosis. Report of a WHO Study Group. WHO Tech Rep Ser. 1994; **843**: 1–129.

Whyte MP. Enzyme defects of the skeleton. In: Rosen CJ (ed.), *Primer on the Metabolic Bone Diseases and Disorders of Mineral Metabolism*. Oxford: Wiley-Blackwell, 2013, pp. 838–42.

Whyte MP, Greenberg CR, Salman NJ, *et al.* Enzyme-replacement therapy in life-threatening hypophosphatasia. *N Engl J Med* 2012, **366**(10): 904–13.

Whyte MP, Reinus WH, Mumm S. High-bone mass disease and LRP5. *N Engl J Med* 2004; **350**: 2096–9.

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Genetic disorders, skeletal dysplasias and malformations

8

Fergal Monsell, Martin Gargan, Deborah Eastwood, James Turner & Ryan Katchky

There can be few diseases in which genetic factors do not play a role – if only in creating a background favourable to the operation of some more proximate pathogen. Sometimes, however, a genetic defect is the major – or the only – determinant of an abnormality that is either present at birth (e.g. achondroplasia) or evolves over time (e.g. Huntington's chorea). Such conditions can be broadly divided into three categories: *chromosome disorders, single gene disorders* and *polygenic* or *multifactorial disorders*. Various anomalies may also result from *injury to the formed embryo*. Many of these conditions affect the musculoskeletal system, producing cartilage and bone dysplasia (abnormal bone growth and/or modelling), *malformations* (e.g. absence or duplication of certain parts) or *structural defects of connective tissue*. In some, a specific *metabolic abnormality* has been identified.

Genetic influences also contribute to the development of many *acquired disorders*. Osteoporosis, for example, is the result of a multiplicity of endocrine, dietary and environmental factors, yet twin studies have shown a significantly closer concordance in bone mass between identical twins than between non-identical twins.

Before considering the vast range of developmental disorders, it may be helpful to review certain general aspects of genetic abnormalities.

THE HUMAN GENOME

Each cell (apart from germ cells) in the human body contains within its nucleus 46 *chromosomes*, each of which consists of a single molecule of *deoxyribonucleic acid (DNA)*; unravelled, this life-imparting molecule would be several centimetres long, a double-stranded chain along which thousands of segments are defined and demarcated as *genes*. A small amount of DNA is also found within the mitochondria of the cell and this is termed the mitochondrial DNA.

Each gene consists of a group of nucleotides and every nucleotide contains a deoxyribose sugar, a phosphate molecule and either a purine base (adenine or guanine) or a pyrimidine (thymine or cytosine) base. Some genes are comparatively large and some much smaller. They are the basic units of inherited biological information, each one coding for the synthesis of a specific protein. Working as a set (or *genome*), they 'tell' the cells how to develop, differentiate and function in specialized ways.

Chromosomes can be identified and numbered by microscopic examination of suitably prepared blood cells or tissue samples; the *cell karyotype* defines its chromosomal complement. *Somatic (diploid) cells* should have 46 chromosomes: 44 (numbers 1–22), called *autosomes*, are disposed in 22 homologous pairs – one of each pair being derived from the mother and one from the father, both carrying the same type of genetic information; the remaining 2 chromosomes are the *sex chromosomes,* females having two X chromosomes (one from each parent) and males having one X chromosome from the mother and one Y chromosome from the father. *Germ-line cells* (eggs and sperm) have a *haploid* number of chromosomes (22 plus either an X or a Y). This is the *euploidic* situation; abnormalities of chromosome number would lead to an *aneuploidic* state.

Gene studies are complicated and involve the mapping of molecular sequences by specialized techniques after fragmenting the chains of DNA by means of restriction enzymes. Each gene occurs at a specific point, or *locus*, on a specific chromosome. The chromosomes being paired, there will be two forms, or *alleles*, of each gene (one maternal, one paternal) at each locus; if the two alleles coding for a particular trait are identical, the person is said to be *homozygous* for that trait; if they are not identical, the individual is *heterozygous*. Some chromosomes contain only a few genes (e.g. chromosomes 13, 18 and 21) whereas others contain many more (e.g. 17, 19 and 22).

The full genetic make-up of an individual is called the *genotype*. The finished person – a product of inherited traits and environmental influences – is the *phenotype*. An important part of the unique human genotype is the *major histocompatibility complex (MHC)*, also known as the *HLA system* (after human leucocyte antigen). This is a cluster of genes on chromosome 6 that is responsible for immunological specificity. The proteins for which they code are attached to cell surfaces and act as 'chaperones' for foreign antigens which have to be accompanied by HLA before they are recognized and engaged by the body's T-cells. HLA proteins can be identified by serological tests and are registered according to their corresponding genetic loci on the short arm of chromosome 6. HLA typing is particularly important in tissue transplantation: acceptance or rejection of the transplant hinges on the degree of matching between the HLA genes of donor and recipient.

Genetic mutation

A mutation is any permanent change in DNA sequencing or structure. Such changes in a somatic cell are characteristic of malignancy. In a germ-line cell, mutations contribute to generational diversity. Some genes have many forms (or mutations) and the Human Genome Project has identified thousands of single nucleotide polymorphisms (SNPs).

POINT MUTATIONS

The substitution of one nucleotide for another is the most common type of mutation. The effect varies from production of a more useful protein to a new but functionless protein, or an inability to form any protein at all; the result may be compatible with an essentially normal life or it may be lethal.

DELETIONS/INSERTIONS

Deletion or insertion of a segment in the gene chain can result in an unusual protein being synthesized, perhaps a more advantageous one but maybe one that is non-functional or one that has a dire effect on tissue structure and function (e.g. production of a shortened dystrophin protein in the Becker variant of muscular dystrophy).

GENETIC DISORDERS

Any serious disturbance of either the quantity or the arrangement of genetic material may result in disease. Three broad categories of abnormality are recognized: chromosome disorders, single gene disorders, and polygenic or multifactorial disorders.

Chromosome disorders Additions, deletions and changes in chromosomal structure usually have serious effects; affected fetuses are either stillborn or become infants with severe physical and mental abnormalities. In live-born children there are a few chromosome disorders with significant orthopaedic abnormalities: *Down's syndrome*, in which there is one extra chromosome 21 (trisomy 21), *Turner's syndrome*, in which one of the X chromosomes is lacking (monosomy X), and *Klinefelter's syndrome*, in which there is one Y but several X chromosomes.

Single gene disorders Gene mutation may occur by insertion, deletion, substitution or fusion of amino acids or nucleotides in the DNA chain. This can have profound consequences for cartilage growth, collagen structure, matrix patterning and marrow cell metabolism. The abnormality is then passed on to future generations according to simple mendelian rules (see below). There are literally thousands of single gene disorders, accounting for over 5% of child deaths, yet it is rare to see any one of them in a lifetime of orthopaedic practice.

Polygenic and multifactorial disorders Many normal traits (body build, for example) derive from the interaction of multiple genetic and environmental influences. Likewise, certain diseases have a polygenic background, and some occur only when a genetic predisposition combines with an appropriate environmental 'trigger'. *Gout*, for example, is more common than usual in families with hyperuricaemia: the uric acid level is a polygenic trait, reflecting the interplay of multiple genes; it is also influenced by diet and may be more than usually elevated after a period of overindulgence; finally, a slight bump on the toe acts as the proximate trigger for an acute attack of gout.

Non-genetic developmental disorders

Many developmental abnormalities occur sporadically and have no genetic background. Most of these are of unknown aetiology, but some have been linked to specific teratogenic agents which damage the embryo or the placenta during the first few months of gestation. Suspected or known teratogens include viral infections (e.g. rubella), certain drugs (e.g. thalidomide) and ionizing radiation. The clinical features are usually asymmetrical and localized, ranging from mild morphological defects to severe malformations such as spina bifida or phocomelia ('congenital amputations').

PATTERNS OF INHERITANCE

The single gene disorders have characteristic patterns of inheritance, which may be *autosomal* or *X-linked*, and *dominant* or *recessive*.

Autosomal dominant disorders Autosomal dominant disorders are inherited even if only one of a pair of alleles on a non-sex chromosome is abnormal; the condition is said to be *heterozygous*. A typical example is hereditary multiple exostoses. Either parent may be affected and half the children of both sexes develop exostoses. The pedigree shows a 'vertical' pattern of inheritance, with several affected siblings in successive generations (Figure 8.1a).

Sometimes both parents appear to be normal: the patient may be the first member of the family to suffer the effects of a mutant gene; or (as often happens) the disease shows variable expressivity, some members of the family (in the above example) developing many large exostoses and severe bone deformities, while others have only a few small and well-disguised nodules.

Autosomal recessive disorders These disorders appear only when both alleles of a pair are abnormal – i.e. the condition is always *homozygous*. Each parent contributes a faulty gene, though if both are heterozygous they themselves will be clinically normal. Theoretically 1 in 4 of the children will be homozygous and will therefore develop the disease; 2 out of 4 will be *heterozygous carriers* of the faulty gene. The typical pedigree shows a 'horizontal' pattern of inheritance: several siblings in one generation are affected but neither their parents nor their children have the disease (Figure 8.1b).

X-linked disorders These conditions are caused by a faulty gene in the X chromosome. Characteristically, therefore, they never pass directly from father to son because the father's X chromosome inevitably goes to the daughter and the Y chromosome to the son. *X-linked dominant disorders* (e.g. hypophosphataemic rickets) pass from an affected mother to half of her daughters and half of her sons, or from an affected father to all of his daughters but none of his sons. Not surprisingly, they are twice as common in girls as in boys. *X-linked recessive disorders* – of which the most notorious is haemophilia – have a highly distinctive pattern of inheritance (Figure 8.1c): an affected male will pass the gene only to his daughters, who will become unaffected heterozygous carriers; they, in turn, will transmit it to half of their daughters (who will likewise be carriers) and half of their sons (who will be affected individuals).

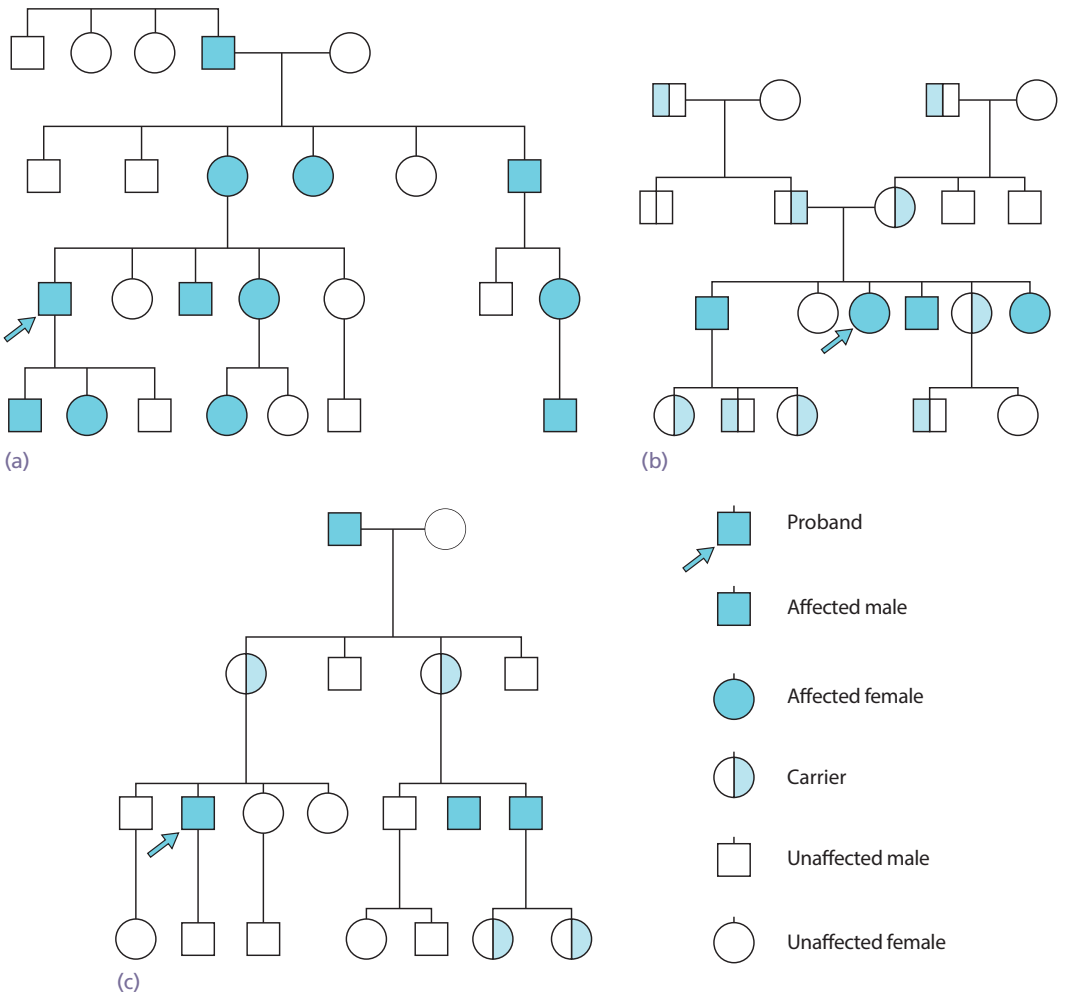

Figure 8.1 Patterns of inheritance (a) Autosomal dominant; (b) autosomal recessive; (c) X-linked recessive.

Inbreeding

All types of genetic disease are more likely to occur in the children of consanguineous marriages or in closed communities where many people are related to each other. The rare recessive disorders, in particular, are seen in these circumstances, where there is an increased risk of a homozygous pairing between two mutant genes.

Genetic heterogenicity

The same phenotype (i.e. a patient with a characteristic set of clinical features) can result from widely different gene mutations. For example, there are four different types of osteogenesis imperfecta (brittle bone disease), some showing autosomal dominant and some autosomal recessive inheritance. Where this occurs, the recessive form is usually the more severe. Subtleties of this kind must be borne in mind when counselling parents.

Genetic markers

Many common disorders show an unusually close association with certain blood groups, tissue types or other serum proteins that occur with higher than expected frequency in the patients and their relatives. These are referred to as genetic markers; they arise from gene sequences that do not cause the disease but are either 'linked' to other (abnormal) loci or express some factor that predisposes the individual to a harmful environmental agent. A good example is ankylosing spondylitis: over 90% of patients, and 60% of their first-degree relatives, are positive for HLA-B27. In this case (as in other autoimmune diseases) the HLA marker gene may provide the necessary conditions for invasion by a foreign viral fragment.

Gene mapping

With advancing recombinant DNA technology, the genetic disorders are gradually being mapped to specific loci. In some cases (e.g. Duchenne muscular dystrophy) the mutant gene itself has been cloned, holding out the possibility of effective treatment in the future.

PRE-NATAL DIAGNOSIS

Many genetic disorders can be diagnosed before birth, thus improving the chances of treatment or, at worst, giving the parents the choice of selective abortion. Ultrasound imaging is harmless and is now done almost routinely. On the other hand, tests that involve amniocentesis or chorionic villus sampling carry a risk of injury to the fetus and are therefore used only when there is reason to suspect some abnormality. Indications are:

- maternal age over 35 years (increased risk of Down's syndrome) or an unduly high paternal age (increased risk of achondroplasia)
- a previous history of chromosomal abnormalities (e.g. Down's syndrome) or genetic abnormalities amenable to biochemical diagnosis (neural tube defects, or inborn errors of metabolism) which will benefit from prompt neonatal treatment
- to confirm non-invasive tests suggesting an abnormality.

Maternal screening

Fetal neural tube defects are associated with increased levels of alpha-fetoprotein (AFP) in the amniotic fluid and, to a lesser extent, the maternal blood. Women with positive blood tests may be given the option of further investigation by amniocentesis. It has also been noted that abnormally low levels of AFP are associated with Down's syndrome.

Fetal cells may be present in maternal plasma and in the near future it is possible that genetic testing of these cells will be possible.

Amniocentesis

Under local anaesthesia, a small amount (about 20 mL) of fluid is withdrawn from the amniotic sac with a needle and syringe. (It is best to determine the position of the fetus beforehand by ultrasonography.) The procedure is usually carried out between the 12th and 15th weeks of pregnancy. The fluid can be examined directly for AFP and desquamated fetal cells can be collected and cultured for chromosomal studies and biochemical tests for enzyme disorders. It is well to remember that this procedure carries a small risk (0.5–0.75% of cases) of losing the fetus.

Chorionic villus sampling

Under ultrasound screening, a fine catheter is passed through the cervix and a small sample of chorion is sucked out. This is usually done between the 10th and 12th weeks of pregnancy. Mesenchymal fibroblasts can be cultured and used for chromosomal studies, biochemical tests and DNA analysis. Rapid advances in DNA technology have made it possible to diagnose sickle-cell anaemia and haemophilia (among other disorders) during early pregnancy, but spina bifida cannot be tested for. The procedure-related fetal loss rate is about 1%.

Pre-implantation genetic diagnosis

With assisted reproductive technologies such as *in vitro* fertilization (IVF), genetic abnormalities in the

embryos can be detected prior to implantation, thus allowing only 'healthy' embryos (as far as technology can tell) to be implanted into the mother.

Fetal imaging

High-resolution ultrasonography should provide images of all the long bones and joint movements by 11 weeks of gestation. Bone lengths increase linearly with time and by 18–23 weeks all three segments of each limb are clearly visible; a single measurement of one bone can be used to estimate growth. A femoral length that is normal for the fetal age is very significant in excluding many of the skeletal dysplasias or malformations; even with mesomelic abnormalities where the lower leg is most affected, the femur is likely to be short. By the 18th week of pregnancy anatomical abnormalities such as open neural tube defects and short limbs should be visible.

DIAGNOSIS IN CHILDHOOD

Clinical features

Tell-tale features suggesting skeletal dysplasia are:

- retarded growth and shortness of stature
- disproportionate length of trunk and limbs
- localized malformations (dysmorphism)
- soft-tissue contractures
- childhood deformity.

All the skeletal dysplasias affect growth, although this may not be obvious at birth. Children should be measured at regular intervals and a record kept of height, length of lower segment (top of pubic symphysis to heel), upper segment (pubis to cranium), span, head circumference and chest circumference. Failure to reach the expected height for the local population group should be noted, and marked shortness of stature is highly suspicious.

Bodily proportion is as important as overall height (Figure 8.2). The normal upper segment:lower segment ratio changes gradually from about 1.5:1 at the end of the first year to about 1:1 at puberty. *Shortness of stature with normal proportions is not necessarily abnormal*, but it is also seen in endocrine disorders which affect the different parts of the skeleton more or less equally (e.g. hypopituitarism). By contrast, *small stature with disproportionate shortness of the limbs is characteristic of skeletal dysplasia*, the long bones being more markedly affected than the axial skeleton.

The different segments of the limbs also may be disproportionately affected. The subtleties of dysplastic growth are reflected in terms such as *rhizomelia* – unusually short proximal segments (humeri and femora), *mesomelia* – short middle segments (forearms and legs) and *acromelia* – hands and feet.

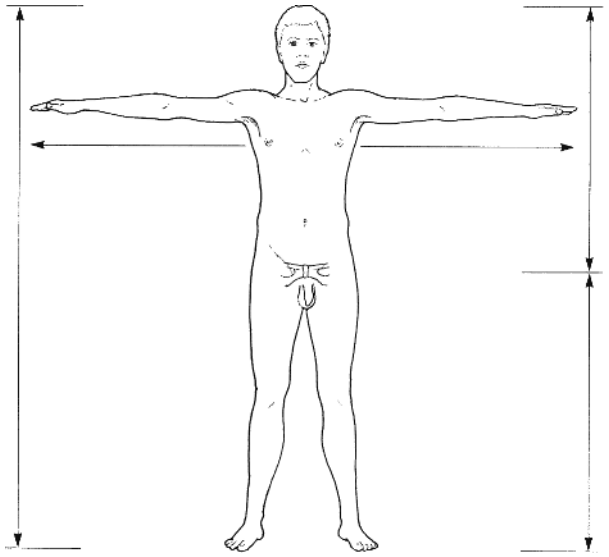

Figure 8.2 Normal proportions Upper segment = lower segment; total height = span.

Dysmorphism (a misshapen part of the body) is most obvious in the face and hands. There is a remarkable consistency about these changes, which makes for a disturbing similarity of appearance in members of a particular group.

Local deformities – such as kyphosis, valgus or varus knees, bowed forearms and ulnar deviated wrists – result from disturbed bone growth.

X-rays

The presence of any of the above features calls for a limited radiographic survey: a posteroanterior view of the chest, anteroposterior views of the pelvis, knees and hands, additional views of one arm and one leg, a lateral view of the thoracolumbar spine and standard views of the skull. Fractures, bent bones, exostoses, epiphyseal dysplasia and spinal deformities may be obvious, especially in the older child. Sometimes a complete survey is needed and it is important to note which portion of the long bones (epiphysis, metaphysis or diaphysis) is affected. With severe and varied changes in the metaphyses, periosteal new bone formation or epiphyseal separation, always consider the possibility of non-accidental injuries – the 'battered baby' syndrome.

Special investigations

In many cases the diagnosis can be made without laboratory tests; however, routine blood and urine analysis may be helpful in excluding metabolic and endocrine disorders such as rickets and pituitary or thyroid dysfunction. Special tests are also available to

identify specific excretory metabolites in the storage disorders, and specific enzyme activity can be measured in serum, blood cells or cultured fibroblasts.

Bone biopsy is occasionally helpful in disorders of bone density.

Direct testing for gene mutations is already available for a number of conditions and is rapidly being extended to others. It is a useful adjunct to clinical diagnosis. Still somewhat controversial is its application to pre-clinical diagnosis of late-onset disorders and neonatal screening for potentially dangerous conditions such as sickle-cell disease.

Previous medical history

Always ask whether the mother was exposed to teratogenic agents (X-rays, cytotoxic drugs or virus infections) during the early months of pregnancy.

The family history

A careful family history should always be obtained. This should include information about similar disorders in parents and close relatives, previous deaths in the family (and the cause of death), abortions and consanguineous marriages. However, the fact that parents or relatives are said to be 'normal' does not exclude the possibility that they are either very mildly affected or have a biochemical defect without any physical abnormality. Many developmental disorders have characteristic patterns of inheritance which may be helpful in diagnosis.

Racial background is sometimes important: some diseases are particularly common in certain communities; for example, sickle-cell disease in people of African descent and Gaucher's disease in Ashkenazi Jews.

DIAGNOSIS IN ADULTHOOD

It is unusual for a patient to present in adulthood with a condition that has been present since birth but in milder cases the abnormality may not have been recognized, particularly when several members of the family are similarly affected.

In the worst of the genetic disorders the fetus is stillborn or survives for only a short time. Individuals who reach adulthood, though recognizably abnormal, may lead active lives, marry and have children of their own. Nevertheless, they often seek medical advice for several reasons:

- short stature – especially disproportionate shortness of the lower limbs
- local bone deformities or exostoses
- spinal stenosis
- repeated fractures
- secondary osteoarthritis (e.g. due to epiphyseal dysplasia)
- joint laxity or instability.

The clinical approach is similar to that employed with children.

PRINCIPLES OF MANAGEMENT

Management of the individual patient depends on the diagnosis, the pattern of inheritance, the type and severity of deformity or disability, mental capacity and social aspirations. However, it is worth noting some general principles.

Communication

Once the diagnosis has been made, the next step is to explain as much as possible about the disorder to the patient (if old enough) and the parents without causing unnecessary distress. This is a skill that the orthopaedic surgeon must develop. Nowadays, with quick and easy access to the internet, it is relatively easy to obtain useful information about almost any condition, which the clinician can pass on in simple language. Rare developmental disorders are best treated in a centre that offers a 'special interest' team consisting of a paediatrician, medical geneticist, orthopaedic surgeon, psychologist, social worker, occupational therapist, orthotist and prosthetist.

Counselling

Patients and families may need expert counselling about (1) the likely outcome of the disorders; (2) what will be required of the family; and (3) the risk of siblings or children being affected. Where there are severe deformities or mental disability, the entire family may need counselling.

Maintaining an independent lifestyle

Parents are often anxious about having their child grow up as 'normal' as possible, yet 'normality' may mean something different for the child. For example, it is expected that children will become independently mobile only by learning to walk in a safe and effective manner, but some children with genetic disorders may be equally independently mobile with the use of a wheelchair. Management must be influenced by goals for adult life and not just the short-term goals of childhood.

Intrauterine surgery

The concept of operating on the unborn fetus is already a reality and is likely to be extended in the

future. At present, however, it is still too early to say whether the advantages (e.g. prenatal skin closure for dysraphism) will outweigh the risks.

Prevention and correction of deformities

Realignment of the limb, correction of ligamentous laxity and/or joint reconstruction can improve the stability and efficiency of gait and reduce the risk of secondary joint degenerative change.

Anomalies such as coxa vara, genu valgum, club foot, radial club hand or scoliosis (and many others outside the field of orthopaedics) are amenable to corrective surgery. In recent years, with advances in methods of limb lengthening, many short-limbed patients have benefited from this operation; however, the risks should be carefully explained and the expected benefits should not be exaggerated.

Several developmental disorders are associated with potentially dangerous spinal anomalies: for example, spinal stenosis and cord compression in achondroplasia; atlantoaxial instability, due to odontoid aplasia, in any disorder causing vertebral dysplasia; or severe kyphoscoliosis, which occurs in a number of conditions. Cord decompression or occipitocervical fusion are perfectly feasible, but surgical correction of congenital kyphoscoliosis carries considerable risks and should be undertaken only in specialized units. When considering the need for surgery, it must be remembered that some of these patients have a significantly reduced walking tolerance and hence improvements in limb alignment or length, for example, may not bring about any significant functional change. Conservative measures such as physiotherapy and splinting still have an important role to play.

Gene therapy

Gene therapy is still at the experimental stage. A carrier molecule or vector (often a virus that has been genetically modified to carry some normal human genetic material) is used to deliver the therapeutic (i.e. normal) material into the abnormal target cells where the DNA is 'uploaded' allowing, for example, functional protein production to be resumed. There have been considerable concerns that the viral 'infection' may trigger an immune reaction and this is one of several factors affecting the development of this line of therapy in the human 'model'.

CLASSIFYING THE SKELETAL DYSPLASIAS

Skeletal dysplasias are a spectrum of approximately 800 conditions that are loosely defined by abnormalities of bone and cartilage growth. Contemporary classification is based on the specific genomic abnormality, but clinical phenotype and common radiological features continue to provide a robust and repeatable taxonomy.

For the purpose of this discussion, the following groups will be described:

1 *Osteochondrodysplasias*
   - Conditions primarily affecting the physis
     - Achondroplasia
     - Hypochondroplasia
   - Conditions primarily affecting the epiphysis
     - Multiple epiphyseal dysplasia
     - Spondyloepiphyseal dysplasia
   - Conditions primarily affecting the meta-diaphysis
     - Metaphyseal chondrodysplasias
   - Conditions with mixed abnormalities
     - Pseudoachondroplasia
     - Diastrophic dysplasia
     - Cleidocranial dysplasia
     - Nail–patella syndrome
   - Dysplasias with altered bone density
     - Osteogenesis imperfecta
     - Osteopetrosis
2 *Storage diseases*
   - Mucopolysaccharidoses
   - Gaucher's disease
3 *Tumour-like conditions*
   - Multiple hereditary exostosis
   - Dysplasia epiphysealis hemimelica
   - Enchondromatosis
   - Fibrous dysplasia
4 *Connective tissue disorders*
   - Ehlers-Danlos syndrome
   - Larsen's syndrome
   - Marfan's syndrome

BOX 8.1 THE SKELETAL DYSPLASIAS

Osteochondrodysplasias

- Conditions primarily affecting the physis
  - Achondroplasia
  - Hypochondroplasia
- Conditions primarily affecting the epiphysis
  - Multiple epiphyseal dysplasia
  - Spondyloepiphyseal dysplasia
- Conditions primarily affecting the meta-diaphysis
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- Conditions with mixed abnormalities
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  - Cleidocranial dysplasia
  - Nail–patella syndrome
- Dysplasias with altered bone density
  - Osteogenesis imperfecta
  - Osteopetrosis

(Cont.)

BOX 8.1 (Cont.) THE SKELETAL DYSPLASIAS

Storage diseases

- Mucopolysaccharidoses
- Gaucher's disease

Tumour-like conditions

- Multiple hereditary exostosis
- Dysplasia epiphysealis hemimelica
- Enchondromatosis
- Fibrous dysplasia

Connective tissue disorders

- Ehlers-Danlos syndrome
- Larsen's syndrome
- Marfan's syndrome

OSTEOCHONDRODYSPLASIAS

CONDITIONS PRIMARILY AFFECTING THE PHYSIS

Achondroplasia

Achondroplasia is the most common skeletal dysplasia, with an approximate incidence of 1/25 000. The inheritance pattern is autosomal dominant but the majority (>80%) occur secondary to *de novo* mutation in the fibroblast growth factor receptor 3 (*FGFR3*) that is identical in 95% of patients with this condition. The mutation causes gain in function affecting many tissues, but the inhibitory effect on the proliferative zone of the physis, with reduction of the thickness of the hypertrophic zone, results in decreased endochondral ossification that produces the characteristic phenotype.

In infancy, the most significant features are due to proximal spinal cord compression, due to narrowing of the foramen magnum and/or proximal cervical spinal canal presenting as sleep apnoea, mandating sleep studies and MRI in the first 6 months of life. Hydrocephalus may also develop, due to expression of *FGFR3* in the choroid plexus. Kyphoscoliosis is common in infancy, but resolves after independent sitting and standing.

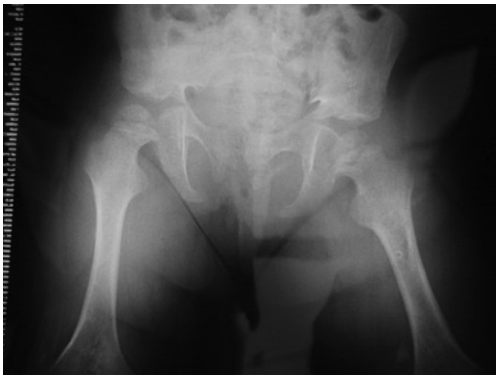
The most striking clinical features *in the older child are* disproportionate short limbs and characteristic facial features. The limb shortening is rhizomelic, with more significant shortening of the proximal segments. The average adult height is approximately 125 cm in females and 132 cm in males, intelligence is normal and lifespan is unaffected.

Facial features are characteristic and include button nose, frontal bossing, macrocephaly, mid-face hypoplasia and small nasal bridge. Joint laxity and hypotonia are also common and lead to postural issues including flat feet, lower limb coronal malalignment, fixed hip flexion, lumbar lordosis and radial head subluxation.

Adults with achondroplasia are at significant risk of spinal stenosis, secondary to progressive distal shortening of the pedicles. This presents as spinal claudication with progressive lower limb pain, weakness, numbness and paraesthesia.

The phenotypic and radiological features of achondroplasia are characteristic, and confident diagnosis is possible on a clinical basis but may be confirmed by genetic testing in selected cases.

The epiphysis is usually spared, with involvement confined to the metaphysis. Characteristic radiological features include short iliac wings, producing the 'champagne glass pelvis', horizontal acetabulae and coxa valga, with bulbous proximal femora (Figure 8.3).



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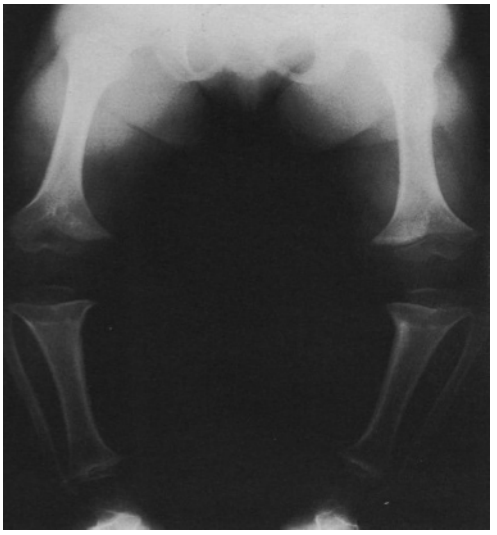


(b)

Figure 8.3 Achondroplasia (a) AP pelvis: note abnormal iliac wings, horizontal acetabular roof and bulbous proximal femur. (b) Standing alignment view: note medial axis deviation, inverted 'V' distal femoral metaphysis, fibular overgrowth and rhizomelic segmental involvement.

The distal femoral epiphysis has an inverted 'V' appearance, the fibula is long and there is often genu varum. Metacarpal shortening and splaying are responsible for 'trident' hands. Spinal radiographs demonstrate age-dependent changes with posterior vertebral scalloping and shortened pedicles in adolescent and adult patients.

Key developments in the management of achondroplasia include the recent introduction of natriuretic peptide as a therapeutic agent. This has produced encouraging results in small mammal models, and is entering the initial stages of human trials. The previous pharmacological approach to short stature was with growth hormone but the results of this approach have been very disappointing.

Patients with achondroplasia and their families frequently seek advice about the surgical management of short stature. Advances in external fixator design and improvements in surgical technique have made this a realistic but difficult option with a high but not prohibitive risk.

Significant complications include non-union, infection, iatrogenic neurologic injury and limb length discrepancy. The appearance of short arms and long legs may be cosmetically unacceptable and cause poor patient satisfaction. It is important that patients and their families have good understanding of the procedure, recovery, goal and limitations prior to embarking on a programme of limb lengthening.

Symptomatic lower limb malalignment, refractory to conventional non-operative treatment modalities, may be managed with guided growth techniques in the skeletally immature patient. Skeletally mature patients may be treated with realignment osteotomies in conjunction with limb lengthening or total knee replacement.

Symptomatic foramen magnum or upper cervical spine stenosis mandates urgent investigation with radiographs and an MRI. Significant stenosis should be managed with urgent decompression and stabilization.

Infantile kyphoscoliosis usually resolves after the initiation of independent weight-bearing. Patients with persistent deformity may require bracing to correct the curve. Patients with symptomatic, residual kyphoses of >40 degrees may rarely require anterior strut corpectomy and posterior fusion.

Lumbar stenosis in the young adult is initially managed with standard non-operative measures, including weight loss, physical therapy and activity modification. If these treatments fail, patients may be candidates for spinal decompression.

Hypochondroplasia

Hypochondroplasia is similar to achondroplasia, with a comparable limb and segmental involvement (Figure 8.4). This is also caused by a mutation in *FGFR3* and the inheritance pattern is autosomal dominant. The clinical features tend to be less marked and include broad hands and feet, loss of elbow extension and lumbar lordosis. Affected families have similar facial features but these are less pronounced and may appear to include macrocephaly due to a normal head in comparison to an involved body.

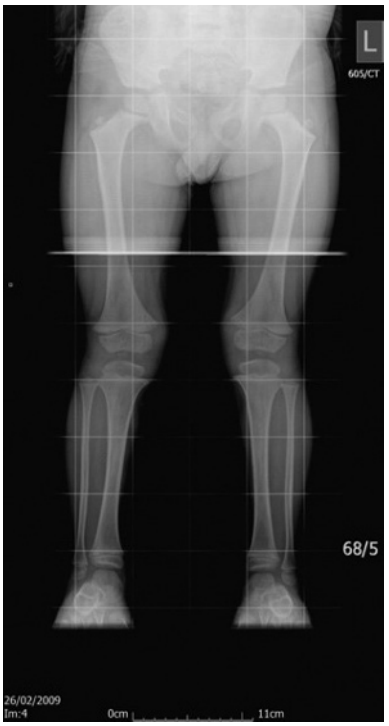


Figure 8.4 Hypochondroplasia Standing alignment view: note the rhizomelic involvement and very subtle epimetaphyseal changes.

The average adult height ranges from 135 to 165 cm in men and 125–150 cm in women and orthopaedic advice may be sought to discuss limb lengthening. While technically possible, this is associated with the same complications profile as in achondroplasia. The reality of surgical lengthening in this context is described from the patient and family perspective at https://lukeslongerlegs.wordpress.com.

Conditions primarily affecting the epiphysis

Multiple epiphyseal dysplasia

Multiple epiphyseal dysplasia (MED) describes a form of short-limb dysplasia with a broad phenotype. The estimated incidence is approximately 1:10 000, but this is probably an underestimate, as the milder forms may elude formal diagnosis. The clinical presentation ranges from asymptomatic patients with subtle anatomical abnormalities to severe pain and joint stiffness

requiring arthroplasty in adolescence or as a young adult.

The most common inheritance pattern is autosomal dominant, but there is a less common and clinically distinct autosomal recessive form. The majority of individuals affected with dominant MED have mutations of the *COMP* (cartilage oligomeric protein) gene, with approximately 10% presenting with abnormalities of the *MATN3* gene, both affecting matrix production and causing abnormalities of the physical and material properties of joint cartilage. Mutations of *COL9A1*, *COL9A2* and *COL9A3* are uncommon and cause accumulation of type IX collagen and also lead to abnormalities of articular cartilage.

The recessive form is associated with mutation of *SLC26A2* and frequently involves scoliosis, talipes and cleft palate, with clinodactyly and ear swelling making differentiation from diastrophic dysplasia difficult. The characteristic radiographic appearance of a 'double-layered patella' is common and assists clinical diagnosis.

The face, spine and skull are usually normal in this condition. Children affected with the dominant form are generally shorter than their peers and present with joint pain that may not present until the onset of puberty.

This is frequently associated with an abnormal gait pattern due to loss of hip movement, but in milder cases function can be nearly normal. The diagnosis in patients in whom the hips are predominately affected can be mistaken for Perthes' disease. A diagnosis of MED should be considered in the presence of bilateral, simultaneous hip involvement and broadly symmetrical radiological features. The obvious discriminating feature is the presence of epiphyseal changes in other joints in MED.

Joint pain at rest or associated with moderate exercise is a usual presenting complaint and involvement of multiple joints is common in more severe forms of this condition. Abnormalities of the knees are an occasional feature and the hands and feet are frequently short and broad.

Common *radiographic features* of dominant MED include delayed epiphyseal ossification. When the epiphysis appears, the shape is abnormal, but this may be subtle. There is a gradual deterioration of epiphyseal shape, with progressive flattening leading to articular incongruity at skeletal maturity (Figure 8.5).

Management is generally symptomatic, with advice on activity modification, avoiding repetitive loading, physiotherapy and analgesic and anti-inflammatory medication. Weight control is also very important and this in isolation is an efficient method of reducing pain and possibly prolonging native joint function.

As patients near skeletal maturity, those with more severe forms of the condition may be considered for osteotomies to correct deformity and improve the mechanical environment. Consideration for realignment osteotomies of the hip should be approached cautiously, as pain relief is unpredictable and joint stiffness is common. Arthroplasty is the conventional method for managing deteriorating joint pain and may be required at a young age in severely affected individuals.

Spondyloepiphyseal dysplasia

Spondyloepiphyseal dysplasia (SED) is an uncommon condition with an approximate incidence of 1:100 000. Affected individuals have significant shortening of the neck, spine and limbs.

The condition presents with two distinct phenotypes, due to a mutation in *COL2A1* leading to impaired synthesis of type II collagen. *SED congenita* is the more severe form, characterized by an autosomal dominant inheritance pattern and average adult height of 90–165 cm. *SED tarda* is characterized by an X-linked recessive inheritance pattern and is associated with milder phenotypic changes.

Since the onset of high definition antenatal ultrasonography, patients with SED congenita are often diagnosed in the perinatal period due to the degree of spinal and limb involvement. Maxillary flattening, facial abnormalities and cleft palate are common associations diagnosed in the postnatal period. Severe myopia, vitreous abnormalities and retinal detachment cause visual impairment, and hearing loss becomes obvious in early childhood.

Atlantoaxial instability is a very significant association in patients with a diagnosis or suspected

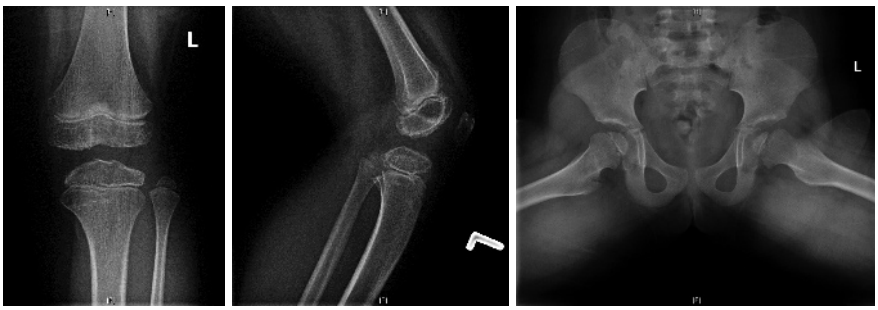
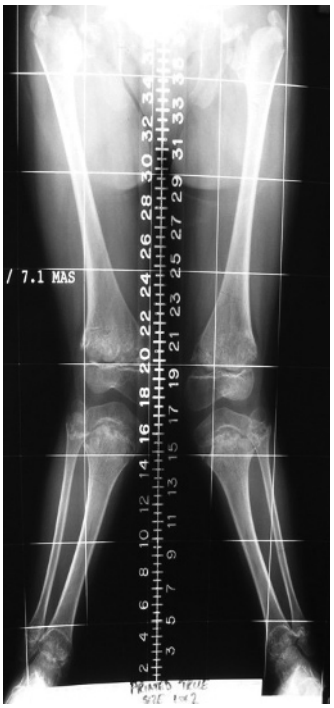


Figure 8.5 Multiple epiphyseal dysplasia (MED) Limited skeletal survey: note subtle alterations of epiphyseal morphology at multiple joints.



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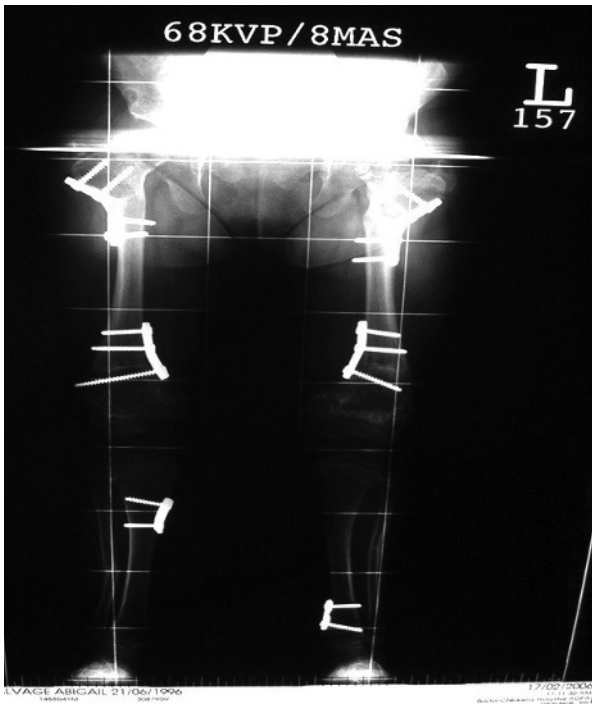


(b)

Figure 8.6 Spondyloepiphyseal dysplasia (SED) (a) Standing alignment view: note multisite epimetaphyseal abnormality with significant lateral axis deviation and coxa vara. (b) Postoperative standing alignment view following multilevel osteotomy.

diagnosis of SED congenita. Patients should undergo radiological screening of the cervical spine and may require cervico-occipital fusion in the presence of significant instability to prevent cervical myelopathy. Thoracolumbar kyphoscoliosis, chest wall abnormalities with respiratory insufficiency and lumbar lordosis are commonly seen.

Radiographic findings include platyspondyly, odontoid hypoplasia and narrowed intervertebral discs. The pubic bones are unossifed at birth with coxa vara and delayed ossification of the femoral head in later childhood (Figure 8.6).

The eventual consequence of abnormal collagen and an unfavourable mechanical environment is the premature onset of osteoarthritis. Joint deformity can be managed with multilevel corrective osteotomies and the small size and complex anatomy introduces a dimension of complexity that requires advanced arthroplasty solutions.

SED tarda is less severe and is generally diagnosed after the age of 5. Males are more commonly affected than females and present with chest wall and spinal abnormalities or concerns about stature. The radiographic features are similar to SED congenita, with similar vertebral and epiphyseal changes.

The initial approach is symptomatic improvement, using physiotherapy, core strengthening, analgesic and anti-inflammatory medication and activity modification for back pain. Joint deformities may require corrective osteotomies, and arthritis of the hips or knees is managed with total joint arthroplasty.

CONDITIONS PRIMARILY AFFECTING THE META-DIAPHYSIS

METAPHYSEAL CHONDRODYSPLASIAS

The metaphyseal chondrodysplasias are a heterogeneous group of disorders with similar radiographic abnormalities but caused by a number of unrelated genetic mutations. They have been historically subclassified according to the predominant clinical features with long bone metaphyseal irregularities being a consistent radiological abnormality.

Type Schmid This is the most common metaphyseal chondrodysplasia. It is inherited in an autosomal dominant pattern, is caused by a mutation in the *COL10A1* gene and affects the hypertrophic zone of the physis.

Patients present in early childhood, usually because of mild short stature, genu varum, excessive lumbar lordosis and a waddling gait due to coxa vara. The tubular bones are short with metaphyseal flaring and a widening of the growth plate (Figure 8.7). Coxa vara may be progressive, requiring an inter-trochanteric valgus osteotomy in severe cases. Lower limb malalignment may improve with growth during childhood, but progressive or severe cases may require multiple osteotomies.

Type McKusick Also known as cartilage hair hypoplasia, Type McKusick is an autosomal recessive disorder, common in the Amish population. It becomes

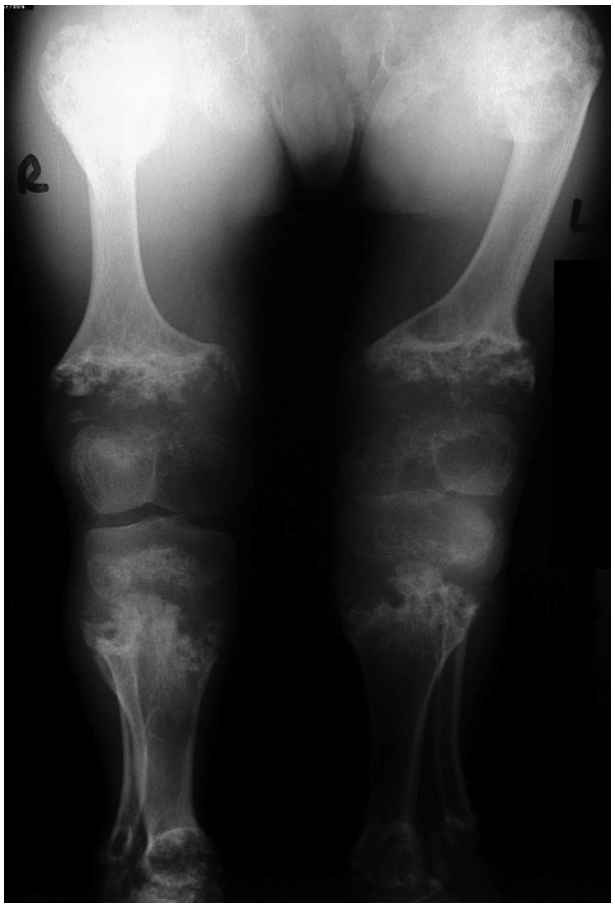


Figure 8.7 Metaphyseal chondrodysplasia type Schmid Standing alignment view: note the obvious metaphyseal flaring, physeal widening and relative sparing of the epiphyses.

apparent in early childhood and is associated with progressive short stature in association with very fine hair growth, which is usually blond or light compared to relatives. Abnormalities of T-cell function and neutropenia lead to an increased susceptibility to infection. Skeletal manifestations include lower-limb malalignment, severe ligamentous laxity, shortened digits and broad feet and hands. Metaphyseal cupping and flaring is usually obvious at the knees, with relative sparing of the proximal femur.

Type Jansen This is a very rare, severe autosomal dominant disorder caused by a mutation of the PTH-related peptide receptor. It is usually recognized at birth because of severe limb shortening with micrognathia and a prominent forehead. The metaphyses are flared, with irregular ossification and significant shortening of the tubular bones. Flexion contractures develop at the hips and knees, with progressive deterioration of limb alignment and periarticular enlargement causing major difficulties with walking.

CONDITIONS WITH MIXED ABNORMALITIES

Pseudoachondroplasia

Pseudoachondroplasia is a disproportionate, short limb dysplasia that is associated with significant short stature, limb malalignment and severe ligamentous laxity. It is an autosomal dominant disorder, affecting the cartilage oligomeric matrix protein gene (*COMP*) on chromosome 19. Secretion of COMP is disrupted and accumulates in chondrocytes, causing cell death and inhibiting epiphyseal cartilage growth and development.

Neonatal development is not obviously affected and the diagnosis is difficult to make in infancy. There is a delay in acquisition of motor milestones, including crawling and walking and a wide-based gait.

The average adult height in this condition is approximately 120 cm and the most striking clinical feature is ligamentous laxity. In older patients, poly-axial knee instability impacts on walking distance and standing tolerance, and gradual deterioration with time results in pain and loss of range in early adult life.

The principle abnormality affects epiphyseal and articular cartilage formation and patients with pseudoachondroplasia have underdeveloped, flattened epiphyses and widened metaphyses (Figure 8.8). The tubular bones are shortened and the vertebral bodies may be oval-shaped. In adolescence and adulthood, the combination of ligamentous laxity, mechanical axis abnormality, epiphyseal irregularity and articular cartilage attrition results in irregular epiphyses leading to joint deformity and early-onset arthritis.

Orthopaedic management is initially directed at preserving joint function, and physical therapy has a role in preserving range of motion and maintaining muscle strength. Initial management of joint pain involves analgesic and anti-inflammatory drugs.

Joint deformities may be managed with corrective osteotomies, but this is difficult because of the conflicting effect of joint incongruity and ligamentous laxity. It is occasionally necessary to deliberately produce geometric abnormalities to provide stability under load, improving standing and walking function. Symptoms generally deteriorate with time and patients frequently require joint replacement, which is technically challenging for the reasons outlined above.

Diastrophic dysplasia

Diastrophic dysplasia is an autosomal recessive disorder, occurring secondary to a mutation in the *SLC26A2* gene. This results in defective production of a sulphate transporter protein, resulting in

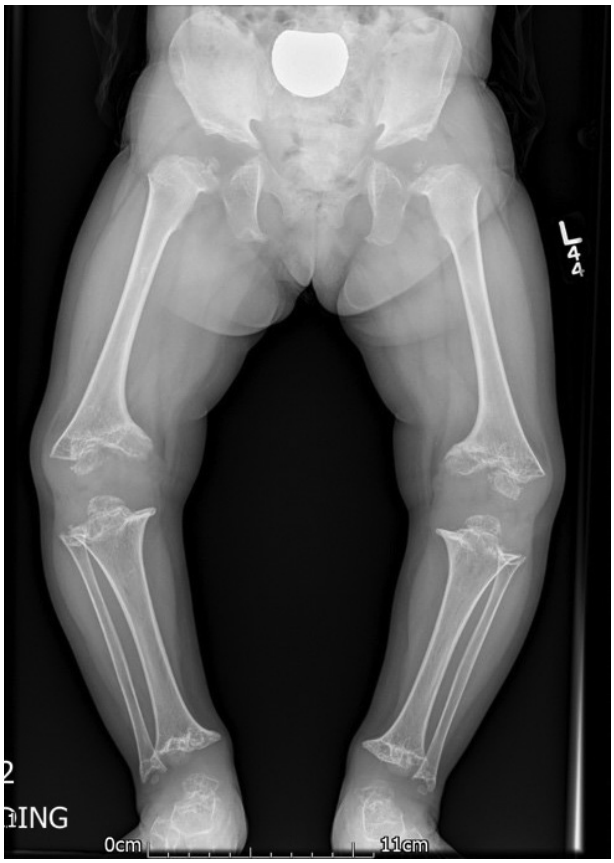


Figure 8.8 Pseudoachondroplasia Standing alignment view: note the obvious epimetaphyseal involvement. The significant medial mechanical axis deviation is a combination of bone morphology and significant ligament laxity that is characteristic of this condition.

abnormalities of cartilage proteoglycan. The genetic mutation is present in approximately 1 in 70 in the indigenous population of Finland, but is rare in the rest of the world, with an estimated incidence of 1:1 000 000.

Patients are usually diagnosed in infancy due to visible abnormalities including severe short stature and characteristic facial features such as cleft palate and cauliflower ears and with hitchhiker's thumb and talipes.

Spinal manifestations include atlantoaxial instability, which may present as spinal cord compression, and cervical kyphosis and thoraco-lumbar scoliosis. Limb involvement includes rhizomelic or mesomelic shortening, genu valgum, hip and knee contractures, and foot deformities including rigid clubfoot or skewfoot.

Radiographs demonstrate generalized epiphyseal hypoplasia and flattening, metaphyseal widening, genu valgum, scoliosis and widening of the first metacarpal space.

Treatment of diastrophic dysplasia is directed at early correction of joint contractures, initially with physical therapy, but surgical release of contractures at the hip, knee and elbow may be necessary. Foot deformities are often severe and rigid and require extensive open releases to produce a plantigrade foot. Patients with atlantoaxial instability with neurological symptoms require occipital–cervical fusion. Cervical kyphosis usually resolves spontaneously, but in refractory cases it may require corrective fusion. Patients with progressive kyphoscoliosis may also require posterior spinal fusion.

Cleidocranial dysplasia

Cleidocranial dysplasia is a skeletal dysplasia which affects bones formed by intramembranous ossification. It is inherited in an autosomal dominant pattern and is due to a mutation in the *RUNX2* gene causing an abnormality in the transcription factor regulating differentiation of osteoblasts.

Affected individuals have proportionate short stature, with dysplastic or absent clavicles and shoulder hypermobility. Coxa vara and shortening of the middle phalanges of the fingers are common features. The skull is also involved, with late closure of the fontanelles, frontal bossing and Wormian bones, which are intrasutural bones, commonly seen at the lambdoid suture. Patients also frequently have dental abnormalities, with delayed eruption of the adult dentition.

Radiographs demonstrate the characteristic absence or hypoplasia of the clavicles, delayed ossification of the pubis, coxa vara and late closure of cranial sutures.

Most patients are asymptomatic and the absence or hypoplasia of clavicles is of no functional consequence although advice may be sought about the appearance. No sensible surgical treatment is, however, available. Patients who develop severe coxa vara may require a realignment osteotomy, which can be performed using standard planning and conventional osteosynthesis. Specialist dental treatment is often required.

Nail–patella syndrome

Nail–patella syndrome (NPS) is an autosomal dominant disorder caused by random mutations of the *LMX1B* gene which results in hypoplastic or absent nails and patellae. There is a spectrum of commonly associated abnormalities including knee instability, limitation of elbow movement, subluxation of the radial head, exostoses at the posterior aspect of the iliac bones (iliac horns), scoliosis and scapular hypoplasia. Important extraskeletal manifestions include nephropathy and glaucoma.

Affected individuals present with a broad spectrum of involvement, and diagnosis is made on the basis of family history and the hallmark digital and patellar abnormalities. Plain radiographs and CT/MRI are

useful to determine the size and position of the patellae. Elbow radiographs confirm subluxation or dislocation of the radial heads, with associated remodelling of the elbow joint. Pelvic radiographs demonstrate iliac horns in 80% of cases and this is considered to be pathognomonic for this condition.

Orthopaedic intervention is based on symptomatic modification, with analgesics and physiotherapy to manage joint pain. Patellar realignment is complex due to the anatomical abnormalities, but it may be necessary to treat significant and deteriorating anterior knee pain. Limited forearm rotation is not usually associated with significant functional loss, but pain due to a subluxed radial head may be sufficiently severe to require radial head excision.

DYSPLASIAS WITH ALTERED BONE DENSITY

Osteogenesis imperfecta

Osteogenesis imperfecta (OI) is a relatively common connective tissue disorder with an incidence of approximately 1:20 000 live births. Mutations of *COL1A1* and *COL1A2* genes cause quantitative and qualitative abnormalities of type I collagen production. All tissues that contain type I collagen are affected, including bone, ligament, teeth and sclera. This results in structurally incompetent bone, vulnerable to fracture, secondary deformity and joint laxity. Abnormalities of non-skeletal collagen produce alterations in the sclera, dentinogenesis imperfecta and deafness.

Sillence described four separate types according to scleral involvement, natural history and the perceived inheritance. Conventional wisdom was that there were clear autosomal dominant and recessive types. Contemporary understanding is that the majority of cases are caused by spontaneous mutation and that all familial mutations are inherited in an autosomal dominant fashion. Mutations previously considered to be recessive are more likely to be due to mosaicism.

Diagnosis tends to be made on clinical and radiological grounds, but DNA analysis can be used and has identified over 800 separate collagen mutations in patients with a typical phenotype. Collagen analysis of dermal punch biopsies is also used to confirm diagnosis in some cases.

The *clinical manifestations* are protean and are determined by the individual pattern of involvement. Quantitative abnormalities of type I collagen formation lead to a milder form of the disease while qualitative defects can cause severe abnormalities in which there is complete absence of lamellar bone.

Recurrent fractures at multiple sites, often with trivial trauma, are a common finding and are a consequence of bone fragility (Figure 8.9). Normal bone healing is present in the majority of fractures, but progressive long bone deformity is a frequent consequence of either fracture mal-union or progressive deformity due to the underlying abnormalities of collagen.

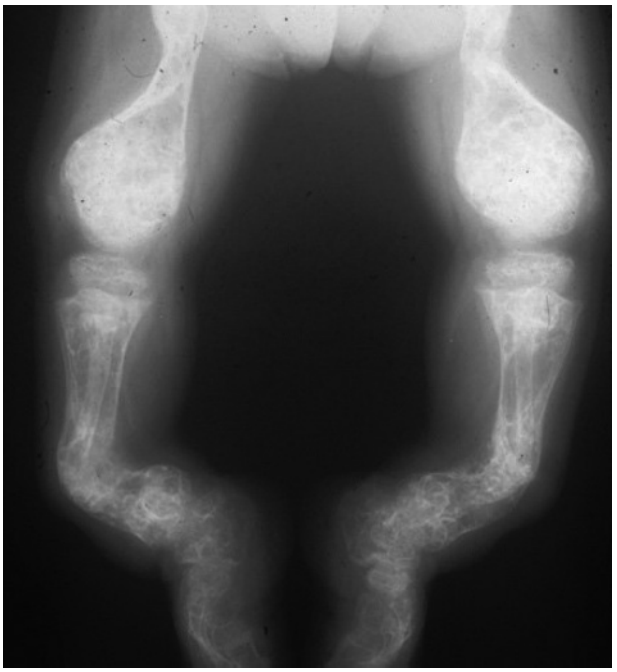


Figure 8.9 Osteogenesis imperfecta AP tibia: note the reduction of bone density that is associated with the clinically severe forms of this syndrome. In addition, there is a mid diaphyseal, apex anterior deformity associated with sequential fragility fractures.

Dental abnormalities, blue sclera, scoliosis and kyphosis associated with flattened vertebral bodies and non-specific bone pain are frequent causes of orthopaedic referral in childhood. In later life, joint degeneration secondary to long-standing malalignment is a common consequence. A combination of protrusio, fracture and abnormal hip mechanics frequently results in painful osteoarthritis, requiring total joint replacement.

Plain radiographs are used to demonstrate the presence, site and configuration of suspected fractures. These are usually associated with low energy transfer, are commonly transverse, and tend to heal with abundant callus formation.

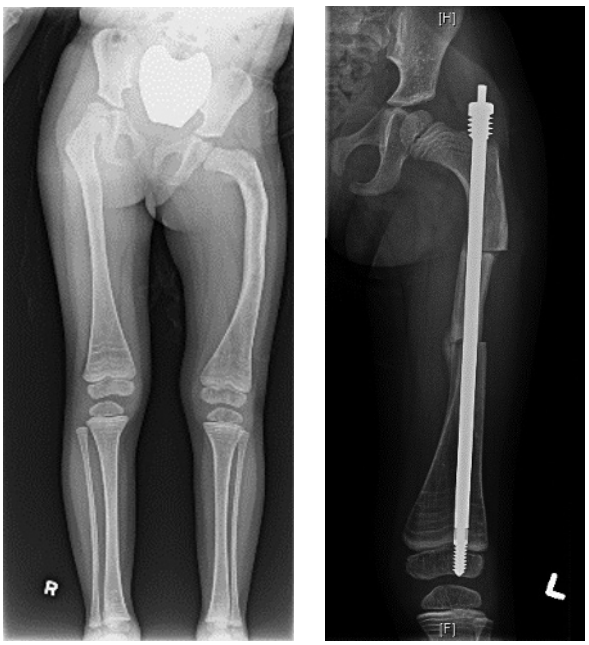
Radiological features are generally non-specific and include Wormian bones, which are present in the normal population and are associated with a number of skeletal abnormalities, including osteogenesis imperfecta. Enlargement of frontal and mastoid sinuses are also seen in some patients with osteogenesis imperfecta. The cortices of long bones are often thin and demonstrate features of general demineralization,

previous fracture and previous pharmacological intervention, particularly bisphosphonates.

Protrusio acetabuli and proximal femoral 'shepherd's crook' deformities of the femurs are common findings. Multiple areas of radiolucent scalloping with radio-dense rims with the appearance of 'popcorn' can be seen in the metaphysis of some patients with osteogenesis. Bone densitometry is frequently used to assess bone health in patients with osteogenesis, and is useful in quantifying the effect of bisphosphonate therapy on metrics including bone mineral density.

Management involves physiotherapy, walking aids and orthotics to maximize mobility. Pharmaceutical agents are used to enhance bone strength, and bisphosphonates have been demonstrated to increase cortical thickness by inhibiting osteoclastic bone resorption. Cycles of intravenous bisphosphonates are used to decrease bone pain and reduce the incidence of fractures.

Surgical intervention to correct deformity and stabilize load-bearing bones generally utilizes intramedullary fixation systems (Figure 8.10). Spinal deformity may require surgical correction but instrumented fusion relies on sufficient bone quality.



(a) (b)

Figure 8.10 Osteogenesis imperfecta (a) AP femora: note multiple radiolucent metaphyseal lines secondary to sequential administration of bisphosphonates. There is a multilevel diaphyseal deformity, secondary to previous fractures, with an overall coxa vara. (b) Postoperative radiograph, following multilevel realignment osteotomies, stabilized with a Fassier–Duval intermedullary rod.

Osteopetrosis (Albers–Schönberg disease)

Osteopetrosis is characterized by abnormally dense bone with poor or absent medullary canal formation. This results in alterations in the material properties, resulting in bone fragility and abnormalities of haemopoiesis.

It is caused by a defect in genes controlling osteoclastic activity with an incidence estimated to be 1:100 000–500 000. It is classified according to age of onset and clinical features into an autosomal recessive infantile (congenital) and autosomal dominant adult (tarda) type.

The infantile form presents in early life as growth retardation. Failure to thrive and genu valgum are common findings. Abnormal skull development is associated with hydrocephalus, sinus blockage, hearing loss and cranial nerve entrapment. Obliteration of medullary canals causes pancytopenia with hepatosplenomegaly due to extra-medullary haemopoesis. Osteitis of the mandible is relatively common due to an alteration in vascularity.

Defective osteoclasts are responsible for raised creatinine kinase and acid phosphatase. Parathyroid hormone is elevated due to secondary hyperparathyroidism. Plain radiographs are usually diagnostic (Figure 8.11). The bones appear sclerotic, but streaks of radiolucency can occur at the bone ends. The skull base is uniformly radio-dense and radiolucent bands through sclerotic vertebrae are often seen and referred to as the 'rugger-jersey' sign.

Regular administration of vitamin D stimulates osteoclastic activity. γ-interferon enhances leucocyte function and increases the bone marrow volume. Erythropoietin can be used to treat the anaemia.

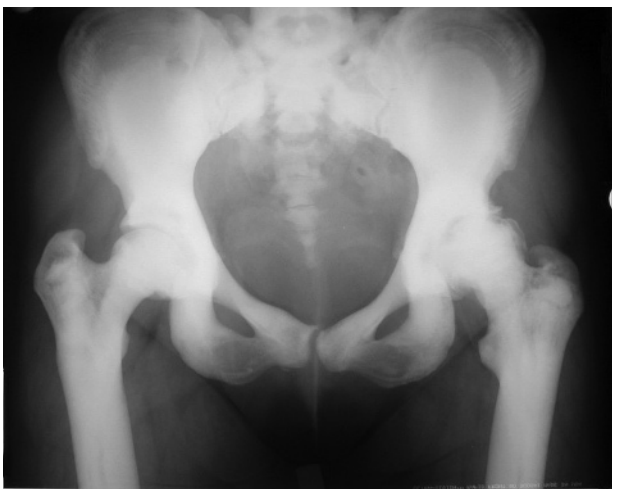


Figure 8.11 Osteopetrosis AP pelvis: note the significant sclerosis and absence of long-bone medullary canal.

Corticosteroids are used to increase the haemoglobin levels and increase the rate of bone resorption.

The adult form is asymptomatic in approximately 50% of patients and is identified as an incidental finding or during investigation for non-specific bone pain. Compressive neuropathies and recurrent fractures also occur in the adult, but haemopoiesis is unaffected. Osteopetrotic long-bone fractures are challenging to manage due to the unusually dense bone. Difficulties arise with attempts to drill bone during plate osteosynthesis. Delayed union is common due to fundamental abnormalities of bone biology and mechanics.

STORAGE DISEASES

Mucopolysaccharidoses

Mucopolysaccharidoses (MPS) are a group of storage disorders characterized by absence or defects of lysosomal enzymes that are responsible for the breakdown of glycosaminoglycans (GAGs). GAGs are long-chain polysaccharides, which accumulate in the liver and spleen in addition to bone, cartilage and connective tissues, leading to a spectrum of skeletal abnormalities.

Hurler (MPS type 1) and Sanfilippo (MPS type III) are the most common, with an overall prevalence of approximately 1:100 000 live births. Hunter (MPS type II) is inherited with an X-linked recessive pattern and all the other types are inherited in an autosomal recessive pattern.

This condition presents in early childhood with short stature, and affected individuals tend to be less than the third percentile for height. Other generic features include characteristic 'coarse' facial features, thick inelastic skin, hepatosplenomegaly, neurological abnormalities and delayed intellectual development. Spinal abnormalities are common and include atlantoaxial instability with abnormal vertebrae producing a rigid kyphoscoliosis.

Hurler (MPS I) (α–L-iduronidase deficiency causing heparan and dermatan sulphate accumulation)

MPS type I is a heterogeneous condition with significant variability of life expectancy. The rate of growth and mental development slows in the second year of life. Affected individuals walk late and develop severe joint stiffness, coxa valga, femoral head irregularities and genu valgum. Anterior vertebral wedging leads to kyphosis or thoracolumbar gibbus and in common with most MPS odontoid hypoplasia may develop.

Non-skeletal features include big tongues (macroglossia), hearing loss and cardio-respiratory insufficiency which can result in death in late childhood.

Hunter (MPS II) (iduronate sulphatase deficiency causing heparan and dermatan sulphate accumulation)

MPS type II occurs only in males due to the inheritance pattern. The features are variable and are similar to those seen in type I. Macrocephaly and coarse facial features are characteristic, with marked joint contractures developing in the pre-school period leading to joint stiffness and hip arthritis in older patients. Cardio-respiratory compromise is also common, which in severe cases leads to death in the second decade.

Sanfilippo (MPS III) (heparan sulphamidase deficiency causing heparan sulphate accumulation)

Children with MPS type III have slowing of their rate of growth after 2–3 years, at which point they also begin to develop intellectual impairment. They may become wheelchair-bound at a young age.

Morquio–Brailsford (MPS IV) (β-galactosidase deficiency causing keratin and chondroitin sulphate accumulation)

Individuals affected with MPS IV have a normal face and intellect in contrast to most MPS subtypes and affected individuals often survive into adulthood.

The most striking features are short stature with disproportionate spinal involvement (Figure 8.12), barrel chest with pectus carinatum and a short neck. Limb abnormalities include coronal knee malalignment and joint stiffness.

Scheie syndrome (MPS V) This has been reclassified as a subtype of MPS I.

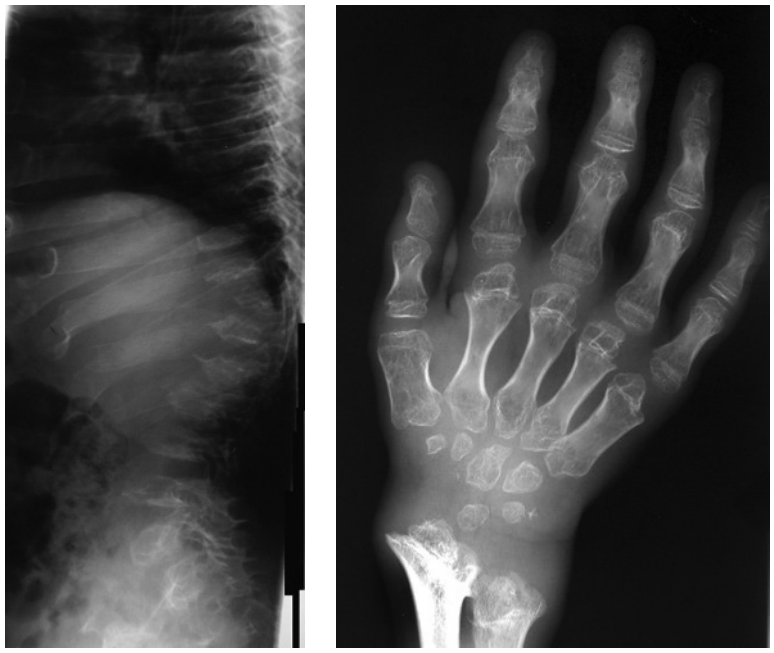
Maroteaux Lamy syndrome (MPS VI) (N-acetylgalactosamine-4-sulphatase deficiency causing dermatan sulphate accumulation)

MPS VI presents with a similar but milder clinical picture than type I. Intellect is unaffected and the prognosis tends to be better with longer life expectancy.

There is no known cure for MPS. *Treatment* is supportive and deals with specific symptomatic areas. *Effective management* requires early, accurate biochemical diagnosis, so that the prognosis for an individual patient can be predicted. Enzyme replacement is currently available for types I and IV and has been effective in reducing non-neurological symptoms and pain. Bone marrow transplantation and umbilical cord blood transplantation have been used in selected cases with variable and unpredictable outcomes.

Surgery to remove tonsils and adenoids may improve airway obstruction. Corneal transplantation can improve vision in patients with significant corneal clouding.

Orthopaedic surgery has a role in correction of deformity in the spine, hips, knees and feet. Conditions with a slow rate of progression and a good



(a) (b)

Figure 8.12 Mucopolysaccharidoses (a) AP lateral spine. (b) Hand in a patient with MPS type IV. Note the degree of platyspondolyly and rigid kyphoscoliosis, shortening of the metacarpals and phalanges and differential radio-ulnar involvement.

life expectancy may benefit from corrective surgery. Surgical stabilization of the spine may be necessary to protect cardio-respiratory function and preserve sitting balance. Standing and walking function may require release of joint contractures or surgical release or produce plantigrade feet.

Gaucher's disease

Gaucher's disease is an autosomal recessive disease caused by mutation of the glucocerebrosidase gene responsible for the breakdown of lipid-rich cell membranes in red and white blood cells. The condition is characterized by accumulation of glucocerebroside in macrophages. The estimated global prevalence is 1:40 000 live births and is substantially more common in the Ashkenazi population.

Clinical features include hepatosplenomegaly, pancytopenia and recurrent infections. Orthopaedic manifestations include stiff joints, osteopenia with vertebral compression and femoral fractures. Osteonecrosis is a frequent feature and affects femoral and humeral heads, femoral condyles and the talus. Chronic bone pain is common and affected individuals can present acutely with a 'bone crisis' characterized by severe pain, pyrexia, leukocytosis and elevated inflammatory markers, which is difficult to discriminate from septic arthritis and osteomyelitis.

Radiological features include areas of patchy radiolucency within cancellous bone. Flaring of the femoral condyles produces the 'Erlenmeyer flask appearance'. Plain radiographs are used to identify complications including fractures, and femoral head osteonecrosis is identified and quantified with MRI.

Enzyme replacement therapy is an effective treatment, although the skeletal abnormalities are the slowest to respond and may not recover. Hip arthroplasty is often required to manage osteonecrosis of the femoral head.

TUMOUR-LIKE CONDITIONS

Multiple hereditary exostosis (multiple osteochondromatosis, diaphyseal aclasis)

This condition is characterized by cartilage-capped, benign bone tumours that appear in childhood at the site of active bone growth. They can occur as a single entity, but commonly they are multiple, inherited and part of a generalized skeletal abnormality, with an approximate incidence of 1:100 000 live births.

The condition is due to a group of mutations of the *EXT1*, *EXT2* and *EXT3* genes on chromosomes 8, 11 and 19, with an eventual phenotype that is independent of the site and type of mutation. The majority are inherited in an autosomal dominant pattern but it is estimated that 30% arise as new mutations.

Uncontrolled transverse growth of the growth plate leads to the formation of discrete peripheral cartilaginous masses at the periphery that undergo endochondral ossification. If the abnormal proliferation ceases at this point and the bone continues to grow in length, the exostoses migrate with the metaphysis, leading to a characteristic, pedunculated appearance.

This condition usually presents in childhood, with the discovery of an incidental bony lump which may be associated with local symptoms. Although these lumps can arise in any growing bone, the characteristic sites are knee, ankle, wrist, shoulder, pelvis, scapula and spine.

Physeal tethering can lead to shortening and angulation of long bones. In paired bone systems, differential growth can result in significant joint deterioration and this is particularly obvious in the forearm and lower leg. Progressive ulnar shortening produces volar and ulnar deviation of the wrist, radial bowing and subluxation of the radial head. Differential tibiofibular growth produces progressive ankle valgus, with an associated loss of joint range. Local symptoms are due to pressure effects on surrounding tissues, impingement of muscle–tendon units and neurovascular compression.

Growth tends to cease at skeletal maturity and the most significant long-term complication is malignant transformation of the cartilaginous cap. Chondrosarcoma has been reported in 0.5–40% of affected individuals over the age of 21. This is more common in proximal and flat bones and in patients with a family history of malignant transformation, and continued growth or pain after skeletal maturity should raise suspicion.

Clinical suspicion is readily confirmed with plain radiographs, and characteristic bony lesions are usually found in the metaphysis of the distal femur (70%), proximal tibia (70%) and proximal humerus (50%) (Figure 8.13).

There is continuity of the cortex of normal bone and the lesion, which is either sessile (broad-based) or pedunculated, grows away from the physis. A mottled appearance overlying the lesion suggests calcification of the cartilaginous cap.

Alignment radiographs are necessary to quantify deformities at the ankles and knees, and plain radiographs of the elbow are necessary in the presence of loss of forearm rotation. CT scans are often necessary to delineate exostoses on the deep surface of the ilium and scapula and to identify spinal exostoses. MRI imaging is required to determine the extent of the cartilaginous cap. If this is greater than 2 cm in depth, it is more likely to be associated with chondrosarcoma transformation.

Excision is considered if the local pressure effect causes pain or restriction of joint movement or to improve the appearance of the limb. Symptoms of neurological or vascular compression are also a relative indication to remove selected lesions.

Angular deformity, limb shortening and joint malalignment also require exostosis removal with the addition of guided growth using staples or corrective osteotomies.

Lesions with suspected malignant transformation are managed with wide excision, but they require initial staging and to be coordinated under the supervision of a multidisciplinary team.

Dysplasia epiphysealis hemimelica (Trevor's disease)

This is a rare condition that is characterized by epiphyseal osteochondromata, which have a characteristic but unexplained anatomical arrangement. The bony

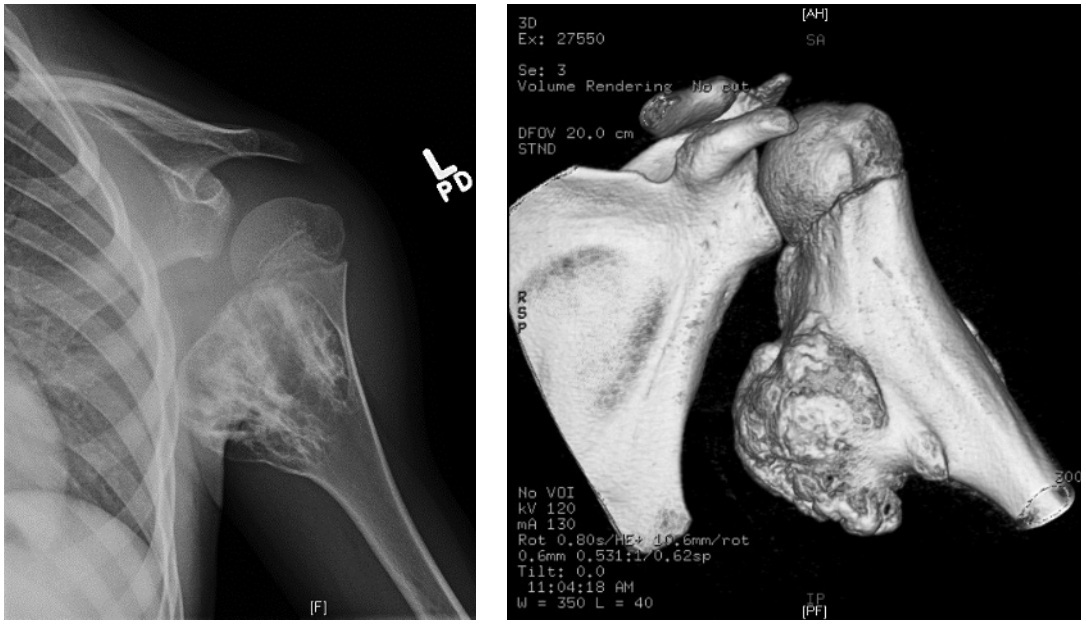


Figure 8.13 Multiple hereditary exostosis

lesions appear on the medial or lateral aspect of the epiphysis and are localized to a single side of the body, with the ankle and knee most frequently involved.

It is a sporadic disorder with no known inheritance pattern and is caused by a cartilage overgrowth arising from the groove of Ranvier. Lesions are histologically similar to an osteochondroma and their effect on articular congruity can lead to progressive deterioration of the affected joint.

The most common presenting symptom is restriction of joint movement or pain, a palpable intra-articular lump or asymmetric limb deformity. Plain radiographs demonstrate asymmetry at the involved physis, with enlargement of the affected side and a visible exostosis. MRI/CT may be useful in assessing the size and position of the lesion in addition to demonstrating cortico-medullary continuity.

Definitive management of this condition is difficult as excision of the intra-articular lesions in a relatively asymptomatic patient may cause a deterioration of function. Treatment is therefore based on the level of symptoms and involves excision, avoiding damaging the uninvolved articular cartilage. In patients with large lesions, with secondary deformities, corrective osteotomies may be undertaken without excision.

Enchondromatosis (dyschondroplasia, Ollier's disease)

Enchondromatosis is characterized by discrete islands of hyaline cartilage within the metaphyses and diaphyses of long bones.

It is a sporadic condition, which is usually unilateral with a reported prevalence of 1:100 000, but there are occasional reports of a familial tendency. There is failure of bone formation in the cartilaginous columns arising from the physis. This causes expansion of unossified cartilage within the bone and leads to physeal damage, with shortening and angular deformity of affected bones and a risk of pathological fracture.

Involvement of the hands and feet is common and, in severe cases, leads to severe disability. Malignant transformation to chondrosarcoma occurs, particularly with multiple digital lesions, but the prevalence of this significant complication is unknown.

Multiple enchondromata, in association with soft tissue haemangiomas, is termed *Maffuci's syndrome*. The skeletal manifestations are generally more severe and the risk of malignant transformation is substantially higher. The estimated risk of malignant change in either tissue is of the order of 50%, and these patients should be under lifelong surveillance.

Radiographs of long bones have characteristic radiolucent streaking, extending from the physis into the metaphysis. This is the appearance of the cartilaginous columns that have failed to ossify and calcification within these areas has the appearance of 'popcorn'. Alignment views are required to assess shortening and angular deformity, and radiographs of the hands demonstrate characteristic bone expansion of the phalanges and metacarpals (Figure 8.14).

Involvement of load-bearing bones leads to pain and progressive deformity and may require *treatment* in the form of corrective osteotomy. Patients may present with pathological fractures in previously asymptomatic areas and require conventional fixation and biological augmentation with autologous bone graft or bone substitutes.

Patients with extensive involvement may require limb equalization surgery involving a combination of lengthening and growth modulation with guided growth or formal epiphysiodesis. Debulking and grafting of lesions within the hand are often required to improve function and any suspicious lesion requires appropriate staging followed by wide local excision.

Fibrous dysplasia

This condition affects bone development and involves replacement of bone with fibrous tissue. The incidence is unknown as it is thought that many lesions are asymptomatic. A single bone (monostotic) is usually involved, but involvement may be more extensive (polyostotic) in 20% of cases.

The disease is caused by a sporadic mutation in the *GNAS1* gene which is located on chromosome 20. This gene codes for the G protein and the abnormality causes stimulation of cAMP and leads to the excessive DNA synthesis in affected osteoblastic cells.

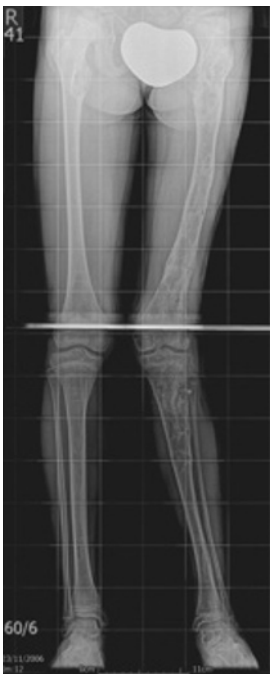


Figure 8.14 Enchondromatosis (Ollier's disease) Standing alignment view: note the unilateral, multi-apical deformity with the characteristic 'popcorn' appearance of the proximal left tibia.

Production of disorganized fibrous material prevents normal mineralization and maturation into lamellar bone.

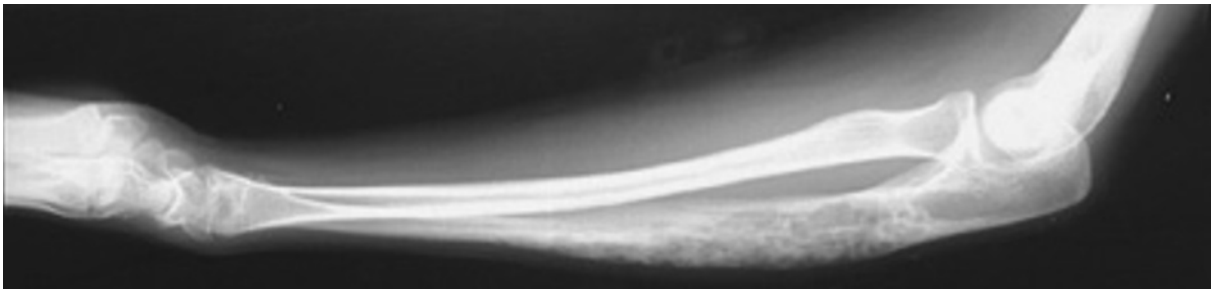
Patients often present in the second decade with progressive limb deformity or following a fracture through a previously asymptomatic lesion. Non-specific bone pain, swelling and tenderness are also common presenting symptoms.

Proximal femur, tibia, pelvis and foot are frequently involved, but ribs, skull and bones of the upper limbs are also affected.

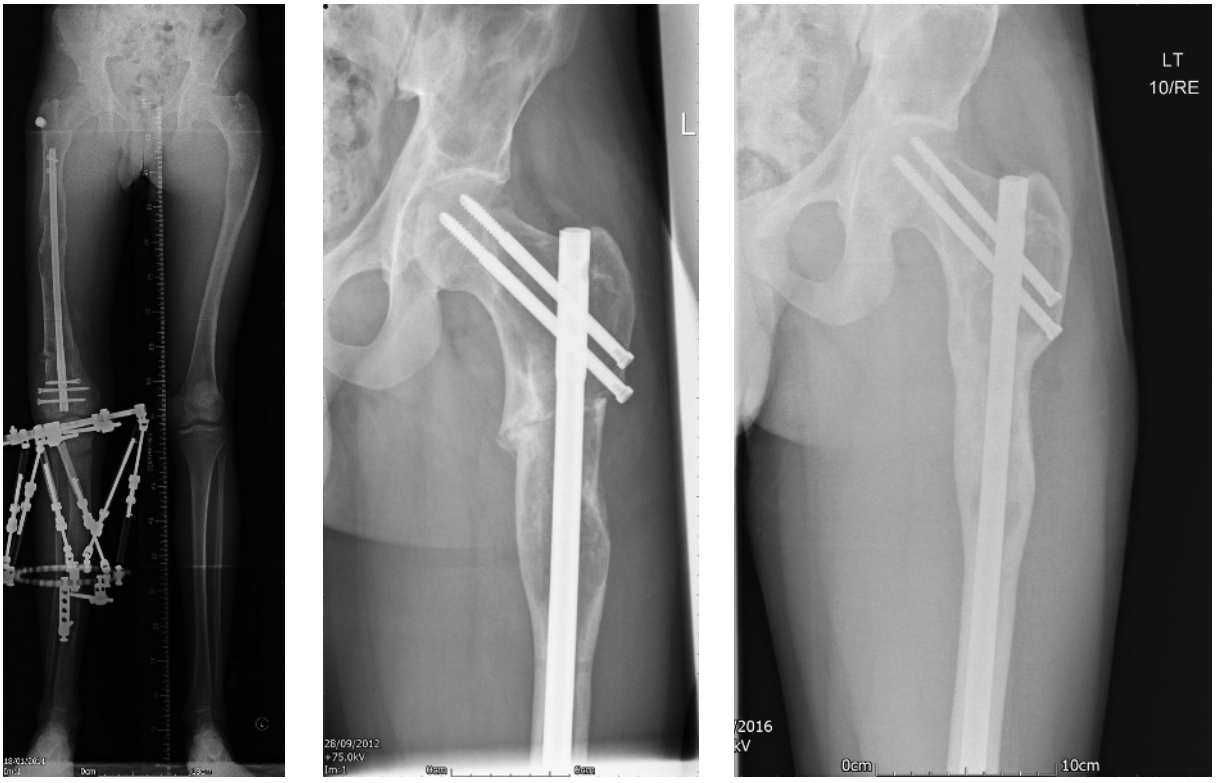
Polyostotic fibrous dysplasia, in association with precocious puberty and café-au-lait spots with irregular borders is called *Albright McCune syndrome*. Malignant transformation to osteosarcoma, fibrosarcoma or chondrosarcoma occurs in 0.4–4% of cases.

Plain radiographs demonstrate well-defined, expansile intra-medullary lesions that vary in appearance (Figure 8.15a). The majority demonstrate a hazy 'ground glass' appearance but fibrous dysplasia may be radiolucent or sclerotic. The characteristic appearances in the proximal femur are expansion with marked varus (the 'shepherd's crook' deformity). Bone scan or whole-body MRI is indicated at first presentation to determine the extent of the disease.

It is often possible to secure a confident diagnosis on the plain radiographic features, but if the diagnosis is not clear, open biopsy is necessary. Incidental



(a)



(b)

Figure 8.15 Fibrous dysplasia (a) Lateral forearm: note extensive proximal meta-diaphyseal ulnar involvement. (b) AP femur pre- and post-op: characteristic 'shepherd's crook' deformity, managed with osteotomy and intramedullary fixation. Note the improvement in ossification following normalization of the proximal femoral anatomy.

lesions in areas that are not subject to load can be managed on a symptomatic basis, and regular clinical and radiological follow-up is recommended.

Bisphosphonates have been used to alleviate bone pain and have useful but unpredictable effect. Symptomatic areas with progressive deformity or pathological fracture require realignment and surgical stabilization (Figure 8.15b). Bone grafting is frequently used as an adjunct but the effect on union is unpredictable.

CONNECTIVE TISSUE DISORDERS

Collagen is the most common protein found in the body and accounts for 90% of non-mineralized bone matrix and approximately 70% of the structure of ligaments and tendons. There are over 20 types of human collagen: type I, the most common, is found in bone, skin, ligaments and tendons, type II in cartilage and type III muscle, skin and the walls of blood vessels.

Disorders of collagen synthesis lead to a diverse group of clinical conditions and this section discusses common disorders encountered in orthopaedic practice.

Ehlers-Danlos syndrome

Ehlers-Danlos syndrome is a group of inherited disorders caused by abnormalities of collagen formation or proteins responsible for normal collagen function. Affected individuals have fragile skin, joint hypermobility, vascular fragility and severe myalgia.

The inheritance pattern is variable and most cases have an autosomal dominant pattern. There are often abnormalities of collagen formation, commonly involving mutations of *COL5A1* or *COL5A2* with *COL1A1*, *COL1A2* and *COL3A1* also reported.

Diagnosis is often based on the constellation of clinical signs, particularly the degree of ligamentous laxity. Biochemical and genomic analysis of collagen are useful in some cases, but they cannot be guaranteed to identify an individual cause.

Affected individuals may develop cardiac anomalies including aortic root dilatation and mitral valve prolapse. Significant ligamentous laxity, elastic skin and a bruising tendency are common. Structural consequences, including congenital talipes equinovarus, progressive kyphoscoliosis and developmental dysplasia of the hips are also present. Shoulder, ankle and patella–femoral instability are common, and approximately 50% of individuals develop chronic musculoskeletal pain and are prone to early-onset osteoarthritis.

Patients with a suspected diagnosis should undergo an echocardiogram to evaluate the aortic root and all patients being considered for surgery should have a detailed cardiovascular assessment.

Specific *orthopaedic management* is directed at addressing painful or unstable joints, and physiotherapy is a central component of treatment. Persistent instability and severe and deteriorating joint pain, refractory to non-operative treatment, may require a surgical solution.

The *complications* of surgery, particularly those associated with wound healing, are increased in this condition due to fragile skin, excessive bleeding and vascular fragility. Soft tissue procedures are ineffective and arthrodesis is occasionally necessary. Instrumented spinal fusion is required in patients with progressive scoliosis and the level is determined to prevent junctional degeneration secondary to hypermobility.

Larsen's syndrome

Larsen's syndrome is a heterogeneous condition caused by an abnormality in the gene encoding filamin B, a cytoskeletal protein. The clinical features include joint hypermobility, multiple joint dislocations and distinct facial features including nasal bridge flattened, prominent forehead and hypertelorism. The majority of cases are inherited in an autosomal dominant pattern but a more severe autosomal recessive subtype is also recognized and is associated with cleft palate, short stature and cardiovascular anomalies.

Cervical kyphosis and talipes are often present in infancy and affected individuals develop dislocations of hips, knees, shoulders and radial heads in childhood.

Cervical spine *radiology* is important in the first year of life to identify cervical kyphosis. Neonatal ultrasound of the hips is necessary to identify acetabular dysplasia or hip dislocation. Tactical radiology is often necessary to evaluate the structure of joints that are prone to dislocation.

Management of patients with significant cervical kyphosis involves surgical stabilization in the first 18 months of life. If the patient has clinical or MRI evidence of neurological involvement, anterior decompression and posterior fusion are often necessary. In the absence of neurological signs, *in situ* posterior fusion provides sufficient stability.

The management of infantile hip dislocation is controversial. Closed reduction should initially be attempted, but it is frequently unsuccessful. Patients with unilateral involvement are usually managed with open reduction and realignment osteotomy. The indications for surgical treatment of bilateral dislocation are less clear and the general approach is

to attempt surgical reduction and stabilization at an early age but not to attempt revision if reduction is unsuccessful.

Knee dislocation presenting at birth can be treated with closed reduction and serial casting. If this is unsuccessful or there is recurrence, open reduction with femoral shortening and soft-tissue reconstruction may be required.

Marfan's syndrome

Marfan's syndrome is an autosomal dominant disorder, which is caused by a mutation in the fibrillin-1 gene (*FBN1*). It has an approximate incidence of 1:10000 and affects the musculoskeletal, ocular and cardiovascular systems.

Affected individuals are tall and hypermobile, with long limbs and digits.

Scoliosis is present in 50% of patients and is often associated with pectus excavatum. Abnormalities of acetabular development (protrusio) occur in 25% of cases. Severe ligamentous laxity produces a planovalgus foot deformity and recurrent joint dislocations. Extraskeletal manifestations include superior lens dislocation, spontaneous pneumothorax and aortic root dilatation leading to dissection. Dural ectasia is present in approximately 60% of patients.

The *diagnosis* is usually made on the basis of the clinical findings and, when suspected, should prompt an echocardiogram to assess aortic root involvement. All patients in whom surgery is planned should undergo preoperative cardiovascular examination and echocardiogram.

Orthopaedic surgical *treatment* frequently involves scoliosis correction, and *complications* including wound infection, hardware failure and pseudarthrosis are not uncommon. Hip pain, secondary to acetabular abnormalities, may require arthroplasty but abnormalities of the feet and joint instability is generally managed without surgery. This is due to the unpredictable and generally unsatisfactory outcome that accompanies attempts at soft-tissue reconstruction.

Tumours

9

Jonathan Stevenson & Michael Parry

Benign and malignant tumours of bone and soft tissue and tumour-like conditions are often considered together as they share a common clinical, radiological and pathological presentation. Given the rarity of their presentation, and the consequences of their misdiagnosis, these lesions are often diagnosed and managed in specialist centres by a multidisciplinary team with expertise in the imaging and pathology of such lesions, as well as their subsequent management. The classification and diagnosis of lesions of mesenchymal origin continues to evolve as developments in imaging, histopathology and genetics continue. Many tumours comprise a spectrum of disease from a latent benign disorder to a destructive, malignant neoplasm, and this overlap demands an appreciation of the principles of diagnosis and management of musculoskeletal tumours.

EPIDEMIOLOGY

Bone and soft tissue sarcomas comprise a family of tumours derived from mesenchymal tissue. Even when considered together, they are rare comprising less than 1% of all new cancer diagnoses. Bone sarcomas account for only 0.2% of all new cancer diagnoses in the UK, an incidence of only 8.2 cases per million of the population.

Soft-tissue sarcomas occur comparatively more frequently, with an incidence of 45 cases per million of the population. The incidence of the more common tumours is given in Table 9.1 along with the average number diagnosed in a year in the UK (population 62 million).

The incidence of bone and soft-tissue sarcomas is marginally higher in males than females. In the case of soft-tissue sarcomas, the incidence increases with age, with the highest incidence occurring in males over 85 years. A female preponderance is seen between 50 and 60 years of age due to the higher incidence of gynaecological sarcomas in this age group.

Table 9.1 The incidence of the common primary bone tumours within the UK population, and the number of new cases diagnosed per year in the UK

	Incidence per million population	Number diagnosed per year
Osteosarcoma	2.5	150
Chondrosarcoma	2.5	150
Ewing's sarcoma	2	120
Soft-tissue sarcoma	45	3300

In contrast, bone sarcomas demonstrate a bimodal distribution in both males and females, with peaks of incidence seen in both teenage/adolescent years and the elderly (Figure 9.1).

Primary bone tumours may arise in all sites but have preponderance for certain anatomical locations. More than 60% of tumours of bone will arise from the long bones of the lower limb, particularly around the knee. A further 18% will arise from the bones of the pelvis, sacrum or coccyx, and a further 13% from the upper limb or shoulder girdle (Table 9.2).

While the patho-aetiology remains unclear in the majority of cases of bone and soft-tissue sarcoma, certain DNA mutations or genetic associations are seen. In the case of osteosarcoma, 70% will demonstrate mutations in DNA helices or tumour suppressor genes. Such tumours are more common in patients with Li–Fraumeni syndrome, involving the *p53* suppressor gene, and those with retinoblastoma, involving the *Rb1* gene. In the case of Ewing's sarcoma, more than 95% of tumours demonstrate *EWS-ETS* fusion gene rearrangements, most commonly the result of translocations between chromosome 22 and chromosome 11 (*t22;11(q24;q12)* translocation) or 21 (*t21;22(q22;q12)*). The ETS transcription factor family is responsible for regulating cellular differentiation, the cell cycle, cell migration and apoptosis. Therefore mutations will result in deregulation of programmed cell death and disruption of cell-cycle control.

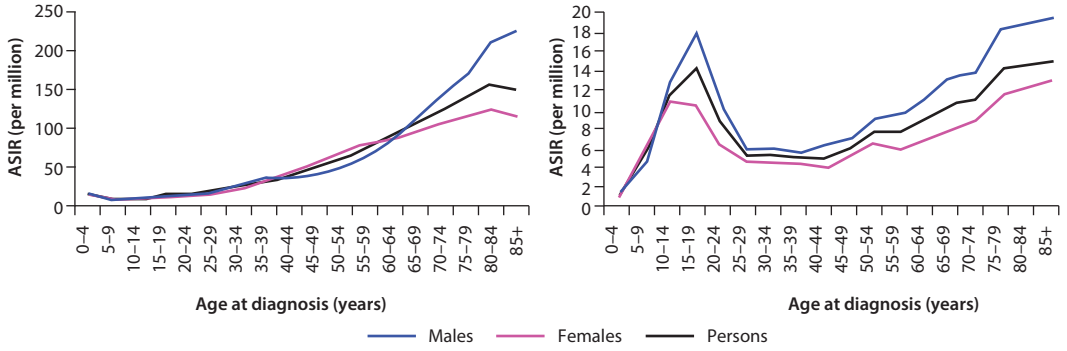


Figure 9.1 Age-specific incidence rates (ASIR) for soft-tissue sarcomas and primary sarcomas of bone (a) Soft-tissue sarcomas in the UK 1996–2010; (b) primary bone sarcomas in the UK for the same period (National Cancer Intelligence Network (NICN) data).

Table 9.2 Location of bone tumours split by the three most common diagnoses with the others being categorized together as UPS (undifferentiated pleomorphic sarcoma). Data from Royal Orthopaedic Hospital Oncology Service, Birmingham, UK

Site/Diagnosis	Chondrosarcoma	Ewing's	Osteosarcoma	UPS	Grand total	% of total
Distal femur	108	79	733	158	1078	27.8
Pelvis	264	190	134	89	677	17.5
Proximal tibia	45	78	349	56	528	13.6
Proximal femur	182	92	103	56	433	11.3
Proximal humerus	103	63	175	27	368	9.5
Trunk	96	44	33	19	192	5.0
Forearm	81	29	41	17	168	4.3
Ankle and foot	36	46	59	19	160	4.1
Fibula	8	61	72	10	151	3.9
Shoulder girdle	52	31	23	12	118	3.0
Grand total	**975**	**713**	**1722**	**463**	**3873**	**100%**

CLASSIFICATION

Tumours of bone and soft tissue are classified on the basis of their principle cell type, and have been classified by the World Health Organization (WHO). Tissue diagnosis is essential to predict the natural history and treatment of the lesion. In the case of soft-tissue sarcomas, the histological grade of the lesion is classified according to the Federation Nationale des Centres de Lutte Contre le Cancer (FNCLCC) system which has demonstrated correlation between grade and outcome for soft-tissue sarcomas (Table 9.3).

In contrast to soft-tissue sarcomas, tumours of bone can vary widely in their behaviour. Their histological subtype often determines the grade of bone tumour as there is no universally accepted grading system. Ewing's sarcoma and dedifferentiated chondrosarcoma are high-grade lesions, whereas parosteal osteosarcoma, though still a malignant lesion of bone, is considered a low-grade lesion.

A *benign lesion* of bone is defined as one that does not invade surrounding tissue or spread elsewhere in the body. Most benign bone tumours have a limited capacity for recurrence and when this does occur, it does so in a non-destructive manner. Surgical resection, therefore, is often curative. Benign lesions can demonstrate a wide variety of behaviours with some benign lesions being *latent* or inactive (e.g. non ossifying fibroma), while others are *active*, with a higher risk of recurrence after treatment (e.g. aneurysmal bone cyst).

Intermediate (locally aggressive) lesions of bone can destroy bone and surrounding tissue (e.g. osteoblastoma). These lesions often recur and are associated with an infiltrative and locally destructive growth pattern. Recurrence is frequent following limited surgical treatment and sometimes en bloc resection is required to completely remove the lesion.

Intermediate (rarely metastasizing) lesions often behave in a similar way to locally aggressive lesions but occasionally demonstrate the ability to spread to

Table 9.3 Definition of histopathological parameters in the Federation Nationale des Centres de Lutte Contre le Cancer (FNCLCC) grading system (HPF = high-power field)

Histological parameter	Definition	
Tumour differentiation	Score 1	Tumour closely resembles normal adult mesenchymal tissue and is difficult to distinguish from the counterpart benign tumour
	Score 2	Sarcoma for which histological typing is certain (e.g. myxoliposarcoma)
	Score 3	Embryonal and undifferentiated sarcomas, synovial sarcomas, sarcomas of doubtful type
Mitotic count (based on 10 HPF)	Score 1	0–9 mitoses per 10 HPF
	Score 2	10–19 mitoses per 10 HPF
	Score 3	>19 mitoses per 10 HPF
Tumour necrosis	Score 0	No necrosis
	Score 1	<50% necrosis
	Score 2	>50% necrosis
Histological grade	Grade 1	Total score 2, 3
	Grade 2	Total score 4, 5
	Grade 3	Total score 6, 7, 8

distant sites. The risk of such spread is <2%, is often not fatal and is not reliably predictable from the histological appearance. The classic example is the giant-cell tumour of bone.

Malignant tumours are truly aggressive with the potential for both local extension and metastases to distant sites. The aggressiveness of a tumour is defined by the histological grade. Low-grade tumours (e.g. chordoma and parosteal osteosarcoma) have a slow rate of growth and metastases are less common but can arise many years after initial diagnosis. High-grade tumours, conversely, have a very high risk of metastasizing, ranging from 20% to 100%, and are locally invasive (e.g. osteosarcoma and Ewing's sarcoma). Some histologically low-grade lesions have a metastatic rate of only 2–10%, though they may acquire a higher grade at the time of local recurrence and so a higher risk of metastasizing (e.g. chondrosarcoma).

When necessary, the grade of primary malignant tumours of bone is based on the cellularity and nuclear features of the cells. Generally, the higher the grade, the more cellular the tumour. Higher-grade lesions have a >25% risk of local recurrence and distant spread, whereas low-grade lesions a <25% risk of local recurrence and metastases (Table 9.4).

Table 9.4 The classification of tumours of bone (based on the World Health Organization (WHO) histological classification of tumours)

	Benign	Intermediate	Malignant
Osteogenic tumours	Osteoid osteoma	Osteoblastoma	Osteosarcoma
Chondrogenic tumours	Enchondroma Osteochondroma Periosteal chondroma Synovial chondromatosis	Chondroblastoma	Chondrosarcoma Clear-cell chondrosarcoma Dedifferentiated chondrosarcoma Mesenchymal chondrosarcoma
Fibrogenic tumours		Desmoplastic fibroma	Fibrosarcoma
Giant cell-rich tumours		Giant-cell tumour	Malignant giant-cell tumour
Notochordal tumours	Benign notochordal tumour		Chordoma
Vascular tumours	Haemangioma	Epithelioid haemangioma	Angiosarcoma Epithelioid haemangioendothelioma
Myogenic tumours	Leiomyoma of bone		Leiomyosarcoma of bone
Lipogenic tumours	Lipoma of bone		Liposarcoma of bone
Undefined tumours	Simple bone cyst Fibrous dysplasia Osteofibrous dysplasia	Aneurysmal bone cyst Langerhans cell histiocytosis Erdheim–Chester disease	Ewing's sarcoma Adamantinoma Undifferentiated pleomorphic sarcoma (UPS) of bone

STAGING

Staging is the process of assessing the extent of a tumour both locally and distantly. As a consequence of most bone and soft-tissue sarcomas metastasizing via the bloodstream, the lungs are the most common site for metastases, although other sites may include bone, lymph nodes, liver and other soft-tissue locations. The basis of all staging systems relies on knowledge of the grade of the tumour (a measure of the aggressiveness of the tumour – high, intermediate or low), the size of the tumour, the local extent, and the presence of metastases.

The classic staging system for primary tumours of bone is the *Enneking system*. Developed as a reference to aid the extent of surgical resection in primary tumours of bone, the Enneking staging system classifies tumours according to whether they are high or low grade, whether there are metastases present or not, and whether the tumour has grown out of its original compartment or remains confined to a compartment. A compartment, for the purposes of this system, is defined as an enclosed tissue space, such as a bone, a joint space or a muscle group confined by its fascial envelope.

The *Enneking stages* are:

Stage 1A	Low-grade, intra-compartmental tumour
Stage 1B	Low-grade, extra-compartmental tumour
Stage 2A	High-grade, intra-compartmental tumour
Stage 2B	High-grade, extra-compartmental tumour
Stage 3	Any of the above with metastases

Thus, according to the Enneking system, a low-grade central osteosarcoma completely within a bone would be defined as stage 1A, operable by wide excision of the bone, whereas a high-grade conventional, osteoblastic osteosarcoma with extension from the bone to the soft tissues would be defined as stage 2B and would need careful consideration with regards to limb salvage or sacrificing surgery.

More recently, the more conventional *tumour–node–metastasis (TNM) staging* has been applied to primary sarcomas of bone. This takes into consideration the histological subtype, size, continuity, grade, and local and distant spread of the tumour. Limitations of this application to bone tumours exist, for example due to the rarity of lymph-node metastases. This system has, however, gained favour for the staging of soft-tissue sarcomas and has been adopted by the American Joint Committee on Cancer (AJCC), and the Union for International Cancer Control (UICC). In this system, size is dichotomized to small (<5 cm for soft-tissue sarcomas, <8 cm for bone sarcomas) or large (>5 cm for soft-tissue sarcomas, >8 cm for bone sarcomas).

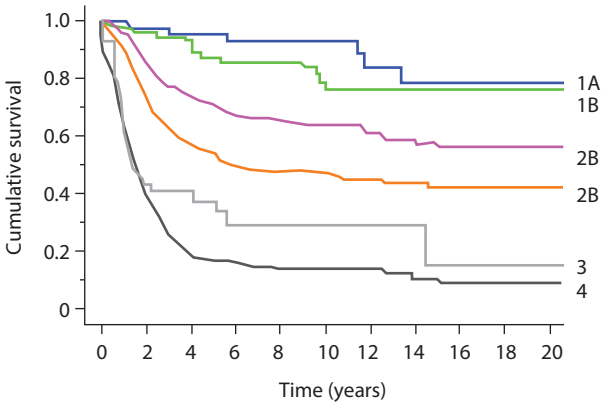


Figure 9.2 Kaplan–Meier survival curve The graph shows the effect of stage on prognosis for all bone sarcomas (Royal Orthopaedic Hospital Oncology Service data).

The *TNM stages* are:

Stage 1A	Low-grade, small, no metastases
Stage 1B	Low-grade, large, no metastases
Stage 2A	Intermediate- or high-grade, small, no metastases
Stage 2B	Intermediate-grade, large, no metastases
Stage 3	High-grade, large, no metastases
Stage 4	Any with metastases

Regardless of the system adopted, the stage of disease has clinical significance to both the patient and the clinician due to the association of poorer prognosis with advanced-stage disease (Figure 9.2). Staging allows prediction of prognosis which improves communication and classification between units and between patients and clinicians.

CLINICAL PRESENTATION

Despite improvements in clinician education, and advances in referral pathways for specialist investigation, the diagnosis of bone and soft-tissue sarcomas is often delayed. Physicians, particularly those dealing with children and adolescents, must maintain a high index of suspicion for malignancy. Patients may be asymptomatic until the lesion is discovered incidentally on radiographs, common for benign lesions, or for malignant lesions arising in areas where there is room for innocuous expansion, such as the pelvis, where tumours can achieve a very large size prior to presentation. In an attempt to rectify these delays in diagnosis, the National Institute for Health and Care Excellence (NICE) in the UK issued guidelines for the improvement in care for patients presenting with sarcomas.

Age is often a consistent feature for primary bone tumours; osteosarcoma and Ewing's sarcoma have preponderance for children, adolescents and young adults, while chondrosarcoma typically occurs in older patients. With increasing age, the likelihood of a lesion of bone being the result of metastatic disease increases and the investigation of any pathological lesion of bone in an elderly patient must include an attempt to identify a potential primary malignancy elsewhere. Indeed, in patients over 70 years of age, the incidence of metastatic bone disease is greater than that of all primary bone tumours in any age group.

The most common symptom experienced by a patient with a bone tumour is *pain*. The pain is initially mild but then gets worse as the tumour increases in size. Night pain is a particularly worrying symptom that should always cause concern. Worrisome features from the history include pain, particularly night pain, pain not responding to simple analgesia, persistent pain following injury, as well as prior benign or malignant lesions, family history and previous radiotherapy. All too often, young patients are not referred for investigation, as the diagnosis is not considered. A recent injury does not rule out a malignant pathology and must not thwart investigation. The duration of symptoms prior to diagnosis is often measured in months and for slow-growing tumours such as chondrosarcoma or chordoma in years. Swelling arises only when a tumour has extended outside the bone and may initially be difficult to detect. Referred pain is not uncommon, particularly with pelvic tumours which can present with abdominal, back or leg pain. While paraesthesia or numbness are suggestive of compression of a nerve by an expanding mass, progressive neurological dysfunction is far more worrisome and is suggestive of direct tumour invasion.

Presentation with a *pathological fracture* has been reported in between 5 and 12% of osteosarcomas and up to 21% of chondrosarcomas, and in the case of benign lesions is suggestive of a locally aggressive lesion. Prodromal symptoms of worsening functional pain are common and, in children in particular, a fracture with a disproportionate level of injury (e.g. fall from standing height) should stimulate further investigation. Certain fracture patterns should raise concern for an underlying lesion, such as supracondylar femoral fractures in children, and avulsion fractures of the lesser trochanter in adults. In elderly patients with diaphyseal long-bone fractures, the possibility of a pathological fracture should be considered.

Clinical findings in the case of primary bone tumours are often non-specific. *Swelling and tenderness* over the affected bone are the most common findings but there will be limitation of joint movement if there has been irritation of the joint by the tumour, or the tumour has grown into the joint. Systemic findings are often rare except in extreme cases where presentation is very late and dissemination has already occurred.

In the case of soft-tissue sarcomas, the majority present as a painless, enlarging mass. The rapidity of enlargement is often suggestive of a malignant process, though this does not accurately differentiate benign from a more sinister pathology. All superficial soft-tissue lesions measuring greater than 5 cm and all deep-seated lesions should be considered a sarcoma until proven otherwise.

INVESTIGATIONS

X-Rays

All patients with the suspicion of a lesion of bone should be investigated with plain X-rays. There may be an obvious abnormality in the bone, such as cortical thickening, a 'cyst', or ill-defined destruction (Table 9.5). The location within the bone and whether the lesion is solitary or multiple should be noted (Figure 9.3). The periphery of the lesion, whether it is well-defined or ill-defined, should also be noted.

When assessing a suspicious lesion on X-ray, there are a number of questions that should be asked (see Box 9.1).

In the majority of cases, the stimulus for further investigation and an initial assessment of the nature of the lesion can be made from the plain X-rays. In certain lesions of bone, the X-ray may illustrate pathognomonic features of the diagnosis (e.g. osteochondroma, non-ossifying fibroma, osteoid osteoma).

Radionuclide scanning

Scanning with ^{99m}Tc-methyl diphosphonate (^{99m}Tc-MDP) shows non-specific reactive changes in bone which can be helpful in revealing the site of a small tumour (e.g. an osteoid osteoma). Skeletal scintigraphy is also useful for detecting skip lesions and evidence of metastatic disease as part of the initial staging process.

Computed tomography

CT extends the range of X-ray diagnosis; it shows more accurately both intraosseous and extraosseous extension of the tumour and the relationship to surrounding structures. CT is essential to complete systemic staging and re-staging in bone and soft-tissue sarcomas and metastatic disease to identify pulmonary metastases.

Magnetic resonance imaging

MRI allows further characterization of lesions and defines the local extent of the lesion. Its greatest value

Table 9.5 Possible diagnosis based on the radiographic appearances, divided by age group

Age (years)	Well-circumscribed lesion	Ill-defined lesions	Sclerotic lesions
0–10	Eosinophilic granuloma Simple bone cyst	Eosinophilic granuloma Ewing's sarcoma Leukaemia	Osteosarcoma
10–20	Non-ossifying fibroma Osteoblastoma Fibrous dysplasia Eosinophilic granuloma Simple bone cyst Aneurysmal bone cyst Chondroblastoma Chondromyxoid fibroma	Ewing's sarcoma Eosinophilic granuloma Osteosarcoma	Osteosarcoma Fibrous dysplasia Eosinophilic granuloma Osteoid osteoma Osteoblastoma
20–40	Giant-cell tumour Enchondroma Low-grade chondrosarcoma Brown tumour Osteoblastoma	Giant-cell tumour	Enchondroma Bone island Parosteal osteosarcoma Burnt-out lesion: • Non-ossifying fibroma • Eosinophilic granuloma • Simple bone cyst • Aneurysmal bone cyst • Chondroblastoma
40+	Metastases Myeloma Geode	Metastases Myeloma High-grade chondrosarcoma	Metastases Bone island
All ages	Infection	Infection	Infection

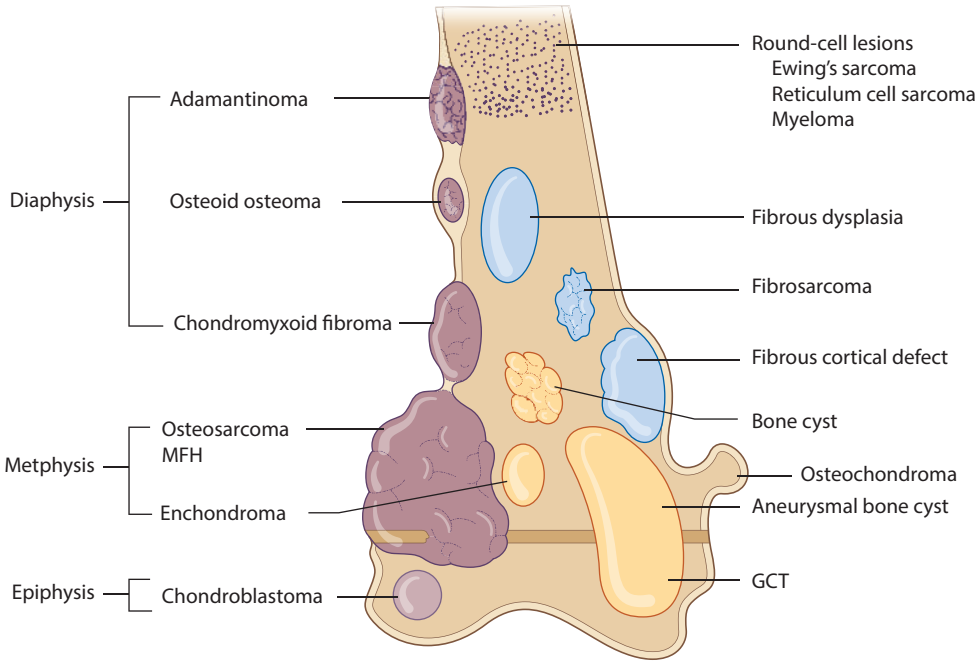


Figure 9.3 Potential diagnoses The location of the lesion within the bone is significant.

is in the assessment of tumour spread: (a) within the bone, (b) into a nearby joint, and (c) into the soft tissues. Blood vessels and the relationship of the tumour to the perivascular space are well-defined, which aids greatly in preoperative assessment and the prediction of resection margins for limb-salvage surgery. MRI is also useful in assessing soft-tissue tumours and cartilaginous lesions. It may demonstrate features consistent within certain lesions (e.g. fluid-fluid levels within an aneurysmal bone cyst (ABC) or telangiectatic osteosarcoma). All primary tumours of bone should be investigated with an MRI of the entire

BOX 9.1 QUESTIONS TO ASK WHEN ASSESSING AN X-RAY

What is the age of the patient?

Which bone is affected?

Where in the bone is the lesion?

What is the lesion doing to the bone: is it osteolytic, osteoblastic or mixed?

What is the bone doing in response?

Is there a periosteal reaction?

Is there an associated soft-tissue mass?

What is the nature of the matrix: is it osteoid, chondroid or fibroid?

Is the lesion solitary or are there multiple lesions?

bone to assess for the presence of satellite lesions (a lesion distant from the tumour but within the reactive zone of the tumour) or skip lesions (a lesion outside the reactive zone of the tumour but within the same bone).

INVESTIGATING THE 'SUSPICIOUS BONE LESION'

In many cases an abnormality will be detected radiologically but the diagnosis will not be clear. These patients require more specific investigation.

The differential diagnosis of a bone abnormality will depend on the age of the patient, the location of the lesion and the radiographic or MRI characteristics of the lesion. Some lesions have absolutely typical presentations: for example, in a child an epiphyseal lesion is likely to be either a chondroblastoma or infection while at the same location in an adult it is more likely to be a clear cell chondrosarcoma or a giant-cell tumour. Well-demarcated lesions tend to be benign while ill-defined lesions are more likely to be malignant or metastatic. Metastases to bone are increasingly likely after the age of 35, particularly if the patient has a past history of malignancy. However, 15% of patients with a bone lesion and a past history of cancer are found to have a different cancer to the primary. Therefore, it is vital to assume nothing and keep an open mind.

Investigating a suspicious bone lesion should follow well-established steps. Always start with a thorough history and examination, looking, for instance, in an older patient at possible symptoms or signs of other malignancy. The usual primary sites that metastasize to bone are bronchus, breast, prostate, kidney and thyroid, and all of these need to be considered as possible primary sites in an older patient with a destructive bone lesion.

If the diagnosis is not clear (to an experienced musculoskeletal radiologist) on plain X-rays, then further investigation is warranted. Usually, this will consist of an MRI scan to delineate the extent of the lesion in the bone and other tissues. If there is a suspicion of malignancy, then tests should be directed at identifying either a metastatic lesion or an occult primary. CT scans of the chest, abdomen and pelvis will be helpful to identify occult primary carcinomas. Biochemical tests will help to exclude prostate cancer (PSA) or myeloma (serum electrophoresis and urinary Bence-Jones proteins) as possible causes. In older patients with the possibility of the lesion being metastatic, a bone scan is indicated to assess if the lesion is solitary or multiple. Other blood tests may give useful information (e.g. raised calcium may indicate hyperparathyroidism, or raised alkaline phosphatase may indicate Paget's disease, which is a prognostic factor in osteosarcoma). In the case of a suspected brown tumour, serum parathyroid hormone will be elevated.

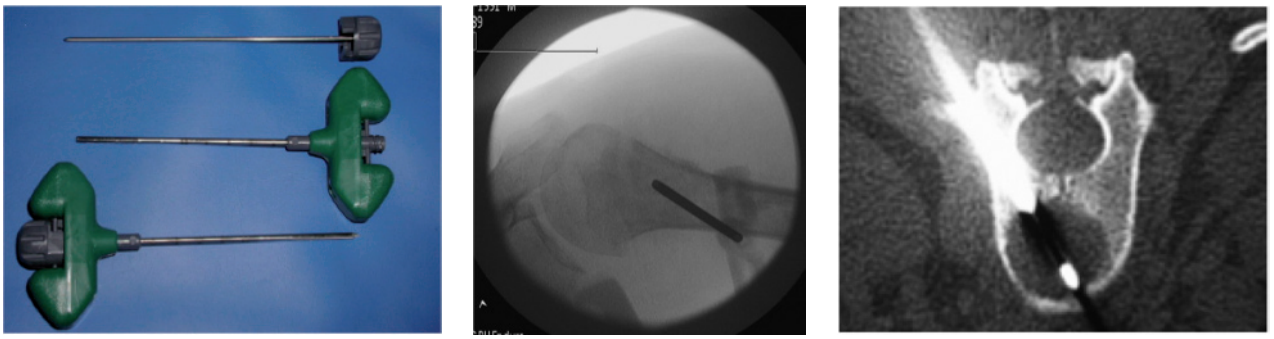
Finally, it must be remembered that infection can be a great mimic of tumours and it is good practice to assess the CRP and ESR in all these patients. Interestingly, these inflammatory markers have also been shown to be of prognostic value in a variety of bone and soft-tissue tumours.

Biopsy

Biopsy remains the gold standard for obtaining a diagnosis in an abnormal lesion of bone. There are several principles in carrying out a biopsy that must be considered:

- The biopsy tract must be sited to minimize potential contamination of normal tissues and should be planned in conjunction with the surgeon who will carry out any definitive surgery.
- The biopsy must be taken from representative tissue.
- Image-guided biopsies should be used to reduce the risk of a sampling error.
- Complete haemostasis must be achieved.
- Samples should always be sent for microbiology as well as histology.
- The pathologist reporting the biopsy must have an appropriate level of experience.
- If there is a risk of fracture following biopsy, the bone must be appropriately splinted.

In general, most bone biopsies are now done with either fluoroscopic or CT guidance and in most large



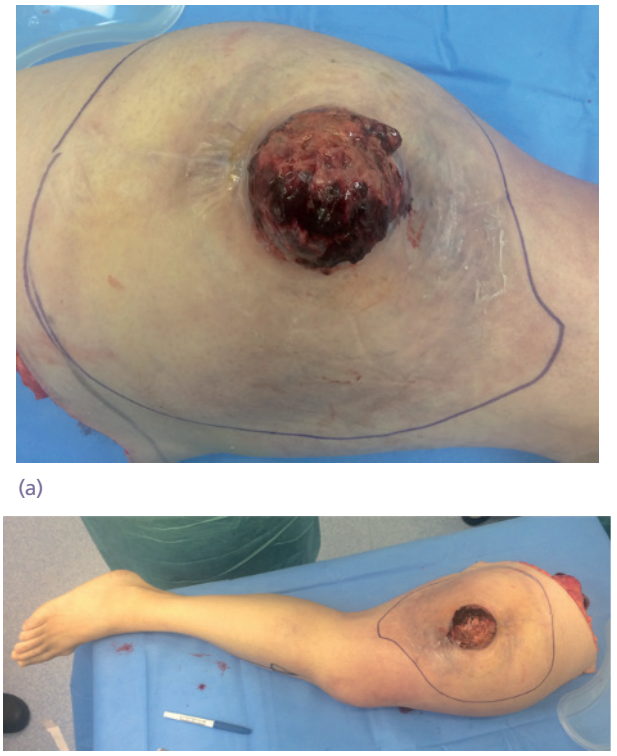
(a) (b) (c)

Figure 9.4 Biopsy (a) The majority of biopsies are now performed with a core needle, such as a Jamshidi needle. The procedure is often performed with radiographic guidance, either as fluoroscopy (b), or under CT guidance (c).

centres *needle biopsies* will provide adequate tissue for diagnosis (Figure 9.4).

In the case of a non-diagnostic needle biopsy or if a needle biopsy could place neurovascular structures at risk, an *open biopsy* may be considered. The site is selected so that it can be included in any subsequent operation. As little as possible of the tumour is exposed and a block of tissue is removed, ideally in the boundary zone, so as to include normal tissue, pseudocapsule and abnormal tissue. If bone is removed, the raw area is covered with bone wax or methylmethacrylate cement, to reduce bleeding and contamination from the cut bone edge. If a tourniquet is used, it should be released and haemostasis achieved before closing the wound. Drains should be avoided, so as to minimize the risk of tumour contamination, but where used they should be placed in line with the wound to allow excision at the time of definitive tumour resection.

Taking a biopsy at the time of fixing a pathological fracture can be condoned only if the patient has known metastatic cancer. In a patient with an apparent solitary lesion and a past history of cancer, then in general biopsy should be carried out to confirm the diagnosis before treatment is instigated.



(a) (b)

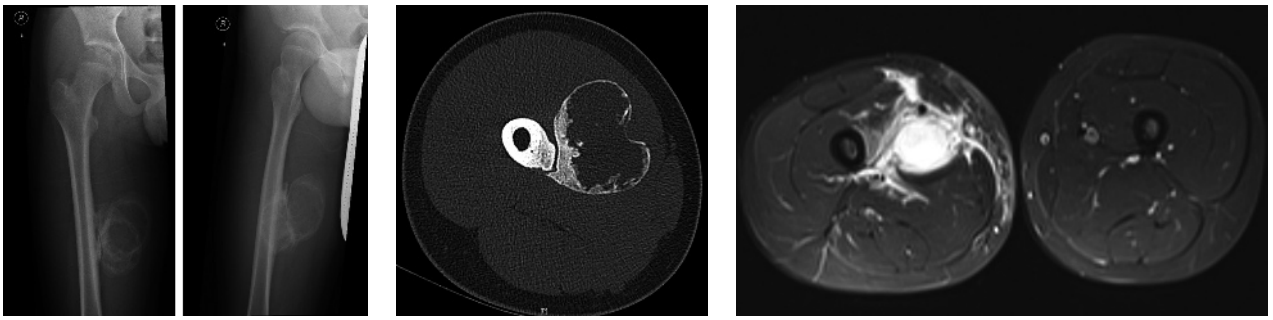
Figure 9.5 (a,b) A 48-year-old woman who was otherwise fit and well presented with a mass on the outer aspect of the upper right thigh. This was presumed to be a haematoma, despite the absence of use of anticoagulants or a history of trauma. The lesion was evacuated and debrided. Histology following repeated evacuation for reaccumulation eventually confirmed a high-grade soft-tissue sarcoma. (b) Unfortunately, due to the degree of soft-tissue involvement following these procedures, this could only be treated by hip disarticulation.

BONE TUMOUR MIMICS

There are a variety of bone and soft-tissue lesions which can cause great concern clinically, radiologically and sometimes pathologically. If in doubt, the case should be discussed with a specialist centre where expertise will be available to obtain the correct diagnosis and advice on treatment.

Soft-tissue haematoma

A large, clotted sub-periosteal or soft-tissue haematoma may present as a painful lump in the arm or lower limb. Sometimes the X-ray shows an irregular surface on the underlying bone. Important clues are the history and the rapid onset of symptoms. However, it must be remembered at all times that haematomas do not just happen and, if the history does not correlate with the X-ray appearance, a malignant process must be considered (Figure 9.5).



(a) (b) (c)

Figure 9.6 Myositis ossifcans A 13-year-old boy presented with a painful mass in the adductor compartment of the right thigh. On questioning, the boy described receiving a dead leg while playing rugby a couple of months earlier, which he felt had never really settled. (a) X-rays demonstrate a large opacification within the medial thigh but separate from the femur. There is evidence of periosteal calcification. (b) CT confirms the calcific mass with a clear plane between the lesion and the femur. (c) MRI in the initial phases demonstrates marked soft-tissue oedema, which corresponds to the patient's symptoms of pain. The patient was managed with careful observation. The lesion was seen to consolidate and symptoms settled without intervention.

Myositis ossificans

Although rare, this may be a source of confusion. Following an injury, the patient develops a tender swelling in the vicinity of the area of injury. Radiographs demonstrate fluffy density in the soft tissue adjacent to bone (Figure 9.6). Unlike a malignant tumour, however, the condition soon becomes less painful and the new bone better-defined and well-demarcated. Where surgical removal is considered, this should occur only once the bone has matured, as seen on bone scintigraphy, to reduce the risk of recurrence.

Stress fracture

Stress fractures present a particular diagnostic minefield. The patient is often a young adult with localized pain near a large joint. Radiographs often demonstrate a dubious area of cortical 'destruction' and overlying periosteal new bone formation, especially if the presentation has been delayed. If a biopsy is performed, the healing callus seen in a stress fracture may be confused with the osteoid production often seen in cases of osteosarcoma. If the pitfall is recognized, and there is adequate consultation between surgeon, radiologist and pathologist, misdiagnosis can be prevented, but this relies on a thorough history from the patient and their family.

Tendon avulsion injuries

Children and adolescents, especially those engaged in vigorous sports, are prone to avulsion injuries at sites of tendon insertion, particularly around the hip and knee (e.g. tibial apophyseal stress lesion, Osgood-Schlatter disease). Less common sites of avulsion, such as the iliac crest, the ischial tuberosity, the lesser trochanter of the femur, hamstring insertions, attachment of adductor magnus and longus, and the distal humeral apophyses), have been confused as tumorous lesions arising from bone.

Infection

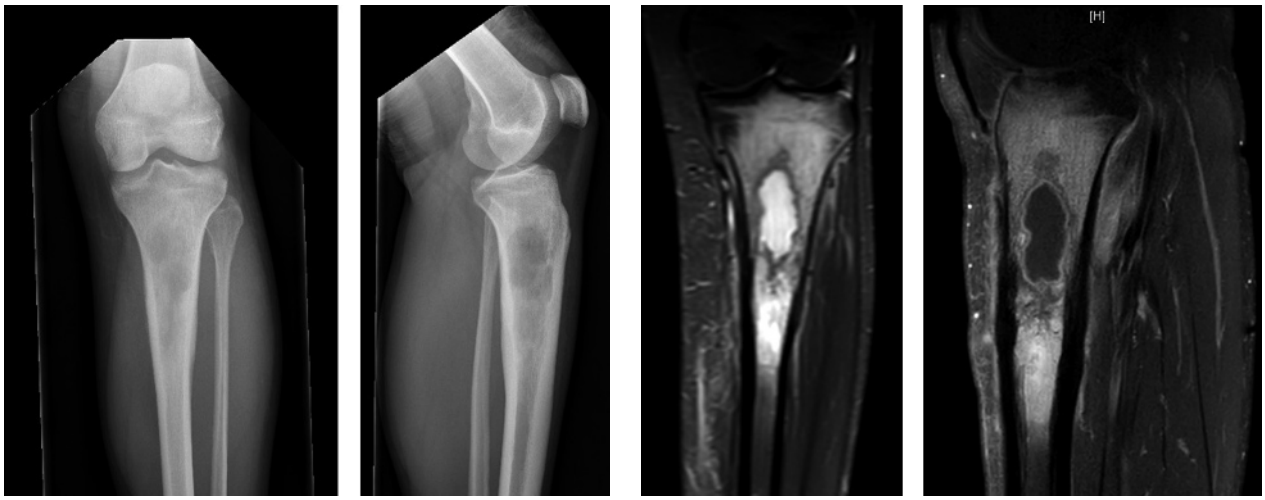
Osteomyelitis typically causes pain and swelling in the metaphysis of long bones in children and young adults, as is the case in primary tumours of bone. Systemic features may be mild. Radiographs may show bone destruction and periosteal new bone formation. While cross-sectional imaging may demonstrate a localized, intraosseous collection (Brodie's abscess), and haematological investigations may point to the diagnosis, often radiographic features may be misleading and blood tests largely normal (Figure 9.7). Biopsy should be considered, sending material for both histopathological and microbiological investigation which can also help guide antimicrobial therapy if infection is identified.

Gout

Occasionally, a large gouty tophus can cause a painful swelling at one of the bone ends. Radiographs may show a large, poorly defined excavation on the bone. In the majority of cases, a diagnosis of gout has already been given, but where doubt remains, a biopsy will be diagnostic.

Osteopetrosis

Known as marble bone disease, or Albers-Schönberg disease, this is a rare inherited disorder characterized by an increase in bone density, although bones



(a) (b)

(c) (d)

Figure 9.7 Infection (a,b,c) A 29-year-old chef presented with rapidly progressive pain in the left proximal tibia with feelings of general malaise and fever. He had previously been treated for osteomyelitis at the same site at the age of 15 with antibiotics alone. (a) X-rays demonstrate a lytic expansile lesion in the proximal tibia. Blood tests in this case revealed a marked elevation in both CRP and ESR. (b) MRI demonstrates a fluid-filled cavity in the proximal tibial metaphysis with rim enhancement and florid perilesional oedema but without expansion into the periosteum or soft tissues. This was successfully treated with extended intralesional curettage and packing of the cavity with antibiotic laden synthetic bone graft (c), together with prolonged antibiotic therapy. (d) X-ray of left femur of 12-year-old girl experiencing progressive pain. Note the imaging features of lamellar periosteal reaction ('onion-skin periostitis') with a permeative pattern in the femoral diaphysis and cortical expansion. These features are remarkably similar to those seen in femoral diaphyseal Ewing's sarcoma but in this case biopsy confirmed osteomyelitis.

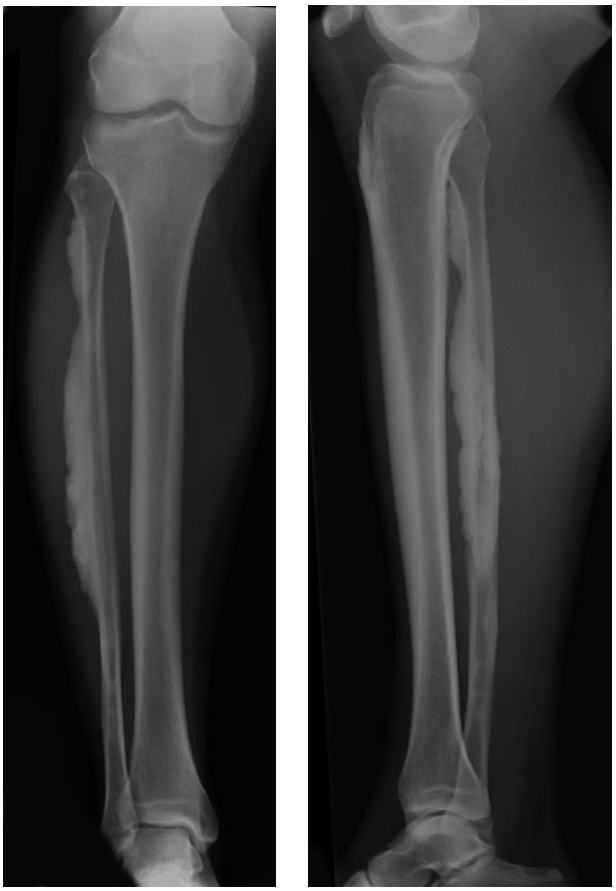
in osteopetrosis are very brittle. The radiographic appearances are often confused with sclerotic metastases from breast or prostate carcinoma, Paget's disease, or osteoblastic primary bone malignancies.

Osteopoikilosis

An autosomal dominant sclerosing bone dysplasia characterized by the formation of multiple bone islands. Classically occurring in patients in their twenties, it can be differentiated from multiple osteoblastic metastases seen in more elderly patients.

Melorheostosis

A mesenchymal dysplasia characterized by widening and sclerosis of cortices in a sclerotomal distribution. Radiographs demonstrate a classic dripping candle wax appearance. The presentation is usually monostotic and unilateral (Figure 9.8).



(a) (b)

Figure 9.8 Melorheostosis Radiographs for a 35-year-old man who presented with pain in the right knee incidentally identified a lesion in the fibula (a) which demonstrates the characteristic 'dripping candle wax' features (b) of melorheostosis.

PRINCIPLES OF MANAGEMENT OF PRIMARY TUMOURS OF BONE

The management of patients with bone sarcomas must be carried out by a multidisciplinary team including clinicians, pathologists, radiologists and oncologists together with appropriate expertise governed on a case-by-case basis, for example vascular, general, urological, spinal, thoracic and plastic surgical specialists, as well as dedicated specialist nurses and rehabilitation experts. All intermediate lesions and some benign lesions are also best managed in these centres. Knowledge of the natural history of all these lesions is essential to avoid both under- and over-treatment.

Latent benign lesions may not require any treatment unless symptomatic. Active benign lesions usually require intervention to halt the active process and allow healing. This can range from limited biopsy and curettage to detailed curettage and bone grafting. Again, knowledge of the lesion and its likely behaviour can strongly influence the management approach.

Intermediate lesions present considerable challenges in management as recurrence is more common, and if this arises, en-bloc removal of the lesion may be required to achieve local control. Close follow-up is also required because of the risk of recurrence.

Malignant tumours require the full skills of the MDT to decide on optimum management. Many tumours will respond to chemotherapy (e.g. osteosarcoma and Ewing's sarcoma) and most will require surgical resection. Patients with a Ewing's sarcoma or chordoma may also require highly specialized radiotherapy.

PRINCIPLES OF SURGICAL TREATMENT AND DEFINITION OF MARGINS

The resection of all detectable tumours, including metastases, is the cornerstone of treatment of the majority of sarcomas of bone. *For primary operable tumours, the options are tumour excision with limb salvage, or amputation.* In the case of tumour resection, this should encompass resection with a clear margin including the pseudocapsule of the tumour and a cuff of normal tissue. The margin of resection will be governed by preoperative local staging imaging. In general, more aggressive tumours mandate wider excisions to ensure complete removal.

In 1980, Enneking defined the different margins of excision that can be used when dealing with bone tumours (Figure 9.9). Many benign lesions can be

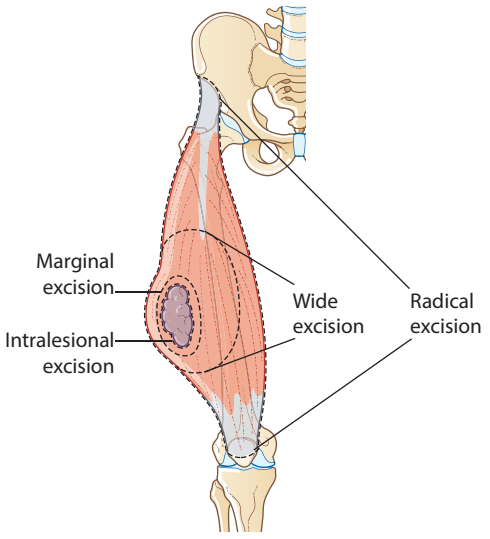


Figure 9.9 Margins of excision The margins of tumour excision defined by Enneking (soft-tissue sarcoma). When considering tumours of bone, the bone can be considered as a compartment.

managed by *intralesional excision*. This implies that the lesion has been removed by debulking. This would apply to curettage of a benign lesion but would also apply if an attempt was made to excise a malignant lesion and the tumour was actually cut across during the operation, allowing tumour spill.

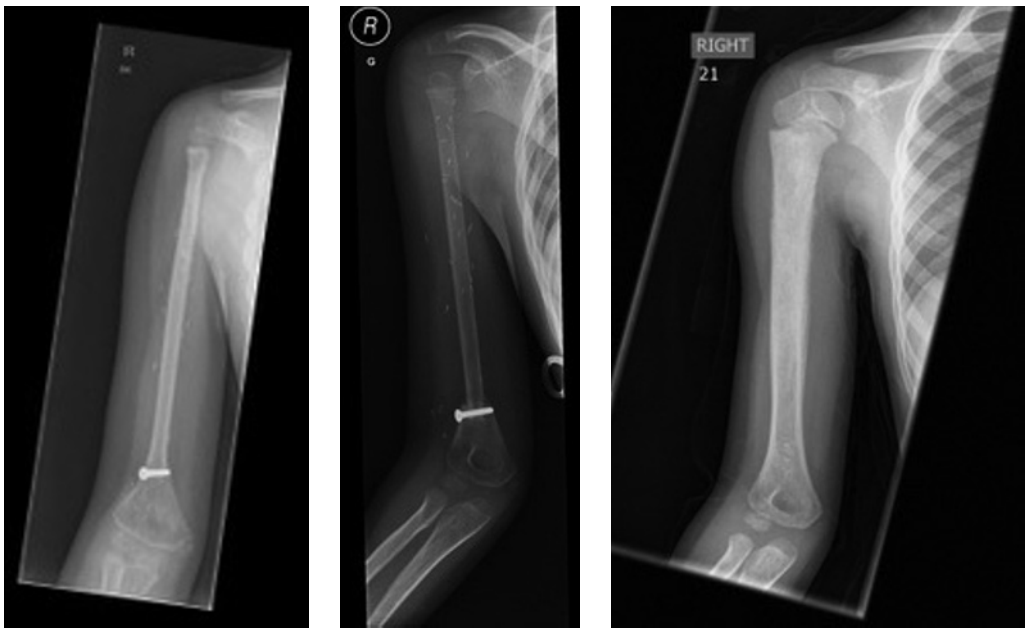
A *marginal excision*, going around the edge of the lesion, will usually be sufficient to completely remove and control a benign or intermediate lesion but will usually be insufficient for a malignant tumour as there are likely to be tumour cells in the tissues left behind. This zone around a malignant tumour is known as the reactive zone and consists of tissues compressed by the expansion of the tumour, also known as the pseudocapsule. While it may appear to be a natural plane of dissection, it will often contain tumour cells. Surgical resection encroaching on the area of oedema around a tumour will also result in a high risk of local recurrence. This risk can be reduced with an effective adjuvant such as chemotherapy or radiotherapy in selected tumours, allowing a planned marginal excision of a malignant tumour (e.g. if the tumour is close to a critical structure such as a nerve or blood vessel).

A *wide excision* implies that the surgery has been carried out through completely normal tissue, well away from the tumour.

There is no doubt that neoadjuvant chemotherapy can change the significance of the margin, with a good chemotherapy response dramatically reducing the risk of local recurrence for the same margin achieved. The key factor affecting the width of the margin is usually the closest critical structure – commonly the neurovascular bundle.

The decision between carrying out *limb salvage or amputation* is governed by the expected postoperative limb function, potential for complications, psychological acceptance and oncological outcomes. Quality of life studies suggest that patients adapt equally well to amputation as limb salvage. Most importantly, no significant difference exists between the survival rates for amputation compared to limb salvage surgery, however, as survival is affected by the margins achieved at resection, limb salvage must not be favoured over amputation where the potential to compromise a complete resection exists. Limb salvage is possible for approximately 85% of appendicular tumours. Local recurrence rates for amputation and limb salvage are similar and approximately 5% for osteosarcoma, 9% for Ewing's sarcoma, and 25% for chondrosarcoma.

Options for reconstruction in limb salvage include endoprosthetic replacement, allograft-prosthetic composite, allograft using donated bone extracorporeal



(a) (b) (c)

Figure 9.10 The case of a 5 year old girl with an osteosarcoma of the proximal humerus. This was treated by neoadjuvant chemotherapy followed by resection of the tumour and reconstruction using a vascularised proximal fibular transfer, utilising the perforating artery to the epiphysis to reconstruct the neo-humeral head (a). Over time, the neo-humerus hypertrophies (b) and after 36 months (c) has achieved a comparable dimension to the contralateral humerus.

sterilization and reimplantation of the patient's own bone, vascularized and non-vascularized autograft and arthrodesis. Each option has its advantages, and application is governed on a case-by-case basis. Endoprosthetic replacement offers mobilization and stability although it incurs the risks of mechanical failure (18%) and infection (11%). Allograft–prosthetic composite affords the advantages of early stability with the potential biological benefits of allograft bone. Allografts, particularly osteoarticular allografts, can be used to reconstruct complex structures including joints, allowing reattachment of soft tissues. However, the complications are potentially high including fracture, non-union and infection rates greater than 20%.

Limb salvage can be particularly challenging in the skeletally immature as resection often requires excision or compromise of the physes, particularly those around the knee, with subsequent implications for limb length discrepancy. This can be counteracted with the use of an extendible endoprosthesis, allowing interval lengthening comparable to the contralateral limb. Lengthening can be achieved either minimally invasively, necessitating multiple surgical interventions and the concurrent increased risk of peri-prosthetic infection, or non-invasively through extracorporeal magnetic distraction of a motor within the implant. In the paediatric upper limb, vascularized epiphyseal transfers allow axial growth and hypertrophy following humeral or radial excisions.

For tumours involving the distal femur or proximal tibia in skeletally immature patients, intercalary resection and 180-degree rotation of the distal leg, recreating the knee joint with the prior ankle joint, is an option. Following rotationplasty, prosthesis is worn at the knee for ambulation, and gait analysis demonstrates improved kinematics when compared to above-knee amputation. However, the procedure is technically challenging and some patients and families cannot tolerate the cosmetic disfigurement of the neo-knee joint.

Neoadjuvant and adjuvant therapies

Neoadjuvant chemotherapy has been used for most patients with osteosarcoma and Ewing's sarcoma for well over 35 years. In osteosarcoma the principle drugs used are doxorubicin, cisplatin, ifosfamide and high-dose methotrexate, in various combinations to avoid chemo-resistance and increase the rate of tumour necrosis. The aim of this chemotherapy is to try to shrink the tumour while also treating the micrometastatic disease. Patients without radiologically detectable metastases are considered to have micrometastases and so are treated with the same neoadjuvant regimen. Neoadjuvant chemotherapy allows the assessment of the tumour's sensitivity to chemotherapy following definitive surgical resection, itself a predictor of local recurrence and disease-free survival. An improved survival is seen for those with a good response to neoadjuvant chemotherapy, defined as a tumour necrosis rate >90% in the case of osteosarcoma.

In cases of Ewing's sarcoma, most patients with apparently localized disease will have subclinical micrometastases and so any treatment must include systemic therapy in combination with local control through radiotherapy and/or surgery. As Ewing's sarcoma is often sensitive to chemotherapy, most patients will receive neoadjuvant chemotherapy, regardless of the extent of the local disease. Current regimens rely on vincristine, doxorubicin and cyclophosphamide, alternating with ifosfamide and etoposide. Multimodal neoadjuvant chemotherapy in Ewing's sarcoma will often produce a dramatic shrinkage of the tumour and reduction in symptoms.

Local control is achieved by surgical resection whenever possible. The main principle is that all of the initially involved tissue must be sterilized either by surgical excision or by radiotherapy, no matter how effective the chemotherapy, and the two are often used in combination. This will result in the lowest risk of recurrence.

High-energy irradiation has long been used to destroy radiosensitive tumours or as adjuvant therapy before operation. Nowadays the indications are more restricted. For highly sensitive tumours (such as Ewing's sarcoma), it offers an alternative to amputation or as an adjunct to surgery and chemotherapy for tumour locations where achieving an adequate margin may be difficult. The same combination can be used as adjunctive treatment for high-grade tumours, for tumours in inaccessible sites, lesions that are inoperable because of their size, proximity to major blood vessels or advanced local spread, for marrow-cell tumours such as myeloma and malignant lymphoma, for metastatic deposits and for palliative local tumour control where no surgery is planned. Radiotherapy may also be employed postoperatively when a marginal or intralesional excision has occurred.

In recent years, a number of focused conventional radiotherapy strategies, including intensity-modulated radiotherapy (IMRT), and non-conventional particle therapies (carbon ion and proton therapy) have demonstrated promising results in the treatment of primary tumours particularly in unresectable regions (especially chordomas), but also in the management of local recurrence. As these modalities become more freely available, evidence for their efficacy and applications will no doubt expand.

Locally advanced disease

As recurrence and hence survival are significantly affected by attaining a clear margin at local control, *amputation* must be considered for tumours which are unresectable by any other means. Such tumours will often be extending out of the primary bone, violating a number of compartments, and will be encircling the neurovascular bundle. While it is, of course, possible to resect all of these, the reconstruction options or the oncological margins that can be achieved may mean that amputation is the preferred option both for oncological reasons and for better function. Amputation is required in approximately 15% of patients with primary bone tumours. The need for amputation will depend on the location and extent of the tumour, the capabilities of the operating team and the wishes of the patient.

For tumours arising around the ankle and hindfoot, amputation may be considered for local control above limb salvage as reconstruction options are limited and function is often excellent following below-knee amputation. For tumours around the knee, primary amputation may be considered if tumour not only involves the knee joint but also involves surrounding soft tissues and encircles the neurovascular bundle.

The surgical treatment of pelvic tumours presents a unique set of challenges, due to the proximity of neurovascular and visceral structures and the complex anatomy of the pelvis. Achieving surgical margins in pelvic resection is often difficult. With advances in imaging and non-surgical treatment modalities, the use of limb salvage as opposed to amputation has demonstrated reliable outcomes in the treatment of osseous pelvic sarcoma. Several reconstruction options have been proposed including iliofemoral or ischiofemoral arthrodesis, massive allografts, endoprosthetic reconstruction, and bone irradiation and reimplantation. However, for all these reconstruction options, significant rates of early and late complications have been reported. Early complications such as infection or instability may delay adjuvant chemotherapy with a subsequent impact on oncological outcomes. One of the simplest procedures is an internal hemipelvectomy without reconstruction, which can lead to a surprisingly good level of function. There is still a role for hindquarter amputation for extensive tumours of the pelvis. This operation can offer a reasonable chance of survival and although there is major functional loss, improvements in prosthetics increasingly allow patients to become mobile again with many returning to work.

For tumours of the upper limb, limb salvage should be undertaken when at all possible, as all amputations in the upper limb will result in significant loss of function. Fortunately, upper-limb tumours tend to present at an earlier stage than those in the pelvis and lower limb. However, survival after upper-limb amputation is often poor as it tends to be reserved for large or multifocal tumours, or for palliation. Microsurgical, allograft and prosthetic options have advanced significantly with amputation reserved for only unresectable tumours.

BENIGN LESIONS OF BONE

Osteochondroma

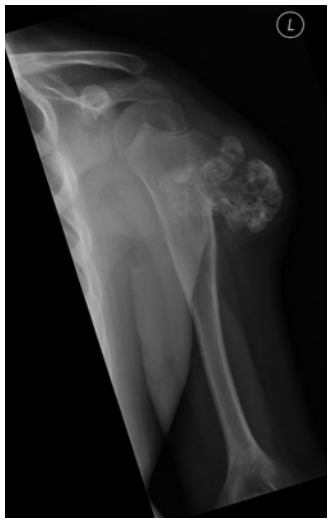
An osteochondroma is a benign, cartilaginous neoplasm derived from an aberrant subperiosteal nest of physeal cartilage which grows and matures according to normal enchondral ossification. Osteochondromas are very frequent and more common in males. They typically present in adolescent years, during the final growth spurt, and most commonly occur in long bones, particularly the femur and humerus.

They normally present as a painless mass though can cause symptoms secondary to formation of an overlying bursa due to friction, or to activity-related discomfort. Very occasionally, the lesion may cause neuropathic symptoms due to compression of a nearby nerve or may fracture producing sudden pain.

The lesion appears as a bony protuberance with well-defined limits, thin outer cortex and an inner cancellous structure. The pathognomonic feature is that the host bone flares from the cortex into the osteochondroma (Figure 9.11). Some are pedunculated with a cauliflower-like summit; others have a broad, sessile base. Pedunculated osteochondromas typically point away from the joint, towards the diaphysis. MRI demonstrates the classic cartilaginous cap which ranges from a few millimetres to a centimetre or more. The cap is typically thicker in children and diminishes with age.

Osteochondromas tend to grow with skeletal maturity and stop once growth stops. The risk of malignant transformation is rare, possibly 1%, but as the exact incidence of these lesions is not known, the exact malignant transformation rate can only be estimated. The risk of transformation is dependent on the size, with the cartilaginous cap being the source of neoplasia into chondrosarcoma. Malignant transformation is more common in the trunk, less common around the knee and exceptional in the extremities.

In asymptomatic lesions, treatment is not indicated. Excision may be warranted in large lesions where local pressure effects may occur or in adults where the risk of malignant transformation warrants removal.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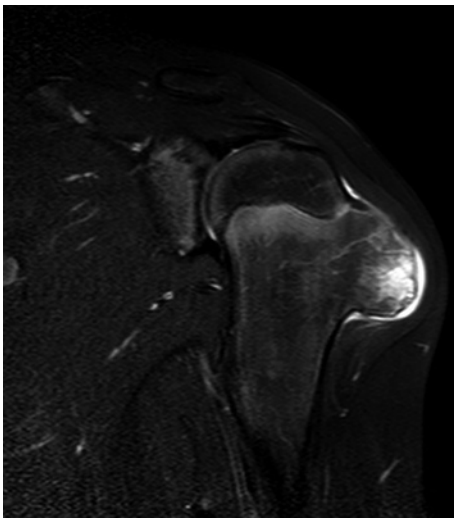
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Figure 9.11 Osteochondroma
A 14-year-old boy presented with a slowly enlarging mass over the left shoulder. Note the pedunculated, osseous lesion arising from the physis of the proximal humerus (a). MRI demonstrates the characteristic features of a thin cartilage cap overlying a disorganized cancellous outgrowth which merges with the metaphyseal bone of the proximal humerus (b). As the boy went into his final growth spurt, the mass increased significantly, causing pain. The lesion was removed without complication and never recurred.

Enchondroma

This is an intramedullary neoplasm made of well-differentiated hyaline cartilage. The exact incidence is unknown as the majority are asymptomatic and discovered incidentally. The commonest location is the tubular bones of the hand, followed by the femur and humerus.

Radiographically, the lesion is most commonly central with rounded, well-defined, lobulated edges and a thin rind of reactive sclerosis. It contains glandular, popcorn, ring-like opacities. They can reach considerable size but rarely exceed 6 cm. MRI demonstrates the black signal voids of internal calcification and isotope bone scan is hot in most lesions.

Histologically, the lesion contains lobules of cartilage with areas of calcification. The chondrocytes are usually sparse with small, round, dense nuclei. The real challenge is differentiating an enchondroma from a well-differentiated or grade 1 chondrosarcoma. Often, a grade 1 chondrosarcoma will demonstrate a higher cellularity, more plump nuclei and more than four or five double-nucleated cells per high-power field, though this is very subjective. Indeed, many of these features will be meaningless in the hand, or in a child. However, if there is evidence of chondroma permeating within the marrow spaces and haversian system, this is highly suggestive of chondrosarcoma.

The diagnosis of an enchondroma can usually be made with X-rays (Figure 9.12). Occasionally, biopsy will be required if the diagnosis is unclear. Treatment is usually not required although, occasionally in the hand, the lesion may be removed through curettage, particularly if there is pain or pathological fracture. Serial X-rays may be helpful if there is a suspicion of a grade 1 chondrosarcoma, as enchondromas in skeletal maturity do not grow.

Periosteal chondroma

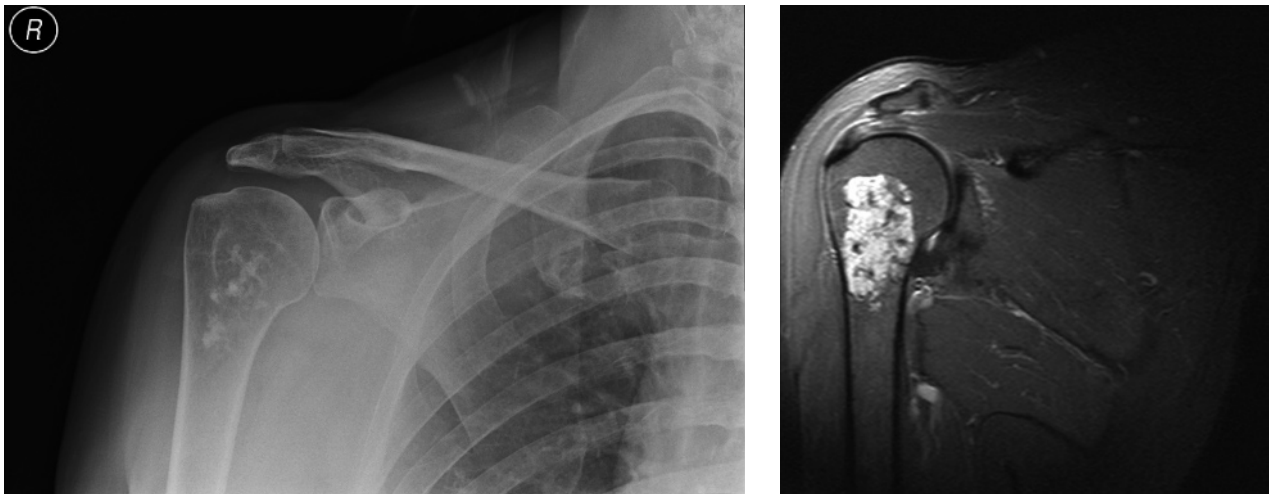
This is a rare, benign cartilage neoplasm occurring at the surface of the bone. It is most frequently seen in children or young adults and prefers the metaphysis of long bones, particularly the humerus. It most commonly presents with pain and occasionally a palpable lump.

Radiographs demonstrate a superficial erosion of the bone cortex with occasional scalloping (Figure 9.13). When large, the lesion demonstrates a popcorn matrix with areas of calcification. Histologically, it resembles an enchondroma. The majority are effectively treated by curettage; though large lesions may require more aggressive resection.

Multiple chondromas and associated conditions

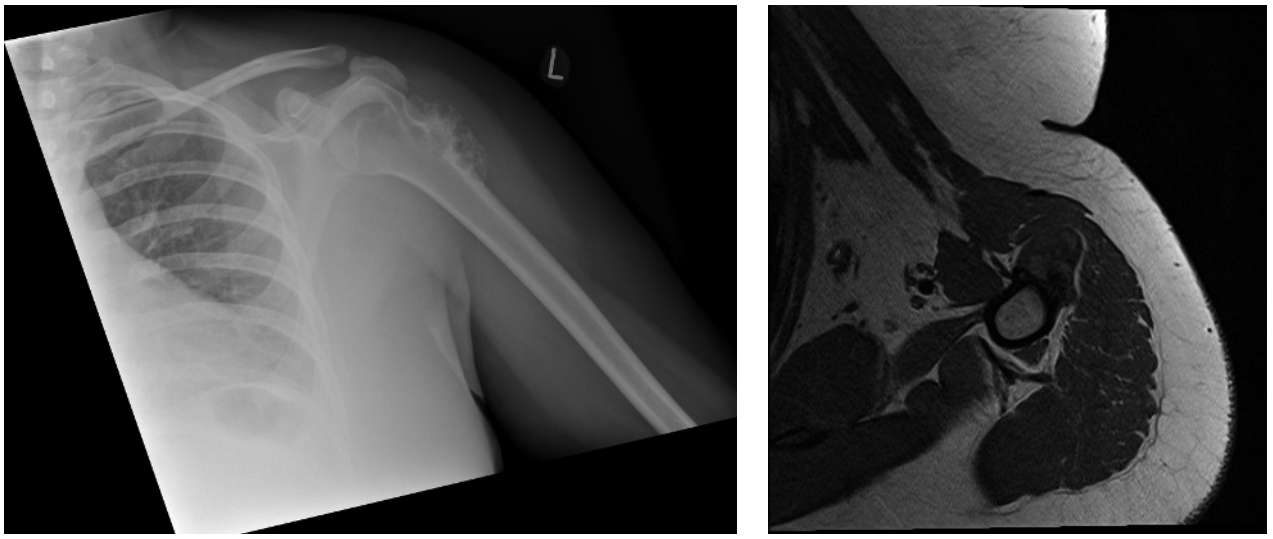
Multiple chondromas are infrequent. In *Ollier's disease*, multiple chondromas may be found within the hand of one limb, or have a much wider, hemisomic distribution, or affect the entire body with a hemisomic prevalence. The disease is non-hereditary and sporadic. It most commonly affects the tubular bones of the hand or foot. Chondromas normally present as bony swellings in childhood which may cause deformities and limb length discrepancy due to epiphyseal fusion anomalies.

In *Maffucci syndrome*, multiple chondromas are associated with multiple cutaneous or deep haemangiomas. On X-ray, the chondromas can be very large with consequent expansion of the bone, thinning of the cortex or, indeed, no cortex at all (Figure 9.14). In Maffucci syndrome, the presence of haemangioma may be seen on imaging by phleboliths. Histologically, the chondromas appear more cellular than solitary chondromas, with more proliferative potential.



(a) (b)

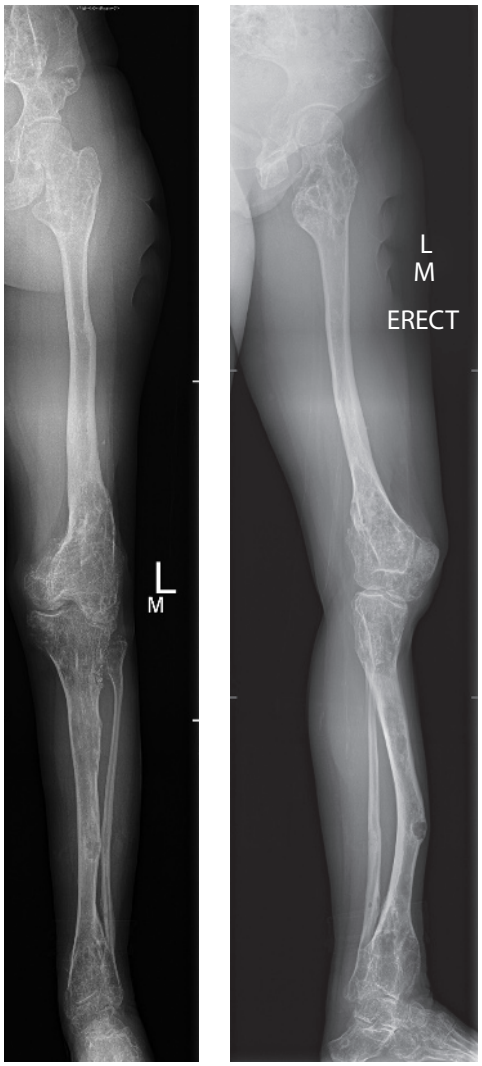
Figure 9.12 Enchondroma X-rays taken of a 65-year-old woman who presented with right shoulder pain demonstrate a 4 cm intraosseous chondroid lesion within the right proximal humerus (a). (b) Subsequent MRI demonstrates typical features of an enchondroma with a mixed cartilaginous lesion with areas of calcification but no evidence of endosteal scalloping or permeation. The shoulder pain was attributed to a full-thickness rotator cuff tear.



(a) (b)

Figure 9.13 Periosteal chondroma In the case of a 22-year-old woman with a painless, incidental mass arising from the left proximal humerus, X-rays demonstrate a pedunculated mass similar to an osteochondroma (a). (b) MRI shows the lesion to be arising from the surface of the bone rather than merging with the underlying metaphysis.

Transformation to a secondary sarcoma is seen in both these conditions. In Ollier's disease, this may occur in 20–30% of patients; in Maffucci syndrome, this is much more common and is likely to be greater than 50%. Malignant transformation may be heralded by an increase in size of a lesion or the development of symptoms. Histologically, there will the appearance of a permeative growth pattern, whilst radiologically, serial X-rays will demonstrate a change in size with a new soft-tissue mass seen on MRI. Both conditions are associated with an increased risk of extra skeletal malignancies such as breast, liver, ovarian and central nervous system tumours.



(a) (b)

Figure 9.14 Maffucci syndrome A young woman, now in her 30s, with Maffucci syndrome with hemimelic involvement affecting virtually all the bones of her left side (a,b). Over the years she has had a number of low-grade malignancies removed with upper and lower limb ray amputations. She now presents with terrible pain from her knees and ankles due to the significant deformity resulting from her multiple chondromas.

Chondromyxoid fibroma

This is a benign cartilaginous tumour comprising lobules of fibromyxoid and chondroid tissue. It is rare, accounting for only 0.5% of all tumours of bone and is slightly more common in males. Common sites of presentation are the long bone metaphyses, particularly the proximal tibia, and the pelvis. It most frequently presents in teenage and young adult years. As these lesions tend to be slow-growing, they present with insidious, mild to moderate pain. Occasionally, they are discovered incidentally.

On X-ray, they appear as small, metaphyseal, eccentric lytic lesions. They lie parallel to the long axis of the bone and, in small bones, can cause fusiform swelling. The tumour often erupts out through the cortex but causes little or no periosteal or soft-tissue reaction. They may be hot on isotope bone scanning and homogeneous on MRI. Histologically, the tumour appears as stellate cells on a myxoid background. Mitotic figures are not common though cellular atypia may occur. Treatment is principally in the form of intralesional curettage with a low risk of recurrence.

Chondroblastoma

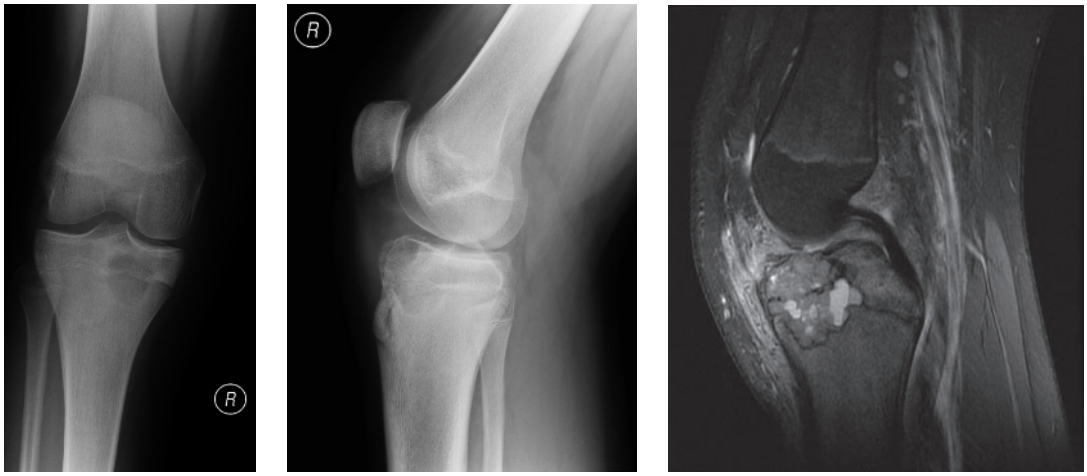
These are benign tumours of childhood. They occur most commonly at the epiphyses, usually at the ends of long bones. They account for less than 1% of all bone neoplasms and are more common in males. The peak age of incidence is in teenage years, rare after the age of 35 and exceptional before the age of 10. They typically present with pain and can occasionally cause a joint effusion or stiffness. They appear as a round or oval lytic lesion ranging in size from 1 to 7 cm on X-ray. They appear within the epiphysis or apophysis and can cross the physis. The cortex may be expanded but often is not breached (Figure 9.15). MRI demonstrates a homogeneous high signal lesion.

Histologically, they appear as 'wet-sawdust' with areas of chondroid matrix, calcification and haemorrhage. The presence of 'chicken-wire' calcification is pathognomonic. Areas resembling an ABC may be seen in 35% of cases. The majority of cases can be treated with simple curettage with or without bone grafting to the defect to support the subchondral plate. Smaller lesions may be amenable to radiofrequency ablation under CT guidance. Recurrence is not infrequent and can be seen in about 10% of cases. Violation of the joint should be avoided to prevent contamination. More aggressive, en-bloc resection may be required for recurrent disease.

Osteoid osteoma

This is a small, benign tumour formed of osteoid and woven bone surrounded by a halo of reactive bone. These lesions are most common in young patients but are rare below 5 years of age and equally rare over 30. They are more common in men than women. It is most commonly seen in the long bones, particularly the proximal femur. It is rare in the trunk with the exception of the spine where it is most often seen in the posterior arches. It is more often diaphyseal than metaphyseal.

Osteoid osteomas most often present with pain, which classically is worse at night and relieved by



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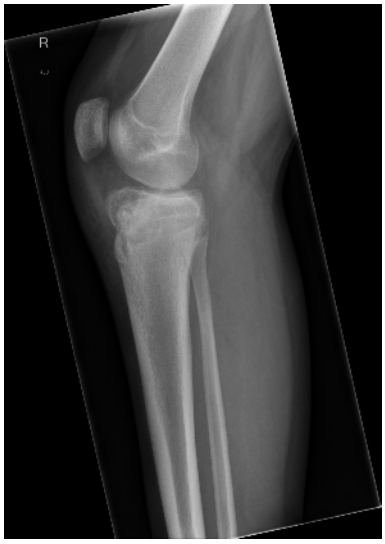


(d)

Figure 9.15 Chondroblastoma Radiographs from an 18-year-old male with a short history of pain in the knee demonstrate a lytic lesion in the proximal tibial physis crossing the physeal scar and abutting the subchondral plate (a,b). MRI demonstrates a typical chondroblastoma with secondary ABC transformation (note fluid-fluid levels) (c). This was treated by extended intralesional curettage, packing the defect with bone graft (d), which resulted in a dramatic reduction in pain and a quick restoration of function.

non-steroidal anti-inflammatories. When in close proximity to a joint, they can result in stiffness and an effusion. In the spine, they can cause muscle spasm and scoliosis.

Plain X-ray demonstrates an area of dense sclerosis with a small, rounded area of osteolysis which is often obscured by the surrounding sclerosis. Isotope bone scan is positive and the central nidus of the lesion is best seen on CT scan (Figure 9.16).

Histology demonstrates a packed mesh of thin, woven bone with osteoblastic rimming, osteoclasts and dilated capillaries. The mature osteoid corresponds to the nidus, which is surrounded by areas of dense, reactive bone with surrounding soft tissue also showing features of inflammation.

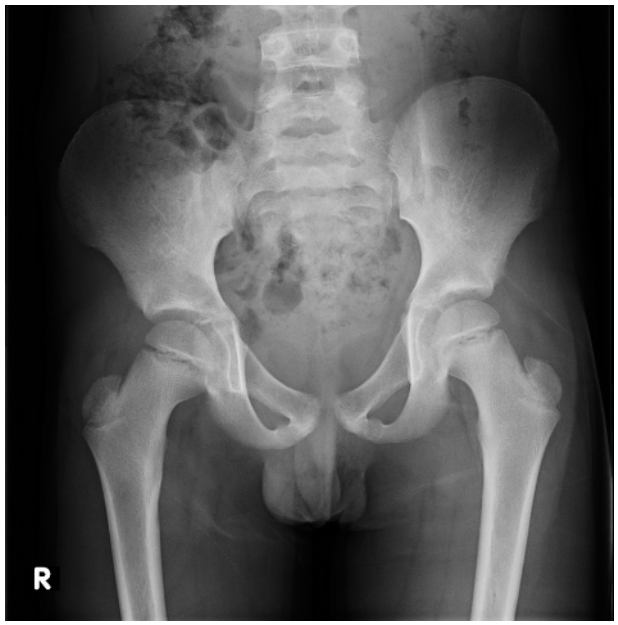
Without treatment, the lesion will slowly increase but over time will regress and usually burns out over a variable period of a number of years. The preferred method of treatment is by CT-guided radiofrequency ablation, which has proven to be highly successful in eradicating the lesion with a dramatic resolution of symptoms.

Osteoblastoma

This is again a benign tumour of osteoblasts producing osteoid and woven bone. Similar in appearance to an osteoid osteoma, osteoblastomas are characteristically larger than osteoid osteomas. These are rare lesions most commonly seen in children and teenagers, rarely seen below the age of 8 or older than 40. Osteoblastoma have a predilection for the spine, again in the posterior arch, but can occur anywhere in the skeleton.

As with osteoid osteoma, osteoblastomas usually present with pain at the site of the tumour. In the spine, they can present with nerve root compression.

Plain X-ray demonstrates a lytic lesion, typically 2–5 cm in size, although they can be much bigger. The lesion is usually central but can be eccentric or



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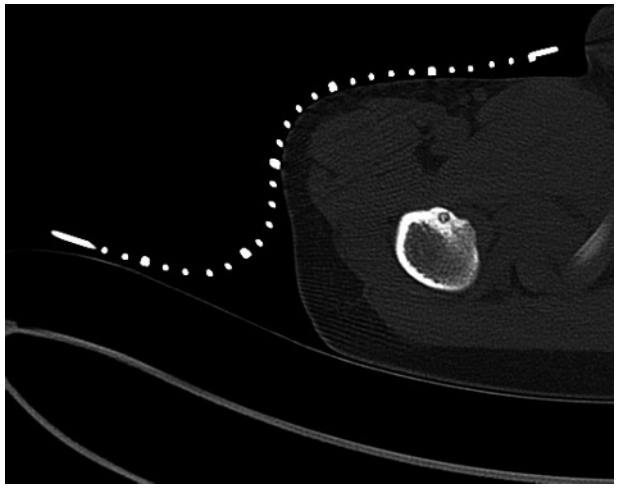

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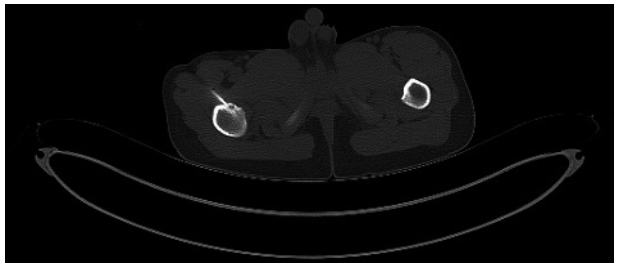


(c)

Figure 9.16 Osteoid osteoma A 9-year-old boy with a short history of worsening pain in the right groin which progressed to night pain. The pain was very responsive to non-steroidal analgesia. (a) X-rays demonstrate coarse trabecular thickening of the medial proximal femoral metaphysis. (b) CT scan demonstrates the characteristic nidus of an osteoid osteoma within the area of trabeculation. This was very effectively treated by CT-guided radiofrequency ablation (c). The pain completely resolved within a matter of days.

periosteal. Often there is a rind of sclerosis with an area of periosteal reaction if there has been breach of the cortex. Occasionally, the lesion may have features of ABC due to rapid expansion. The lesion is hot on bone scan and MRI often shows intense peritumoural oedema.

Histologically, the lesion consists of large osteoblasts producing osteoid and woven bone. Cytological activity may also be present. Of note, however, is the interface between the tumour and the surrounding normal bone which is sharp without evidence of permeation.

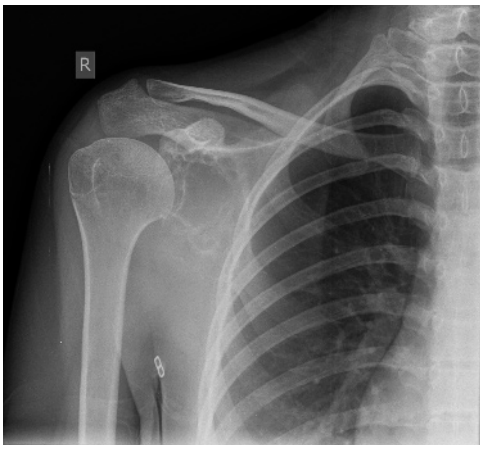
The majority of osteoblastomas are intraosseous at presentation although, in advanced disease, soft tissue extension is not uncommon. Reports of metastatic spread of osteoblastomas are most likely undiagnosed osteoblast-like osteosarcomas. These can be differentiated by the finding of osteoid permeating the marrow spaces and trapping the host lamellar bone.

In the majority of cases, treatment comprises extended intralesional curettage with radiofrequency ablation for smaller volume lesions. Rarely, en-bloc resection may be required. Consideration may be given to preoperative embolization in the case of larger lesions to reduce intraoperative haemorrhage.

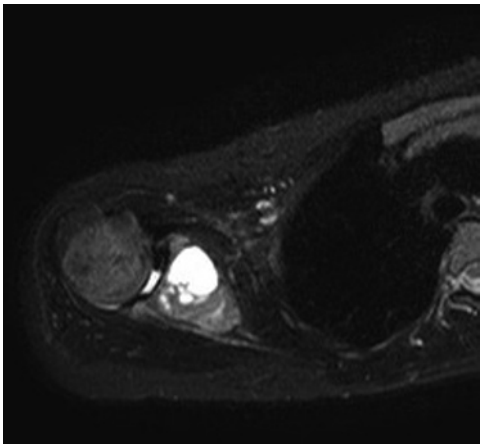
Desmoplastic fibroma of bone

This is a benign neoplasm composed of spindle cells and collagen which resembles a desmoid tumour of soft tissues. They are slow-growing lesions with a high rate of recurrence following excision. These are rare tumours which are more common in men and rarely occur after the age of 30. They most frequently occur in the mandible and pelvis as well as the long bones. When in the long bones, they most frequently affect the metadiaphysis though can extend to the epiphysis. As these lesions tend to grow slowly, symptoms are often vague. Most frequently, they present with pain and can be a cause of pathological fracture.

Plain X-rays most often demonstrate a lytic, expansile lesion with the cortex replaced by a thin shell of new bone (Figure 9.17). The lytic portion classically has a finely reticulated or bubbly appearance. Bone scan is often cold and MRI shows a low signal lesion on both T1 and T2, resembling cartilage. Histology classically shows a hypocellular spindle cell lesion with large amounts of collagen and bland nuclei without features of atypia. The lesion tends to grow very slowly and has often been present for many years prior to presentation. Traditionally, if symptomatic, these lesions were treated by en-bloc resection although, more recently, thorough curettage has been advocated as yielding similar rates of recurrence with improved functional outcomes.



(a)



(b)

Figure 9.17 Desmoplastic fibroma of bone Plain radiographs (a) and MRI (b) of an 18-year-old girl with a short history of pain in the right shoulder with progressive weakness of arm elevation demonstrate an expansile, lytic mixed cystic lesion within the scapula. Biopsy confirmed features consistent with a desmoplastic fibroma of bone.

Non-ossifying fibroma

Non-ossifying fibroma (NOF), the commonest benign lesion of bone, is a developmental defect in which a nest of fibrous tissue appears within the bone and persists for some years before ossifying. It is asymptomatic and is almost always encountered in children as an incidental X-ray finding. The commonest sites are the metaphyses of long bones; occasionally there are multiple lesions. There is a more-or-less oval radiolucent area surrounded by a thin margin of dense bone (Figure 9.18). Views in different planes may show that a lesion that appears to be 'central' is actually adjacent to or within the cortex, hence the alternative name 'fibrous cortical defect'.

Although the lesion looks cystic on X-rays, it is a solid lesion consisting of unremarkable fibrous tissue with a few scattered giant cells. As the bone grows, the defect becomes less obvious and it eventually heals spontaneously. However, it sometimes enlarges to several centimetres in diameter and there may be a pathological fracture. There is no risk of malignant change.

Treatment is usually unnecessary. If the defect is very large or has led to repeated fractures, it can be treated by curettage and bone grafting; recurrences are rare.

Giant-cell tumour of bone

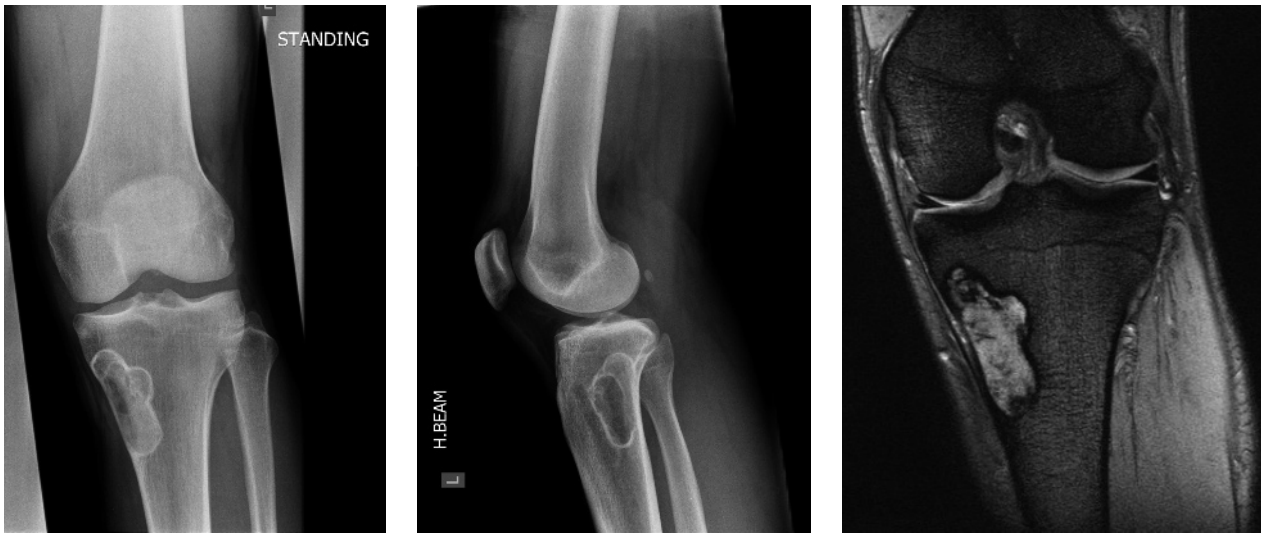
This is a benign but locally aggressive tumour of bone composed of a proliferation of mononuclear cells with scattered macrophages. Giant-cell tumours (GCTs) account for approximately 5% of all primary bone lesions and are most common between 20 and 45 years of age. Though they can appear in teenage years, they are rare in the immature skeleton. GCTs can occasionally be seen in conjunction with Paget's disease of bone and can arise in association with focal dermal hypoplasia (Goltz syndrome). Malignant transformation can occur in GCTs though this is rare (<1%) and is marginally more common in females.

GCTs typically affect the metaphyses of long bones with preponderance for the distal femur, proximal tibia, distal radius and proximal humerus. When affecting the spine, they most commonly arise in the vertebral bodies of the sacrum with reduction in frequency as the spine is ascended. While flat bones are not commonly affected, GCTs affecting the pelvis are most commonly seen in the ilium. GCTs are rarely multicentric and rarely affect the tubular bones of the hands. In such cases, hyperparathyroidism (brown tumour) must be excluded.

Patients typically present with pain and, less frequently, an increasing mass, particularly around the knee. In a small number of cases (5–10%), pathological fracture is the presenting feature.

Plain X-ray classically demonstrates an eccentric, expansile, lobulated lytic lesion with a narrow zone of transition. Tumours have extended into the soft tissues at presentation and a soft-tissue mass, sometimes covered by a thin layer of sclerosis, can be seen on X-ray (Figure 9.19). GCTs usually have little or no discernible matrix calcification and little new bone formation or periosteal reaction. They are typically located within the metaphysis and are one of the few lesions to involve the physis, abutting the subchondral plate. CT gives an accurate estimation of cortical bone involvement, and MRI demonstrates low signal on T1 and intermediate to high signal on T2 with areas of heterogeneity.

Although it is rare, malignant transformation in GCT can occur. Typically, this presents with pain



(a) (b) (c)

Figure 9.18 Non-ossifying fibroma Plain radiographs (a,b) demonstrate a well-circumscribed lesion of the proximal tibia of a 29-year-old man referred with knee pain. MRI (c) confirms a partially consolidated benign lesion of the proximal tibia, which, given the patient's age, is entirely in keeping with a non-ossifying fibroma.

many years after initial treatment. Radiologically, this appears as a lytic destructive lesion with a sclerosing destructive tumour adjacent to the GCT. Histology is diagnostic. Secondary malignant GCTs can occur many years after radiotherapy for treatment of a non-malignant GCT.

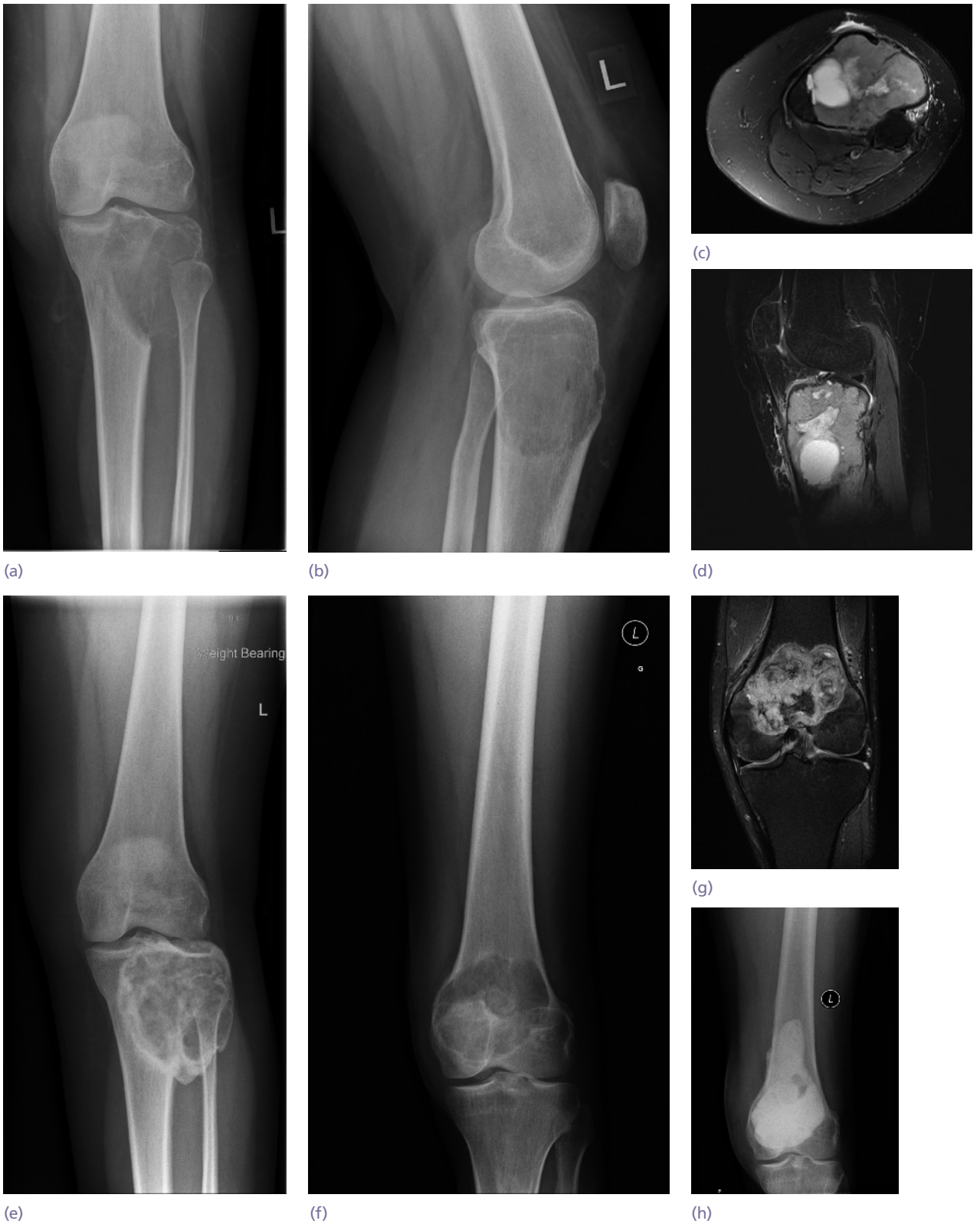
Histologically, GCTs appear as meaty, reddish-purple tissue merged with soft yellow areas. The lesion is characterized by the presence of a large number of osteoclast-like giant cells among scattered round or spindle-shaped mononuclear cells. The tumour stroma is often well vascularized and may contain bands of cellular or collagenous fibrous tissue. Interposed are areas of haemorrhage, haemosiderin and collections of foamy macrophages. Secondary ABC-like changes are seen in approximately 10% of cases. Soft-tissue extensions, and metastatic lesions within the lung, resemble the primary lesion.

The intercellular signalling of giant cells in GCTs has been the area of much research in recent years. The giant cells in GCTs are osteoclast-like cells originating from haematopoietic stem cells. These cells are stimulated by, among other signalling pathways, the soluble receptor activator for nuclear factor κB (RANKL) which interacts with RANKL expressed on monocyte lineages and osteoclast precursors which induce osteoclast differentiation resulting in osteolysis. This understanding of the pathways of osteoclast activation in GCT has resulted in the application of novel pharmacological techniques to reduce the activity of GCTs and shift the balance of bone remodelling towards osteoblast function.

GCTs are staged according to the classification system described by Campanacci:

Stage 1 Completely intraosseous
Stage 2 Demonstrates cortical erosion without destruction
Stage 3 Characterized by cortical destruction with a soft-tissue component

The majority of GCTs are stage 2 or 3 at presentation. In the majority of stage 1 and 2 lesions, extended intralesional curettage with a detailed debridement of the lesional wall will be effective. Recurrence rates vary depending on the use of adjuvant treatments at the time of curettage (including phenol, bone cement, liquid nitrogen) but a commonly accepted rate of local recurrence is in the order of 15%. Stage 3 disease can often be treated by thorough curettage though in some cases en-bloc resection is required. Historically, radiotherapy was used following curettage and still has a role in difficult locations such as the spine. In recent years, the use of pharmacological manipulation has gained favour. As the lesion is driven by osteoclasts responding to RANKL, anti-RANKL antibodies such as denosumab can be used to stop the osteolytic process and switch the balance towards bone formation. The indications for denosumab remain to be defined but it has a role in advanced stage 3 lesions or lesions in inoperable locations. The response of the tumour can be dramatic but the side-effect profile can result in significant morbidity, including hypocalcaemia, osteonecrosis of the jaw and atypical fracture patterns.



(a) (b) (c) (d) (e) (f) (g) (h)

Figure 9.19 Giant-cell tumour of bone (GCT) (a–e) A 29-year-old female presented with left-sided knee pain and a mass. Radiographs (a,b) demonstrate a geocentric lesion in the proximal tibia with marked destruction of the lateral proximal tibial metaphysis. MRI confirms a heterogeneous lesion in the proximal tibia which abuts the subchondral plate (c,d), with a large extraosseous, soft-tissue mass (Campanacci stage 3) and focal areas of cystic/haemorrhagic change. Biopsy confirms a giant cell-rich lesion in keeping with a giant-cell tumour of bone. The same patient after 4 months' treatment with the RANKL antagonist Denosumab (e). (f–h) A similar case in the distal femur as demonstrated by X-ray (f) and MRI (g). In this case, given the lack of an extraosseous component, the patient was treated by extended intralesional curettage and cementation (h).

Haemangioma of bone

Haemangioma are benign lesions of bone composed of capillary blood vessels of small or large calibre. These are common lesions, often asymptomatic and often identified incidentally. Post-mortem studies have identified them to be present in the vertebrae of 10% of the adult population. Haemangiomas can be identified at any age but most commonly present in the fifth decade. They are slightly more common in females than males. They are most commonly seen in the vertebral bodies, followed by the craniofacial bones. When seen in the long bones, they most commonly affect the metaphysis. Polyostotic disease is not uncommon.

The majority of haemangiomas are identified incidentally. If large, they can cause symptoms particularly in the spine, where they may present with cord compression, pain and neurological symptoms.

Radiologically, they appear as a radiolucent lesion often with coarse trabeculae. On MRI, they appear to contain fat, and trabeculae are evident. Histology varies but typically these lesions demonstrate thin-walled blood-filled vessels lined by a thin layer of fat. Treatment is often not required but, when indicated, curettage and stabilization are often effective with a low incidence of recurrence.

Epithelioid haemangioma

Compared to haemangioma, epithelioid haemangioma are considerably more locally aggressive. Patients are typically in the fourth decade, though these lesions can present at any age. They are more frequently seen in women. In comparison to haemangioma, these lesions are most commonly seen in the long bones, affecting flat bones, vertebrae and tubular bones of the extremities to a lesser extent. In up to 25% of cases, they may be multifocal and regional.

Patients most frequently present with pain. X-ray demonstrates a well-defined, lytic, sometimes expansile, septated lesion which often erodes the cortex and results in a soft-tissue mass. MRI demonstrates a hypointense lesion on T1 and hyperintense lesion on T2.

Histologically, they have a lobulated architecture which replaces marrow and infiltrates the pre-existing bony trabeculae. The centre of the lesion often contains epithelioid cells formed into solid sheets while the periphery often contains small arteriolar-like vessels lined by epithelioid cells.

The majority of lesions require treatment, most often with curettage although occasionally en-bloc resection is required. The prognosis is usually good with a local recurrence rate of 9%. Occasionally, radiotherapy may be required, particularly for inaccessible locations.

Simple bone cyst

This is a solitary, usually unilocular cystic bone cavity lined by a fibrous membrane and filled with serous or serosanguinous fluid. Males are more frequently affected and the majority occur within the first two decades of life. Although they can arise at any location, the vast majority of simple bone cysts (SBCs) occur in the proximal humerus, the proximal femur or the proximal tibia, most commonly affecting the metaphyseal areas close to the physis.

In the majority of cases the lesion is asymptomatic, being discovered incidentally. However, fracture through the lesion is not uncommon and often this is the presenting feature. Occasionally, mild pain or swelling may be present. X-rays demonstrate a well-outlined, lytic centrally placed, metadiaphyseal lesion expanding and thinning the cortices (Figure 9.20). It often abuts but does not cross the physis. Bone septa are often present which give the impression of a multiloculated cyst. When fracture occurs, a small fragment may be seen within the cavity, the classic 'fallen leaf' sign. MRI will demonstrate the homogeneous fluid-filled cavity.

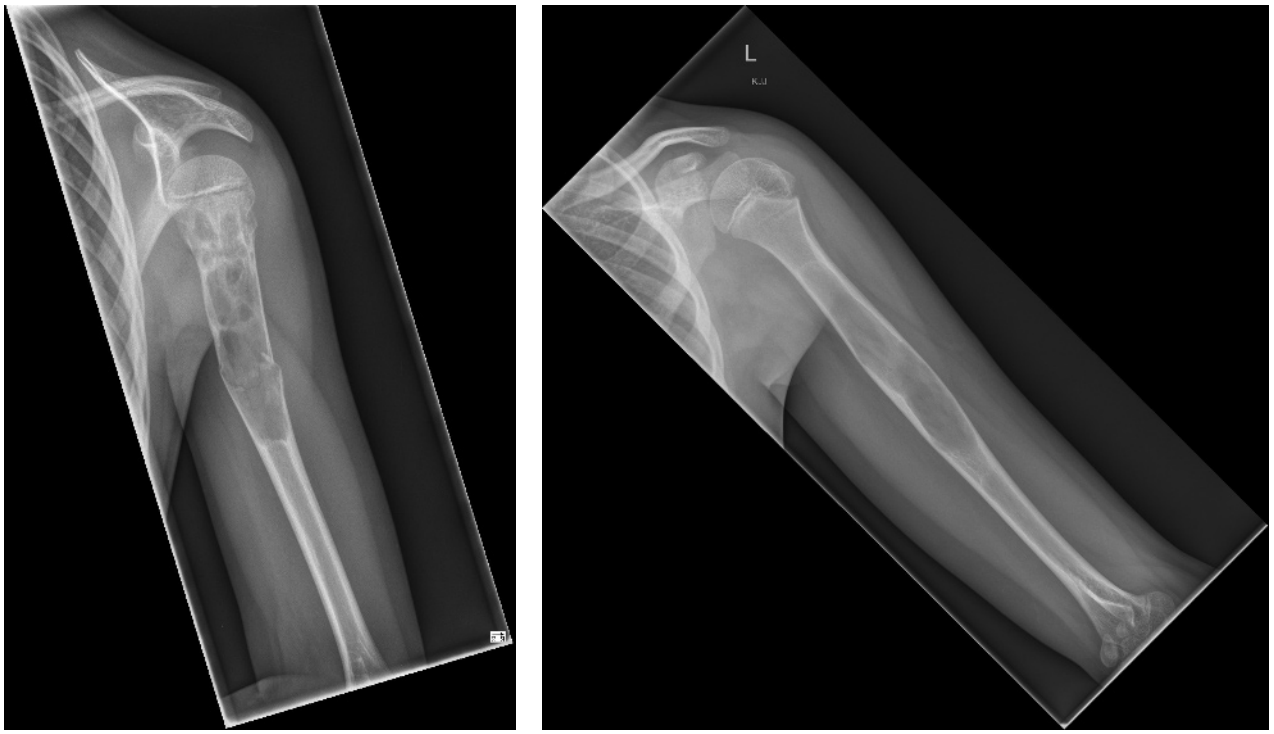
Histologically, the cyst lining demonstrates connective tissue with foci of reactive bone. Following fracture, there will be features of fracture callus and new bone formation.

Treatment is often supportive as lesions will regress following skeletal maturity. Percutaneous injection of steroid has been reported with varying success but is best reserved for 'active' lesions, i.e. those abutting the physis in young children. Curettage and bone grafting may be required in areas at risk of fracture, and pathological fractures of the proximal femur in particular will often require fixation and stabilization. Recurrence is reported in 10–20% of cases. Large cysts may result in limb shortening, and avascular necrosis of the femoral head can occur following fracture through proximal femoral lesions. Spontaneous resolution following fracture has been reported.

Aneurysmal bone cyst

Aneurysmal bone cysts (ABCs) are benign, expansile lesions of bone composed of blood-filled cystic spaces. They are destructive lesions and, while they are histologically benign, they can result in significant disability. They predominantly affect children and teenagers with an equal sex distribution. ABCs can affect any bone but are most commonly seen in the metaphyses of long bones, particularly the femur, tibia and humerus. They may occur in the spine where they typically affect the posterior elements.

Patients typically present with pain and swelling. When affecting the spine, presentation can be with nerve root impingement and neurological impairment.



(a) (b)

Figure 9.20 Simple bone cyst (SBC) A 10-year-old boy with a spontaneous fracture through the left humerus. Plain radiographs (a) demonstrate an expansile, lytic lesion in the humeral metadiaphysis with a pathological fracture through the base of the lesion. The imaging features are in keeping with a simple bone cyst. The fracture was managed conservatively with a humeral brace and careful observation. Over time, the lesion consolidated and united (b).

ABCs appear as a subperiosteal, poorly defined osteolytic lesion, elevating and progressively eroding the cortex. MRI demonstrates the typical cystic features with multiple intralesional septations and fluid levels (Figure 9.21). Angiography demonstrates persistence of contrast and a blush of flow within the lesion.

Histologically, ABCs are composed of blood-filled, cystic spaces separated by fibrous septae which may comprise woven bone in chronic lesions. Necrosis is rare unless there has been a pathological fracture. An ABC can appear as a solid lesion, which resembles a giant-cell lesion. ABC-like areas can be seen in other benign tumours and occasionally malignant lesions where there has been haemorrhage within the tumour (secondary ABC). Secondary ABCs are most commonly seen in association with GCTs, chondroblastoma, osteoblastoma and fibrous dysplasia, though can also be seen with osteosarcomas.

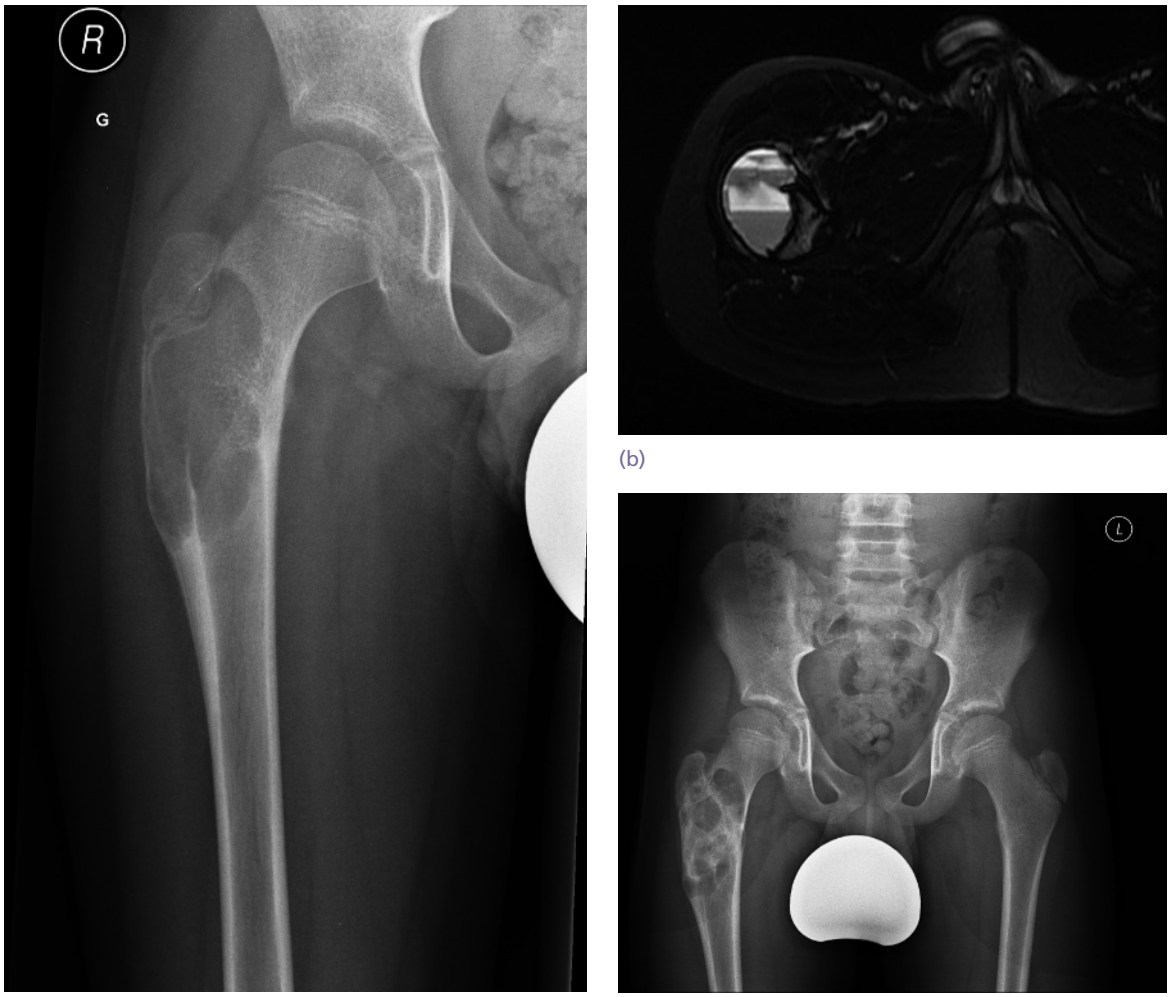
The lesion may demonstrate rapid progression, but equally it may resolve spontaneously following trauma, either fracture or biopsy. Curettage of the lesion at the time of biopsy ('curopsy'), debriding the cystic cavity wall, is often effective though recurrence can occur in up to 20% of cases. Radiation is effective at stimulating cyst calcification but must be offset by the risk of secondary sarcoma or of growth arrest due to damage to the nearby physis.

Fibrous dysplasia

This is a benign, medullary fibro-osseous lesion which may affect one bone (monostotic) or a number of bones (polyostotic). Fibrous dysplasia can affect children and adults with equal sex and race distribution. The monostotic form is considerably more frequent than the polyostotic. The craniofacial bones and the femur are the most frequently affected bones, but any bone can be affected by the monostotic form. In the polyostotic form, the pelvis, femur and tibia are commonly involved. Multiple sites of fibrous dysplasia may be seen in the same bone, particularly in the monostotic form.

The monostotic form is often discovered incidentally and is largely asymptomatic. Not infrequently, pain and fracture are the presenting features. Bony expansion in superficial bones, deformity and growth disturbance with deformity and lower-limb length discrepancy can also occur.

Fibrous dysplasia can be associated with endocrinopathies and café-au-lait spots (coast of Maine) in



(a) (b) (c)

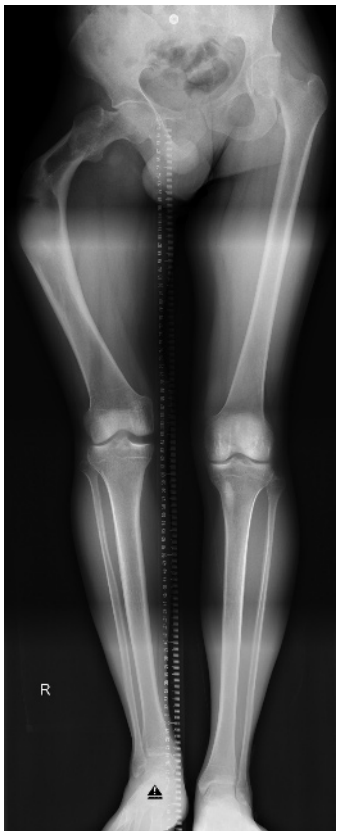
Figure 9.21 Aneurysmal bone cyst (ABC) A 9-year-old boy presented with progressive right-sided hip and thigh pain on exertion. (a) Radiographs demonstrated an expansile, lytic, cystic lesion of the right proximal femur. (b) MRI demonstrated a characteristic expansile, loculated lesion of the proximal femur with thinning of the cortices but no soft-tissue component. Fluid-fluid levels are seen within the lesion which is highly suggestive of an aneurysmal cyst. This was treated by curopsy (detailed curettage of the inner membranous lining of the cyst). Over a year, the lesion progressively corticated (c) and eventually was replaced with normal bone.

McCune–Albright syndrome, and with intramuscular myxomas in Mazabraud syndrome.

X-rays often demonstrate a non-aggressive, well-circumscribed lesion with a characteristic ground-glass matrix (Figure 9.22). More mature lesions may show cyst formation and secondary changes. The characteristic deformity resulting from proximal femoral fibrous dysplasia is the shepherd's crook deformity. There is seldom a soft-tissue component or periosteal reaction, except in the case of fracture. MRI reveals a homogeneous low signal lesion on T1. There may be associated cystic degeneration within the lesion.

Histology demonstrates a characteristic mixture of benign proliferating fibroblastic cells within islands of woven bone which characteristically appear in a 'Chinese letter' formation. Benign giant cells are not infrequently seen and islands of cartilage may dominate the appearance. Secondary ABC features are often seen.

Treatment is often not needed. Correction of deformities is sometimes required but should be undertaken with caution. Occasionally, in the case of fracture or impending fracture, intralesional curettage and bone grafting may be required and, for very large lesions, augmentation with internal fixation may be required. However, recurrence following treatment is not uncommon. Malignant transformation, to fibrosarcoma of bone, is rare but more frequently seen in McCune–Albright syndrome.



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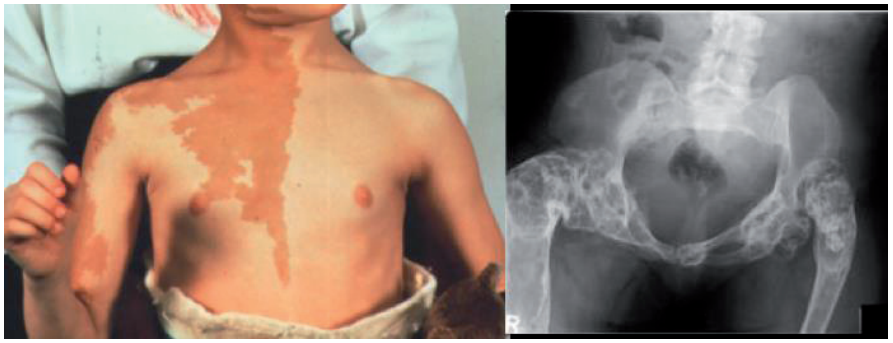


(b)

Figure 9.22 Fibrous dysplasia Plain radiographs of a 73-year-old male from India who presented with a long history of progressive bowing of the right thigh (a). He eventually sought medical advice when the pain suddenly became severe. Radiographs show a varus deformity of the proximal femur with a stress fracture on the tension side. The lesion demonstrates the typical features of mixed lytic and blastic appearances with thinning of the cortices and progressive deformity. When associated with café au lait spots and polyostotic fibrous dysplasia, as well as hyperfunctional endocrine disease (b), this is known as McCune–Albright syndrome.

Osteofibrous dysplasia

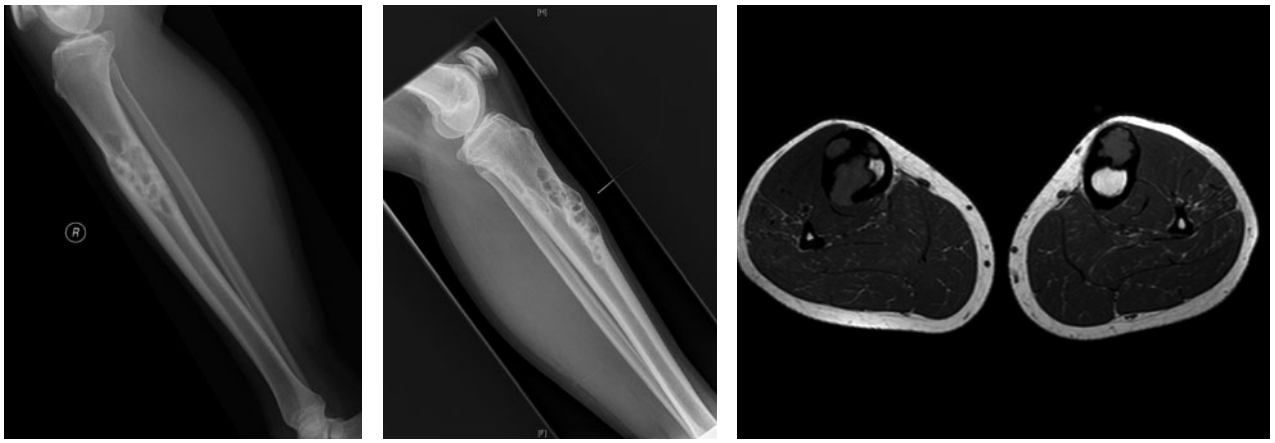
Osteofibrous dysplasia (OFD) is a benign, non-osseous lesion of bone typically seen in the anterior cortex of the tibia, particularly in children or teenagers. It is commonly seen in children in the first decade and can on rare occasions be polyostotic. It is almost exclusively seen in the tibia but can on rare occasion affect the ipsilateral fibula or be bilateral at presentation.

It presents with moderate expansion of bone which is manifest as a palpable lump or progressive bowing of the tibia. It is usually painless but stress fractures and pathological fractures can occur.

The lesion classically presents radiographically as intracortical osteolysis, most commonly affecting the anterior cortex of the tibia (Figure 9.23). There is expansion of the cortex to accommodate the lesion which typically has a soap bubble appearance. There is a narrow zone of transition between the lesion and normal bone with a surrounding rim of sclerotic bone on the medullary side, which may obscure the medullary canal. MRI demonstrates a cortical lesion with a surrounding area of sclerosis which demarcate the lesion from its comparable malignant variant, adamantinoma, which typically demonstrates a 'moth-eaten' margin with medullary involvement.

Histologically, osteofibrous dysplasia appears as irregular fragments of woven bone rimmed by layers of lamellar bone laid down by osteoblasts. The fibrous component consists of bland spindle cells with collagen production and a matrix that may be myxoid or fibrous. The histological appearance can be differentiated from fibrous dysplasia by the absence of keratin-positive epithelial cells in OFD, and from adamantinoma by the absence of epithelial cell clusters. An intermediate lesion, OFD-like adamantinoma, is described when there are abundant keratin-positive cells but in a smaller number and less easily identifiable than seen in classic adamantinoma. Thus, OFD, OFD-like adamantinoma and adamantinoma can be differentiated histologically.

These three comparable lesions behave very differently in their natural history, however: OFD tends to progress through the first decade of life, stabilizing during the second and often healing or resolving spontaneously thereafter. Treatment, therefore, is rarely indicated except in exceptional circumstances of severe progressive deformity. The progression of OFD-like adamantinoma (OFD with small clusters of keratin-positive cells) to adamantinoma has been reported but OFD-like adamantinoma seldom, if ever, results in metastases, which differentiates it from adamantinoma.



(a) (b) (c)

Figure 9.23 Osteofibrous dysplasia (OFD) A palpable mass over the anterior right tibia (a) of a 15-year-old boy who also complained of pain and a mass in the contralateral left tibia (b). MRI confirms bilateral osteofibrous dysplasia (c). Note the characteristic soap bubble ossification seen on plain X-ray and the narrow demarcation between the dysplastic regions and the native bone, without medullary invasion.

Langerhans cell histiocytosis

This is a clonal proliferation of pathological Langerhans cells. It may affect just the skeleton where it may be solitary or polyostotic, and may manifest as a multi-system disease. Histiocytosis refers to an abnormal proliferation of cells of the reticulo-endothelial system. The disease has previously gone by several names, including eosinophilic granuloma (commonest form, results in osseous punched-out lesions), Hand–Schüller–Christian disease (chronic disseminated form), Letterer–Siwe disease (rare, severe, progressive form seen in young children, often fatal), and histiocytosis X (generalized disorder of the reticulo-endothelial system with multiple osseous granulomas). Langerhans cell histiocytosis (LCH) accounts for <1% of all osseous lesions. It has a wide age distribution but the majority of cases are diagnosed in those under 30. It is more frequently seen in men than women. LCH can affect any bone but is most commonly seen in the skull, the pelvis and femur, and the mandible.

LCH most commonly presents with pain and swelling in the affected area. When affecting the calvarium, it can result in otitis media and mastoiditis. Spinal involvement may result in compression fractures and neurological involvement. X-rays demonstrate a purely lytic, well-demarcated lesion with thick periosteal new bone formation (Figure 9.24). Skull lesions are classically described as a 'hole within a hole'. Lesions can sometimes result in 'onion-skin' periosteal reaction which mimics the features of Ewing's sarcoma. Vertebral involvement can result in rapid flattening of the vertebral body (vertebra plana). Bone scan or whole-body MRI will identify polyostotic forms.

Histologically, LCH is characterized by the appearance of intermediate-sized Langerhans cells with indistinct cytoplasm intermixed with inflammatory cells. Langerhans cells contain distinctive 'tennis-racket' intracytoplasmic inclusion bodies, Birbeck granules, which have arisen from the cell membrane.

In the majority of cases, spontaneous resolution occurs. Systemic anti-inflammatories may be effective at controlling symptoms and reducing osteolysis. Mortality is rare and is associated with a multi-organ visceral variant, most frequently in individuals under 2 years of age.

Primary malignant bone tumours

Osteosarcoma

The most common primary malignant bone tumour, osteosarcoma has a bimodal age distribution peaking in adolescence (10–14 years) and again in the seventh decade. In its conventional form, osteosarcoma is a high-grade, medullary osteoid-producing tumour spreading rapidly outwards through the periosteum and into surrounding tissues. Typically metaphyseal or metadiaphyseal, the majority present at the distal femur, proximal tibia, proximal femur and humerus (i.e. where the majority of long-bone growth occurs). Patients complain of worsening pain and swelling, particularly suffering night pain, non-mechanical pain or joint restriction. Pathological fracture is rare. Fever, elevated alkaline phosphatase (ALP) and lactate dehydrogenase (LDH) may be

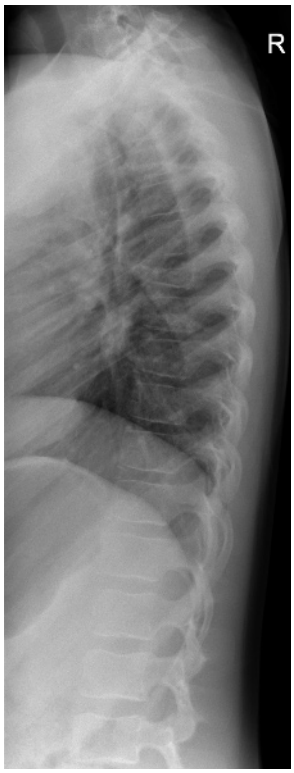


Figure 9.24 Langerhans cell histiocytosis (LCH)
Plain radiographs of an 8-year-old girl who presented with insidious onset of pain in the mid-thoracic spine demonstrate vertebra plana at the level of T6. Biopsy revealed features consistent with LCH.

useful aids to clinical assessment. The skin may be warm and erythematous with venous engorgement. The development of osteosarcoma is associated with Li–Fraumeni (p53 tumour suppressor gene mutation), hereditary retinoblastoma syndromes and Rothmund–Thomson syndrome.

Osteosarcomas may be described as primary (arising *de novo*) or secondary (arising in abnormal bone). These secondary osteosarcomas may be related to previous irradiation, Paget's disease, fibrous dysplasia, bone infarcts, liposclerosing myxofibrous tumour, chronic osteomyelitis or in dedifferentiated chondrosarcomas and have a less favourable prognosis. This explains the secondary elevation in the bimodal distribution of osteosarcoma.

X-rays are generally diagnostic showing an ill-defined, permeative bone-forming lesion causing cortical destruction, periosteal reaction and expansion into the soft tissues. The tumours can be variably mineralized, and there may be a Codman's triangle where new bone forms in response to periosteal elevation and a 'sunburst' appearance when the periosteum does not have enough time to lay down a new layer and instead the Sharpey's fibres stretch perpendicular to the periosteum (Figure 9.25). Classically, 80% are extra-compartmental at presentation, i.e. extending through the cortex. MRI of the whole bone will delineate the medullary and extra-osseous extent of the tumour. Nuclear medicine bone scintigraphy is intensively hot and may identify bone skip lesions, distant bone metastases or bone-producing chest metastases. CT chest is mandatory as lung metastases are common (up to 15%).

A biopsy should always be carried out before commencing treatment; it must be carefully planned according to the principles of performing a biopsy to allow for complete removal of the biopsy tract at definitive surgery. The neoplastic cells demonstrate severe anaplasia and pleomorphism, producing primitive woven bone and osteoid and display a permeative pattern replacing host bone.

Conventional osteosarcoma is subclassified according to the predominant extracellular matrix evident in the tissue, such as osteoblastic osteosarcoma, chondroblastic and fibroblastic variants.

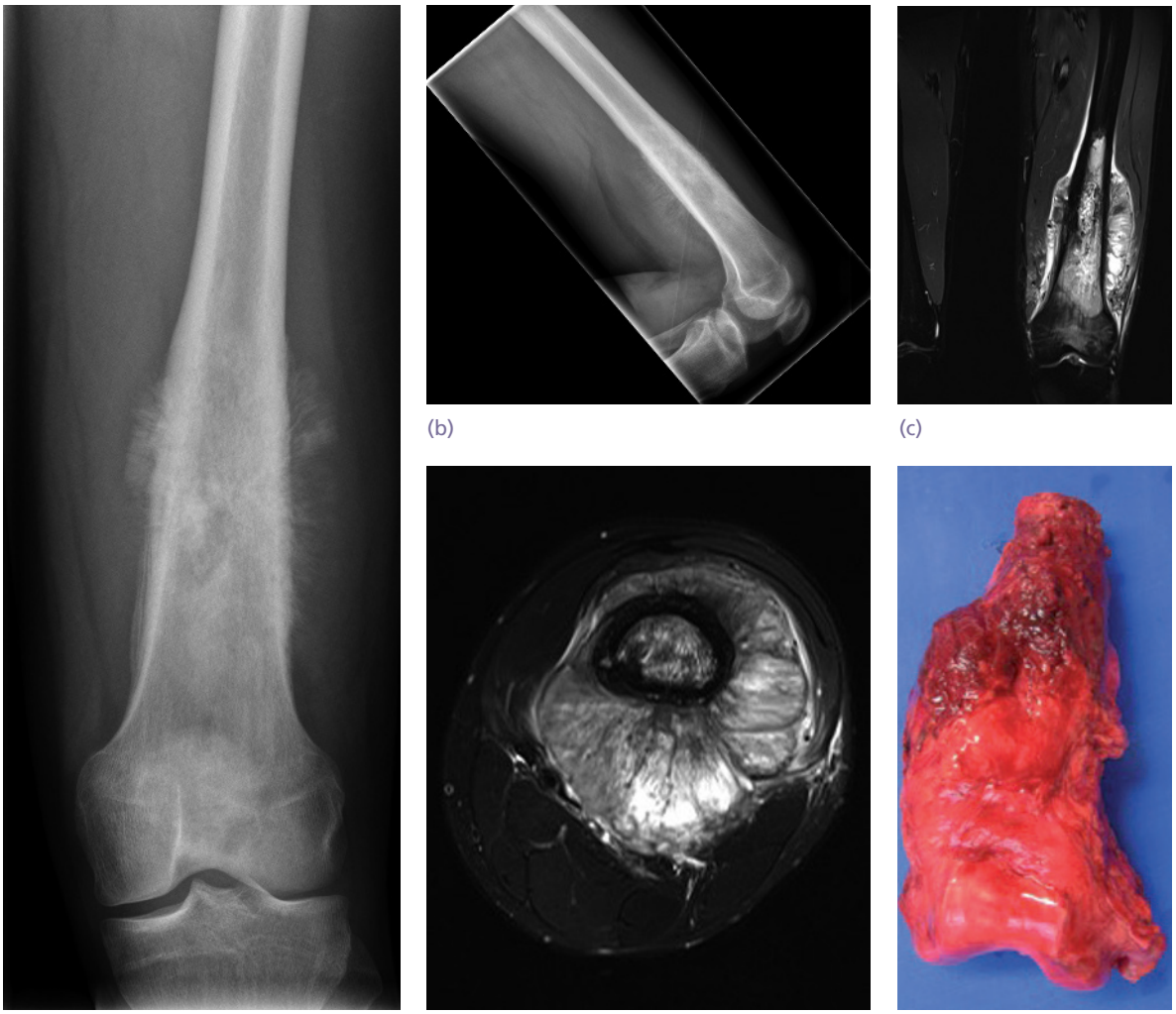
Treatment

Prior to modern chemotherapy and reconstruction techniques, amputation was the standard surgical treatment for musculoskeletal sarcomas of the extremities for the majority of the twentieth century, because of the high risks of local recurrence and associated poor survival. Advances in multi-agent chemotherapy, diagnostic imaging and surgical reconstruction enabled limb-salvage surgery, which constituted the major advancement of the last century in bone sarcoma treatment. Limb salvage has become the preferred method of treatment for osteosarcomas, such that an amputation is considered only when attempted tumour excision would compromise safe surgical tumour margins.

The principal first-line chemotherapeutic agents used in osteosarcoma are doxorubicin, cisplatin, ifosfamide and methotrexate. Multi-agent neoadjuvant chemotherapy is started immediately after diagnosis for 8–12 weeks and then, after re-staging to evaluate response to induction chemotherapy and provided the tumour is resectable and there are no skip lesions, a wide resection and limb reconstruction are performed.

The resected specimen is subjected to histopathological analysis to determine if satisfactorily wide margins have been achieved and the response to chemotherapy is judged by the degree of tumour cell necrosis. These can predict the risk of local recurrence and survival. At least 30 microscope fields within the tumour are inspected for the extent of necrosis, and a minimum 90% necrosis rate is required for a good response. Factors other than resection margin and chemotherapy response that are independently predictive of a worse outcome are large tumours, ablative surgery, age under 14 years, male gender, high ALP, local recurrence, p-glycoprotein expression, and absent *Erb2* expression.

If tumour necrosis is marked, chemotherapy is continued for another 6–12 months. If, however, the



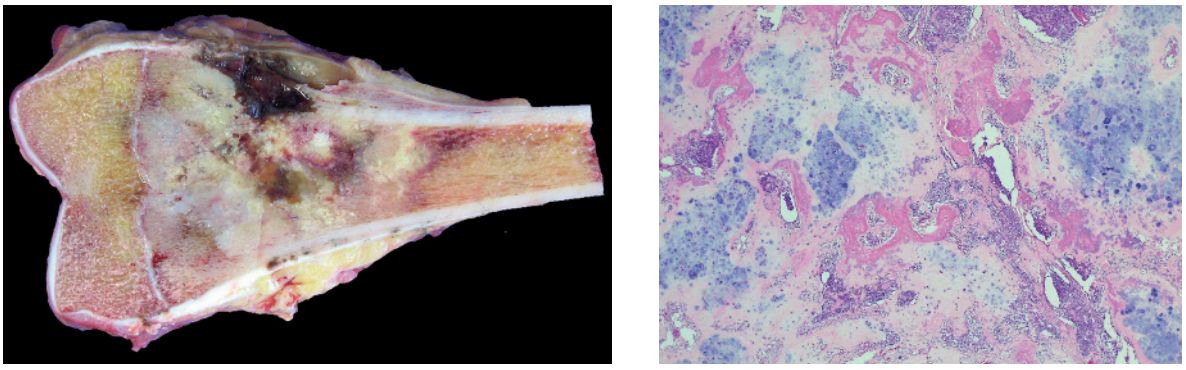
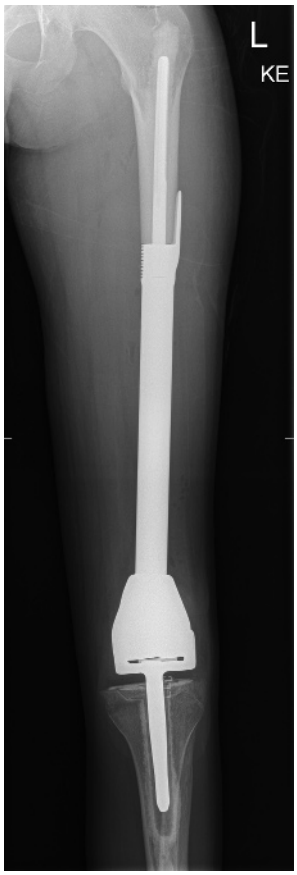


Figure 9.25 Osteosarcoma The characteristic features of a distal femoral osteosarcoma as seen on plain radiographs (a,b). The lesion, located in the metadiaphysis, demonstrates a mixed lytic blastic appearance. There is elevation of the periosteum resulting in a Codman's triangle. The soft-tissue extension of the tumour results in bone formation within the surrounding soft tissues and the appearance of sunray spiculations. On MRI (c,d), the true extent of the tumour can be seen erupting from the bone and extending into the posterior soft tissues. The patient received neoadjuvant chemotherapy with a dramatic response in the tumour volume (i, j) followed by resection of the tumour (e). The resection specimen demonstrates a pale tumour occupying the distal femur and extending through the cortex (f). The dominant histological features are those of malignant stromal tissue with areas of osteoid formation with interspersed areas of chondroid differentiation in keeping with a chondroblastic osteosarcoma (g) (x100). The tumour was excised in its entirety and reconstruction was with a distal femoral endoprosthetic replacement (h). *(Continued)*



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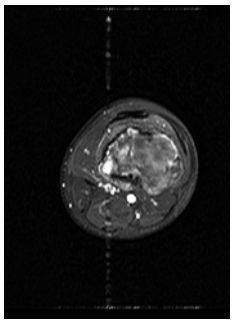

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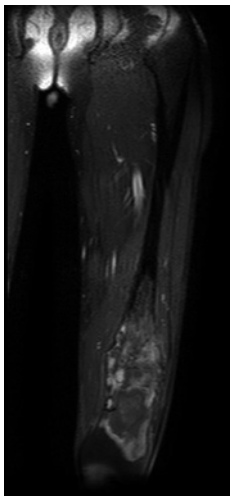


(j)

Figure 9.25 Osteosarcoma (Continued)

response is poor, a different chemotherapeutic regime is substituted. Pulmonary metastases, especially if they are small and peripherally situated, may be completely resected with a wedge of lung tissue.

Five-year survival after wide resection and chemotherapy has improved from around 50% in 1984 to over 60%. The possibility of downgrading tumours before surgery facilitated the evolution of reconstruction methods after tumour resection. Until the beginning of neoadjuvant chemotherapy in 1978, 80% of patients with an extremity osteosarcoma underwent amputation; today, limb-salvage surgery is possible in 90% of cases.

Variants of osteosarcoma

Rare subtypes

Rare histological subtypes of conventional medullary osteosarcoma include *telangiectatic* and *small-cell osteosarcoma*. The clinical presentation is similar to medullary osteosarcoma, although pathological fracture may occur in 25% of telangiectatic osteosarcomas. Radiographically, small-cell osteosarcomas have a classic appearance; telangiectatic osteosarcomas are purely lytic lesions with cortical disruption and soft-tissue extension and may be mistaken for benign giant-cell tumours of bone. MRI may show multiple fluid-fluid levels similar to aneurysmal bone cysts but with nodular and solid components. Histologically, scant osteoid matrix is found in telangiectatic osteosarcoma, which correlates with the radiological appearance. Small-cell osteosarcoma contains sheets of small, round blue cells.

Small-cell osteosarcoma requires a different chemotherapeutic regime, similar to the agents used in other round blue-cell tumours (e.g. Ewing's sarcoma) and carries a slightly worse prognosis. Telangiectatic osteosarcoma is treated in the same way as conventional osteosarcoma with similar outcomes.

Parosteal osteosarcoma

This is a low-grade osteosarcoma arising on the surface of almost exclusively long bones, classically at the posterior distal femoral or proximal tibial metaphysis. More common in females, peak incidence is in the third decade and patients present with little pain but a slowly enlarging hard mass and joint restriction, potentially for years prior to presentation.

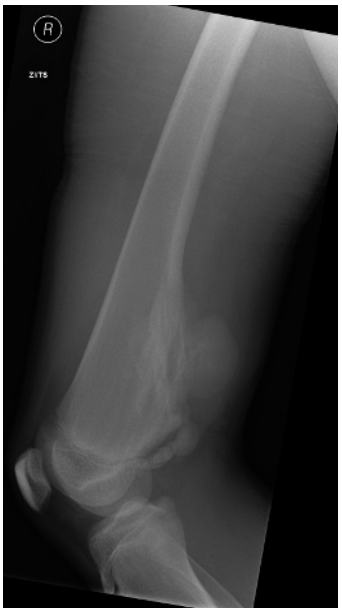
Radiographs show a radio-dense mass with a broad base enveloping the bone (Figure 9.26). CT and MRI are useful to identify medullary involvement which is needed for operative planning. Histologically, the lesion consists of well-formed bone but lacks regular trabecular arrangement. The spaces between trabeculae are filled with cellular fibroblastic tissue; a few atypical cells and mitotic figures can usually be found. In 15% of cases, more aggressive areas of dedifferentiation to high-grade will be seen.

Surgical excision should aim to achieve wide excision margins, as local recurrence is associated with dedifferentiation to high-grade in 80% and consequently worse survival. The role of chemotherapy in parosteal osteosarcoma is not well established and overall survival is greater than 90% at 5 years.

Periosteal osteosarcoma

Periosteal osteosarcoma is a rare tumour and is quite distinct from parosteal osteosarcoma. This predominantly chondroblastic intermediate-grade tumour arises from the periosteum, more often in the diaphysis than metaphysis of long bones, especially the femur and tibia (80% of cases), typically arising earlier in the second decade of life, causing a painful mass and 6–12 months of symptoms at time of diagnosis.

X-rays show a fusiform periosteal radiolucent mass sometimes with a sunburst appearance and Codman's triangle. MRI excludes medullary involvement and highlights the extent of the soft-tissue component. The tumour may involve almost the entire circumference



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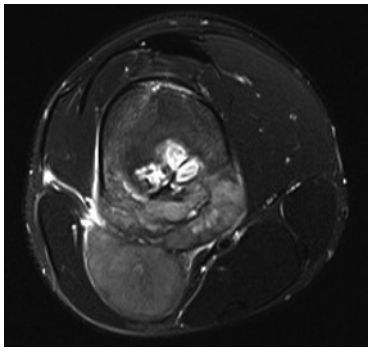


(b)

Figure 9.26 Parosteal osteosarcoma A 12-year-old male presented with a rapidly enlarging mass behind the knee. Plain X-rays (a) demonstrate the characteristic features of a parosteal osteosarcoma with a bone-forming surface lesion arising from the cortex and merging with the bone. MRI demonstrates the true extent of the tumour (b) with a clear line of separation between the surface lesion and the bone but merging with the cortex and, in this case, intramedullary involvement.

of the affected bone. The differential diagnosis would include periosteal chondroma. Histologically this is a true osteosarcoma of intermediate grade with osteoid and chondroid matrix deposition.

The management is the same as for conventional medullary osteosarcoma with neoadjuvant chemotherapy and wide excision; overall survival is reported to be 89% at 5 years and 83% at 10 years, with survival rates related to the development of local recurrence.

Secondary osteosarcoma

Paget's disease affects approximately 2% of Western Europeans. Although malignant transformation is a rare complication of this disease, most osteosarcomas appearing after the age of 40 years fall into this category. The incidence of osteosarcomatous change in Paget's disease is approximately 1% and most common in the polyostotic form, usually involving the femur, pelvis, humerus and skull and may be multifocal in up to 20%. Warning signs are the appearance of pain or swelling in a patient with long-standing Paget's disease. Mean age at presentation is 71 years. Pathological fractures may occur, commonly in femoral lesions. X-rays show typical pagetic bone with a lytic destructive mass extending into the soft tissues.

The risk of post-radiation sarcoma is dose-dependent and is most common in the pelvis and scapula (reflecting radiotherapy for cervical/ovarian and breast carcinomas respectively). The time lag between radiotherapy and subsequent development of osteosarcoma can range from 6 to 23 years. Clinically and radiologically the features are similar to conventional osteosarcoma, although post-radiation changes (trabecular coarsening and cortical lysis) may be evident.

The treatment principles for secondary osteosarcoma are the same, however elderly patients poorly tolerate the aggressive chemotherapeutic and surgical treatments offered to young osteosarcoma patients. Chemotherapeutic dose reductions may be possible if there are concerns regarding cardiac and renal toxicity. Consequently, osteosarcoma in elderly patients older than 65 years has a worse prognosis than that of the younger population. Paget's osteosarcoma specifically has an abysmal prognosis; median survival is 9 months post-diagnosis.

Chondrosarcoma

Chondrosarcomas are the second most common primary malignant bone tumours after osteosarcoma, with heterogeneous clinical, radiological and histological features. This group of aggressive malignant cartilage tumours represent a spectrum of the same disorder that arises in enchondromas. This is suggested by the observation that enchondromatosis could evolve into a low-grade chondrosarcoma and that, after excision, radiologically low-grade chondrosarcomas may have areas of higher grade.

Chondrosarcomas can present in adults from the third to the eighth decades, peaking between 40 and 70 years of age, and men are affected more often than women. These tumours are slow-growing and are usually present for many months before being discovered. They produce deep pain and/or a gradually enlarging mass and arise in any bone derived from enchondral ossification. Most frequently located in the proximal femur, pelvis, proximal humerus, distal femur, scapula and proximal tibia, these metaphyseal lesions can

extend across large segments of the involved bone. Despite the relatively frequent occurrence of benign cartilage tumours in the small tubular bones of the hands and feet, malignant lesions are rare at these sites, representing <1% of chondrosarcomas.

Chondrosarcomas take various forms, usually designated according to: (a) their location in the bone (*central* or *peripheral*); (b) whether they develop without benign precursor (*primary chondrosarcoma*) or by malignant change in a pre-existing benign lesion (*secondary chondrosarcoma*); and (c) the predominant *cell type* in the tumour. Approximately 85% are *primary central* chondrosarcomas occupying the medullary cavity. Radiographically these appear as large, intraosseous, osteolytic tumours with a narrow zone of transition and irregular, granular calcifications within the matrix described as 'honeycomb' or 'popcorn' (Figure 9.27). Endosteal scalloping of the cortex and eventual cortical destruction can occur, and there may be a faint periosteal reaction. These tumours grow along the path of least resistance i.e. along the medullary canal, and soft-tissue extension is more common in pelvic chondrosarcomas. CT is useful for demonstrating the matrix calcifications and permeative destruction of the tumour.

MRI is useful to define tumour extent and identify more biologically active parts of the tumour which may have undergone further malignant change and become higher-grade or even dedifferentiated. These avascular tumours reproduce the high signal of hyaline cartilage on T2 weighted sequences. Accurate differentiation of low-grade from high-grade chondrosarcomas is essential before surgery, and MRI can reliably differentiate high-grade from low-grade chondrosarcomas of long bone. Differentiating features are bone expansion, periostitis, soft-tissue mass and tumour length (mean intramedullary extent 11.8 cm in high-grade tumours; 5.5 cm in low-grade tumours), and the presence of these four MRI features has a diagnostic accuracy of 96%. This has significant prognostic importance for the patient, as more aggressive tumours have higher mortality rates.

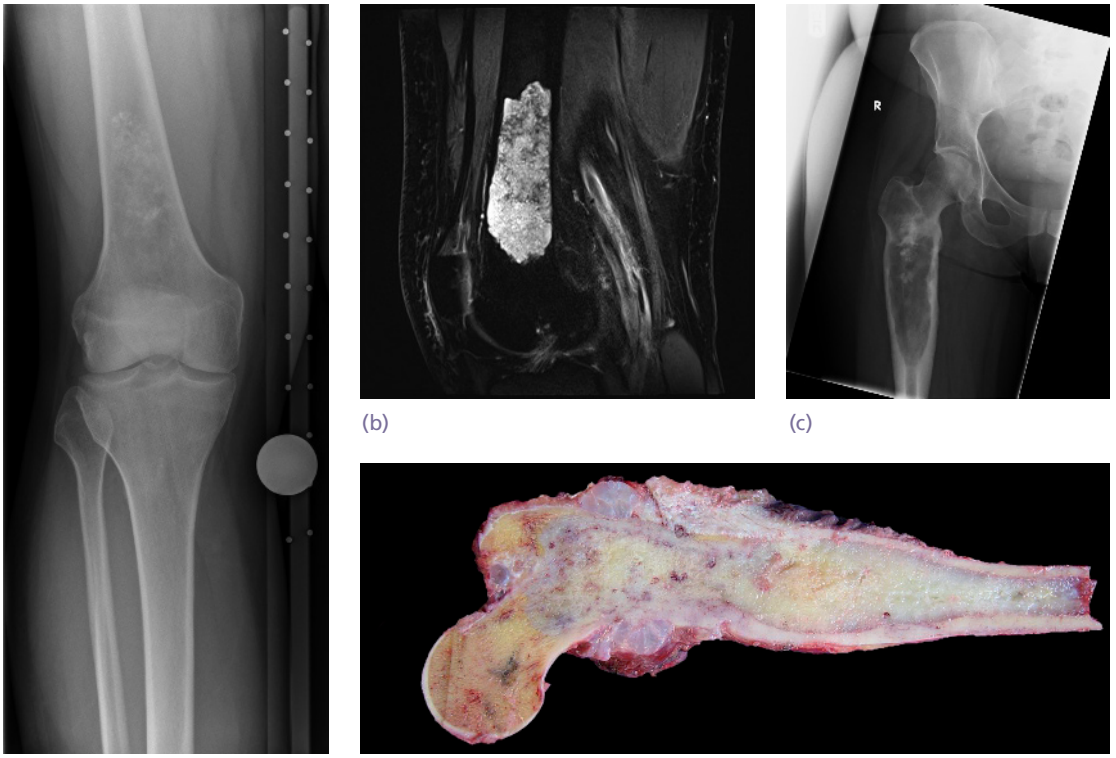
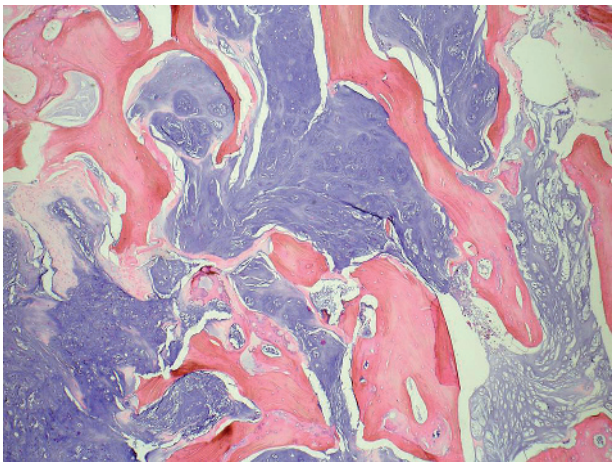


Figure 9.27 Chondrosarcoma–central chondrosarcoma The challenge of the central chondroid lesion. Plain X-rays (a) demonstrate an 8.5 cm chondroid lesion in the distal femur which remained unchanged on radiographs over a period of 12 months. However, on MRI (b), minor endosteal scalloping of the anterior femoral cortex can be seen which, combined with the advent of pain in the thigh, warranted resection of the tumour. Histology post resection confirmed a low-grade chondrosarcoma. (c–e) Often, the differentiation between benign chondroid lesion and chondrosarcoma can be a little easier, as in the case of a 76-year-old woman who presented with worsening pain in the right thigh: radiographs (c) demonstrate a destructive, expansile lesion in the proximal femur with the characteristic features of a chondroid matrix. Resection histology demonstrates a pale, glistening cartilage lesion in the medullary cavity which spreads beyond the cortex (d). *(Continued)*



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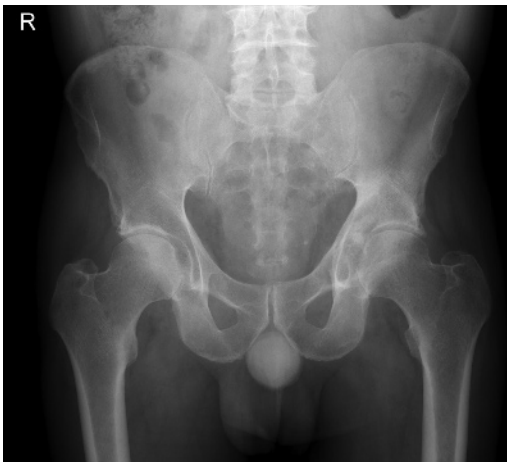

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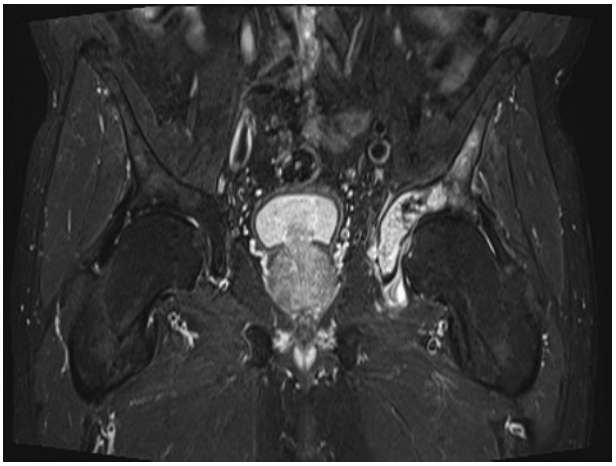

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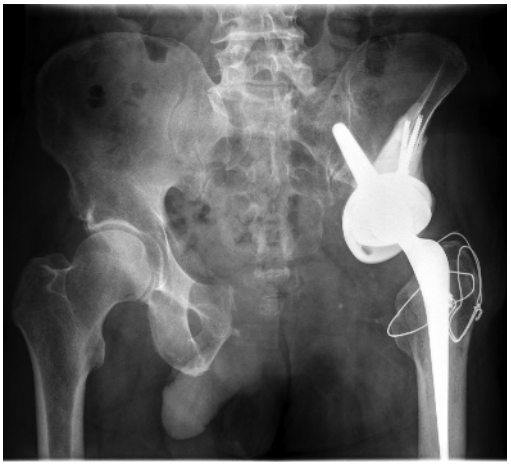


(h)

Figure 9.27 Chondrosarcoma–central chondrosarcoma (Continued) Microscopy (x100) demonstrates lobules of highly atypical cartilage cells, including binucleate cells permeating through the surrounding bone matrix (e). (f–h) For a 70-year-old man who presented with an 18-month history of left-sided hip pain, plain X-rays (f) and MRI (g) demonstrate a chondroid lesion in the periacetabulum which extends down the anterior column. The tumour was biopsied and found to be a chondrosarcoma. Treatment was by resection of the periacetabulum and pubis and reconstruction with an ice-cream cone prosthesis and hip replacement (h).

Dedifferentiated chondrosarcomas represent the highly malignant end of the chondroid tumour spectrum, developing in 10–15% of central chondrosarcomas, in which a high-grade undifferentiated sarcoma or osteosarcoma coexists with a lower-grade chondroid tumour. The median age is 59 years, with a slight predominance of males, and the most common sites are the femur and pelvis. They have a very poor prognosis (5-year survival 7–24%), improved only by wide surgical resection, worsened by pathological fracture (29% of cases), advancing age and metastasis at diagnosis. Chemotherapy and radiotherapy have not been shown to improve survival.

Full staging mandates X-rays and MRI of the entire bone affected, prior to biopsy, plus distal staging including bone scintigraphy and chest CT. A biopsy is essential to confirm the diagnosis. Histopathology reveals macroscopically lobular white hyaline cartilage, areas of mineralization and cystic changes, erosion of the cortex and soft-tissue expansion. Microscopically the high cellularity, atypia, mitoses and permeation into host bone distinguish a chondrosarcoma from an enchondroma. They are classified according to grade (grade I/atypical cartilaginous tumour and high-grade II and III) which is useful in predicting prognosis. Grade I tumours are usually locally aggressive with low metastatic potential, unlike high-grade lesions. Not infrequently, areas of low-grade and high-grade chondrosarcoma may be seen in the same tumour. Over 60% of chondrosarcomas show

mutation in either (isocitrate dehydrogenase 1) *IDH1* or *IDH2* genes.

Chondrosarcoma is resistant to both chemotherapy and radiation, making surgical excision the only treatment. In high-grade tumours, only wide excision margins are oncologically acceptable to minimize local recurrence. Prognosis is determined by the cellular grade, stage, tumour size, (axial versus appendicular) site and the resection margin. Overall survival at 5 years for low-grade tumours is 90–100%, for grade II tumours approximately 60% and for grade III approximately 30–40%. A pathological fracture of the femur has a negative prognostic influence in grade I chondrosarcoma and increases the risk of local recurrence in dedifferentiated femoral chondrosarcomas. In some cases isolated pulmonary metastases can be resected and adjuvant chemotherapy may be contemplated in dedifferentiated or mesenchymal chondrosarcomas.

In low-grade chondrosarcomas, some surgeons advocate intralesional curettage or radiofrequency ablation in selected tumours, although this may be associated with higher rates of local recurrence and it is believed that recurrences may transition to higher-grade tumours associated with worse prognosis. For this reason, others recommend wide resection and reconstruction, balancing the worse physical function with reduced risk of local recurrence.

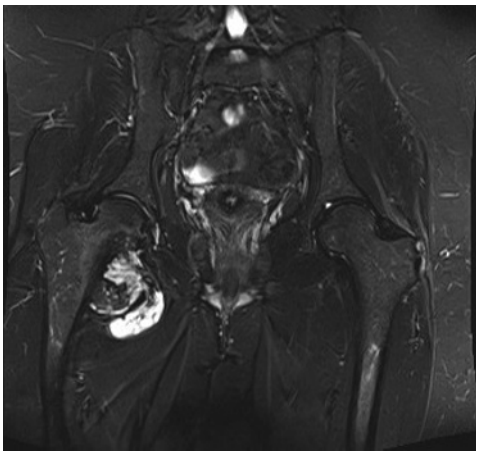
Secondary central chondrosarcomas These arise in previously benign enchondromas, although the risk cannot be quantified as enchondromas are frequently asymptomatic incidental findings on X-rays. Ollier's disease or multiple enchondromatosis patients present in childhood with enchondromas, most numerous in the phalangeal bones of the hands and feet. Clinical findings include asymmetric limb shortening, swelling of the fingers and toes, and disturbed movements of the interphalangeal joints. They may occur unilaterally, but frequently they are bilateral. After puberty no new enchondromas develop, and tumour growth in adulthood is considered to be malignant degeneration of these tumours. Pathological fractures in the affected bones may occur. Maffucci syndrome combines multiple enchondromas and cutaneous or visceral haemangiomas, and may present at birth or develop later. Ollier's disease and Maffucci syndrome have a significant risk of developing secondary chondrosarcomas of approximately 20–30% and 40–50% respectively, typically in the third or fourth decade, although in Maffucci syndrome, patients are at increased risk of also developing carcinomas (e.g. breast, liver, ovary), which suggests an underlying genetic predisposition.

Secondary peripheral chondrosarcomas These arise in the cartilage cap of an exostosis (osteochondroma) that has been present since childhood. Exostoses of the pelvis and scapula seem to be more susceptible than others to malignant change, because these sites permit undetected growth. Malignant transformation is associated with 1% of solitary osteochondromas and in 4% of patients with multiple osteochondromas (MO) (diaphyseal aclasia), again typically in the fourth decade, much earlier than central medullary chondrosarcomas. MO is an autosomal dominant disorder; in almost 90% of MO patients germline mutations in the tumour suppressor genes *EXT1* or *EXT2* are present. Any increase in size or onset of new pain is suspicious in patients after skeletal maturity. A cartilage cap thicker than 1.5 cm when measured with MRI may indicate malignant change and surgical excision (Figure 9.28).

Periosteal chondrosarcomas These are among rarer types of chondrosarcomas. They arise from the periosteum of the metaphysis of the femur or humerus, often in the second to fourth decades. Pain and swelling may be evident, and imaging reveals a lobular mass adjacent to the cortex often containing flecks of calcification, as well as 'sunray' streaks and new bone formation at the margins of the stripped periosteum and medullary involvement rare. Histologically these are usually grade I or II chondrosarcomas. Local recurrence depends upon the margin of surgical excision, with up to 29% recurrence rates reported, and metastasis-free survival depends on histological grade.

Clear-cell chondrosarcomas Making up 2% of all chondrosarcomas, these classically have an epiphyseal location of the proximal or distal femur or humerus and present more commonly in males between the third and fifth decades of life. Considered slow-growing, low-grade tumours, they present with pain and joint restriction and appear as osteolytic lesions with narrow transition, a narrow sclerotic rim and small matrix calcifications. Histologically they can be difficult to differentiate from chondroblastomas, except that they develop in adults and lack the IDH mutations of other chondrosarcomas. Radiological correlation is mandatory, as clear-cell chondrosarcomas tend to lack per-tumoural oedema and to be higher signal on T2-weighted sequences. Long-term survival is good, although there are reports of late metastasis mandating long-term surveillance after wide surgical excision.

Mesenchymal chondrosarcomas These highly malignant and poorly differentiated chondrosarcomas unusually present in most age groups (median 30 years) in a wide variety of anatomical sites (limb, pelvis, vertebrae, craniofacial) including extraskeletal sites in one-third. The radiographic appearances are similar to those of the common types of chondrosarcoma but the clinical behaviour



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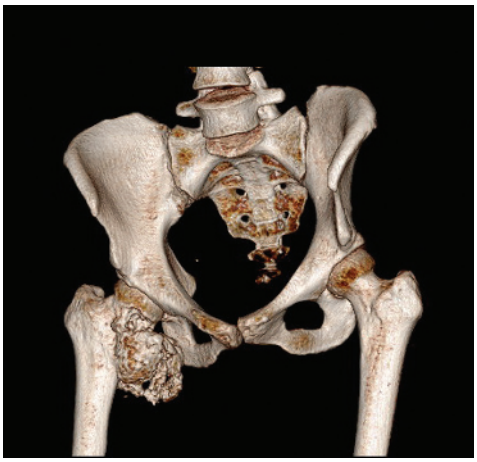

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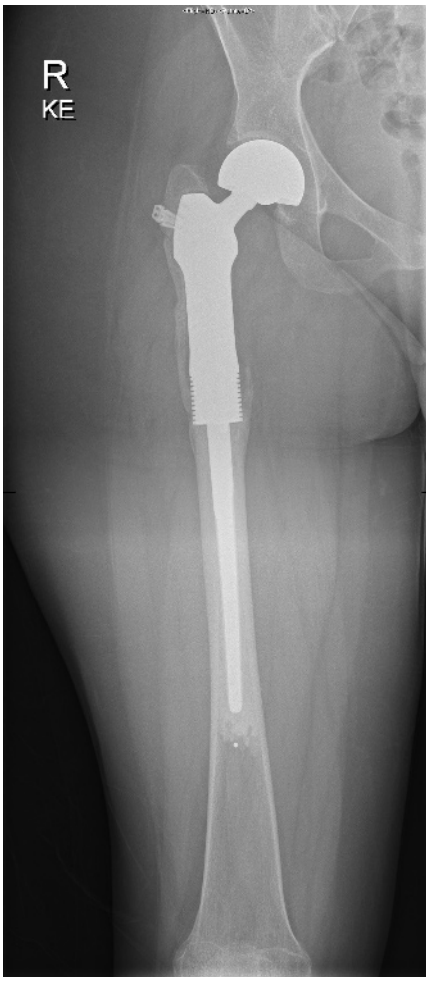


(c)

Figure 9.28
A long-standing osteochondroma arising from the proximal femur
(a) MRI demonstrates a thick cartilage cap overlying the osteochondroma, which measures 3.5 cm. (b) 3D-CT reconstruction demonstrates the calcification and speculation within the cartilage cap. The tumour was removed in its entirety and reconstructed with a proximal femoral endoprosthetic replacement (c). The resection histology confirmed a low-grade chondrosarcoma.

of the tumour is usually more aggressive. Prognosis varies considerably, with metastatic disease at diagnosis conferring the worst prognosis. Overall 5-year survival is approximately 50%. Wide resection and adjuvant chemotherapy are considered the standard of care, and late metastasis is a feature of this rare disease.

Ewing's sarcoma

Ewing's sarcoma is believed to arise from mesenchymal stem cells in the bone marrow and is closely related to other tumours such as primitive neuroectodermal tumour of soft tissues, peripheral neuroepithelioma and Askin tumour. This highly malignant tumour occurs more frequently in males, typically between 10 and 20 years of age, and has a diaphyseal long-bone location but is equally as common in flat bones. The femur is the most common long-bone site, followed by the tibia, humerus and fibula, but overall the pelvis accounts for the majority of cases.

Pain is the earliest symptom, followed by swelling (although soft-tissue masses may not be apparent in pelvic sites) and a low-grade fever. Serologically, the ESR and WCC may be elevated and haemoglobin may be reduced. Differential diagnoses may therefore include osteomyelitis.

Radiographically, an aggressive, permeative, poorly defined osteolytic lesion with cortical destruction, periosteal reaction and large, radiolucent soft-tissue mass may be found. Periosteal reaction is common in young patients with the lamellar 'onion-skin' appearance causing fusiform bone enlargement (Figure 9.29). This may mimic infection or eosinophilic granuloma in young patients; subtle bone involvement with a large soft-tissue mass could mimic primary bone lymphoma in older patients.

Routine local and distal staging includes bone scintigraphy and chest CT. MRI demonstrates the intra- and extra-osseous tumour extent, and lesions are 'hot' with bone scintigraphy. Lung, skeletal and lymph node (rare) metastases may be evident at presentation in up to 20% of cases. Bone marrow involvement is a unique feature of Ewing's sarcoma

among other bone sarcomas and is associated with a worse prognosis.

Unlike other bone sarcomas which produce extracellular matrix that resembles bone or cartilage, Ewing's sarcoma is composed of primitive, undifferentiated, small, round blue cells with large nuclei and scant cytoplasm that bears no resemblance to normal tissue. The characteristic translocation between chromosomes 11 and 22 results in a fusion gene, *EWSR1-FL11*, which is present in 90% of cases. Other translocations have been discovered, most frequently *t(21:22)*. The presence of the translocation may be supported by a split fluorescence in situ hybridization (FISH) signal.

Preoperative chemotherapy frequently results in extensive tumour necrosis and shrinkage. Agents used in Ewing's sarcoma include vincristine, ifosfamide, doxorubicin and etoposide. Wide excision may be combined with adjuvant radiotherapy if surgical margins are poor. Definitive radiotherapy rather than surgical excision, although associated with worse overall survival, may be advocated if non-resectable or metastasis has occurred during chemotherapy, to spare the surgical morbidity. A more favourable outcome is expected with younger age, distal appendicular sites, small tumour volume, normal ESR and greater chemotherapy necrosis response. Five-year survival greater than 60% can be expected for appendicular tumours.

Rare bone sarcomas

These are generally known as 'spindle-cell sarcomas of bone'. Most spindle-cell sarcomas producing collagen, but not osteoid matrix, were initially identified as fibrosarcomas, latterly as malignant fibrous histiocytomas, and currently as spindle-cell sarcomas of bone. There have been a number of differing diagnostic descriptions for these tumours, although the recent World Health Organization (WHO) classification has retained the characterizations for all types.

Age at the time of diagnosis ranges from the third to the sixth decade, with equal sex distribution. The long bones of the lower limb are most commonly affected, and the clinical presentation does not differ from that of the classic bone sarcomas, with pain, swelling, restriction and pathological fracture. Treatment is similar to that recommended for osteosarcoma, including chemotherapy and wide excision and limb salvage, if chemotherapy is not contraindicated in an elderly or unfit patient. The typical radiological appearance of a spindle-cell sarcoma is a poorly defined lytic lesion without osteoid formation or punctate calcification, and with very little periosteal reaction.

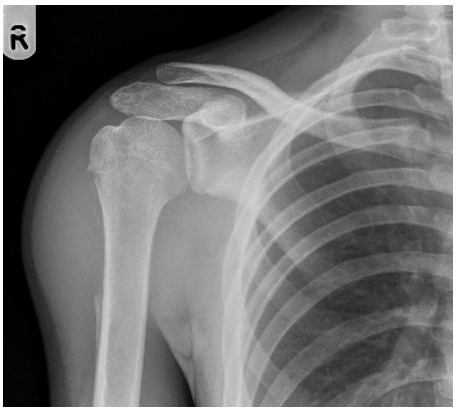
The prognostic factors associated with worse survival are similar to those of patients with an osteosarcoma: large tumour size, older age, pathological fracture, need for amputation and poor response to preoperative chemotherapy are all significant. Risk factors for local recurrence were also similar to those in patients with an osteosarcoma: close surgical margins and poor response to preoperative chemotherapy were the two most significant factors. The 5-year survival rate for non-metastatic spindle-cell sarcomas of bone is 67%.

Haematopoietic tumours – multiple myeloma

Myeloma is a malignant proliferation of neoplastic plasma cells of B-cell lineage within the bone marrow, leading to increased production of plasma paraprotein and immunoglobulin. It most commonly arises in the marrow containing bones of the vertebrae, pelvis and femur. The median age at diagnosis is 70 years and it is infrequently diagnosed before the age of 40 years. The commonest primary malignant lesion arising in bone, it has an estimated incidence of 3000 per annum in the UK.

Multiple myeloma arises as a result of multiple genetic mutations of plasma cells and the immunoglobulins they produce. Multiple myeloma tumours may arise *de novo* or from the premalignant condition monoclonal gammopathy of undetermined significance (MGUS). Diagnosis of myeloma depends on the detection of para-proteins in the plasma or urine, bone marrow biopsy, and evidence of end-organ/bone damage. Serum or urinary electrophoresis measures immunoglobulins that are overproduced by the malignant plasma cells. Multiple myeloma has several effects on bone: focal osteolytic lesions, generalized bone loss (osteopenia), and elevated bone turnover via the receptor activator of nuclear factor-κB (RANKL) system. End-organ damage and disease severity are assessed by haemoglobin levels, ESR, renal function and serum calcium.

Traditionally, a skeletal survey required X-rays of the spine, skull, pelvis, ribs/sternum, and the humerus/femurs to screen for osseous lesions, and assess tumour burden and fracture risk. Classic 'punched-out', osteolytic lesions with cortical thinning can be seen, which in the skull may cause a 'pepper-pot' appearance (Figure 9.30). More often found in the axial skeleton (particularly spine, skull and pelvis), long-bone lesions are typically metaphyseal. CT is superior for demonstrating fractures, osteolytic lesions, and soft-tissue masses and may aid in distal staging of disease. MRI scanning is useful to stage the extent of marrow infiltration, visualization of focal masses and areas most at risk of fracturing, and to highlight response to treatment. Bone scintigraphy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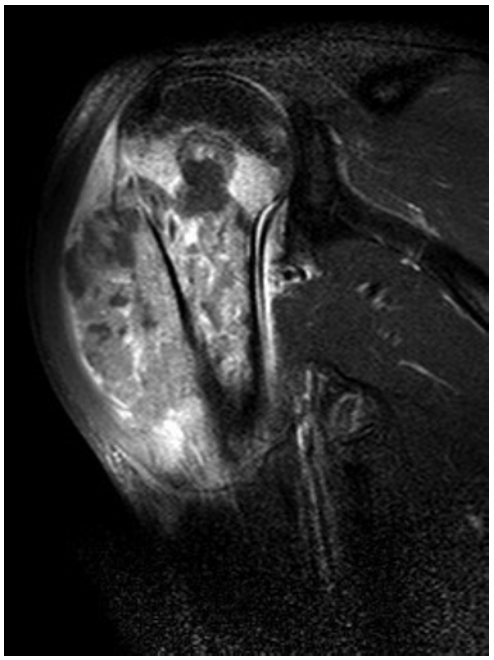
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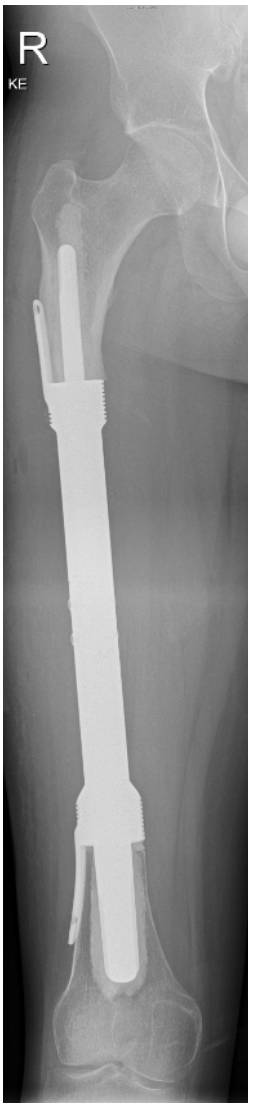

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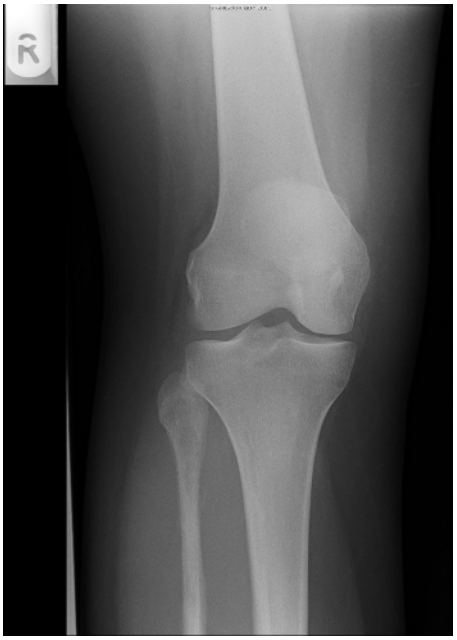
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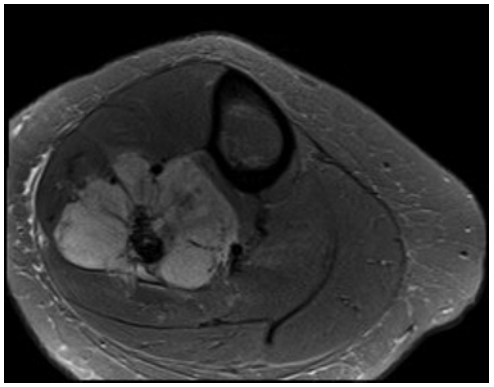
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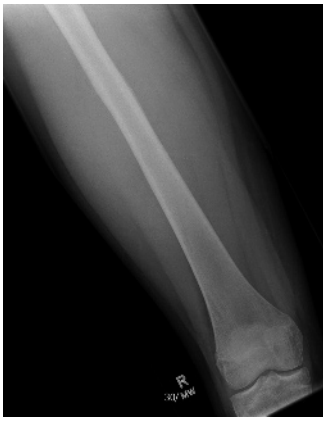
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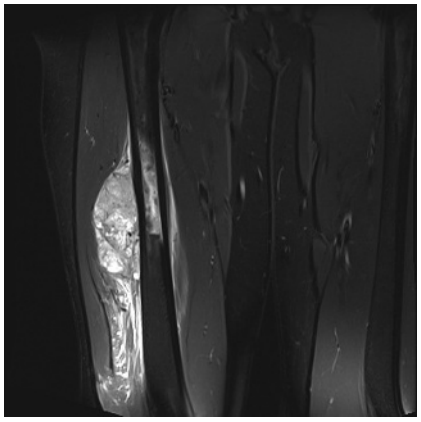
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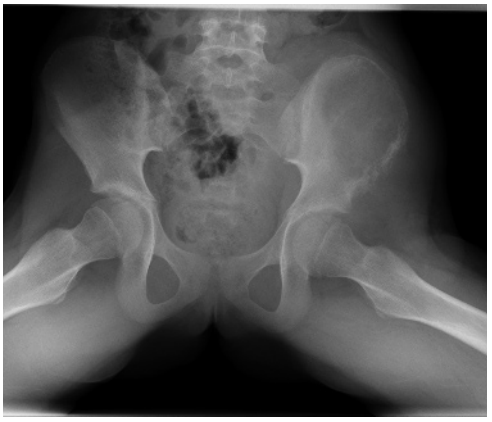
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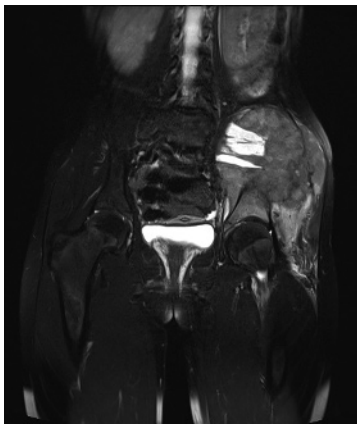
Figure 9.29 Ewing's sarcoma (a–g) Examples of Ewing's sarcoma, affecting the humerus (a,b), the fibula (c,d), and the femoral diaphysis (e,f), treated by neoadjuvant chemo-radiotherapy and subsequent reconstruction with a diaphyseal endoprosthesis (g). Note the evidence of bone destruction, which can sometimes be subtle on plain radiography, but the extensive soft-tissue component which is arising from the intramedullary compartment seen on MRI. (h–n) A 14-year-old girl who presented with a short history of pain and subsequently a mass in the left hemipelvis. X-rays (h) demonstrate a destructive lesion with a wide zone of transition in the left ileum. MRI (i) shows a very large soft-tissue mass with associated bone destruction arising from the left ilium extending down to the upper border of the acetabulum. *(Continued)*



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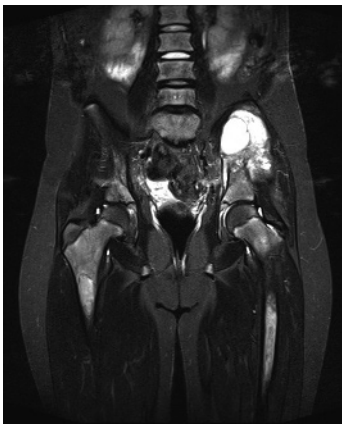

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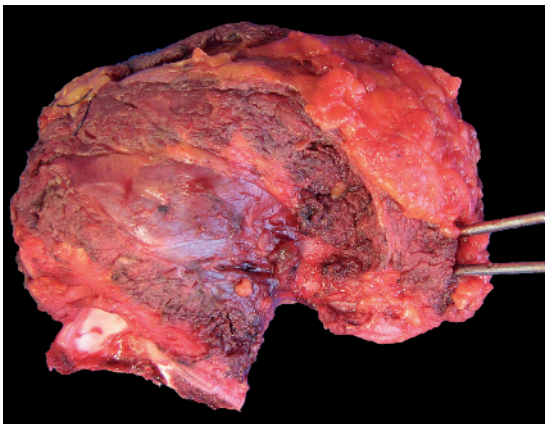

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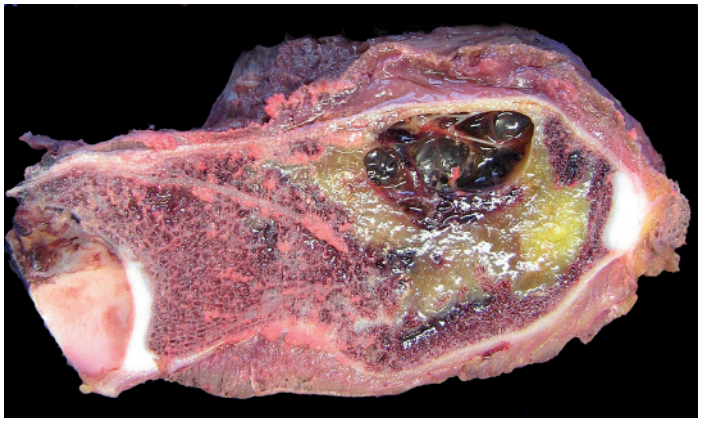

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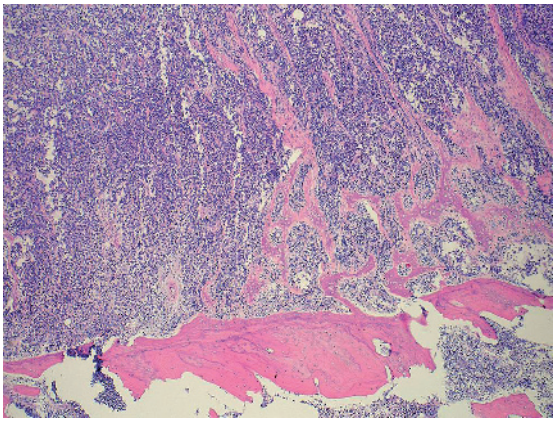

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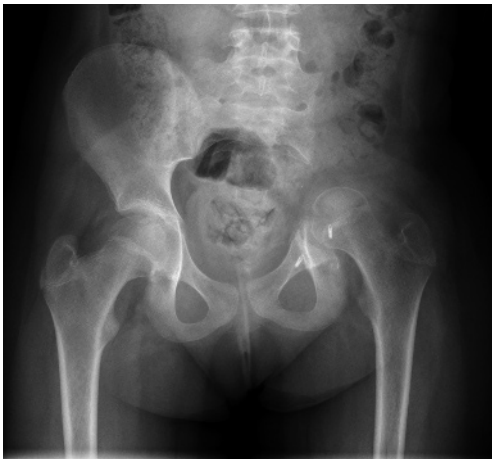
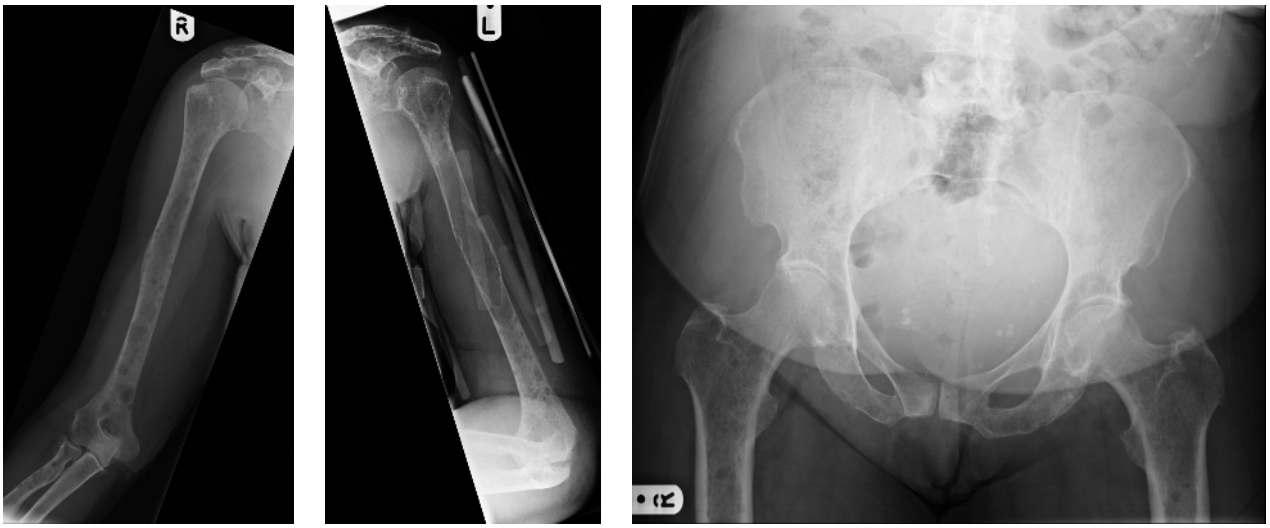


(n)

Figure 9.29 Ewing's sarcoma (Continued) The patient was treated with neoadjuvant chemotherapy and high-dose proton radiotherapy to the left ilium. Post-treatment MRI (j) demonstrates a dramatic response with significant reduction in the soft-tissue component. The patient underwent resection of the ilium to the level of the triradiate cartilage. The resection was assisted by computer navigation, which allowed very accurate planning of the level of the osteotomies. Note the navigation pins in the resection specimen (k) for mounting the navigation tracker. At sectioning, the necrotic, cystic core of the tumour can be seen (l). Histology of the initial diagnostic biopsy (x100) confirms a small, round blue-cell tumour (m) which is confirmed as Ewing's sarcoma by fluorescence *in situ* hybridization and the presence of the *EWS/FLI-1* translocation. Due to the use of pre-operative radiotherapy, no implant was inserted into the pelvis due to the perceived risk of infection. Rather, a collagen matrix graft was hung from the residual acetabulum over the femoral head to form a neo-hip joint (n). After a period of rehabilitation, the patient can mobilize unaided with a Trendelenburg gait. The resection histology demonstrated no evidence of viable tumour, suggesting an excellent response to neoadjuvant therapy and an excellent prognosis.



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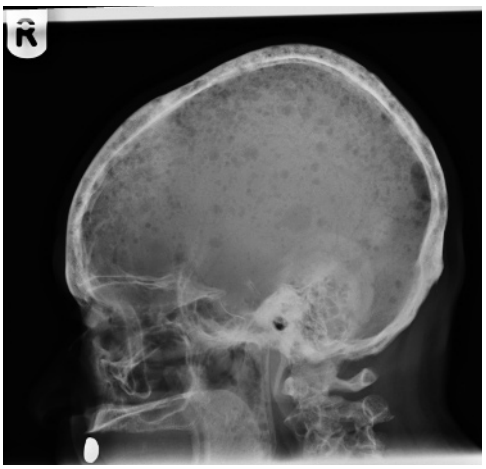


(d)

Figure 9.30 Multiple myeloma Skeletal survey (a–c) of a 74-year-old woman who presented with a pathological fracture through her left humerus demonstrated multiple deposits throughout the skeleton, with characteristic 'pepper-pot' appearance of the skull (d). The diagnosis is multiple myeloma.

underrepresents the extent of disease as it relies on osteoblastic activity.

Bone disease in myeloma may cause severe bone pain, spinal cord compression, diffuse osteoporosis, hypercalcaemia and pathological fractures, which occur in approximately 40% of patients. Medical complications include anaemia, hypercalcaemia, hyperviscosity, immunosuppression and renal dysfunction.

Although incurable, long-term remission can be achieved in some patients. Management comprises combined alkylating chemotherapy, a steroid such as dexamethasone or prednisolone and an immunomodulatory agent such as thalidomide. Autologous stem-cell transplantation (ASCT) involves bone marrow harvest, high-dose chemotherapy to produce myelosuppression and transfusion with stem cells, and it is the standard of care in patients up to 65 years. Bisphosphonates are used in all symptomatic patients to prevent fractures and control hypercalcaemia. Surgery has a supportive effect on the management, long-term survival and quality of life in multiple myeloma patients. The aim is usually to decompress or stabilize vertebral fractures with instability or neurological compression, to stabilize (impending) pathological fractures and to reduce pain.

Adverse prognostic factors include anaemia, hypercalcaemia, renal failure, hyperuricaemia, hypoalbuminaemia, and elevated beta-2 macroglobulin level. The prognosis is highly variable, but overall estimated survival has improved to 35% 5-year survival and 17% 10-year survival with the advent of new treatments.

Primary lymphoma of bone

Primary malignant lymphoma of bone accounts for 5% of all extranodal lymphomas and is distinct from (the more common) nodal lymphoma with secondary bone involvement (16–20% have osseous disease at presentation). Mean age at presentation is 45 years, but it can occur in all age groups, mostly arising in

metadiaphyseal locations of long bones, particularly the femur, pelvis, humerus and tibia. Presenting features include pain, palpable masses and fracture with permeative, osteolytic lesions eroding through the cortex with a soft-tissue mass, without a periosteal reaction (Figure 9.31). Serum LDH may be elevated. Rarely B-cell symptoms such as fever or night-sweats are a feature.

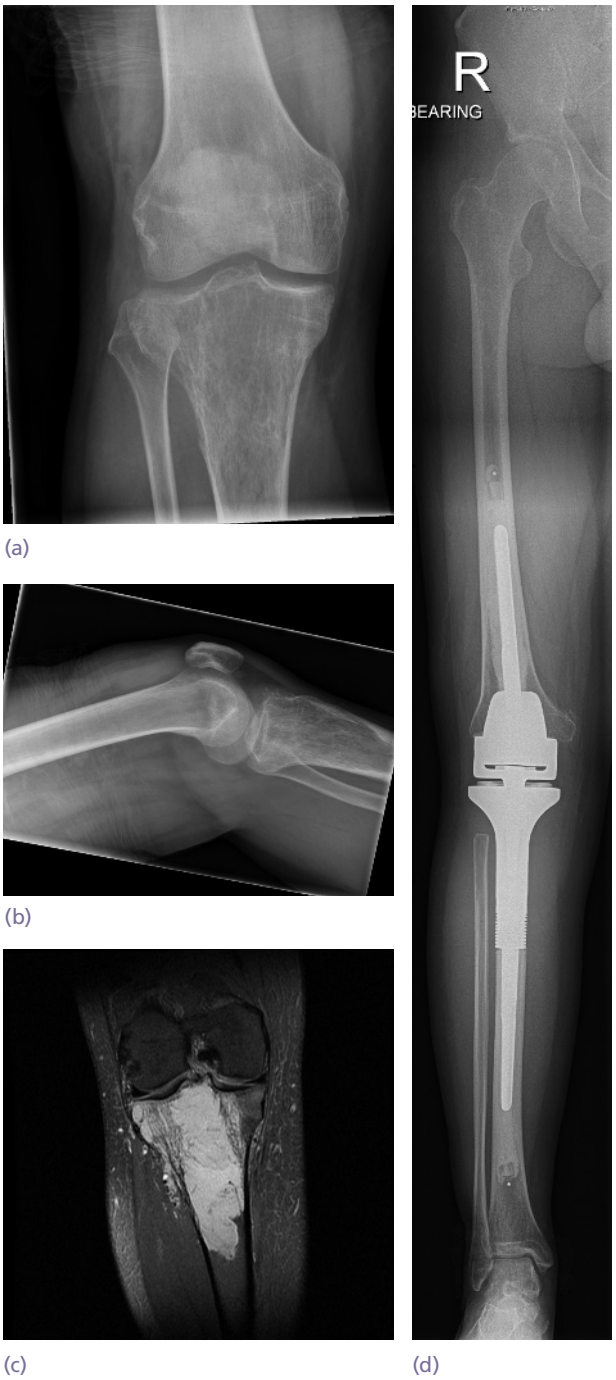
Histologically, this is a small, round-cell tumour of the reticuloendothelial system, although the prominent nuclei are less uniform than Ewing's sarcoma. The majority are intermediate or high-grade and 95% are of B-cell lineage. Distal staging includes imaging of regional and distal lymph nodes, solid viscera and nervous system with CT or PET-CT. Polyostotic disease is not uncommon, and bone scintigraphy shows significant uptake.

The disease is responsive to radiotherapy and chemotherapy. Surgery may be required for pathological fracture stabilization or spinal decompression. Age is the only poor prognosticator; the disease-free 5-year survival for patients with primary lymphoma of bone is 81%, which is more favourable than patients with systemic lymphoma with bone involvement (44% at 5 years).

Chordoma

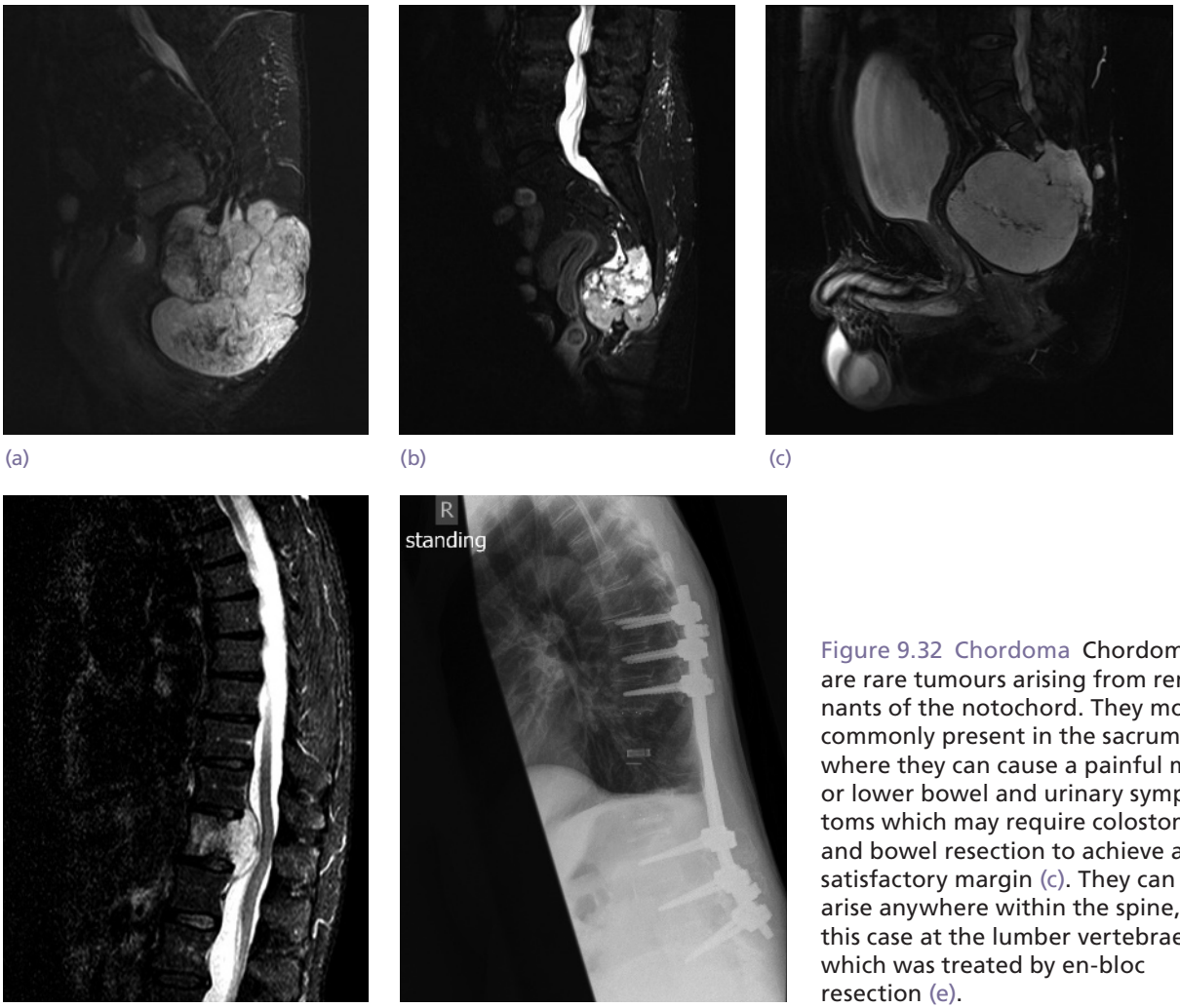
This rare, slow-growing tumour arises from primitive embryonic notochordal remnants along the spinal column, most commonly in the sacrum and less commonly at the occiput or vertebrae. More common in males, the mean age at presentation is 60 years (ranging from the third to the ninth decades). Base-of-skull chordomas present with pain and cranial nerve palsies. Sacral chordoma patients present with typically dull pain (85%), usually present for years, and worse with sitting. The classic symptoms of cauda equina (saddle anaesthesia, bladder or bowel dysfunction) occur in 70% of patients. Sacral chordoma should be considered in cases of back pain with coccydynia, especially with neurological symptoms. The insidious nature of the disease often leads to late presentation and worse prognosis. The local invasion of adjacent neural structures threatens bowel and bladder function loss.

X-rays show ill-defined osteolysis of the sacrum, possibly with tumoural calcifications. Bone scan highlights areas of bone involvement. MRI defines the anatomical level within the sacrum and relationships to the rectum and nerve roots (Figure 9.32). Wide surgical excision is attempted, possibly requiring anterior and posterior surgical approaches plus faecal diversion and vertical rectus abdominal flap



(a) (b) (c) (d)

Figure 9.31 Primary lymphoma of bone A 27-year-old male who presented with a short history of progressive knee pain. (a,b) X-rays confirm an expansile lytic lesion of the right proximal tibia abutting the subchondral plate. (c) MRI demonstrates the true extent of the tumour which, on biopsy, was confirmed as lymphoma. Due to the risk of impending fracture, the tumour was excised in its entirety and reconstructed with an endoprosthetic replacement (d) prior to commencing chemotherapy.



(a) (b) (c) (d) (e)

Figure 9.32 Chordoma Chordomas are rare tumours arising from remnants of the notochord. They most commonly present in the sacrum (a,b) where they can cause a painful mass or lower bowel and urinary symptoms which may require colostomy and bowel resection to achieve a satisfactory margin (c). They can arise anywhere within the spine, in this case at the lumber vertebrae (d) which was treated by en-bloc resection (e).

reconstruction. Bowel and bladder dysfunction relates to the anatomical level of nerve-root resection; navigated resections help to preserve nerve roots. In high sacral chordomas, with predictable surgical morbidity, proton beam and carbon-ion therapies have been used to achieve local control. Surgical margins predict local recurrence (56%) and survival; five-year survival is 82%. Late pulmonary metastasis occurs in up to 9%.

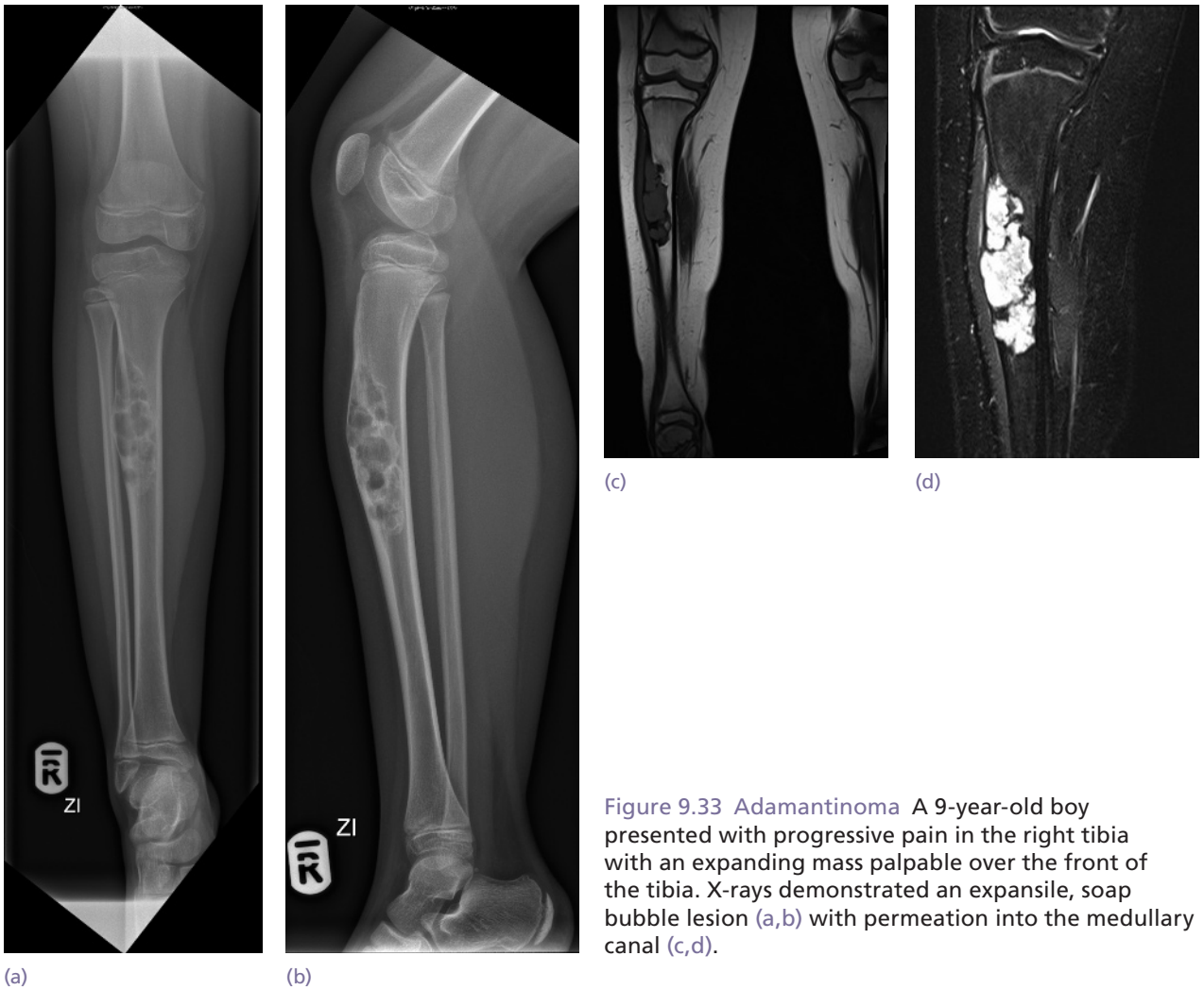
Adamantinoma

This rare, low-grade malignant tumour classically involves the tibial diaphysis in up to 90% of cases. Patients are aged between 20 and 40 years and complain of pre-tibial pain and swelling and occasionally fractures. X-rays show expansion and lobulation of a lucent cystic lesion with anterior cortical disruption (Figure 9.33). This solid, fibrous tumour is regarded as the malignant end of the spectrum of osteofibrous dysplasia (OFD) and OFD-like-adamantinoma which present in children and young adults respectively. Most are low grade but higher-grade areas in the primary tumour may require systemic therapy.

Complete excision is the treatment of choice as local recurrence is common with intralesional or marginal excision, may occur late and is associated with more aggressive histological features. Metastasis may occur in 20% to the lymph nodes, lungs and skeleton. Reconstruction with vascularized autograft or allograft or endoprosthesis are all described, although amputation may need to be considered for recurrence.

Malignant vascular tumours of bone

Vascular tumours arising in bone range from benign haemangiomas, low-grade epithelioid haemangiomas, and intermediate-grade epithelioid haemangioendotheliomas to high-grade angiosarcomas. *Haemangiomas* are common incidental findings of spinal imaging, may be seen in the metaphysis of long bones and are more



(a) (b) (c) (d)

Figure 9.33 Adamantinoma A 9-year-old boy presented with progressive pain in the right tibia with an expanding mass palpable over the front of the tibia. X-rays demonstrated an expansile, soap bubble lesion (a,b) with permeation into the medullary canal (c,d).

common with age. They are well demarcated and usually indolent, requiring no treatment.

Locally aggressive epithelioid haemangiomas are found in young adults in the lower limbs and spine, causing pain, and they may even metastasize. They are well-defined, lytic lesions that can erode the cortex and extend into soft tissues. Intralesional curettage is usually sufficient, and local recurrence is rare.

Epithelioid haemangioendotheliomas are rare, intermediate-grade and present in all age groups in the lower limbs, spine and pelvis causing pain and swelling. X-rays demonstrate expansile, lytic, invasive tumours eroding the cortex. Wide resection is advocated.

High-grade angiosarcomas of bone are very rare. They have a broad age range and wide anatomical distribution, most frequently the femur and pelvis. They can be secondary to previous irradiation of the soft tissues (e.g. breast carcinoma). Significant pain and swelling may be evident and an aggressive, osteolytic lesion with cortical destruction and soft-tissue mass is evident on radiographs and MRI. Wide surgical excision or radiation with chemotherapy is used to attempt disease control. This very aggressive tumour has a 5-year survival of 20%, although this is 0% in patients with metastases.

METASTATIC BONE DISEASE

Metastatic disease is the most common malignancy of bone. It is estimated that 1 in 5 cancer patients will suffer symptomatic bone metastasis, and post-mortem studies have demonstrated skeletal metastasis in 70% of patients. An ageing population combined with an improved survival rate of patients with cancer have led to an increase in the prevalence of osseous metastatic lesions that are symptomatic and require orthopaedic care. Metastasis is defined as follows: 'Transmission of pathogenic micro-organisms or cancerous cells from an original site to one or more sites elsewhere in the body usually via blood vessels or lymphatics.' The most common primary tumours to metastasize to bone are breast, lung, prostate, renal and thyroid

carcinomas, which account for more than 80% of metastatic bone tumours.

In all patients over 40 years of age with suspicious radiological lesions, metastasis should form at least part of the differential because metastases can mimic other bone lesions. In 20% of cases of skeletal metastasis, it will be the first presentation of the disease. Common sites, in order of frequency, are the spine, proximal femur, pelvis, proximal humerus, femur, scapula, distal femur and ribs.

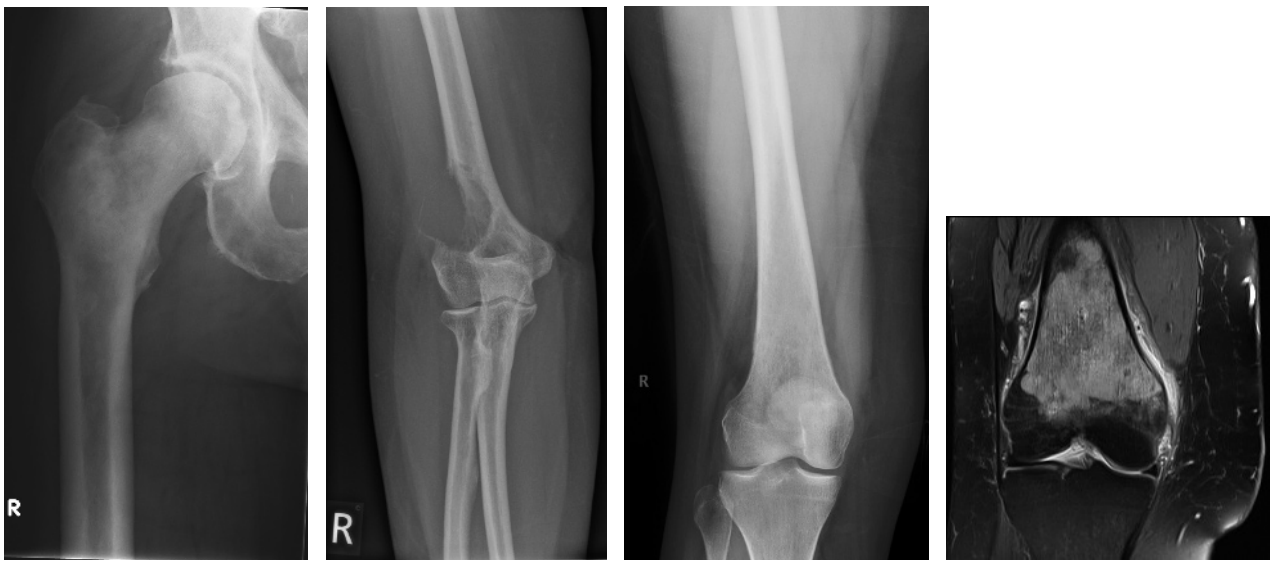
Presenting features include pain, swelling, pathological fracture, spinal cord compression and hypercalcaemia. Symptoms of hypercalcaemia may occur, including anorexia, nausea, thirst, polyuria, abdominal pain, general weakness and depression. In children under 6 years of age, metastatic lesions are most commonly from adrenal neuroblastoma. The child presents with bone pain and fever and an abdominal mass.

Bone metastases result in bone destruction and increased tumour burden. Tumour cells in the bone secrete factors (such as TNFα, IL1, IL6 and PTHrP) which activate osteoclasts responsible for bone resorption. In turn, resorption by osteoclasts releases growth factors from the bone which may stimulate tumour growth.

Typically, prostatic lesions are densely osteosclerotic, in contrast to osteolytic renal and thyroid lesions (Figure 9.34). Lung and breast metastases produce mixed osteoblastic/osteolytic lesions. Routine clinical examination of all systems and screening bloods are required to exclude anaemia, renal failure, hypercalcaemia and screen prostate-specific antigen in males. Local and distal staging includes X-rays and MRI of the whole affected bone, bone scintigraphy and CT thorax, abdomen and pelvis. Bone scintigraphy is widely used to search for further areas of radionucleotide uptake; classically, cold lesions such as renal-cell carcinoma and myeloma do not initiate an osteoblastic response, therefore disease extent may be underestimated. Whole-body MRI has superior sensitivity to scintigraphy for identifying metastatic disease in a range of visceral tumours, particularly renal-cell carcinoma.

Biopsy is indicated in patients with a solitary bone lesion, no known primary cancer, a long disease-free interval following a previous malignancy, or a history of more than one previous invasive cancer. A primary sarcoma must be excluded. Solitary suspicious bone lesions in patients with a prior history of carcinoma have been reported as different pathology in up to 15% of cases. Treatment without making a histological diagnosis should only be made in the presence of known disseminated malignancy by a specialist multidisciplinary team.

The prognosis for many patients with metastatic bone disease, particularly those without visceral disease, has significantly improved due to advances in medical therapy, including hormonal



(a) (b) (c) (d)

Figure 9.34 Metastatic bone disease (a) X-rays of a 75-year-old man with a past history of prostate carcinoma with a progressive, painful right hip demonstrate a sclerotic lesion in the proximal femur which was confirmed as metastatic prostate carcinoma. (b) A 68-year-old woman presented with a rapid deterioration in right elbow function with progressive pain. Radiographs demonstrate a punched-out lesion in the distal humerus with associated soft-tissue component. Histology confirmed metastatic renal cell carcinoma. (c,d) A 71-year-old woman with a past history of breast carcinoma treated 11 years previously represented with progressive pain in the left knee. X-rays demonstrate a mixed blastic and lytic lesion (c), the true nature and extent of which are clearly seen on MRI (d).

treatment, bisphosphonates, chemotherapy and biologically targeted agents. Radiotherapy for metastases is usually delivered in a single fraction by oncologists. Bisphosphonates are useful in metastatic breast and prostate carcinoma and multiple myeloma. Radiofrequency ablation has been shown to be effective, safe and well tolerated by patients.

Surgery is indicated for intractable pain, or impending or pathological fractures. Surgery ranges from osteosynthesis, with or without cement augmentation, to endoprosthetic replacement. The aims of surgery are to provide immediate absolute stability permitting full weight-bearing. Implant failure and re-operation rates in metastatic patients of up to 14% and significant post-surgical complication rates of 20% have been reported, therefore the procedure should assume that pathological fractures do not unite and the fixation/implant should outlive the patient. Inappropriate surgery risks mechanical failure, because the patient outlives the construct, and may hasten death because of the sequelae of surgical intervention (Figure 9.35). Renal and thyroid metastases may be highly vascular lesions necessitating pre-operative embolization to avoid catastrophic haemorrhage.

Mirels' score is commonly quoted to assess risk of fracture in metastatic bone lesions (Table 9.6). A score of 1 to 3 is given for each of four variables. There is a high risk of fracture for total scores of 8 or above, so prophylactic fixation should be carried out prior to radiotherapy.

SOFT-TISSUE TUMOURS

Soft-tissue tumours (STTs) are a heterogeneous group of benign and malignant diseases accounting for <4% of all tumours in adults and <8% of all tumours in children. Soft-tissue sarcomas are rare malignant tumours derived from mesenchymal cells at all body sites. These rare tumours comprise approximately 1% of all newly diagnosed cancers. The incidence of soft-tissue tumours is approximately 2000 cases per annum in the UK. Radiation exposure has long been known to induce sarcomas since the common use of X-rays. Other aetiologies include malignant transformation of neurofibromas in NF-1, Li–Fraumeni syndrome (*p53* mutation) and inherited retinoblastoma mutations.

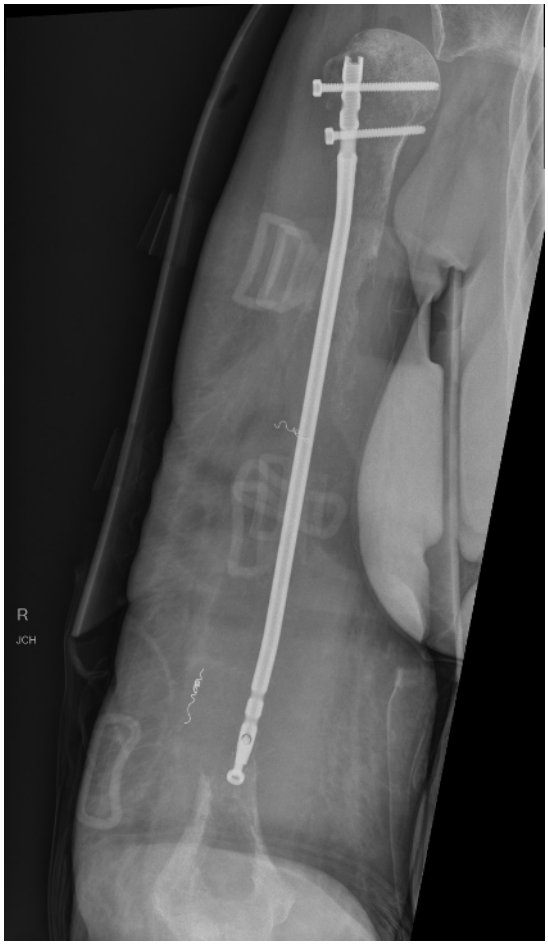


Figure 9.35 Failed fixation of metastatic lesions
This patient with known metastases from a renal cell carcinoma was treated by intramedullary fixation. Due to advances in his oncological management, he survived a significant period. Unfortunately, progression of his metastatic disease resulted in near-complete destruction of his humerus around his intramedullary nail.

Table 9.6 The Mirels' score for quantifying the relative risk of fracture through a pathological lesion of bone

Variable	Score		
	1	2	3
Site	Upper limb	Lower limb	Pertrochanteric
Pain	Mild	Moderate	Severe
Lesion	Blastic	Mixed	Lytic
Size*	<1/3	1/3–2/3	>2/3

* The maximum degree of cortical destruction as defined by the overall width of the bone, as seen on plain radiograph, in any view.

Patients may present with a painful or painless swelling that is growing insidiously. Almost 50% arise in the lower limbs (most commonly the adductor compartment of the thigh) and the median age for presentation is 65 years. There are, however, age-related variations: embryonal rhabdomyosarcomas occur exclusively in children, synovial sarcomas in young adults and liposarcomas in older people. Concern at the biological activity of a lesion should arise if the size is >5 cm, it is painful, deep to fascia, increasing in size, or recurrence of a previously excised lesion. Any lesion presenting with features that are suggestive of a soft-tissue tumour requires referral to a specialist centre, where appropriate evaluation and staging will take place. A clinical assessment and examination are mandated, with attention required to regional lymphadenopathy, distal neurovascular deficit and localized skin changes at the tumour site.

Plain X-rays of soft-tissue lesions are helpful to exclude bone lesions with soft-tissue extension and to assess bone invasion from extra-osseous tumours with a risk of fracture. X-rays should be obtained as the location and presence of any periosteal reaction, erosion (e.g. glomus tumour, pigmented villonodular synovitis (PVNS)) or soft-tissue mineralization (e.g. synovial sarcoma) are useful for characterization. Soft-tissue tumours are more difficult to diagnose specifically from imaging, but there may be characteristics which can help to determine aggressiveness or identifiable tissues such as fat, calcification or haemorrhage. Matrix calcification or ossification may be found in synovial sarcomas and in the rarer mesenchymal chondrosarcomas or soft-tissue osteosarcomas.

MRI sequencing of the affected limb is valuable to define the extent of the lesion and local invasion of critical structures and to characterize the lesion prior to biopsy. If surgery is planned, this will define the margins in relation to neurovascular structures and the involved musculature, joints or tendons. There is extensive overlap of the soft-tissue sarcoma appearances with MRI; the majority have a low signal T1 and heterogeneous high signal T2 appearances. Exceptions to this may include high T1 signals in some liposarcomas and low T2 signal in desmoid fibromatosis and PVNS. The characteristics of particular tumours may be emphasized, such as a serpiginous pattern in vascular tumours, dystrophic calcification in synovial sarcomas or blooming artefact caused by the haemosiderin content of diffuse-type PVNS. MRI should be performed before biopsy as the tissue characteristics may alter. Contrast is reserved for targeting the biologically active areas of tumours for biopsy, although it risks nephrogenic systemic fibrosis in patients with chronic renal impairment.

Systemic staging will require chest CT or PET-CT to identify other biologically active lesions. Bone scintigraphy is not normally required. Systemic staging for soft-tissue sarcomas principally involves chest X-rays and chest CT for pulmonary metastases. In pelvic or retroperitoneal primaries, CT of the abdomen and pelvis would be included; PET-CT is useful to identify other biologically active lesions in neurofibromatosis type 1. Whole-body MRI is reliable in detecting extra-pulmonary metastases in myxoid liposarcomas, which unusually rarely metastasize to the chest.

Lipomatous tumours

Lipomas are the most common soft-tissue tumours encountered in clinical practice, typically presenting in middle to old age but sometimes occurring in children and adolescents. Benign superficial lipomas classically arise in the subcutaneous tissue of the back, shoulder, neck and proximal extremities. Intramuscular lipomas are found within or between muscles and adherent to joints, tendons, bone and nerves. In 5% of cases, lipomas can be multiple and symmetrical across the dorsum and proximal upper limbs.

Examination reveals solitary, soft, painless, mobile and slow-growing lesions. Deep lipomas may be occult and therefore larger at presentation. X-rays may reveal radiolucent swellings, which appear as encapsulated, homogeneous, high signal lesions on T1 MRI sequences with isointense signal to adjacent subcutaneous fat and fully suppress on fluid-sensitive PD sequences. These are avascular lesions with no uptake with angiography, scintigraphy or contrast. After marginal excision, these lobulated yellow tumours demonstrate mature adipoctyes organized in lobules with empty cytoplasm. Excision is curative and recurrences are rare in superficial lipomas but more common with intramuscular lipomas.

Liposarcomas are the most common malignant soft-tissue sarcomas, accounting for 10% of all soft tissue sarcomas typically arising in adults after the third decade. They can arise in any location with fat, but the majority are found deep in the thigh, groin, calf, popliteal fossa and buttock. These insidious lesions can reach large sizes prior to presentation and a few may have pain. Nerve compression or oedema secondary to venous occlusion can occur, particularly if involving the retroperitoneum. MRI sequences reveal heterogeneous and non-specific appearances of high and low signal regions on T2. These tumours can erode adjacent cortical bone, which responds with a modest periosteal reaction. They show diffuse uptake with scintigraphy and angiography shows dense neo-angiogenesis.

Liposarcomas comprise a spectrum of low- to high-grade lesions. Atypical lipomatous tumours are

considered locally aggressive with histological features of cellular atypia (low grade). Atypical lipomatous tumours account for 40% of all liposarcomas and have a recurrence rate of <10% with marginal excision. They are rarely metastatic or lethal (unless arising in retroperitoneal or mediastinal sites where they behave more aggressively). Differentiating lipomas from atypical lipomatous tumours is assisted using cytogenetic murine double minute-2 (MDM2) amplification by fluorescence in situ hybridization (FISH). In studies, the absence of MDM2 amplification has supported the distinction of well-differentiated lipomas.

High-grade subtypes of liposarcoma include myxoid, dedifferentiated (progression to high-grade from an atypical lipomatous tumour) and the rarest pleomorphic liposarcomas. Dedifferentiated liposarcomas occur most commonly in the retroperitoneum. Liposarcomas require wide excision and sometimes radical excision in the higher-grade tumours (Figure 9.36). Radiotherapy is used pre- or postoperatively and chemotherapy may be used. Postoperative radiotherapy is indicated for poor surgical margins. Local recurrence occurs more rapidly and metastases are frequent in these high-grade liposarcomas. Mortality ranges between 30% and 50% at 5 years.

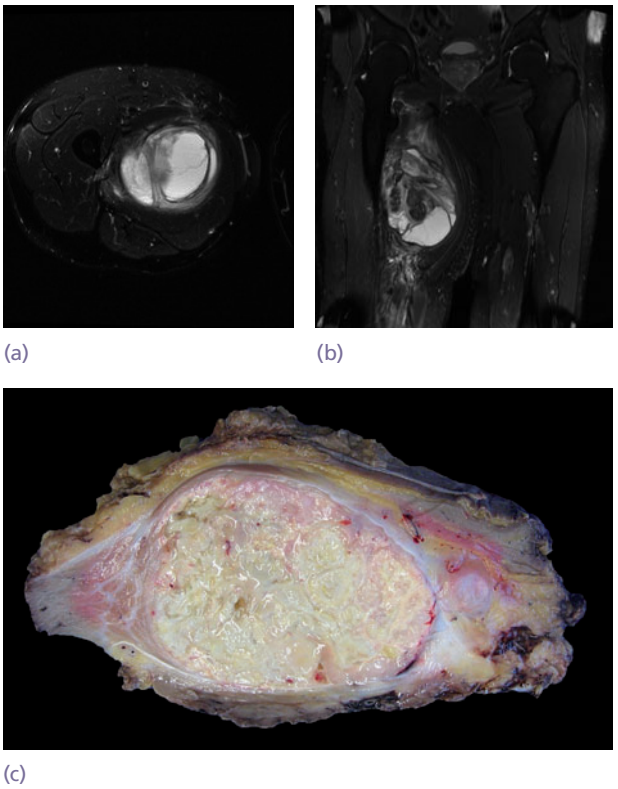


Figure 9.36 High-grade liposarcoma A 68-year-old man who presented with an enlarging mass within the right thigh which rapidly became painful and tethered to the overlying skin. (a) MRI demonstrates a large, heterogeneous mass in the abductor compartment of the right thigh. Biopsy confirmed this to be a high-grade liposarcoma. Due to their potential sensitivity to radiotherapy, preoperative radiotherapy was given. The tumour did not shrink in size but the internal architecture demonstrated a much more cystic, liquefied appearance in keeping with necrosis within the tumour (b). The tumour was removed in its entirety, preserving the nearby vessels and removing with a clear margin (c).

Fibrous tumours

Fibrous tumours also represent a spectrum of disease from benign to high-grade neoplasms with similar microscopic appearances. *Superficial lesions* include Dupuytren's, Ledderhose, Peyronie's and Garrod's pads (Dupuytren's diathesis). They are caused by proliferation of myofibroblasts which secrete collagen and undergo contraction by their interaction with the deposited matrix (similar to their involvement in wound healing). Palmar and plantar fibromatosis causes firm, painless nodules leading to puckering of the skin, fibrous cords and deformity. Many patients have a significant family history and treatment ranges from collagenase injections to dermofasciectomy with a significant risk of recurrence. These do not represent a premalignant condition.

Deep lesions are known as *desmoid-type fibromatosis*, which is a benign, locally aggressive myofibroblastic disorder characterized by infiltrative growth that does not metastasize. It has an association with Gardner-type familial adenomatous polyposis with genetic mutations inactivating the *APC* gene and may manifest as aggressive fibromatosis of the colon; 85% of sporadic mutations contain β-catenin mutations. These insidiously growing and deep masses may arise in any site including the abdominal wall, chest wall, head, neck and mesentery, and they may cause pain, paraesthesia or deformity if adjacent to nerves and joints respectively. Typical appendicular sites include the shoulder and thigh in 60%. CT and MRI are useful to show the extent of this invasive tumour. Histologically these may be difficult to distinguish from malignant tumours. Various treatments exist including non-steroidal anti-inflammatories, radiotherapy, hormone therapy (tamoxifen), chemotherapy and surgery. All are associated with rates of recurrence approaching 50%, and often the recurrences behave more aggressively.

Dermatofibrosarcoma protuberans is a low-grade malignant tumour, more common in males, typically occurring between the third and fifth decades, which arises in the trunk, groin and proximal extremities. These slow-growing, nodular lesions may cause cutaneous erythematous plaques; rapid enlargement may indicate progression to high-grade sarcoma present in 10%. Wide surgical excision may be curative, although the rate of local recurrence may reach 50% and is

associated with incomplete resection margins. Up to 10% may develop distant metastases which are usually high-grade.

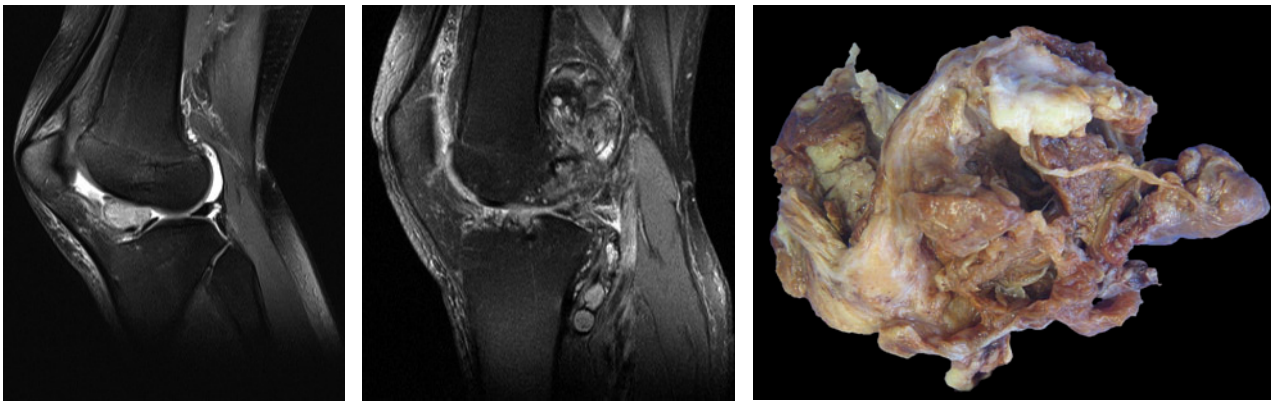
Fibrosarcomas are deep tumours arising in the thigh, trunk, arm and forearm and are most common in adults (mean age 50 years). They present as slowly enlarging masses with and without pain. MRI demonstrates homogeneous, low-signal tumours which may erode into bone. Histologically they reveal a classic 'herringbone' pattern of dark-staining nuclei, but they may contain relatively bland areas mimicking fibromatosis. Greater than 80% are high-grade, necessitating systemic staging and wide excision with adjuvant radiotherapy. Fibrosarcomas metastasize to lung and bone, and chemotherapy may be indicated for systemic treatment.

Myxofibrosarcomas are the more common soft-tissue sarcomas in older patients with a mean age at diagnosis of 60 years, although they can arise in all age groups after skeletal maturity. Most commonly found in the lower limbs, upper limbs and girdles, they are more often subcutaneous than myofascial in origin. They present as painful enlarging masses with infiltrative margins. Low-grade myxofibrosarcomas tend not to metastasize; in intermediate- or high-grade tumours metastases are found in the lungs, bone or lymph nodes. Deeper lesions more commonly metastasize and consequently have a higher mortality rate. Wide excision and radiotherapy are the mainstays of treatment. Overall 5-year survival is approximately 65%.

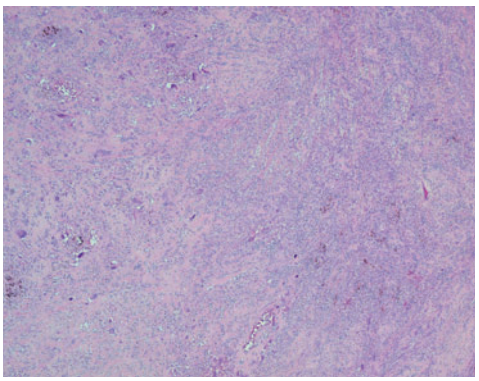
Synovial tumours

Pigmented villonodular synovitis (PVNS) is a rare, benign and aggressive disorder arising from synovial joints and tendon sheaths capable of eroding articular structures and bone. PVNS represents a spectrum of 'fibrohistiocytic' disorders ranging from the most common giant-cell tumour of tendon sheath (GCTTS) to localized and diffuse intra-articular PVNS lesions. The estimated incidence of articular PVNS is 1.8 new cases annually per million population, highlighting the rarity of this disorder. Patients primarily present in the third to fifth decades with swelling and/or pain around a single joint. Articular tumours are found predominantly in the knee, hip and ankle; extra-articular tendon sheath tumours typically involve the digits. Classically the difference in magnetic susceptibility between haemosiderin-laden PVNS and surrounding tissue may cause the lesion to appear larger ('blooming') on gradient echo images (Figure 9.37).

Surgical excision is the gold standard therapy to control symptoms and prevent further joint erosion,



(a) (b) (c)



(d)

Figure 9.37 PVNS (a) MRI of the stiff, painful knee of a 16-year-old girl shows an area of focal PVNS sitting within the notch of the knee. This was removed through an open arthrotomy and the patient regained a full range of movement. (b) In the case of a 66-year-old man who presented with worsening knee pain and a large, painful joint effusion, MRI demonstrates a large synovial proliferation involving the anterior and posterior compartments of the knee with evidence of haemorrhage and secondary joint destruction. The features are in keeping with diffuse PVNS. (c) The typical macroscopic appearances of PVNS showing areas of synovial proliferation with evidence of haemorrhage. (d) Histology demonstrates mononuclear stromal cells infiltrating the synovium with lipid laden histiocytes and haemosiderin stained multinucleated giant cells (x100).

but high recurrence rates are related to incomplete excision. Joint erosion is particularly associated with the diffuse articular form, necessitating arthroplasty in the hip and the knee. Adjunctive therapies include injection of intra-articular radiation materials and external beam radiotherapy. Alternative therapies such as tyrosine kinase inhibitors (e.g. Imatinib) have demonstrated efficacy in recurrent cases although the lesions tend to recur if the therapy stops.

Synovial sarcomas are histologically and genetically separate tumours arising in any location, but they do not arise from synovium, though the histological appearance may resemble synovium. Usually found adjacent to tendons, joint capsules, fascia and muscles, 80% arise in extremities. They have a specific chromosomal translocation *t(X:18)(p11:q11)*. They can occur in all age groups (mean 40 years) with equal sex distribution. Pain is a common feature and many lesions are often present for years before they are diagnosed. Radiographically, multiple small calcifications may be present within the periphery of the tumour. A heterogeneous high signal lesion with septations and fluid-fluid levels may be seen on MRI.

Wide excision and radiotherapy are required but improved survival has been found with chemotherapy. Due to the often ill-defined borders of these tumours, frequently radical resections or amputations are necessary. Five-year survival is in the range of 40–75% although very late metastasis can occur (e.g. 30 years).

Vascular and smooth muscle tumours

Haemangiomas are benign vascular tumours usually seen during childhood but which may be present at birth. They occur in two forms. The *capillary haemangioma* is more common; it usually appears as a reddish patch on the skin, and the congenital naevus or 'birthmark' is a familiar example. A *cavernous haemangioma* consists of a sponge-like collection of blood spaces; superficial lesions appear as blue or purple skin patches, sometimes overlying a soft subcutaneous mass; deep lesions may extend into the fascia or muscles, and occasionally an entire limb is involved.

X-rays may show calcified phleboliths in the cavernous lesions. There is no risk of malignant change and treatment is needed only if there is significant discomfort or disability. Local excision carries a high risk of recurrence, but more radical procedures seem unnecessarily destructive. Embolization of feeding vessels may reduce the symptoms and size of the lesion.

Glomus tumour is a rare mesenchymal perivascular tumour usually occurring around fine peripheral neurovascular structures, particularly the nail beds of fingers or toes in young adults. These small, pea-sized blue nodules cause recurrent episodes of pain in the fingertip that are worse with cold temperatures. X-rays may show erosion of the underlying phalanx. Surgical excision from the fibrous capsule surrounding it is usually successful.

Angiosarcoma of soft tissue is a rare, malignant tumour of cells, morphologically similar to normal endothelium, the majority of which develop as cutaneous lesions. Often they are associated with previous radiotherapy and chronic lymphoedematous tissue (e.g. breast carcinoma). They may also occur adjacent to synthetic or foreign material, adjacent to arterio-venous fistulae and in Maffucci syndrome. They usually arise in the deep muscles of the thigh, calf, arm and trunk as an enlarging mass. Associated symptoms may include coagulopathy, anaemia, haematoma or bruising.

Although X-rays are usually normal, MRI demonstrates a serpentine lesion described as a 'bunch of grapes'. Fluid-fluid levels may be appreciated between fibrous septae. Histologically high-grade, complete wide excision is required as local recurrence is 20% and first-year mortality approaches 50% due to metastasis to distant bones, soft tissues and lymph nodes.

Leiomyosarcoma is a malignant tumour of spindle cells originating from smooth muscle, which can occur in retroperitoneal, cutaneous and vascular locations. It is the predominant sarcoma arising from larger blood vessels, most commonly the vena cava, iliac and femoral veins, causing occlusion and limb swelling. Typically arising after the fourth decade and peaking at 70 years, the retroperitoneal tumours can involve solid organs and the vertebral bodies. They present with abdominal masses, pain, weight loss, nausea and vomiting. Angiography or duplex ultrasound imaging demonstrates highly vascularized lesions, highlighting the need for vascular reconstruction after resection**.** These highly aggressive tumours are frequently not resectable and metastasize to lung, bone and soft tissue; consequently, survival is 25% at 5 years. Leiomyosarcoma is the commonest sarcoma giving rise to metastases of the skin.

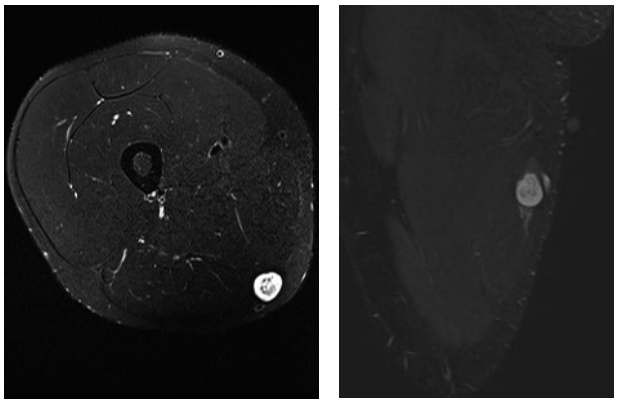
Nerve-sheath tumours

A *neuroma* is not a tumour but an overgrowth of fibrous tissue and randomly sprouting nerve fibrils following injury to a nerve. It is often tender, and local percussion may induce distal paraesthesia, indicating the level of the lesion (Tinel's sign). Treatment can be frustrating. If the neuroma is excised (or as a prophylactic measure during amputation), the epineural sleeve can be freed from the nerve fascicles and sealed with a synthetic tissue adhesive or buried into muscle or bone.

A *schwannoma* is a benign tumour of the nerve sheath. It is seen in the peripheral nerves and in the

spinal nerve roots. The patient complains of pain or paraesthesia; sometimes there is a small palpable swelling along the course of the nerve. MRI demonstrates a homogeneous encapsulated lesion with a 'target sign' (Figure 9.38). With careful dissection the tumour can be shelled out from its capsule without damage to the nerve.

Neurofibromas are benign tumours of the peripheral nerve sheath. They may be solitary (90%) or multiple in neurofibromatosis type 1 (NF1) and are sometimes associated with skeletal abnormalities (scoliosis, pseudarthrosis of the tibia) or overgrowth of a digit or an entire limb, in which there is no obvious neural pathology. These painless nodules can arise in all age groups, usually superficial subcutaneous growths of the lower limbs but other forms may include dumbbell foraminal tumours or plexiform neurofibromas. In contrast to schwannomas, neurofibromas are poorly defined lesions consisting of bland spindle cells. If a nerve root is involved, symptoms can mimic those of a disc prolapse; X-rays may show erosion of a vertebral pedicle or enlargement of the intervertebral foramen. MRI may demonstrate homogeneous high signal on T1 with a 'target-sign' appearance for plexiform tumours. Malignant transformation is rare in solitary neurofibromas but occurs in up to a third of patients with neurofibromatosis. Large painful lesions are excised with marginal margins.



(a) (b)

Figure 9.38 Schwannoma This patient presented with an enlarging mass in the postero-medial aspect of the right thigh. On examination, the lesion was mobile but Tinel's sign resulted in dyasthesia in a dermatomal distribution distal to the lesion. MRI scan demonstrates a well-circumscribed high signal lesion which on axial sections (a) demonstrates a characteristic 'target' sign. The sagittal sections (b) demonstrate the nerve entering above and exiting below the lesion. Resection histology confirmed a benign peripheral nerve-sheath tumour, a schwannoma.

Malignant peripheral nerve-sheath tumours (MPNSTs) arise sporadically from a peripheral nerve or neurofibroma (50%) and account for up to 5% of malignant soft-tissue sarcomas. They present with rapidly enlarging masses plus neurological symptoms including numbness, paraesthesia and weakness. The sporadic MPNSTs typically occur in major nerves of the thigh, brachial plexus and pelvis and may arise in all age groups. Most NF1-associated tumours arise in plexiform neurofibromas in approximately 2–5% of NF1 patients; these have a 10-year mortality of 40%. FDG-PET imaging is sensitive at identifying neurofibromas that have undergone malignant change.

Muscle tumours

Rhabdomyosarcoma is the most common soft-tissue sarcoma arising in children and adolescents (although it can arise in all age groups) and is composed of malignant striated muscle cells. More common in the lower limbs, the most common embryonal subtype displays the rapid, infiltrative growth of an aggressive tumour. Frequently bound to bone, radiographic erosion of bone may be seen, although there may be minimal periosteal reaction.

Treatment comprises neoadjuvant chemotherapy followed by wide excision including regional lymph nodes, chemotherapy and radiation if incompletely excised. The entire muscle from origin to insertion must be excised. Five-year survival ranges from 80% if completely excised to 20% if lung metastases are present at diagnosis.

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Neuromuscular disorders

10

Deborah Eastwood

NERVES AND MUSCLES

Neurons

The neuron (Figure 10.1) is the defining unit of the nervous system. It is a specialized cell, capable of electrical excitation and conduction of electrochemical impulses (*action potentials*) along its thread-like extensions. Its basic structure consists of a cell body, 5–25 μm in diameter, with branching processes, *dendrites*, that are capable of receiving signals from other neuronal terminals. The *axon*, a finer, longer branch, carries the action potentials along its length to or from excitable target organs. Further signal transmission to the dendrites of another neuron, or neuro-excitable tissue like muscle, occurs at a *synapse* where the axon terminal releases a chemical neurotransmitter, typically acetylcholine.

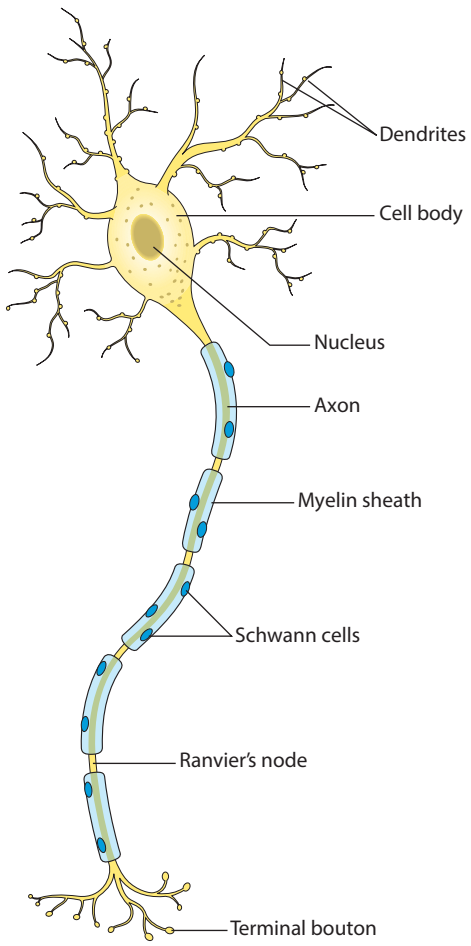


Figure 10.1 Diagram of a typical neuron

All motor axons and the larger sensory axons serving touch, pain and proprioception are covered by a sheath, *the neurilemma,* and coated with *myelin*, a multilayered lipoprotein substance derived from the accompanying Schwann cells (or oligodendrocytes in the central nervous system). Every few millimetres the myelin sheath is interrupted, leaving short segments of bare axon called the *nodes of Ranvier*. In these nerves the myelin coating serves as an insulator, which allows the impulse to be propagated by electromagnetic conduction from node to node, much faster than is the case in unmyelinated nerves. Consequently, depletion of the myelin sheath, as in multiple sclerosis, causes slowing of axonal conduction and eventually a complete block to conduction.

Most axons, in particular the small-diameter fibres carrying crude sensation and efferent sympathetic fibres, are not myelinated but wrapped in Schwann cell cytoplasm. Damage to these axons causes unpleasant or bizarre sensations and abnormal sudomotor and vasomotor effects.

Nervous pathways

Anatomically, neurological structures can be divided into the *central nervous system* (the CNS, comprising the brain and tracts of the spinal cord) and the *peripheral nervous system* (PNS) which includes the cranial and spinal nerves. In terms of physiological function, both the CNS and the PNS have a somatic component and an autonomic component.

The *somatic nervous system* provides efferent motor and afferent sensory pathways to and from peripheral parts of the body serving, respectively, voluntary muscle contraction and sensibility. The *autonomic system* controls involuntary reflex and homeostatic activities of the cardiovascular system, visceral organs and glands. Its two components, sympathetic and parasympathetic divisions, serve more or less opposing functions.

Somatic motor system

Efferent impulses are conducted along axons in the corticospinal or pyramidal tracts (upper motor neurons – UMNs) and along peripheral nerves from cell bodies in the anterior horn of the spinal cord to striated muscle fibres (lower motor neurons – LMNs) (Figure 10.2). The terminal synapses are situated at the neuromuscular junctions. Each large α-motor neuron innervates from a few to several hundred muscle fibres (together forming a *motor unit*) and stimulates muscle fibre contraction. In large muscles of the lower limb, power is adjusted by recruiting more or fewer motor units. Smaller γ-motor neurons connect to sensors (muscle spindles) that control proprioceptive feedback from muscle fibres.

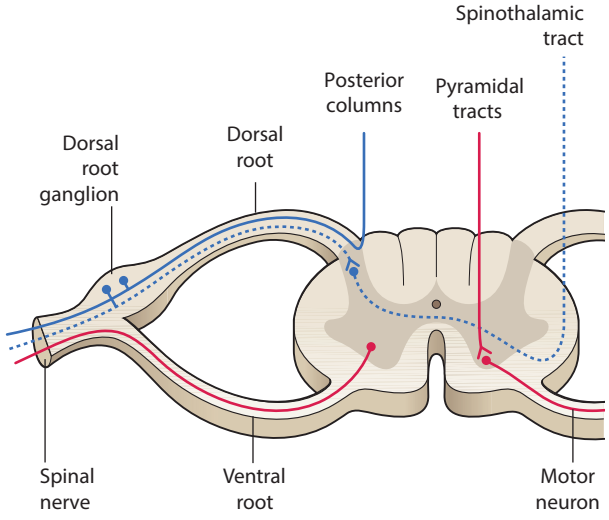


Figure 10.2 Main nerve pathways Simplified diagram showing the main neurological pathways to and from a typical thoracic spinal cord segment. Fibres carrying touch, sharp pain and temperature impulses (-------) decussate, in some cases over several spinal segments, and ascend in the contralateral spinothalamic tracts; those carrying vibration and proprioceptive impulses (——) enter the ipsilateral posterior columns. Motor neurons (——) arise in the anterior horn of the grey matter and innervate ipsilateral muscles.

Somatic sensory system

Axons conveying afferent impulses from receptors in the skin and other peripheral structures enter the dorsal nerve roots, with their cell bodies in the dorsal root (or cranial nerve) ganglia, and end in synapses within the central nervous system. Myelinated fibres carrying sensory stimuli from touch, pressure, pain and temperature (*exteroceptive sensation*) decussate and enter the contralateral spinothalamic tracts running up the spinal cord to the brain. Fibres from sensors in the joints, ligaments, tendons and muscle carrying the sense of movement and bodily position in space (*proprioceptive sensation*) join the ipsilateral posterior columns in the spinal cord.

Sensory areas (dermatomes) corresponding to the spinal nerve roots are shown in Figure 10.3. However, it should be remembered that there is considerable overlap of the boundaries shown in these body maps;

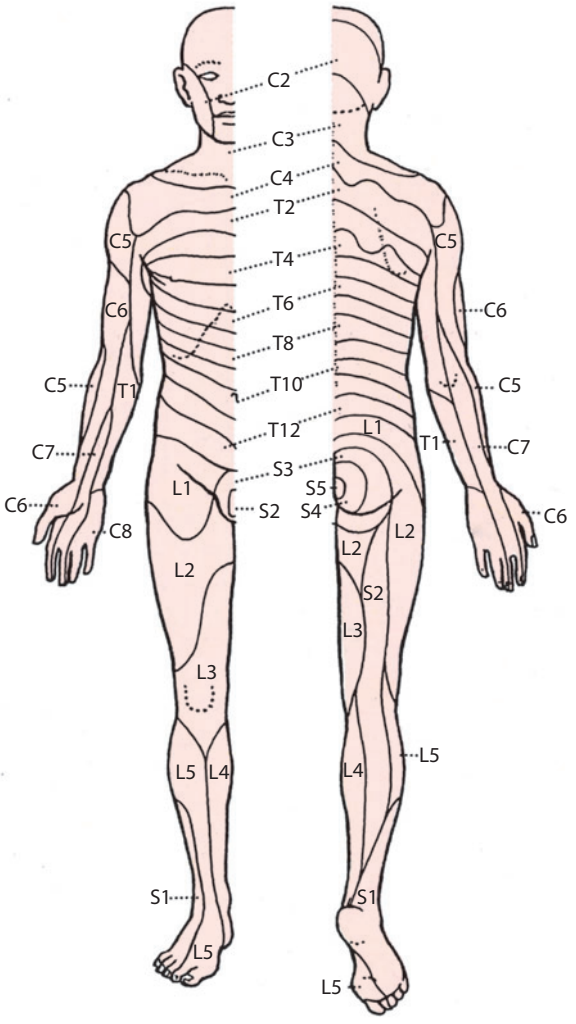


Figure 10.3 Body map The anterior (on the left) and posterior (on the right) dermatomal supply for each spinal nerve root.

furthermore, some parts, such as the hands and lips, are more sensitive and discriminatory than others.

Reflex activity and tone

Sudden stretching of a muscle (e.g. by tapping sharply over the tendon) induces an involuntary muscle contraction, *the stretch reflex*. The sharp change in muscle fibre length is detected by the muscle spindle; the impulse is transmitted rapidly along myelinated afferent neurons which synapse directly with the corresponding segmental α-motor neurons in the spinal cord, triggering efferent signals which stimulate the muscle to contract. This is the basis of the familiar clinical tests for tendon reflexes, and it is also the mechanism for maintaining normal *muscle tone*.

Segmental reflex activity is normally regulated by motor impulses passing from the brain down the spinal cord. Interruption of the UMN pathways results in undamped reflex muscle contraction (clinically hyperactive tendon reflexes) and spastic paralysis. Damage to either afferent or efferent neurons in the reflex arc causes hypotonia; interruption of the LMN pathway results in flaccid LMN paralysis.

Autonomic system

The autonomic system is involved with the regulation of involuntary activities of cardiac muscle and smooth (unstriated) muscle of the lungs, gastrointestinal tract, kidneys, bladder, genital organs, sweat glands and small blood vessels, with afferent (sensory) and efferent (motor) pathways constituting a continuously active reflex arc modulated a little by input from higher centres. In addition, afferent fibres also convey visceral pain sensation.

The system is divided into *sympathetic (thoracolumbar outflow)* and *parasympathetic (craniosacral outflow)* pathways, both of which also contain efferent inputs.

Preganglionic *sympathetic neurons* leave the spinal cord with the ventral nerve roots at all levels from T1 to L1, enter the paravertebral sympathetic chain of ganglia and synapse with postganglionic neurons that spread out to all parts of the body; they may also run up or down the sympathetic chain to synapse in other ganglia or pass on to become splanchnic nerves. Important functions are the reflex control of heart rate, blood flow and sweating, as well as other responses associated with conditions of 'fight and flight'.

Parasympathetic neurons originating in the brain stem leave the CNS with cranial nerves III, VII, IX and X. There are also neurons which leave with the nerve roots of S2, 3 and 4 to reach ganglia where they synapse with postganglionic neurons close to their target organs.

Peripheral nerves

Peripheral nerves are bundles of axons conducting efferent (motor) impulses from cells in the anterior horn of the spinal cord to the muscles, and afferent (sensory) impulses from peripheral receptors via cell bodies in the dorsal root ganglia to the cord (Figure 10.4). They also convey sudomotor and vasomotor fibres from ganglion cells in the sympathetic chain. Some nerves are predominantly motor, some predominantly sensory. The larger trunks are mixed, with motor and sensory axons running in separate bundles. Detailed peripheral nerve structure is described in Chapter 11.

Skeletal muscle

Each skeletal muscle belly, held within a connective tissue *epimysium*, consists of thousands of muscle fibres, separated into bundles (or *fascicles*). Each fascicle is surrounded by a flimsy *perimysium* which envelops anything up to about 100 muscle fibres (Figure 10.5). Large muscles concerned with mass movement, such as the glutei or quadriceps, have a large number of fibres in each fascicle, while muscles used for precision

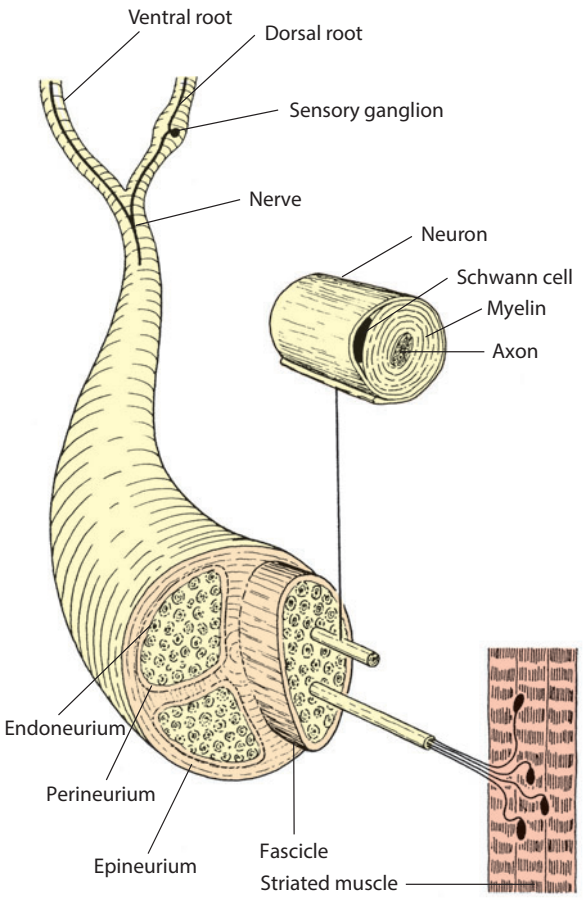


Figure 10.4 Nerve structure Diagram of the structural elements of a peripheral nerve.

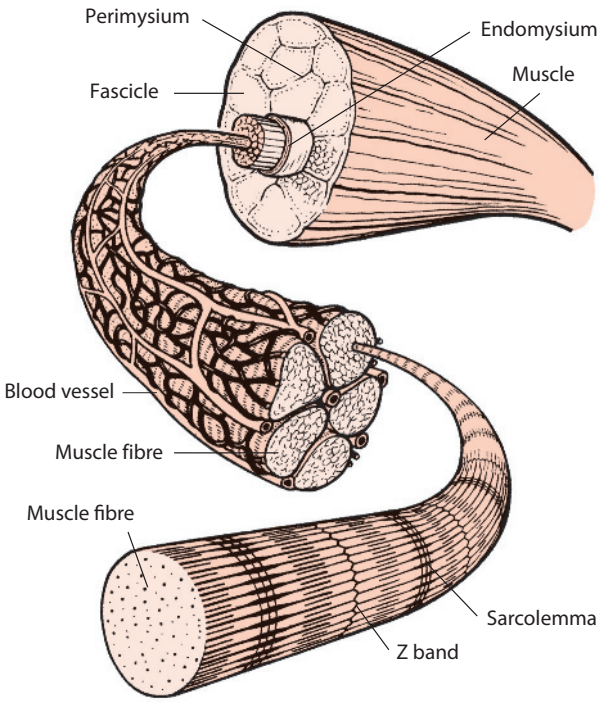


Figure 10.5 Muscle structure Diagram showing the structural elements of striated muscle.

movements (such as those of the hand) have a much smaller number in each bundle.

The muscle fibre is the important unit of all striated muscle. Lying in a barely discernable connective tissue cover, or *endomysium*, it is in actuality a single cell with a cell membrane (the *sarcolemma*), a type of cytoplasm (or sarcoplasm), mitochondria and many thousands of nuclei; its diameter is about 10 µm at birth and 60–80 µm in mature adults.

The fibre itself consists of many tiny (1 µm diameter) *myofibrils*, each of which is striated: dark bands consisting of thick myosin filaments alternate with light bands of thin actin filaments (A and I bands respectively). In the middle of each A band is a lighter H zone and in the middle of the I band there is a dark, thin Z line. The portion of the myofibril between two Z lines is the sarcomere, representing a single contractile unit. The α-motor neuron and the group of muscle fibres it supplies constitute a single motor unit; the number of muscle fibres in the unit may be less than five in muscles concerned with fine manipulatory movements or more than 100 in those employed in gross power movements.

Muscle fibres are also of different types, which can be distinguished by histochemical staining. *Type I fibres* contract slowly and are not easily fatigued; their prime function is postural control. *Type II fibres* are fast-contracting but they fatigue rapidly; hence they are ideally suited to intense activities of short duration. All muscles consist of a mixture of fibre types, the balance depending on anatomical site, basic muscle function, degree of training, genetic disposition and response to previous injury or illness. Long-distance runners have a greater proportion of type I fibres than the average in age- and sex-matched individuals.

Muscle contraction is a complex activity. Individual myofibrils respond to electrical stimuli in much the same way as do motor neurons. However, muscle fibres, and the muscle as a whole, are activated by overlap and summation of contractile responses. When the fibres contract, internal tension in the muscle increases. In *isometric contraction* (against resistance) there is increased tension without actual shortening of the muscle or movement of the joint controlled by that muscle. In *isotonic contraction* the tension within the muscle fibres remains constant; force is generated by changing the length of the muscle: the contraction can therefore be concentric or eccentric.

Muscle tone is the state of tension in a resting muscle when it is passively stretched; characteristically tone is increased in upper motor neuron (UMN) lesions (spastic paralysis) and decreased in lower motor neuron (LMN) lesions (flaccid paralysis).

Muscle contracture (as distinct from contraction) is the adaptive change which occurs when a normally innervated muscle is held immobile in a shortened position for some length of time. If a joint is held flexed for a long time, it may subsequently be impossible to straighten it passively without injuring the muscle. Active exercise will eventually overcome the muscle contracture, unless the muscle has been permanently damaged.

Muscle wasting follows either disuse or denervation; in the former, the fibres are intact but thinner; in the latter, they degenerate and are replaced by fibrous tissue or fat.

Muscle fasciculation is a local involuntary muscle contraction, or twitch, of a small bundle of muscle fibres. It is usually benign but can be due to motor neuron disease or dysfunction.

Clonus is also a series of involuntary but rhythmic muscular contractions (and relaxations) usually triggered by the stretch reflex but, unlike fasciculations, it results in relatively large movements and is often indicative of a UMN lesion.

CLINICAL ASSESSMENT

History

Age at presentation is important. Certain congenital or syndromic neuromuscular disorders are obvious at birth (e.g. spina bifida and arthrogryposis).

Others, while undoubtedly caused by perinatal problems, may not actually manifest themselves until later in childhood; cerebral palsy is the prime example. Conditions such as poliomyelitis may affect anyone although children are most commonly afflicted. In contrast, spinal cord lesions and peripheral neuropathies are more common in adults. The orthopaedic surgeon must be ready to diagnose and treat neuromuscular disease throughout life.

Past medical history may be relevant in terms of previous trauma (accidental or surgical), previous illnesses and their treatment (chemotherapy).

Muscle weakness may be due to upper or lower motor neuron lesions (spastic versus flaccid paralysis) but it may also be due to a primary muscle problem (Table 10.1). The type and degree of weakness, the rate of onset, whether it affects part of a limb, a whole limb, upper or lower limb, one side of the body or both sides are all details which may help give an insight into the aetiology.

Table 10.1 Nerve root supply and actions of main muscle groups

Muscles / Muscle action	Nerve root supply
Sternomastoids	Spinal accessory C2, 3, 4
Trapezius	Spinal accessory C3, 4
Diaphragm	C3, 4, 5
Deltoid	C5, 6
Supra- and infraspinatus	C5, 6
Serratus anterior	C5, 6, 7
Pectoralis major	C5, 6, 7, 8
Elbow flexion extension	C5, 6 C7
Supination	C5, 6
Pronation	C6
Wrist extension flexion	C6, (7) C7, (8)
Finger extension flexion ab- and adduction	C7 C7, 8, T1 C8, T1
Hip flexion extension adduction abduction	L1, 2, 3 L5, S1 L2, 3, 4 L4, 5, S1
Knee extension flexion	L(2), 3, 4 L5, S1
Ankle dorsiflexion plantarflexion inversion eversion	L4, 5 S1, 2 L4, 5 L5, S1
Toe extension flexion abduction	L5 S1 S1, 2

Numbness and paraesthesia may be the main complaints. Dysaesthesia may also be present. It is important to establish the exact distribution of such symptoms to help localize the anatomical nature and level of the lesion accurately (see Figure 10.3). The rate of onset and the relationship to posture may, similarly, suggest the cause. A history of trauma, including recent surgical procedures, and/or the use of a tourniquet must be noted.

Deformity is a common complaint in long-standing disorders. It arises as a result of muscle imbalances that may be very subtle and the deformity (such as 'claw toes' or loss of ankle dorsiflexion) may not be recognized until it is pointed out to the patient.

Non-orthopaedic problems should also be discussed. It can be particularly important to note 'throw-away' comments regarding problems such as headaches, dizziness, falls, feeding problems, hearing difficulties or visual disturbances in addition to the more obvious complaints of cognitive impairment, speech disorders or incontinence. Some symptoms will only be disclosed on direct questioning as the patient may not consider them relevant; other symptoms, such as incontinence or impotence, may be too embarrassing to mention. Symptoms may also have been present for so long that they are considered to be 'normal'.

Family history may reveal clues to the underlying aetiology of the patient's symptoms.

Examination

Neurological examination is described in Chapter 1. Particular attention should be paid to the patient's mental state, natural posture, gait, sense of balance, involuntary movements, muscle wasting, muscle tone and power, reflexes, skin changes, the various modes of sensibility and autonomic functions such as sphincter control, peripheral blood flow and sweating. *The back* should always be carefully examined as it holds the key to many causes of neurological disorder.

GAIT AND POSTURE

A single gait cycle consists of a stance phase (60%) and a swing phase (40%) and each full cycle represents the stride length which is often measured from initial contact at the start of stance to the next initial contact of the same foot. It represents the summation of the length of a right and a left step length. Many parameters of each phase at each joint and in all three planes (coronal, sagittal and transverse) can be analysed, often using a *computerized gait analysis* facility. However, much can be learnt by carefully studying the way the patient walks and moves; clinical gait analysis improves with experience and, these days, simple video analysis can be performed easily by using a mobile phone (or similar device) and playing it

back in slow motion. Distinctive movement patterns can be recognized:

- *Antalgic gait* – A markedly shortened stance phase on one side. Pain makes the patient move off the affected limb as quickly as possible. The step length may be short.
- *Scissoring gait* – A stiff-legged gait with the legs crossing each other is often associated with the muscle imbalance found in cerebral palsy. Often, there is also a crouched posture with flexed hips and knees, feet that are in equinus and both limbs internally rotated.
- *Drop-foot gait* – During swing, there is no 'pick up' of the foot so it effectively 'drops' into equinus; if the foot was not lifted higher than usual to accommodate this, the toes would drag along the floor. This is caused by disorder or damage to the peripheral nerves supplying the foot dorsiflexors.
- *High-stepping gait* – This could be due to a bilateral foot drop or it may signify problems with balance or proprioception.
- *Waddling (Trendelenburg) gait* – The trunk is thrown from side to side with each step. The mechanics are similar to those that produce a positive Trendelenburg test as seen in patients with functionally weak abductor muscles of the hip, perhaps due to dislocation or simply pain.
- *Ataxic gait* – Ataxia produces a more obvious and irregular loss of balance, which is compensated for by a road-based gait, or sometimes uncontrollable staggering.
- *Dystonia* – This term refers to abnormal posturing (focal or generalized) that may affect any part of the body and is often aggravated when the patient is concentrating on a particular motor task such as walking.

MOTOR POWER AND TONE

It is important to examine not only individual muscles but also functional groups. In flaccid paralysis, grading muscle power is important; in spastic paralysis, the spasticity often obscures the inherent weakness and testing specific muscles can be difficult due to the patient's inability to isolate individual movements. Muscle power is usually graded as shown in Table 10.2. Repeated muscle charting allows an objective measure of progressive disease or recovery to be documented.

WEAKNESS

When patients complain of 'weakness', they often fail to distinguish between true loss of muscle power and difficulties due to pain or instability. When testing for muscle power, it is essential to address individual muscles and muscle groups as well as mass movements.

Table 10.2 MRC grading of muscle power

Grade	Description
0	No muscle action – total paralysis
1	Minimal muscle contraction
2	Power insufficient to overcome gravity
3	Anti-gravity muscle power
4	Less than full power
5	Full power

Different patterns of weakness will be encountered. Weakness may be partial (*paresis*) or complete (*paralysis*) although the terms are sometimes used interchangeably.

- *Monoplegia* (weakness of one limb) is usually indicative of a lower motor neuron defect, most commonly a peripheral nerve or nerve root; the movements affected on clinical testing will suggest the likely anatomical location. However, if only the lower limb is affected, the lesion could be in the distal part of the spinal cord.
- *Hemiparesis or hemiplegia* (weakness of either the right or the left side of the body) usually denotes pathology somewhere between the cerebral cortex and the cervical segment of the spinal cord; this will be an upper motor neuron (spastic) type of weakness.
- *Diplegia* (weakness in both upper or both lower limbs) can be due to either UMN or LMN disorder but it is a term classically associated with the UMN lesions seen in cerebral palsy. In some cases the apparently unaffected limbs may show minimal degrees of weakness which could easily be missed.
- *Quadriplegia* (all four limbs affected) could be due to either UMN or LMN pathology, e.g. cerebral palsy, high spinal cord damage or anterior horn cell pathology such as poliomyelitis.

DEFORMITY

In *unbalanced paralysis*, one group of muscles is too weak to balance the pull of the antagonists. At first this produces a deformity that can be corrected passively (*dynamic deformity*); over time the active muscles and the soft tissues of the joints contract and the deformity becomes *fixed* or *structural*.

In *balanced paralysis*, the joint assumes the position imposed on it by gravity and it may feel floppy or flail. In a dynamic deformity, rebalancing of the muscle forces may be possible with a tendon transfer. If the deformity is fixed, soft-tissue releases, and possibly osteotomies, may be needed to correct the deformity before rebalancing can be considered.

Paralysis occurring in childhood seriously affects growth. Bones are thinner and shorter than usual and in the absence of normal mechanical stresses

(imposed by normal muscle pull) bone modelling can be defective.

SENSATION

All sensory modalities must be tested over all dermatomes. Any sensory disturbance must be mapped to see if it fits a particular pattern: dermatomal, distribution of a peripheral nerve or 'glove and stocking'.

AUTONOMIC SYSTEM

A basic assessment of autonomic nervous system function is useful: colour, warmth and sudomotor function can be assessed quickly and easily.

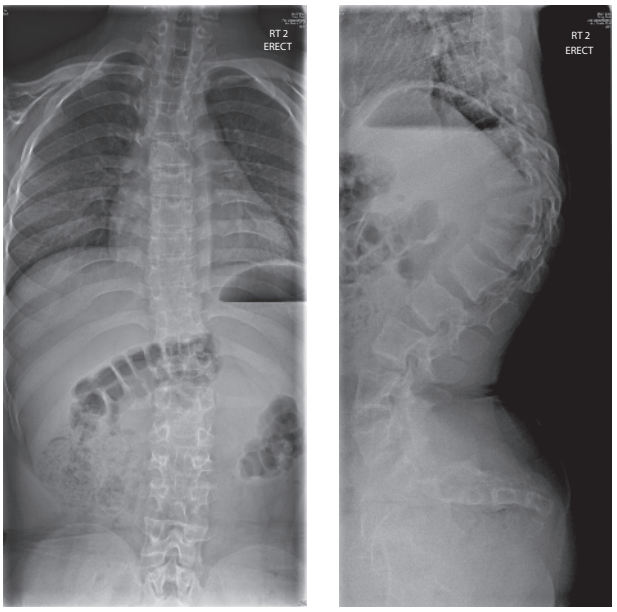
Imaging

Globally plain X-rays of the skull and/or spine may be considered routine in the assessment of disorders of the CNS, but in many centres MRI is the imaging modality of choice.

Spinal imaging is usually directed at identifying cord anatomy, the site and causes of compression of the cord or the nerve roots. *Fractures and dislocations* usually show on the plain X-rays but a CT scan will reveal the exact relationship of bone fragments to nerve structures. A *prolapsed intervertebral disc* is usually diagnosed on clinical examination, but MRI will help to establish the extent of the lesion and its exact site. *Narrowing of the spinal canal* is best demonstrated by CT or MRI; the commonest cause is osteophytic overgrowth following disc degeneration and osteoarthritis of the facet joints. This is even worse when the spinal canal is congenitally narrow or trefoil-shaped (e.g. in the spinal stenosis associated with achondroplasia – Figure 10.6).

Destructive lesions of the bones may be due to infection or tumour (usually metastatic lesions). These may show on plain X-rays but CT and MRI are often helpful.

Imaging of the brain is usually by MRI. Functional scans such as positron emission tomography (PET scan) that can isolate specific areas of brain activity are also gaining popularity and may be used in conjunction with MRI and CT.



(a) (b)

Figure 10.6 Imaging (a) AP and (b) lateral radiographs of the spine of a child with achondroplasia. Note the thoracolumbar kyphosis, the exaggerated lumbar lordosis and the short pedicles on the lateral view. There is posterior vertebral scalloping and the sacrum appears horizontal. On the AP view, the interpedicular distance narrows towards the bottom of the lumbar spine. Spinal stenosis was confirmed on MRI.

Additional investigations

Blood and *cerebrospinal fluid* investigations may be necessary, depending on the working diagnosis and the necessity to exclude other potential diagnoses.

A *muscle biopsy* can provide useful information but great care must be taken with the process if the information gained is to be reliable: the biopsy must be taken from a muscle that is affected but still functioning; local anaesthetic infiltration must be avoided; the specimen must be handled gently; and, depending on the exact tests requested, it must be kept at its resting fibre length. Biopsies must be placed in special transport medium or frozen immediately in liquid nitrogen.

Audiological and *ophthalmic testing* and *assessment of mental capacity* are also helpful in certain cases.

NEUROPHYSIOLOGICAL STUDIES

Nerve conduction studies and needle *electromyography* have an important role in the investigation of peripheral nerve and muscle disorders. Theoretically, any motor or sensory nerve can be studied, but in everyday clinical practice most of these investigations look at the median, ulnar and radial motor and sensory responses in the upper limb, and, in the lower limb, the sciatic nerve including both the posterior tibial and peroneal divisions, motor and sensory modalities.

Needle electromyography (EMG) of individual muscles is used as a complementary technique, which gives information about the nature and number of the activated or denervated motor units from the specific nerve root that innervates the muscle being tested. This can be used for anatomical clarification and separation of radiculopathy from peripheral neuropathy and myopathy.

Nerve conduction studies

Motor nerve conduction

The nerve under study (usually a mixed motor and sensory nerve) is stimulated electrically at an easily accessible subcutaneous site (e.g. the forearm or wrist for the median nerve or behind the medial malleolus for the posterior tibial nerve), until it propagates an action potential which travels to the innervated muscle where a surface electrode records the response (Figure 10.7). Measurements are displayed on an oscilloscope screen, the most informative being the time it takes in milliseconds (ms) for the impulse to reach the muscle, called the *latency*, and the magnitude of the response evoked in millivolts (mV), called the *amplitude* of the evoked compound muscle action potential (CMAP). By measuring the distance from the stimulating electrode to the recording electrode, and setting this against the latency, one can deduce the *nerve conduction velocity (NCV)* in metres per second between those two points (Figure 10.8).

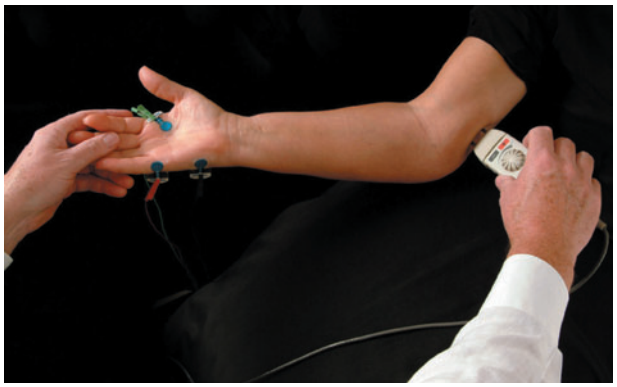
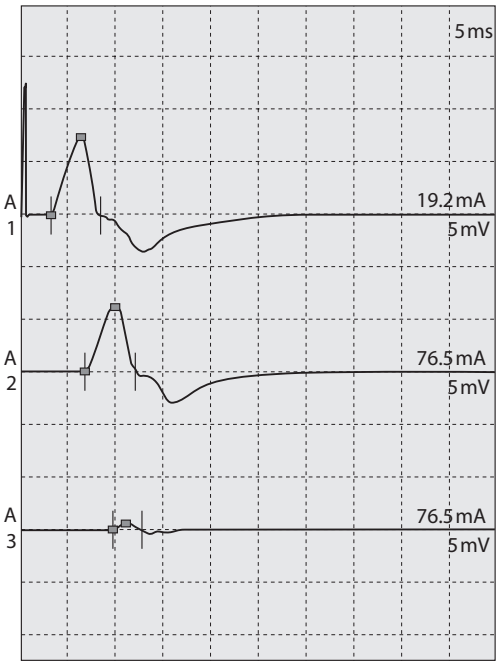


Figure 10.7 Ulnar motor nerve conduction The ulnar nerve is stimulated above the elbow, posterior to the medial epicondyle, and the CMAP is recorded from the abductor digiti minimi.

In practice it is more useful (and more accurate) to stimulate the nerve at two points, first at a distal site and then at a proximal site, and subtract the distal latency from the proximal latency to obtain a truer measurement for the intervening segment of the nerve. Thus, to measure the NCV of the median nerve in the carpal tunnel, one would take readings with the stimulating electrode first distal to the carpal tunnel and then in the upper forearm; this would allow one to deduce the NCV in the particular segment of the nerve at the carpal tunnel.

Similarly with measurement of amplitude, which is proportional to the number of motor units stimulated: if a patient has lost one-half of the nerve fibres in a peripheral nerve (e.g. due to compression, trauma or vascular insufficiency) the size of the elicited CMAP will be reduced by approximately 50% compared to the contralateral normal limb. When a nerve is stimulated at two sites, distally and then proximally, the evoked



Stimulus site	Lat1 ms	Dur ms	Amp mV	Area mVms
A1: Wrist	2.9	5.2	7.7	21.1
A2: Below elbow	6.5	5.3	6.3	17.5
A3: Above elbow	9.5	3.0	0.6	1.1
A4: Axilla				
A5: Erb's point				

Segment	Dist mm	Diff ms	CV m/s
Wrist–Below elbow	220	3.6	61
Below elbow–Above elbow	80	3.0	27
Above elbow-Axilla			
Axilla–Erb's point			

Figure 10.8 Nerve conduction velocity Oscillographic recordings of nerve conduction studies in a case of acute ulnar nerve palsy due to compression of the patient's arm while undergoing surgery under general anaesthesia. These tracings show an acute motor nerve conduction block at the elbow, with normal distal CMAPs when stimulating below the elbow (tracings A1 and A2) and a reduced amplitude CMAP when stimulating above the elbow (A3). There is severe focal conduction slowing across the elbow at 27 m/s, compared to 61 m/s in the segment below the elbow.

CMAPs should be of similar amplitudes. However, if the CMAP on proximal stimulation is observed to be smaller than the CMAP on distal stimulation, one assumes that a reduced number of motor units have conducted the action potential over the intervening segment of the nerve: this is referred to as *conduction block* (Figure 10.8) and is a feature of a potentially recoverable *neuropraxia*.

Common investigations are measurement of the NCV for the median nerve at the wrist or the ulnar nerve at the elbow in suspected cases of carpal tunnel syndrome or cubital tunnel syndrome respectively. In a focal entrapment neuropathy one will find focal slowing with normal velocities on either side of the lesion.

Slowed conduction of uniform degree along the whole length of the nerve suggests a demyelinating neuropathy (e.g. Charcot–Marie–Tooth syndrome).

Sensory nerve conduction

In a similar manner, a sensory nerve action potential (SNAP) may be recorded by stimulating a suitable subcutaneous sensory nerve and recording with surface electrodes on the skin over a measured distance along the same sensory nerve, e.g. from the index and middle fingers of the *median* nerve. SNAP is much smaller in amplitude than CMAP and is measured in microvolts.

NOTE: Clinical nerve conduction studies estimate the population of large myelinated sensory or motor nerves. Type C fibres (small myelinated fibres serving pain and temperature appreciation) have an amplitude below the sensitivity of recording techniques, as well as slowed velocity (5–10 metres/second) and cannot be tested with standard clinical techniques.

Electromyography (EMG)

To record the electrical discharge of motor units in a muscle, a concentric needle electrode, the shape of a small hypodermic needle, is inserted into the muscle and connected to an oscilloscopic screen and a loudspeaker (Figure 10.9). This will provide both a visual pattern on the screen and, simultaneously, crackling sounds from the loudspeaker.

At rest, a normal muscle is silent. As the patient slowly contracts the muscle, there is recruitment of one, then a few and then multiple motor units (a motor unit being defined as the anterior horn cell in the spinal cord, its motor axon and the muscle fibres it innervates in the muscle). This is reflected first as a progressive increase in the number and then also as increased amplitude of motor unit action potentials, with recognizable patterns. A full *recruitment pattern* usually looks and sounds like 'white noise', with so many motor units firing that both the spikes on the screen and the crackles from the speakers overlap each other; a so-called *'interference pattern'*.

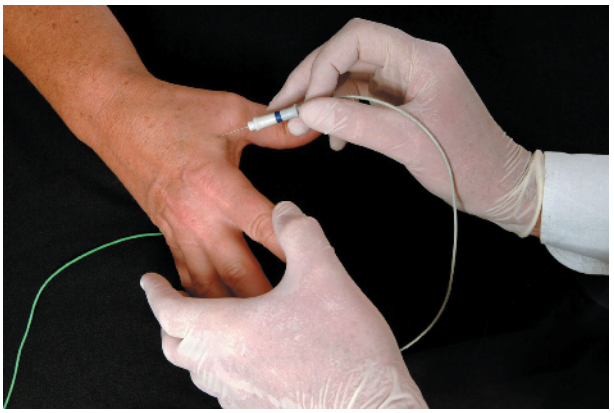


Figure 10.9 Needle electromyography (EMG) The first dorsal interosseus muscle (C8–T1 ulnar nerve) is being sampled during voluntary contraction against resistance.

In nerve disorders the muscle may not be silent at rest and may manifest increased insertional activity (activity during insertion of the needle electrode). There are changes of active denervation, referred to as *fibrillation potentials* and *positive sharp waves*, produced by denervated muscle fibres firing spontaneously. This signifies motor nerve fibre loss or disruption. It takes 7–12 days for the changes of active denervation to develop after axonal disruption. In a denervated muscle (e.g. the result of spinal root entrapment) the number of motor units recruited will be reduced proportional to the number of disrupted axons. Instead of the white noise of full recruitment, one sees a reduced pattern of muscle potentials. In muscle disease similar changes to the above may be seen but the pattern of action potentials differs and the full interference pattern appears at lower levels of active contraction.

A chronic neuropathy, with re-sprouting of remaining viable nerve fibres, results in longer re-innervated motor units with a polyphasic or higher amplitude profile. See Figure 10.10.

Diagnostic evaluation of the patient

Which nerves are studied in any patient, and the interpretation of the electrophysiological findings, will depend upon the clinical presentation and the provisional diagnosis. Appropriate nerve conduction studies and EMG can confirm or refute the clinical diagnosis (see Box 10.1). *Comprehensive study of all nerves without a diagnostic plan is usually unhelpful.*

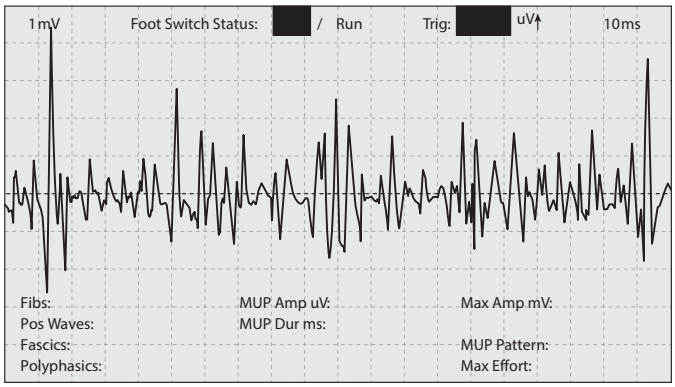
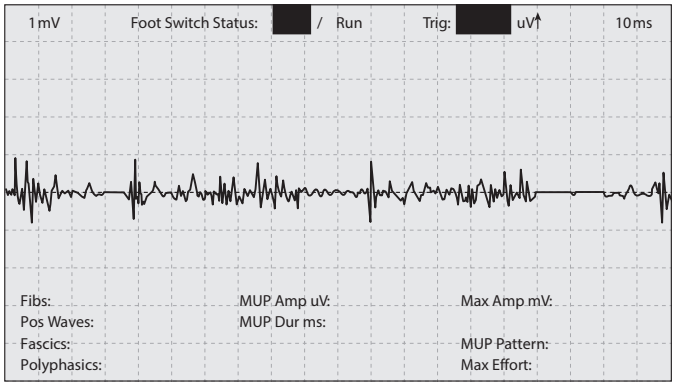
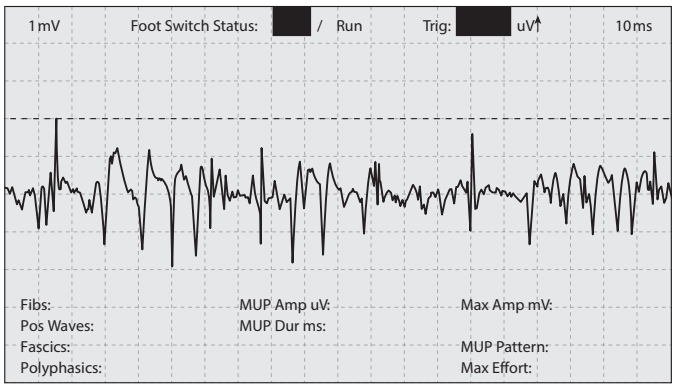


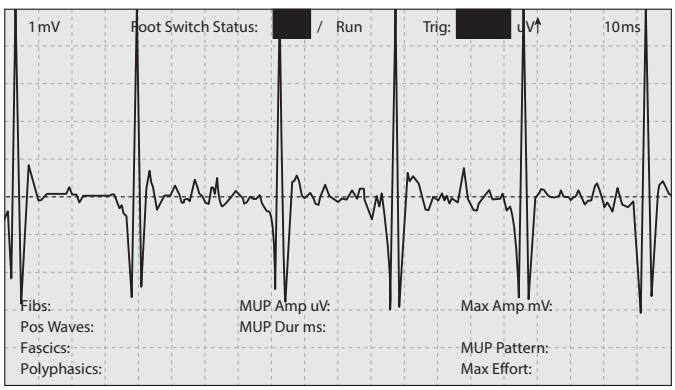
Figure 10.10 Electromyography
(a) Normal recruitment of motor units on needle EMG of the biceps muscle, to full interference/ recruitment pattern. (Amplitude 1 mV/division)



(b) Myopathic recruitment pattern in a patient with polymyositis. There are multiple small-amplitude motor units. (Amplitude 1 mV/division)



(c) Acute denervation pattern, characterized by florid low-amplitude fibrillation potentials recorded from tibialis anterior (resting state)



(d) Severe neurogenic abnormality. Single rapidly firing giant motor potential, typical of severe motor unit loss in a patient with old poliomyelitis. A similar pattern is seen in motor neuron disease. (Amplitude 1 mV/division)

When investigating a specific nerve root syndrome, nerve conduction and EMG studies are concentrated in the appropriate anatomical territory and the findings are compared to those in other nerve root territories in the same limb as well as in the contralateral (usually asymptomatic) limb. For example, in a patient with a weak arm and radial distribution paraesthesiae due to a C5/6 disc prolapse, one would study the median nerve motor and sensory potentials at the carpal tunnel, the radial sensory potentials at the wrist and EMG of C6 innervated muscles (e.g. biceps and brachioradialis). The findings are then compared to

BOX 10.1 NEUROPHYSIOLOGICAL SIGNS OF NEUROPATHIC DISORDER

Reduced motor or sensory potentials reflect non-functioning (perhaps transected) nerves.

Loss of sensory responses (SNAP) reflects a disorder *distal* to the spinal foramen (e.g. in the plexus); intact SNAP in a hypaesthetic limb suggests disease *proximal* to the foramen (e.g. a prolapsed disc).

Conduction block (i.e. intact distal motor response with focal conduction block) implies a neuropraxic recoverable injury.

Denervation changes on EMG more than 10 days after injury confirm significant nerve damage and loss of motor nerve function.

Any recruited volitional motor units in a weak limb imply a potential for recovery.

those in the C7 muscles such as extensor digitorum communis and triceps.

In a *mononeuropathy* or *plexopathy* one needs to compare conduction values (amplitude and velocity) in one limb to those in the other.

In a disorder such as a *focal entrapment* one may demonstrate a reduced amplitude on proximal stimulation compared to distal stimulation, representing conduction block, or significant focal conduction slowing.

Distinguishing nerve root disease from peripheral entrapment

The major anatomical defining characteristic of a proximal root entrapment (e.g. due to a prolapsed disc) is the preservation of the sensory action potential in the involved limb. This is because the lesion interrupts the nerve root proximal to the dorsal root ganglion which is anatomically (and electrically) situated outside the spinal cord where it is in continuity, and maintains the integrity of the distal axon; hence the SNAP remains normal.

The CMAP may be reduced as the motor nerve is separated from the anterior horn cell in the spinal cord. For example, in a wrist drop from a C7 root entrapment, the radial motor potentials are reduced or even absent, there is gross denervation on EMG, but the radial sensory potentials are preserved and entirely normal. *The presence of an intact sensory potential is what distinguishes root and proximal disease from peripheral entrapment and plexus disease.*

Intraoperative neurophysiological techniques

Spinal monitoring: somatosensory evoked responses (SSEP)

Spinal cord monitoring with neurophysiological tests is now considered an essential part of any spinal surgery that includes deformity correction to ensure that no neurological damage occurs. The techniques used are based on the principles defined above but often in combination with techniques employed in electroencephalography (EEG), such as *averaging*. A peripheral nerve in the upper or the lower limb (usually the median or posterior tibial) is stimulated but, instead of recording from the nerve or the muscle twitch, one records from the scalp overlying the patient's sensory parietal cortex.

The evoked responses from the recorded cortex are miniscule and one must therefore *average* the obtained responses from at least 100–200 stimuli in order to differentiate the time-linked evoked response from the background brain EEG activity. Averaging 200 or more responses at a stimulus rate of 3 per second to demonstrate a reproducible response may take 1-2 minutes, assuming all other factors are even and perfect. The surgeon should be aware of this drawback. One can also measure potentials developed in the cervical spinal cord at C7 level and the L1 level as well as distally in the brachial plexus at Erb's point, resulting from peripheral nerve stimulation.

The important measured parameter is usually the *latency* of the response, e.g. the N20 response from median nerve stimulation (a brain response occurring at approximately 20 milliseconds after stimulating the median nerve at the wrist). Accidental nerve injury during surgery around the spinal cord will produce a delay in the latency or a sudden loss of the evoked response.

Other intraoperative techniques

Various techniques are used, tailor-made to the clinical situation and the procedure being undertaken. These may include *nerve or nerve root stimulation* at various sites and measurement of either the distal nerve or muscle impulse. This can demonstrate normal electrical activity, a conduction block or slow conduction along the nerve.

Intraoperative EMG is performed with the needle *in situ* in the appropriate muscle (e.g. the quadriceps for L4 root procedures, abductor hallucis for the S1 root) to assess the muscle contraction when the nerve is stimulated, either intentionally or otherwise.

Cord-to-cord stimulation and *cord-to-cortical potential measurement* are usually resolved as averaged recordings to reveal intraoperative evidence of spinal pathway disruption.

CEREBRAL PALSY

The term 'cerebral palsy' describes a heterogeneous group of permanent disorders of movement and posture which are attributed to non-progressive disturbances in the developing fetal or infant brain which cause limitations in activity. Worldwide cerebral palsy is the commonest cause of motor disability in childhood with an incidence of 2–3 per 1000 live births increasing to 40–100 per 1000 live births in premature babies and those of low/very low birth weight. Multiple births also increase the risk. Other causal factors include maternal infection, peri-partum anoxia, postnatal meningitis, near-drowning or trauma. In the developed world, the majority of cases are due to prenatal causes and, contrary to popular belief, peri-partum hypoxia (hypoxic–ischaemic encephalopathy (HIE)) accounts for only 10% of cases.

The motor disorder results from centrally mediated abnormal muscle tone with spasticity ('high tone') the commonest abnormality. Oro-facial motor incoordination may make speech and swallowing difficult and drooling is a frequent problem; none of these defects implies a poor intellect although far too frequently the wrong conclusions are drawn. However, the factors that damage the motor centres may also cause damage to other areas of the developing brain and thus many children with cerebral palsy have associated problems such as epilepsy (30–40%), impaired vision and/or hearing, significant learning difficulties (30%) and perceptual and behavioural problems.

Classification

Many classification systems are used in cerebral palsy and these fall broadly into two groups: those that describe physical motor abnormalities and those that describe function. The Surveillance of Cerebral Palsy in Europe collaboration recommends describing distribution as unilateral or bilateral and classifies the main motor tone types into one of four groups: spastic, dyskinetic, ataxic and mixed.

TOPOGRAPHIC DISTRIBUTION

Unilateral

- *Hemiplegia* is the commonest form of cerebral palsy. This usually appears as a spastic palsy on one side of the body with the upper limb more severely affected than the lower. Most children can walk and they may respond well to treatment.
- *Monoplegia* occasionally appears in an upper limb; careful examination will often show that other areas are involved as well. True monoplegia is so unusual that other diagnoses should be considered, such as a neonatal brachial plexopathy.

Bilateral

- *Diplegia* involves both sides of the body, with the lower limbs always most severely affected. Some upper limb dysfunction is invariably present but signs may be subtle. Side-to-side involvement may be asymmetrical and the terms asymmetric diplegia and occasionally bilateral hemiplegia are used. Many cases are secondary to prematurity and periventricular leucomalacia (PVL) is seen on brain MRI. Intelligence is often normal. The less severely affected children can have reasonable mobility but the non-walking diplegic patient may be similar to the total body involvement group discussed below.
- *Total body involvement* describes a general and often more severe disorder affecting all four limbs, the trunk, neck and face, with varying degrees of severity. Patients usually have significant associated problems, such as difficulty swallowing and epilepsy, and they are wheelchair-based.

TYPE OF MOTOR DISORDER

- *Spasticity* is the commonest muscle movement disorder and is associated with damage to the pyramidal system in the CNS. It is characterized by a velocity-dependent increased muscle tone and hyper-reflexia. The resistance to passive movement may obscure a basic weakness of the affected muscles.
- *Dyskinetic* movement disorders are characterized by recurring, uncontrolled and involuntary movements that may be stereotyped. The tone varies. There are two main types:
  - Dystonic – characterized by hypokinesia (reduced activity) and hypertonia (increased tone) resulting in stiff movements
  - Choreoathetotic – characterized by hyperkinesia (increased activity) and hypotonia (reduced tone) resulting in uncoordinated writhing and jerky movements. It is caused by damage to the extrapyramidal systems of the CNS. In pure athetoid cerebral palsy, joint contractures are unusual as muscle tone is not increased.
- *Ataxia* is characterized by generalized hypotonia with loss of muscle coordination during voluntary movements showing as movements which are defined by uncontrolled, inaccurate movements of abnormal force and/or rhythm. It is usually due to cerebellar damage. Balance is poor and the patient walks with a characteristic wide-based gait.
- *Mixed forms* in which no one tone abnormality and movement disorder predominates.
  - A combination of spasticity with dyskinesia is the commonest mixed type.

  In addition to these four main types, the following clinical picture is frequently seen:

- *Hypotonia*, which is usually (but not always) a phase, lasting several years during early childhood, before the features of spasticity become obvious.

NOTE: In some types of cerebral palsy there is considerable variability in the 'tone' and 'posture' from day to day or situation to situation. If surgical treatment is being considered, it should never be based on a single assessment when, due to stress, the child appears to have abnormally high tone and muscle contractures: if seen again the following day, the clinical picture could be very different.

Diagnosis before walking age

The full-blown clinical picture may take months or even years to develop. A history of a pre- or peri-natal event such as a premature birth, difficult labour or fetal distress should arouse suspicion. A neonatal ultrasound scan of the head may identify an intraventricular haemorrhage (IVH) due to HIE which may predict later problems.

At birth, almost all motor behaviour is controlled by primitive reflex movements in response to a variety of sensory stimuli; these gradually disappear with development of the CNS and are replaced by a more mature set of protective and postural reflexes. The five reflexes that Bleck suggested we look for are: (1) the primitive neck-righting reflex, (2) asymmetrical and (3) symmetrical tonic neck reflexes, (4) the Moro reflex and (5) the extensor thrust response. Persistence of any two of these past the age of 12 months is a cause for concern.

Early symptoms include difficulty in sucking and swallowing, and excessive drooling. The mother may notice that the baby feels stiff or fidgets awkwardly. Gradually it becomes apparent that the motor milestones are delayed (Figure 10.11). The normal child develops head control at 3 months, sits up at 6 months and begins walking at about 1 year.

NOTE: Children in whom 2/5 primitive reflexes persist after 12 months of age, who cannot sit unaided by 4 years and who cannot walk by 8 years are unlikely ever to walk independently.

Diagnosis in later childhood

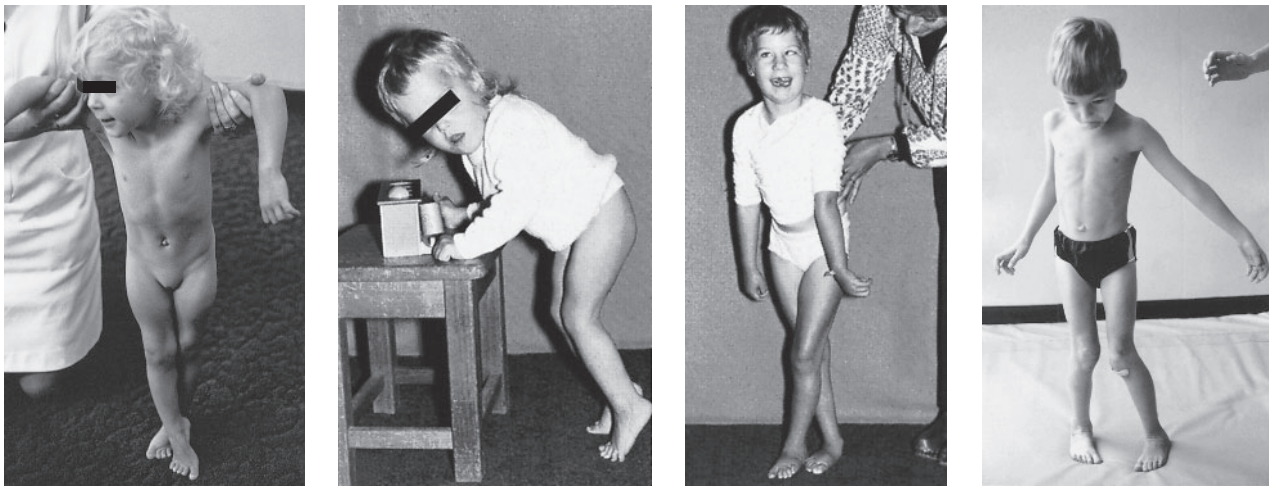
Most children presenting to the orthopaedic surgeon have already had the diagnosis made. Occasionally, for example with a mild hemiplegia or a symmetrical mild diplegia, the diagnosis has not been established and the child is simply referred for advice about their gait

(a)

(b)

(c)

Figure 10.11 Cerebral palsy – early diagnosis By 6 months, these twin brothers demonstrate differences in development with (a) one being obviously smaller than the other and (b) one demonstrating a lack of head control when being pulled in to sitting and (c) an inability to sit unaided when compared to his twin.

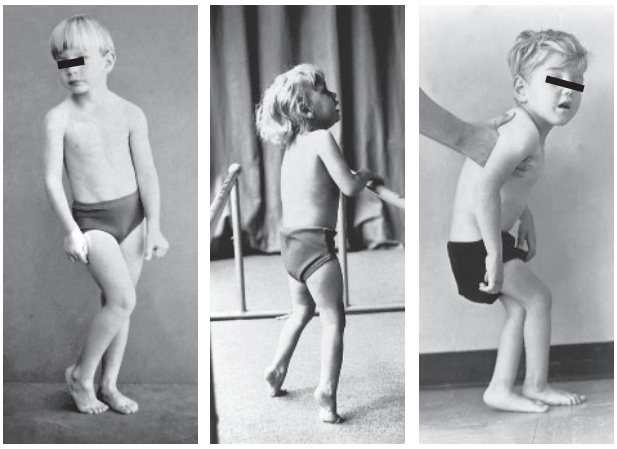


(a) (b) (c) (d)

Figure 10.12 Cerebral palsy (a) Adductor spasm (scissor stance); (b) flexion deformity of hips and knees with equinus of the feet; (c) general posture and characteristic facial expression; (d) ataxic type of palsy.

or their tendency to trip and fall. A familiarity with and knowledge of the normal developmental milestones and gait patterns helps the clinician identify the child who is outside the normal range (Figures 10.12 and 10.13).

Ideally, the child should be reviewed by a multidisciplinary team so that feeding, speech, hearing, visual acuity, intelligence and motivation can also be assessed. Since cerebral palsy is essentially a disorder of posture and movement, the child should be carefully observed sitting, standing, walking and lying. His or her condition should then be evaluated according to the gross motor function classification system (GMFCS) which categorizes the child, relative to their age, in terms of mobility (Figure 10.14), basing this on their average function rather than on the best that they can achieve on any given occasion.



(a) (b) (c)

Figure 10.13 Spastic palsy Common types of spastic palsy: (a) hemiplegic, (b) diplegic, (c) whole body.

The system is reliable and valid; it aids in communication between members of the multidisciplinary team and is a useful guide to prognosis and thus for management.

SITTING POSTURE

The child may find it difficult or impossible to sit unsupported: children with a hypotonic trunk may slump into a kyphotic posture and others may always 'fall' to one side. In attempting to sit, the lower limbs may be thrust into extension. There may be an obvious scoliosis or pelvic obliquity (Figure 10.15). Head control may be poor.

STANDING POSTURE

In the typical case of a spastic diplegia, the child stands with hips flexed, adducted and internally rotated, the knees are also flexed and the feet are in equinus. With tight hamstrings, the pelvis may be tilted posteriorly obliterating the normal lumbar lordosis and the child may have difficulty standing unsupported. Often attempts to correct one aspect of the deformity may aggravate another and it is important to establish which deformity is the primary one and which are compensatory. Many older children show pelvic obliquity and a scoliosis. Asking the child to 'stand tall' and watching their response often gives some insight into the dynamic nature of the posture, muscle strength and, of course, intellectual ability.

Balance reactions are often poor and a gentle push that would force a normal child to 'right themselves' and take a step in the appropriate direction to maintain his or her balance may simply knock over a child with cerebral palsy.

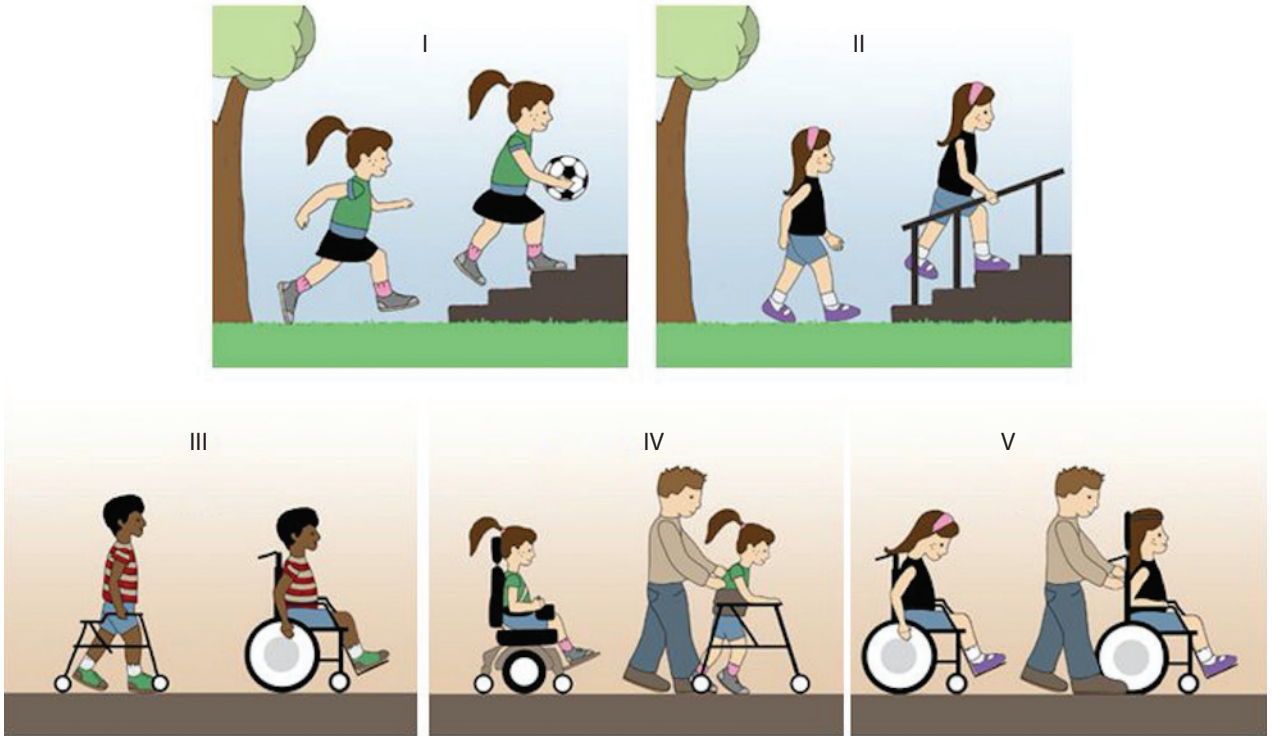


Figure 10.14 Gross motor function scale (GMFCS) A diagram that illustrates the GMFCS levels I (normal) to V (wheelchair-based with no head control). The scale is age-dependent. It has implications for prognosis and for treatment.

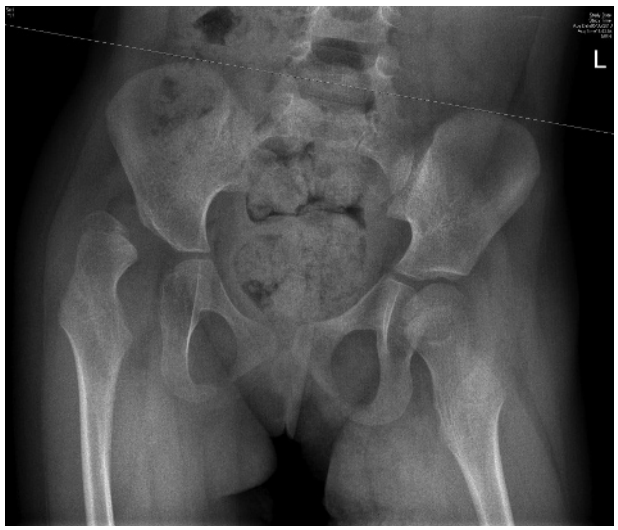


Figure 10.15 Cerebral palsy AP view of the pelvis showing a right hip which is essentially dislocated. Note the pelvic obliquity (green line) with the right side 'high'. The right hip is adducted with respect to the line of the pelvis and the left hip is abducted as part of the 'windswept' posture that is seen in many children with severe cerebral palsy.

GAIT

If a child can walk, the elements of gait are analysed taking note of the use of walking aids and orthotic devices. Gait should be observed with and without shoes or orthotic supports and the differences (if any) noted. Abnormal postures, fluidity of movement, balance and the ability to start/stop easily may all become more noticeable during walking. Every opportunity must be taken to observe gait so that differences between 'normal' and 'best behaviour' walking can be identified. In hemiplegics, best behaviour walking may demonstrate a flat foot pattern with the heel coming down most of the time, while the more normal or representative pattern will highlight the asymmetric flexed knee and toe-walking pattern.

Clinical gait analysis is difficult but improves with practice. Each limb must be observed in both the stance and swing phases of gait and in the coronal, sagittal and transverse planes. In the spastic diplegic patient, the standing posture mentioned above is influential in defining their walking pattern too. The lack of free rotation at the hip means that the trunk has to move from side to side as each leg swings through and with the adduction it leads to a 'scissoring' action (one leg crossing in front of the other).

This results in a narrow walking base and, when combined with the hip and knee flexion and foot equinus, there is a strong tendency to fall; this can be helped by the use of walking aids such as crutches or quad sticks.

Computerized gait analysis ideally supplements observational gait analysis. Kinematics (joint and limb segment movement), kinetics (joint moments

and powers), EMG (identification of the phases in which muscles are firing), pedobarography (foot pressures) and metabolic energy analysis (assessment of the 'cost' of walking) are all part of the analysis, as is a video recording which can be viewed from any direction and at any speed. Interpretation of all this data requires skill and experience and the application of the information to an individual child also requires a degree of common sense. Pattern recognition is important (in both forms of gait analysis). Perhaps its main role is to help the clinician determine the influence of dystonic posturing on gait and to identify the dynamic components that aggravate the fixed tightness seen on clinical examination.

NEUROMUSCULAR EXAMINATION

By the end of this part of the examination the clinician should have a clear idea of overall *muscle tone and muscle power* and the *range of movement* at each joint.

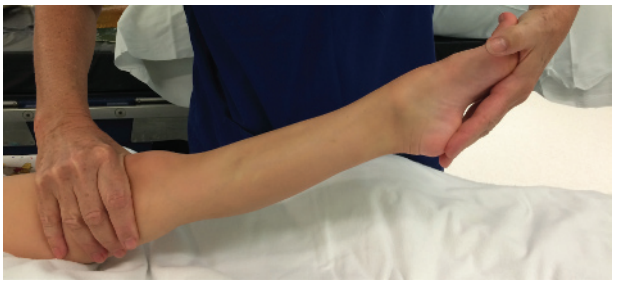
Most commonly the limbs will show the typical features of an upper motor neuron syndrome which include increased tone, brisk reflexes, a positive Babinski response and clonus, as well as important 'negative' signs such as weakness, poor coordination and poor selective muscle control. Spasticity gives the appearance of 'strong' muscles but actually muscle power is invariably relatively poor. It is associated with a loss of muscle excursion, reduced joint movement, secondary contractures, bony deformity, joint dislocations and pain.

In children with cerebral palsy the physical signs often vary from day to day or even minute to minute depending on factors such as the happiness of the child and the room temperature. It takes time to examine a child and get a representative 'feel' for the tone, the muscle strength and the degree of deformity present. The physiotherapist has often seen the child more often and in more relaxed circumstances than the orthopaedic clinic and can therefore identify whether the day's examination is truly representative. Be sure to listen to what they and the family say.

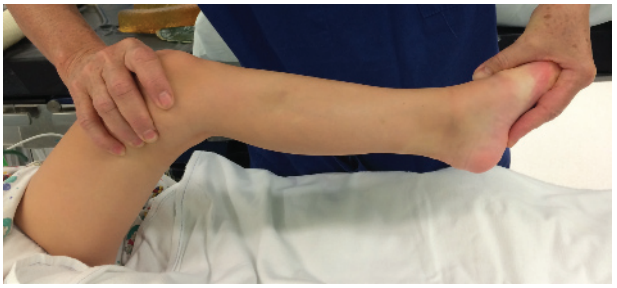
FIXED (STATIC) vs FLEXIBLE (DYNAMIC) CONTRACTURE

It is important to assess the degree of deformity present at each joint and relate it to the length of each specific muscle–tendon unit. Deformity at one level may be markedly affected by the position of the joints above and below.

For example, ankle equinus with the knee extended often disappears when the knee is flexed; thus one can differentiate between tightness in the soleus and tightness in the gastrocnemius muscle. In the *Silfverskiöld* test (Figure 10.16), with the child lying supine on the examination couch, the knee is flexed to a right angle and the ankle is dorsiflexed; this tests soleus tightness. The knee is then fully extended on the couch and ankle dorsiflexion is repeated; now it is mainly gastrocnemius tightness that is being tested. Similarly, tight hamstrings may limit knee extension more with the hips flexed than when the hips are extended and hip adduction may be easier in knee flexion than in extension due to a tight gracilis. If hip abduction in flexion is restricted, especially symmetrically, order an X-ray to look for subluxation of the joint.



(a)



(b)

Figure 10.16 Cerebral palsy, Silfverskiöld test (a) With the knee extended, the hindfoot varus/valgus is corrected and the foot dorsiflexed as far as possible: this picture confirms significant equinus. If part of this equinus is due to tightness in the gastrocnemius muscle which inserts above the knee, then (b) with the knee flexed and the hindfoot varus/valgus controlled, the equinus deformity is improved. If it does not improve and/or there is residual equinus, this is due to tightness in the soleus muscle. This clinical sign is the Silfverskiöld test and it helps direct surgical treatment.

In the upper limb, finger flexors may be tight with the wrist extended but, if the wrist is allowed to flex, the fingers can extend. Children can use these fixed length reactions to manipulate their hand and finger function using 'trick' movements.

A degree of muscle contracture is almost inevitable with all forms of cerebral palsy where a long-standing increase in tone (spastic and dystonic types) leads to relative shortening of the muscles and hence fixed contractures. There is still some debate as to whether the changes are due to a true shortening of the muscle or a failure of the muscle to grow in tandem with skeletal growth. Certainly, most contractures develop during the period of growth; after skeletal maturity the changes in muscle–tendon length and joint contracture are much less progressive.

BONY DEFORMITY AND CHANGES IN JOINT CONGRUITY

Normal bone growth is influenced by muscle pull. Hence in children with persistent abnormal muscle pull there may be abnormal bone growth with both failure of the normal modelling processes of childhood in addition to the development of new deformities. The normal degree of infant femoral neck anteversion persists and sometimes even increases with growth rather than improving, and significant external tibial torsion may also develop (Figure 10.17).

Bony deformities may, in turn, engender new problems. External tibial torsion may give rise to planovalgus deformity of the foot. Persistent flexion and adduction of the hip, in addition to the persistent femoral neck anteversion, leads to acetabular dysplasia and subluxation of the joint. At the knee, shortening in the quadriceps may contribute to the patella alta appearance and the development of patella–femoral pain (Figure 10.18).

SCOLIOSIS

Flexible curves are common, but unfortunately many become structural; this is especially likely in patients with total body involvement cerebral palsy where there is often an associated pelvic obliquity. Kyphosis and lordosis also occur.

SENSORY EXAMINATION

Sensation is often not entirely normal and problems with stereognosis (as well as with perception) may be important factors contributing particularly to upper limb disability.

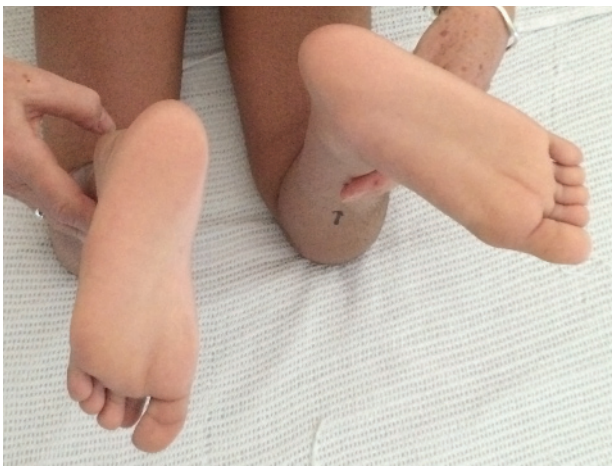


Figure 10.17 Cerebral palsy A child lying prone with knees flexed to 90 degrees: the right foot lies in gross external rotation (compared to the left foot) secondary to unilateral and excessive external tibial torsion.

Management

There is no single 'blueprint' for the holistic management of all patients with cerebral palsy; each patient and his or her family provide a different challenge. This section will aim to discuss first some basic principles that are applicable to all children and then some more specific principles that relate to the various types of cerebral palsy.

GOAL SETTING

It is human nature for a parent to want and indeed expect the best for their child, and it is the role of the healthcare professionals to support them in their wishes. However, it is also important for the professionals to ensure that the difference between hopeful optimism and pragmatic realism is understood by all involved in the child's care. Few patients with total body involvement will ever walk. The prognosis for walking in the patient with spastic diplegia should be assessed early and will be influenced by many factors including motivation. The definition of walking, in terms of ease and distance, must also be conveyed to the parents along with an explanation that many children with cerebral palsy reach their peak of physical function in late childhood (much earlier than their

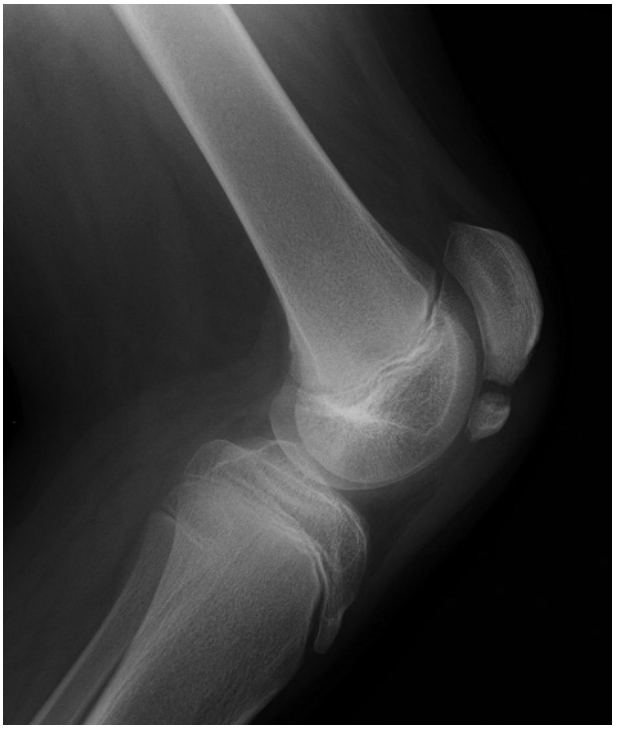


Figure 10.18 Diplegic CP A lateral radiograph of the knee of a child with diplegic CP: the patella is high-riding (patella alta) secondary to spasticity in the quadriceps muscles. This has also led to an avulsion injury at the inferior pole of the patella: this exacerbates the inefficiency of the 'quadriceps mechanism'.

contemporaries) and with the increase in size and weight that comes with puberty, weak muscles may no longer be able to maintain walking ability even with the use of aids.

The WHO has developed the International Classification of Functioning, Disability and Health (ICF) which looks at health and health-related domains but acknowledges that functioning and disability occur in the context of the individual's environment. Thus, quality of life (QoL) may be judged in terms of not only the child's impairment but the extent to which their ability to be active is limited and their ability to participate is restricted.

For all patients with cerebral palsy, the priorities are: (1) an ability to communicate with others; (2) an ability to cope with the activities of daily living (including personal hygiene); and (3) independent mobility – which may mean a motorized wheelchair rather than walking aids.

For the child who from an early age is recognized to be 'non-walking', realistic goals should be: (1) a straight spine with a level pelvis; (2) located, mobile and painless hips that flex to 90 degrees (for comfortable sitting) and extend sufficiently to allow comfortable sleeping and participation in standing/swivel transfers; (3) knees that are mobile enough for sitting, sleeping and transferring; and (4) plantigrade feet that fit into shoes and rest on the footplates of the wheelchair comfortably.

For all children, good medical care including nutrition is also essential as is access to good quality orthotic supports, walking aids and/or wheelchairs as appropriate. Unfortunately, these basic needs are still not met for children in many disadvantaged communities in both developed and less developed nations.

TONE MANAGEMENT

Tone management is one of the most important aspects of patient care and it underpins all other forms of treatment (Figure 10.19). For patients with high tone, the factors shown in influence patient care and medical treatments are often indicated.

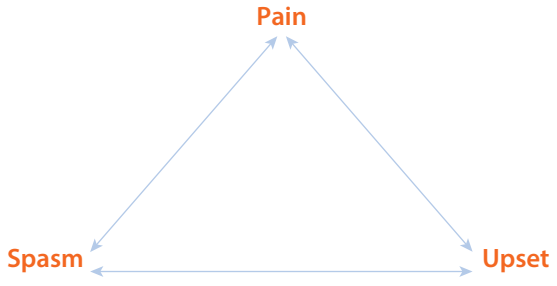


Figure 10.19 Pain, upset and anxiety All of these inter-react to exacerbate the patient's underlying spasticity and adversely affect the outcome of both medical and surgical treatments.

Medical treatment The most generally effective medications are *anticonvulsants* for seizures, *short-term benzodiazepine* use for postoperative pain and *trihexyphenidyl* for dystonia. *Analgesic medication* is needed for the reduction of pain associated with musculoskeletal problems, constipation and gastro-oesophageal reflux in addition to more specific drugs such as antacids or gastric H2 blockers.

Baclofen, a gamma-aminobutyric acid (GABA) agonist, acts by inhibiting reflex activity. In oral form it does not cross the blood–brain barrier well. When effective, it reduces muscle tone/spasticity generally. This may have a negative effect on head and trunk control and that combined with the side effects of drowsiness means that its use may be limited. Intrathecal baclofen (ITB) is administered via a refillable implanted pump that sits subcutaneously in the abdomen with the catheter leading from the pump into the epidural space of the lumbar spine. The dose administered can be titrated according to the child's response. Long-term studies of its use suggest that the technique is most effective in those with severe spasticity or dystonia. It is not effective in all patients and a test dose is used to identify the potential benefits in each prospective patient.

Botulinum toxin (BoNT-A) This potent neurotoxin, produced by *Clostridium botulinum*, acts by blocking acetyl choline release at the neuromuscular junction. The preparation is injected into the 'spastic'/'dystonic' muscle at (or as close as possible to) the motor end point. Ideally, these targeted injections are performed under US control or while using a nerve stimulator. The usual lower limb targets are the hip adductors, hamstrings, gastrocnemius and tibialis posterior. The weakness/paralysis that it causes takes a few days to become obvious; the effect is temporary and as new nerve terminals form there is a return of muscle tone at around 10–12 weeks.

BoNT-A must not be used on its own but rather as part of a package of care within the overall tone management programme. Thus, injections are followed by increased physiotherapy input to obtain an increase in muscle length of the injected muscles and an increase in strength in the antagonistic muscles. Often there is a concomitant alteration in orthotic/splinting support. This means that the overall benefits attributed to the injections may last considerably longer than the 10–12 weeks of true neuromuscular blockade. It is precisely because the toxin is never used on its own that it has been difficult to prove what the true benefits of this form of treatment are, but it is considered useful as a focal treatment for a dynamic muscle imbalance that is interfering with function, producing deformity or causing pain. It is perhaps more effective

in younger children who are less likely to have fixed deformity. Multilevel injections may be required but the overall dose per child must be kept within safe limits.

There is also a role for BoNT-A in the management of postoperative pain and spasm although for optimal effect the injections should be given some days prior to surgery.

Selective dorsal rhizotomy (SDR) Division of selected dorsal nerve roots from L1 to S2 has recently gained increasing acceptance as the indications for its use have been refined and the techniques for performing the procedure have improved. In cerebral palsy, the normal inhibitory influences on muscle tone from the higher centres are deficient. SDR aims to reduce spasticity and rebalance muscle tone by selectively reducing the input from the muscle spindles, thus leading to less excitation of the anterior horn cells. Long-term studies are not yet available but good results have been obtained in children aged 3–12 years who meet the following criteria: they are walking but have significant spasticity; they have the features of PVL on imaging; they have good intellectual function and good voluntary control. The presence of fixed contractures may be a relative contraindication and certainly implies that surgical correction of these will be required around the same time as the SDR to facilitate the rehabilitation programme.

Physical therapy Cerebral palsy affects motor function in several ways. There is a dependence on immature or primitive reflexes and a loss of selective muscle control. Physiotherapy attempts to reduce or prevent the problems arising from abnormal muscle tone, imbalance between opposing muscle groups and abnormal body balance mechanisms. To this end various structured approaches or 'schools' have been popularized. No single method has been shown to be significantly better than another but all have good points and all can work well in individual cases. In addition to these programmed approaches, there is a philosophy that regular 'range of movement' exercises will prevent or (perhaps more realistically) reduce the degree of muscle/ joint contracture.

Physiotherapy is considered to be most helpful in early childhood up to the age of 7 or 8 years but there is surprisingly little evidence to guide us in knowing what type of physiotherapy to prescribe and how often to do so in any particular case. However, postoperative physiotherapy is essential in order to maximize the effects of surgery and overcome the inevitable postoperative problems of pain, stiffness and weakness.

Positioning and splinting Care must be taken at all times to ensure that the child sits and sleeps and works and eats in a good position and with good posture. Adjustments may need to be made to chairs, wheelchair and the child's sleep system so as to limit disadvantageous positions such as hip adduction or ankle equinus. *Splints* are used to prevent muscle contracture, maintain joint position and improve movement and hence function. They also have an important role in maintaining position following surgery. Splints may be corrective in that they aim to hold a passively correctable deformity in the corrected position, or 'accommodative', for example when the splint adopts the shape of the foot and simply aims to prevent further loss of position. Splints must be useful: a badly fitting splint at best does nothing and at worst provokes pain and spasm which in turn often increases deformity.

Dynamic lycra splinting may benefit some children with cerebral palsy by improving their balance, muscle control, proximal stability and movement: it also reduces associated movements. Body suits which extend from above the knee to above the elbow can improve sitting posture and walking balance in certain individuals.

Manipulation and serial casting These methods may have a limited role in improving muscle/joint contractures, but relapse is frequent.

Operative treatment

The indications for surgery might include: (1) a spastic deformity which cannot be controlled by conservative measures; (2) fixed deformity that interferes with function; and (3) secondary complications such as bony deformities, dislocation of the hip and joint instability.

It is important to remember that in cerebral palsy all muscles are weak: thus, muscle-lengthening surgery is also muscle-weakening surgery unless by improving the mechanical alignment of the limb, and hence the muscle, you allow it to work more efficiently. Correction of bony deformity may be important in this respect (Table 10.3) and, although the surgery may seem more aggressive, it may actually be more appropriate, particularly given that internal fixation devices suitable for children are now more widely available.

Weak muscles can be augmented by tendon transfers but the muscle being transferred is weak already and may have a limited ability to function in its new role; on the other hand it may produce an unwanted overcorrection because of its increased tone. The role of gravity plays an important part in guiding the choice of tendon transfers.

The timing of surgical intervention is often crucial. Both CNS development and the gait pattern mature around the age of 7–8 years and thus many orthopaedic surgeons advocate delaying surgery until this age and then doing all the necessary operations at one or two sittings. Earlier operation may be called for if the

Table 10.3 Treatment of the principal deformities of the limbs

	Deformity	Splintage	Surgery
Foot	Equinus Equinovarus	Spring-loaded dorsiflexion Bracing in eversion and dorsiflexion	Lengthen tendo Achillis Lengthen tendo Achillis and transfer lateral half of tibialis anterior to cuboid
Knee	Flexion	Long caliper	Hamstring release
Hip	Adduction	–	Obturator neurectomy Adductor muscle release
Shoulder	Adduction	–	Subscapularis release
Elbow	Flexion	–	Release elbow flexors
Wrist	Flexion	Wrist splint	Lengthen or release wrist flexors; may need fusion or carpectomy
Fingers	Flexion	–	Lengthen or release flexors

hip threatens to dislocate. Our preferred approach is to avoid *'little and often'* surgery ('birthday surgery') in favour of the *'all or none'* philosophy (SEMLS – single event multilevel surgery) but, as always, some patients require the former and some the latter.

Regional survey

Upper limb

Upper-limb deformities are seen most typically in the child with spastic hemiplegia or total body involvement and consist of elbow flexion, forearm pronation, flexion of the wrist with clenched fingers and a flexed and adducted thumb ('thumb-in-palm' deformity). In the mildest cases, spastic postures emerge only during exacting activities. Proprioception is often disturbed, as are two-point discrimination and stereognosis, and all of these may preclude any significant functional improvement, whatever the kind of treatment. Some hemiplegic patients do respond well to constraint-induced movement therapy (CIMT) early in childhood. Operative treatment is usually delayed until after the age of 8 years and is aimed at improving the resting position of the limb and restoring grasp.

Elbow flexion deformity Provided the elbow can relax into an extended position that is below a right angle, no treatment is needed. Occasionally, to facilitate washing and dressing, it may be necessary to treat a more marked flexion contracture by fractional lengthening of the biceps and brachialis tendons with release of the brachialis origin.

Forearm pronation deformity This is fairly common and may give rise to subluxation or dislocation of the radial head. Simple release of pronator teres may improve the position, or the tendon can be rerouted round the back of the forearm in the hope that it may act as a supinator.

Wrist flexion deformity Wrist flexion is usually accompanied by ulnar deviation; it can be improved by lengthening or releasing flexor carpi ulnaris and maintaining the position with a splint. If extension is weak, the released flexor tendon is transferred into one of the wrist extensors where it acts more as a tenodesis than a true tendon transfer. In severe cases, wrist arthrodesis with excision of the proximal carpal row may be of cosmetic rather than functional benefit. *Before operating on the wrist it is essential to consider what effect this will have on finger movements.*

Flexion deformity of the fingers Spasticity of the long flexor muscles may give rise to clawing. Highly selective motor neurectomy has recently regained favour as a potential treatment for spasticity while, for contractures, the flexor tendons can be lengthened individually at the risk of provoking a swan neck deformity. If the deformity is severe, a forearm muscle slide may be more appropriate. Ideally these operations should be undertaken by a specialist in hand surgery. *If the fingers can be unclenched only by simultaneously flexing the wrist, it is obviously important not to extend the wrist by tendon transfer or fusion.*

Thumb-in-palm deformity This is due to spasticity of the thumb adductors or flexors (or both), but later there is also contracture of flexor pollicis longus. In mild cases, function can be improved by splinting the thumb away from the palm, or by operative release of the adductor pollicis and first dorsal interosseus muscles. Resistant deformity may need combined lengthening of flexor pollicis longus and release of the thenar muscles, followed by tendon transfers to reinforce abduction and extension. Here again, the operations should be performed by a specialist in this field.

Lower limb

The functional effects of lower-limb spasticity differ considerably, depending on whether the patient has

hemiplegia, diplegia or total body involvement; this will obviously influence the lines of surgical treatment.

UNILATERAL CEREBRAL PALSY (UCP) – HEMIPLEGIA

Four subtypes of hemiplegia have been identified (Figure 10.20) and the most common lower-limb problem is with foot deformity. As highlighted previously, initial treatment consists of physiotherapy and splintage to prevent fixed contractures and most children will use an ankle–foot orthosis (AFO).

Foot/ankle Tibialis anterior is invariably weak and the patient develops an *equinovarus foot deformity*. Active plantarflexion is required to assist knee extension during the stance phase of gait so care must be taken when considering a lengthening of the gastrocnemius/soleus complex. The trend is to perform a muscle recession rather than a tendon lengthening procedure.

A *dynamic varus deformity* can be treated by a split tendon transfer to the outer side of the foot (only half the tendon is transferred so as to avoid the risk of overcorrection into valgus): both tibialis anterior and tibialis posterior tendon transfers have been advocated depending perhaps on whether the surgeon feels that the varus is originating from the hindfoot or the midfoot/forefoot. In older children with fixed deformity, formal muscle lengthening with or without a calcaneal osteotomy may be required.

Pes planovalgus (with or without a midfoot break) may require subtalar arthrodesis in addition to judicious tendon lengthenings.

Hip/knee Surgery is not usually required but, if it is, it follows the principles outlined below for the walking diplegic patient.

Leg length discrepancy Due to discrepancies in growth, the hemiplegic limb is often short irrespective of any joint contractures. An epiphyseodesis of the contralateral distal femoral and/or proximal tibial physes may be considered. This can improve some aspects of the gait pattern but it will not 'remove' the limp.

BILATERAL CEREBRAL PALSY – SPASTIC DIPLEGIA

Most patients with cerebral palsy have a spastic diplegia where the lower limbs are more affected than the upper limbs. Treatment is concentrated on the lower limbs but upper-limb function is also important particularly if the child requires walking aids. In the young child, treatment consists of physiotherapy, the use of AFOs and tone-reducing medications to prevent fixed contractures. By 3–4 years of age the sitting and walking patterns can be observed, and particular attention should be paid to the interrelationship between the various postural defects, especially lumbar lordosis/hip flexion and knee flexion/ankle equinus. The muscles that cross two joints (iliopsoas, hamstrings and gastrocnemius) are influential in the development of the muscle and joint contractures characteristic of children with a spastic form of cerebral palsy. Surgery is indicated either to correct structural defects (e.g. a fixed contracture or hip subluxation) or to improve gait.

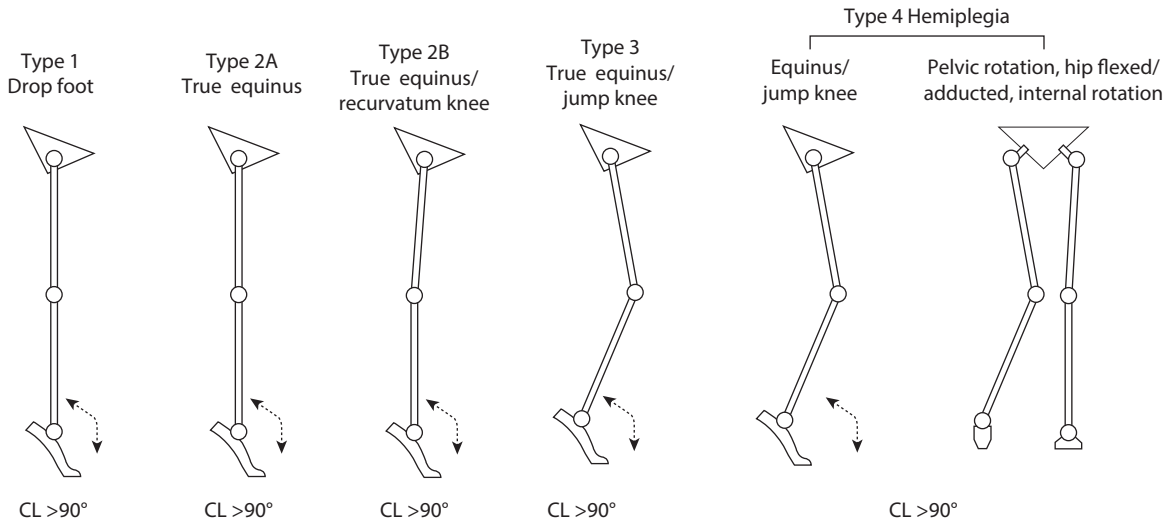


Figure 10.20 Common gait patterns: spastic hemiplegia The diagram illustrates the differing hemiplegic gait patterns and the various positions of the three main joints: hip, knee and ankle. The ankle equinus can be due to a drop foot or a tight gastrocsoleus complex and may be influenced by the presence of a flexible or fixed knee flexion deformity.

Most children will walk but they are delayed in learning to master this; a child who is not walking by the age of 6 or 7 years is unlikely to do so. Non-ambulant children often have orthopaedic problems similar to those with total body involvement (see below).

In walking diplegics, observational gait analysis is important, and computerized gait analysis may have a role in guiding treatment. Affected children are often relatively symmetrical in their gait pattern but in some asymmetry is very marked, with one limb maintaining a hemiplegic posture and one more consistent with a diplegic gait. Each limb must be assessed independently.

Hip adduction deformity The child walks with the thighs together and sometimes even with the knees crossing ('a scissoring gait'). This may be combined with internal rotation at hip/thigh level. Adductor release is indicated if passive abduction is less than 20 degrees on each side. For most patients open tenotomy of adductor longus and division of gracilis will suffice. Only if this fails to restore passive abduction (a rare occurrence) should the other adductors be released. A neurectomy of the anterior branch of the obturator nerve used to be popular but there is a risk of overweakening the adductor muscles and revealing the abductor muscle spasm. A hip stuck in abduction can be more troublesome than an adducted hip.

Hip flexion deformity This is often associated with fixed knee flexion (the child walks with a 'sitting' posture) or else hyperextension of the lumbar spine. Operative correction is indicated if the hip deformity is more than 20–30 degrees. In the walking child, it is important not to weaken hip flexion too much and thus intramuscular lengthening of the psoas tendon at the pelvic brim is advocated. (In the non-walking child, psoas release at the level of the lesser trochanter is allowed.) An associated fixed flexion deformity of the knee may require hamstring lengthening as well.

Hip internal rotation deformity Internal rotation is usually associated with flexion and adduction. If so, adductor release and psoas lengthening will be helpful (and perhaps also medial hamstring lengthening). If, after a few years, rotation is still excessive, a derotation osteotomy of the femur (subtrochanteric or supracondylar) may be considered; however, be warned that this may have to be followed by a tibial derotation osteotomy to correct the compensatory external rotation deformity.

Hip subluxation Subluxation of the hip (Figure 10.21) occurs in about 30% of children with cerebral palsy. Femoral neck anteversion is not remodelled with growth in children with cerebral palsy and, when combined with flexion-adduction deformities, there is a risk of joint subluxation and acetabular dysplasia. This is particularly true in children who are not fully weight-bearing.

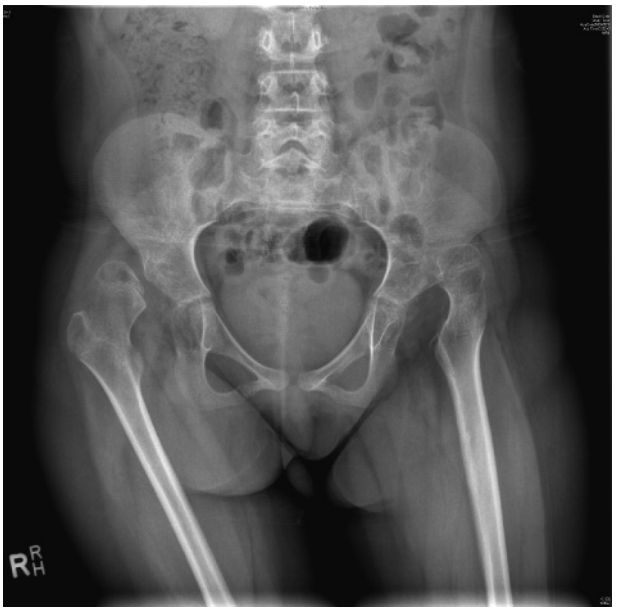
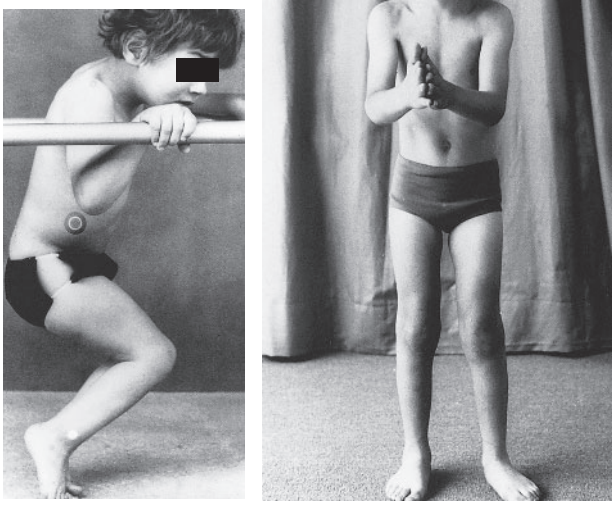


Figure 10.21 Bilateral CP AP pelvic radiograph of a child with bilateral cerebral palsy GMFCS IV: note the severe constipation, the straight spine and the level pelvis. The right leg is adducted and the hip essentially dislocated. The left hip is also dislocated: the femoral neck is valgus, as is often the case in children with an underlying neurological diagnosis.

Correction of flexion and adduction deformities (see above) before the age of 6 years may have a role in *preventing subluxation*. Older children may need a femoral varus derotation osteotomy, perhaps combined with some shortening and with acetabular reconstruction. In the adult walking diplegic patient, total hip replacement can be considered in selected cases where painful degenerative change is affecting function.

Knee flexion deformity This is one of the commonest deformities; it is usually due to functional hamstring tightness (Figure 10.22) but is often aggravated by hip flexion and/or weakness of ankle plantarflexion. The popliteal angle is one measure of hamstring tightness, but it can be difficult to distinguish between a functionally short hamstring muscle and one that is truly contracted; gait analysis can be useful in this regard.

Overlengthening of the hamstrings will result in an anteriorly tilted pelvis and a lumbar lordosis. Capsular contracture of the knee joint is uncommon. Fractional lengthening of the hamstrings (medial more often than medial and lateral combined) reliably improves gait mechanics but risks weakening hip extension and giving rise to an anteriorly tilted pelvis which, in turn, exacerbates the hip flexion/lumbar lordosis posture. Fractional lengthening of semimembranosus can be



(a) (b)

Figure 10.22 Spastic knee flexion deformity (a) This boy has spastic flexion of the knees due to tight hamstrings. (b) Here he is after hamstring release.

combined with detachment and transfer of semitendinosus to the adductor tubercle at the distal end of the femur.

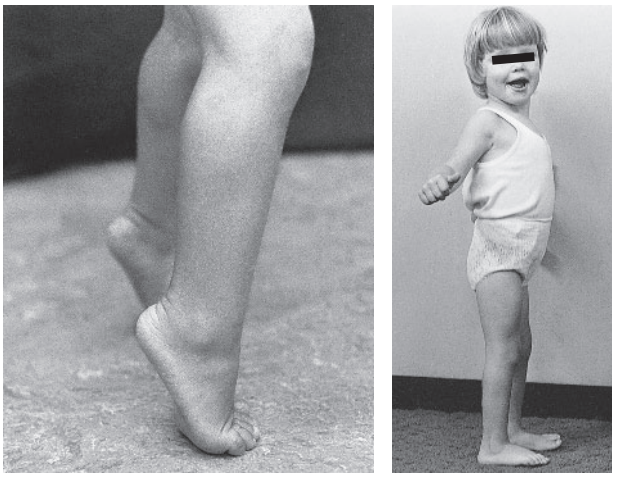
Severe flexion deformities (more than 25–30 degrees) can be treated by extension osteotomy of the distal femur or by physeal plating anteriorly. The risk of sciatic nerve injury can be mitigated by shortening the femur as it is extended. This combined with the pre-existing patella alta means that the quadriceps mechanism is defunctioned and thus the procedure must be combined with a patella tendon advancement or reefing procedure in order that active knee extension can be improved.

Remember that knee extension is aided by plantarflexion of the foot in walking, so it is important not to weaken the triceps surae by overzealous lengthening of the Achilles tendon (see below).

Knee extension deformity This can usually be corrected by simple tenotomy of the proximal end of rectus femoris.

External tibial torsion This is easily corrected by supramalleolar osteotomy, but before doing this it is important to ensure that the deformity is not actually advantageous in compensating for an ankle/hindfoot deformity (see below).

Ankle equinus The child with spastic diplegia usually toe-walks (Figure 10.23). This triggers an excessive plantarflexion–knee extension couple that may be manifested as knee hyperextension. In children with limited dorsiflexion, the gastrocnemius is often more



(a) (b)

Figure 10.23 Spastic equinus (a) Standing posture of a young girl with bilateral spastic equinus deformities. (b) Tendo Achillis lengthening resulted in complete correction and a balanced posture.

affected than the soleus. Selective fractional lengthening of the fascia/muscle is gaining favour but judicious percutaneous lengthening of the Achilles tendon may be required in certain cases. Relative overlengthening is a problem, particularly when associated knee flexion contractures exist.

If a *varus deformity* is present, treatment is as for the hemiplegic patient described above. The more common deformity is, however, one of *equinovalgus and a 'rocker-bottom' foot.* It makes the use of splints difficult and disrupts the plantarflexion–knee extension couple, exacerbating a knee flexion posture. It is important to note whether the hindfoot deformity is reducible or not. Correction can be achieved by either a calcaneal lengthening or displacement osteotomy; in some cases a subtalar fusion may be required. Such surgery must be combined with a release of tight structures (such as the Achilles tendon) and possibly peroneal tendon lengthening and plication of the medial structures when appropriate.

External tibial torsion may be corrected by a supramalleolar osteotomy but remember that an externally rotated gait pattern may be compensating for an inability of the foot to clear the ground when walking because of weak muscles/stiff joints.

Single event multilevel surgery (SEMLS) The diplegic patient usually has problems at all levels and often the most appropriate way to improve gait and overall function is to enhance the mechanical efficiency of all three joints by operating at hip, knee and ankle at the same time. Soft-tissue and bony surgery to both limbs can be performed at one sitting or staged over a few weeks. Postoperative rehabilitation is complex and time-consuming but the results can be very rewarding.

Total body involvement (TBI)

With total body involvement all parts of the body are affected; function is generally poor and the aims of surgical intervention differ significantly from those for the hemiplegic or walking diplegic patient. Patients with TBI are often GMFCS IV and V and therapeutic standers only; they often have significant comorbidities which result in increased complication rates following operative intervention. In developed countries the probability of such children reaching their twenty-first birthday is less than 40%.

HIP

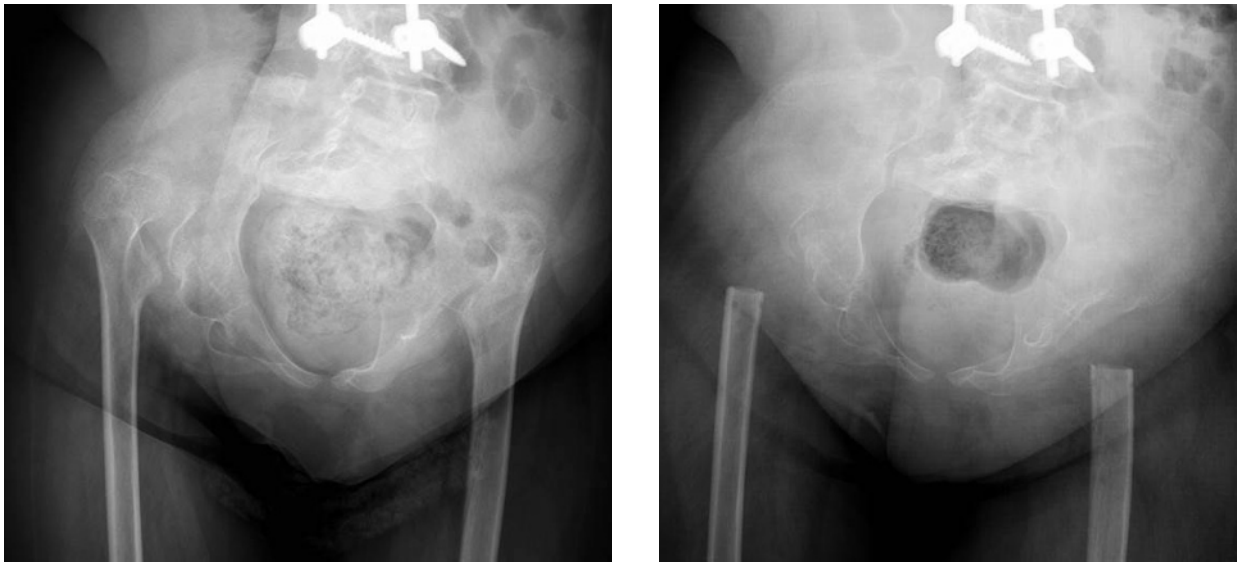
Hip subluxation progressing to dislocation is common. The adduction and flexion contractures outlined above are more frequent and more severe in this group of patients, leaving the hip at risk of developing subluxation with acetabular dysplasia. Hips are often 'windswept' (one hip lying adducted, flexed and internally rotated while the other lies in abduction and external rotation and often more extended).

The hip at risk of subluxation must be watched closely and, if necessary, treated by adductor and psoas releases as outlined above (a psoas tenotomy at the lesser trochanter is appropriate). The migration percentage (MP) measures the proportion of the proximal femoral epiphysis that is uncovered. Hip subluxation, defined as a MP of more than 33%, may require a femoral varus derotation (and shortening) osteotomy as well as an acetabular procedure for correction in addition to the soft-tissue releases. If the hip has dislocated, open reduction, release of soft tissues and bony realignment will be necessary (Figure 10.24). Long-standing dislocation in a non-walker may be impossible to reconstruct; if pain, problems with perineal care and poor tolerance of a sitting position make operation imperative, the proximal end of the femur can be excised.

The opposite hip may require similar surgery or, in the case of a windswept deformity, it may benefit from a release of the hip abductors and extensors, mainly the gluteus maximus and the iliotibial band in addition to a femoral osteotomy.

This is complex surgery and the complication rates are high. Some families, and indeed some surgeons, opt for no active treatment of the subluxed or dislocated hip, particularly if it is relatively pain-free and care of the child is not compromised significantly. Others feel that hip subluxation/dislocation should be prevented at all costs and, although recent reports from Scandinavia suggest that hip dislocation is 'preventable', this is only true with an aggressive regimen of tone management and surgery which many people feel causes unnecessary suffering to the child concerned. Obviously, the management of such cases brings up moral dilemmas which are best dealt with by maintaining good communication with the families and therapists at all stages and being clear about the aims of any intervention.

While 90% of hips that dislocate do so posteriorly, anterior dislocations do occur (Figure 10.25). The X-rays may look surprisingly normal but the clinical picture will show a lump in the groin and the abducted, externally rotated leg cannot be flexed or brought into a neutral sitting position. This, too,



(a) (b)

Figure 10.24 Hip involvement (a) AP pelvic radiograph of a teenage patient with bilateral painful, chronically dislocated hips secondary to CP: the spine had already been fused. With stiff and painful hips, it was difficult to sit the patient comfortably. (b) Following bilateral proximal femoral excisions, the legs were much looser and the legs could be placed wherever necessary for comfortable seating in the wheelchair.

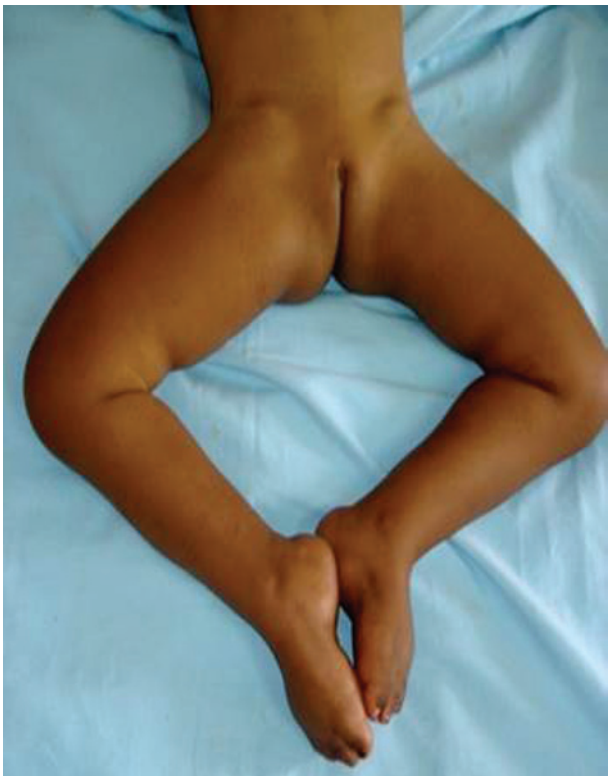


Figure 10.25 Clinical photograph A child with bilateral anterior hip dislocations: the femoral heads are visible in the groin.

may need reconstruction or indeed proximal femoral excision.

SPINE/PELVIS

Scoliosis is common and probably affects more than 50% of this group of patients. The deformity is often a long C-shaped thoracolumbar curve and it frequently incorporates the pelvis, which is tilted obliquely so that one hip is abducted and the other adducted and threatening to dislocate. Of course, the adducted hip may be the primary problem with pelvic obliquity and scoliosis following; in essence, trunk muscle involvement due to the cerebral palsy must be a major determinant of developing deformity.

Various forms of non-operative treatment have been used, and in some cases patients opt for long-term use of an adapted wheelchair.

Where facilities and surgical expertise are available, operative correction and spinal stabilization are often advocated. Indications are a progressive curve of more than 40 degrees in a child over 10 years, inability to sit without support, and a range of hip movement that will allow the child to sit after spinal stabilization. Fixation is achieved with pedicle screws and rods extending from the thoracic spine to the pelvis; there is an attempt to recreate a lumbar lordosis but in so doing it may, at least temporarily, exacerbate hamstring tightness, making sitting more difficult.

Careful preoperative evaluation including nutritional status and bone health is essential to ensure that the child is fit for a long and difficult operation that is known to carry a high complication rate, including neurological defects, problems with wound healing and implant failure. This type of spinal surgery has been shown to increase life expectancy, but demonstrating a concurrent improvement in quality of life has been more difficult to prove.

OTHER JOINTS

Surgery to other joints may be required and follows the principles outlined above for the hemiplegic and diplegic patient.

ADULT-ACQUIRED SPASTIC PARESIS

Cerebral damage following a *stroke* or *head injury* may cause persistent spastic paresis in the adult; this can be accompanied by disturbance of proprioception and stereognosis.

In the early recuperative stage, physiotherapy, splintage and tone-reducing medications, including BoNT-A injections, are used to prevent fixed deformities; all affected joints should be put through a full range of movement every day.

Deformities that are passively correctible should be splinted in the neutral position until controlled muscle power returns; proprioception and coordination can be improved by occupational therapy. Yet even with the best attention, these measures may fail to prevent the development of fixed deformities. Once maximal motor recovery has been achieved, usually by 9 months after a stroke but more than a year after a brain injury, residual deformities or joint instability should be considered for operative treatment. The patient should have sufficient cognitive ability, awareness of body position in space and good psychological impetus if a lasting result is to be expected.

In the lower limbs the principal deformities requiring correction are equinus or equinovarus of the foot, knee flexion and hip adduction. In the upper limb (where the chances of regaining controlled, functional movement are less) the common residual deformities are adduction and internal rotation of the shoulder (often accompanied by shoulder pain), and flexion of the elbow, wrist and metacarpophalangeal joints. Treatment is similar to that outlined for the management of spastic UMN lesions in the child, and is summarized in Table 10.3.

LESIONS OF THE SPINAL CORD

The three major pathways in the spinal cord are the corticospinal tracts (in the anterior columns) carrying motor neurons, the spinothalamic tracts carrying

sensory neurons for pain, touch and temperature, and the posterior (dorsal) column tracts serving deep sensibility (joint position and vibration) (see Figure 10.2).

Clinical features

True lesions of the spinal cord present with a UMN spastic paresis and often a fairly precise sensory level that suggests the level of cord involvement. However, extradural compressive lesions will often involve the nerve roots as well resulting in a combination of UMN and LMN signs.

Patients often complain of weakness and numbness with loss of balance and possibly alteration in bowel or bladder control and, in men, impotence. The symptoms may be of variable severity and the speed of onset is similarly variable, depending largely on the aetiological factor.

Several 'classical' patterns are recognized.

Cervical cord compression The patient typically presents with UMN symptoms in the lower limbs (stiffness and a change in walking pattern) and LMN signs in the upper limbs (complaints of numbness and clumsiness). Pain is a variable feature. Bladder symptoms of frequency and incontinence are more common than retention.

A central cord syndrome is caused classically by a hyperextension injury in a middle-aged patient with long-standing cervical spondylosis, or it may develop with syringomyelia. In these cases there is disproportionately more UMN weakness in the upper limbs compared to the lower limbs with bladder dysfunction and a variable sensory loss below the lesion.

Thoracic cord compression This typically presents as a UMN paralysis affecting the lower limbs, together with variable sensory loss depending on the degree of involvement of the dorsal columns or the spinothalamic tracts.

Lumbar cord compression The spinal cord terminates around the level of L1 so compression here may involve the conus medullaris or the cauda equina or both, giving a mixture of UMN and LMN signs. The typical *cauda equina syndrome* consists of lower limb weakness, absent reflexes, impaired sensation and urinary retention (with overflow perhaps mimicking incontinence).

Brown-Séquard lesion The pure form of this syndrome is very unusual but less pure forms are common and serve as a reminder that careful assessment of the neurological symptoms and signs is important in helping the clinician to localize the pathology and understand its aetiology. The pure lesion is defined as an incomplete hemispherical cord lesion: below the lesion there is ipsilateral UMN weakness and posterior column dysfunction, with contralateral loss of skin sensibility; at the level of the lesion there is ipsilateral loss of sensibility (Figure 10.26).

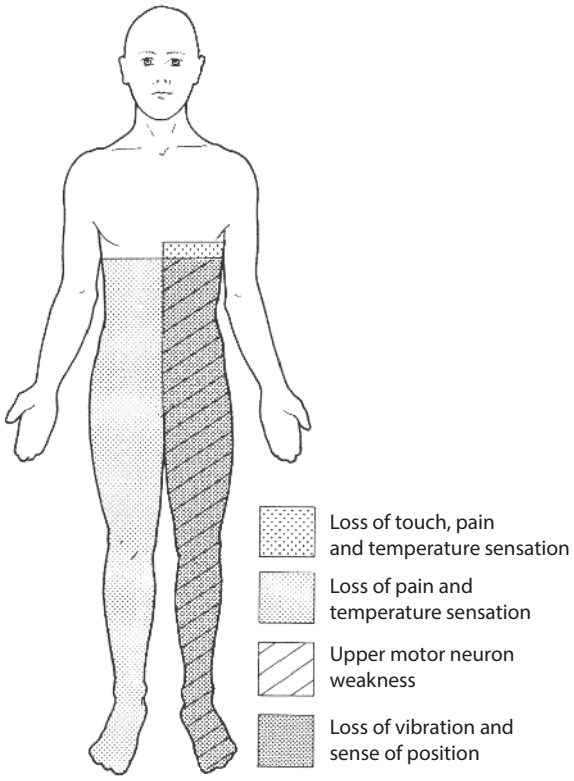


Figure 10.26 The Brown-Séquard syndrome

SPINAL SHOCK: Do not forget that acute cord lesions at any level may present with a flaccid paralysis which resolves over time, usually to reveal the more typical UMN signs associated with cord injury.

Diagnosis and management

The more common causes of spinal cord dysfunction are listed in Box 10.2. Traumatic and compressive lesions are the ones most likely to be seen by orthopaedic surgeons. Plain X-rays will show structural abnormalities of the spine; cord compression can be visualized by MRI as can intrinsic lesions of the cord. Further investigation with blood tests, CSF examination or gadolinium enhanced MRI may be helpful.

Acute compressive lesions These require urgent diagnosis and treatment if permanent damage is to be prevented. Bladder dysfunction is ominous: whereas motor and sensory signs may improve after decompression, loss of bladder control for more than 24 hours is usually irreversible.

BOX 10.2 CAUSES OF SPINAL CORD DYSFUNCTION

Acute injury
- Vertebral fractures
- Fracture-dislocation

Infection
- Epidural abscess
- Poliomyelitis

Intervertebral disc prolapsed
- Sequestrated disc
- Disc prolapse in spinal stenosis

Vertebral canal stenosis
- Congenital stenosis
- Acquired stenosis

Spinal cord tumours
- Neurofibroma
- Meningioma

Intrinsic cord lesions
- Tabes dorsalis
- Syringomyelia
- Other degenerative disorders

Miscellaneous
- Spina bifida
- Vascular lesions
- Multiple lesions
- Multiple sclerosis
- Haemorrhagic disorders

Spinal injury Spinal injury is discussed in detail in Chapter 25 but a few important points deserve mention here.

- Any spinal injury may be associated with cord damage, and great care is needed in transporting and examining the patient.
- In the early period of 'spinal shock' the usual picture is one of flaccid paralysis, with or without priapism.
- Plain X-rays seldom show the full extent of bone displacement, which is much better displayed by CT or MRI. Plain films do not delineate the soft-tissue damage and spinal cord injuries do occur without radiographic abnormality (SCIWORA).
- Unstable injuries usually need operative decompression and/or stabilization; stable injuries can be treated conservatively.
- Many centres consider the use of corticosteroids beneficial in terms of reducing the degree of permanent neurological damage, but the side effects of gastrointestinal haemorrhage and avascular necrosis are potentially serious.

Epidural abscess This is a surgical emergency. The patient develops acute pain and muscle spasm, with fever, leucocytosis and elevation of the ESR and CRP. X-rays may show disc space narrowing and bone erosion. Treatment is by immediate decompression and antibiotics.

Acute disc prolapse This usually causes unilateral symptoms and signs. However, complete central lumbar disc prolapse may present as a cauda equina syndrome with urinary retention and overflow; spinal canal obstruction is demonstrated by MRI. Operative discectomy is urgent.

Chronic discogenic disease This is often associated with narrowing of the intervertebral foramina and compression of nerve roots (radiculopathy), and occasionally with bony hypertrophy and pressure on the spinal cord (myelopathy). Diagnosis is usually obvious on X-ray and MRI. Operative decompression may be needed.

Spinal stenosis This produces a typical clinical syndrome, due partly to direct pressure on the cord or nerve roots and partly to vascular obstruction and ischaemic neuropathy during hyperextension of the lumbar spine. The patient complains of 'tiredness', weakness and sometimes aching or paraesthesia in the lower limbs after standing or walking for a few minutes, symptoms that are relieved by bending forward, sitting or crouching so as to flex the lumbar spine. This symptom complex is known as spinal claudication.

Congenital narrowing of the spinal canal is rare, except in developmental disorders such as achondroplasia, but even a moderately reduced canal may be further narrowed by osteophytes, thus compromising the cord and nerve roots.

Treatment calls for bony decompression of the nerve structures.

Vertebral disease Conditions such as tuberculosis or metastatic disease, may cause cord compression and paraparesis. The diagnosis is usually obvious on X-ray, but a needle biopsy may be necessary for confirmation.

Management is usually by anterior decompression and, if necessary, internal stabilization. However, in metastatic disease, if the prognosis is poor, it may be wise also to use radiotherapy and corticosteroids, plus narcotics for pain.

Spinal cord tumours Tumours are a comparatively rare cause of progressive paraparesis. X-rays may show bony erosion, widening of the spinal canal or flattening of the vertebral pedicles. Widening of the intervertebral foramina is typical of neurofibromatosis. Treatment usually involves operative removal of the tumour.

Intrinsic lesions Lesions within the cord produce slowly progressive neurological signs. Two conditions in particular – *tabes dorsalis* and *syringomyelia* – may present with orthopaedic problems because of neuropathic joint destruction.

Tabes dorsalis Tabes (meaning 'wasting') represents a late manifestation of syphilis, causing degeneration of the posterior columns of the spinal cord. A pathognomonic feature is 'lightning pains' in the lower limbs. Much later other neurological features appear: sensory ataxia, which causes a stamping gait; loss of position sense and sometimes of pain sensibility; trophic lesions in the lower limbs; progressive joint instability; and almost painless destruction of joints (Charcot joints). There is no treatment for the cord disorder.

In *syringomyelia* a long cavity (the syrinx) filled with CSF develops within the spinal cord, most commonly in the cervical region. Usually the cause is unknown but the condition is sometimes associated with tumours, or spinal cord injury in adults and congenital anomalies with hydrocephalus and herniation of the cerebellar tonsils in children.

Symptoms and signs are most noticeable in the upper limbs. The expanding cyst presses on the anterior horn cells, producing weakness and wasting of the hand muscles. Also, destruction of the decussating spinothalamic fibres in the centre of the cord produces a characteristic dissociated sensory loss in the upper limbs: impaired response to pain and temperature but preservation of touch. There may be trophic lesions in the fingers and neuropathic arthropathy ('Charcot joints') in the upper limbs. CT may reveal an expanded cord and the syrinx can be defined on MRI. Deterioration may be slowed down by decompression of the foramen magnum.

SPINA BIFIDA

Spina bifida (Figures 10. 27 and 10.28) is a congenital disorder in which the two halves of the posterior vertebral arch fail to fuse at one or more levels. This neural tube defect, or spinal dysraphism, which occurs within the first month of fetal life, usually affects the lumbar or lumbosacral segments of the spine. In its most severe form, the condition is associated with major neurological problems in the lower limbs together with bowel and bladder incontinence.

Pathology

In the mildest forms of dysraphism, *spina bifida occulta*, there is a midline defect between the laminae and nothing more; hence the term 'occulta' (Figure 10.29a). Most cases are discovered incidentally on spine X-rays, usually affecting the L5 level.

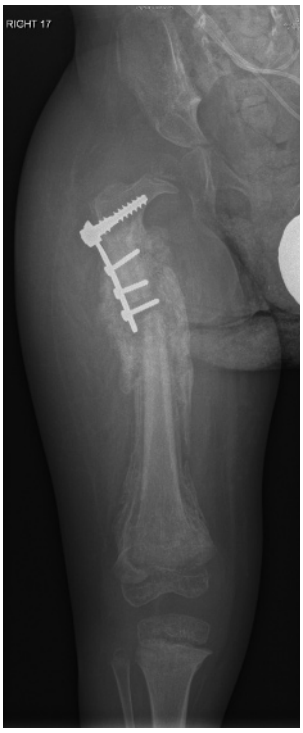


Figure 10.27 Spina bifida AP radiograph of the right femur of a patient with spina bifida who underwent an open reduction and femoral osteotomy for a hip dislocation. Subsequently, when he came out of plaster, he fractured his right femur through the bottom screw hole of his femoral plate and just above the distal femoral physis in two separate incidents during the rehabilitation phase. Both fractures healed with exuberant callus formation which led to problematic knee stiffness. This, in turn, limited the child's floor mobility. Fractures are not uncommon due to the relatively reduced density and the lack of sensation.

Figure 10.28 The same child as in Figure 10.27. He prefers to be wheelchair-based rather than to try to stand and walk. He is a keen athlete and has requested that his knees stay 'flexed'. His stepbrother has Down's syndrome: his issues with balance mean that he has not yet learnt to walk independently.

In the more overt forms of dysraphism the vertebral laminae are missing and the contents of the vertebral canal prolapse through the defect. The abnormality takes one of several forms. The least disabling is a *meningocele*, which accounts for only about 5% of cases of true spina bifida (Figure 10.29b). The dura mater is open posteriorly but the meninges are intact and a CSF-filled meningeal sac protrudes under the skin. The spinal cord and nerve roots remain *inside* the vertebral canal and there is usually no significant neurological abnormality.

By far the most common and most serious abnormality is a *myelomeningocele*, which usually occurs in the lower thoracic spine or the lumbosacral region (Figure 10.29c). Part of the spinal cord and nerve roots prolapse into the meningeal sac. In some cases the neural tube is fully formed and the sac is covered by a membrane and/or skin – the myelomeningocele. In others the cord is in a more primitive state, the unfolded neural plate is effectively open to the environment and there is no sac – an *'open' myelomeningocele* (Figure 10.29d). This is always associated with a neurological deficit distal to the level of the lesion. If neural tissue is exposed to the air, it may become infected, leading to more severe abnormality and even death.

Tethering of the cord distally may cause problems proximally, with herniation of the cerebellum and brainstem through the foramen magnum, resulting in obstruction to CSF circulation and *hydrocephalus*. The ventricles dilate and the skull enlarges by separation of the cranial sutures. A ventriculoperitoneal (VP) shunt may be indicated. Persistently raised intracranial pressure may cause cerebral atrophy and learning difficulties.

Incidence and screening

Isolated laminar defects are seen in over 5% of lumbar spine X-rays but true spina bifida is rare at 2–3 per 1000 live births. However, if one child is affected, the risk for future siblings is significantly higher.

Neural tube defects are associated with high levels of alpha-fetoprotein (AFP) in blood and in the amniotic fluid, and this offers an effective method of antenatal screening during the pregnancy.

Maternal blood testing is performed routinely at 15–18 weeks and followed by an amniocentesis if necessary. A mid-term high-resolution ultrasound scan will detect 95% of cases of spina bifida and, in many countries, counselling regarding a termination is offered. If the pregnancy is continued, arrangements should be made to ensure that appropriate services are available at birth and in the neonatal period to minimize the risk of further neurological damage.

Folic acid, 400 micrograms daily taken before conception and continuing through the first 12 weeks of pregnancy, has been shown to reduce the risk of neural-tube defects in the fetus.

Clinical features

EARLY DIAGNOSIS

The major neural-tube defects can be detected easily on antenatal scans or identified immediately at birth.

Myelomeningocele and *meningoceles* are usually obvious at birth in the shape of a saccular lesion overlying the lumbar spine. It may be covered only with membrane, or with membrane and skin. In open myeloceles the neural elements form the floor of the cyst, and CSF leaks continuously from the open cord.

The baby's posture may suggest some type of paralysis, or even the neurological level of the lesion. Deformities of the lower limbs such as equinovarus or calcaneovalgus of the feet, recurvatum of the knee and hip dislocation are common and probably due to a combination of factors such as muscle imbalance, lack of movement and abnormal limb position *in utero*, or to associated anomalies that are independent of the paralysis.

Muscle charting, although difficult, is possible in the neonate and should be performed so that neurological deterioration can be identified promptly. In about one-third of infants with myelomeningocele

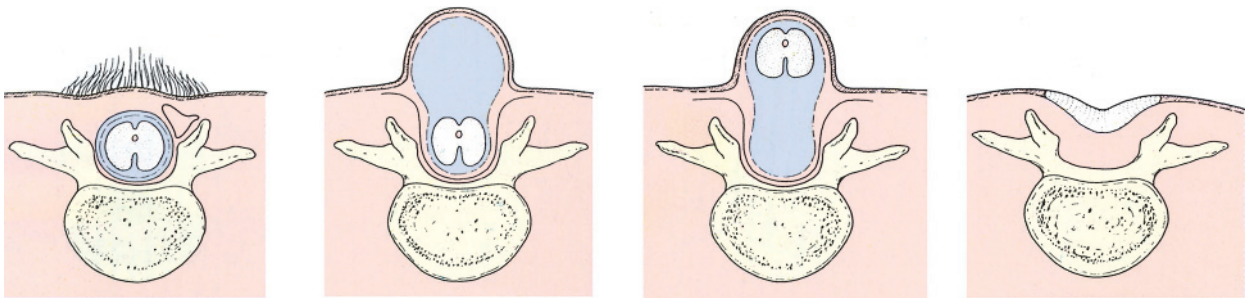
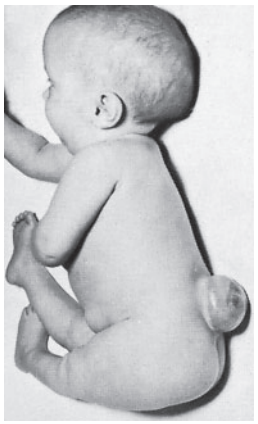


Figure 10.29 Dysraphism (a) Spina bifida occulta. (b) Meningocele. (c) Myelomeningocele. (d) Open myelomeningocele.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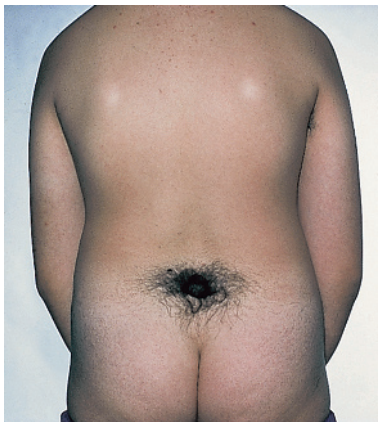
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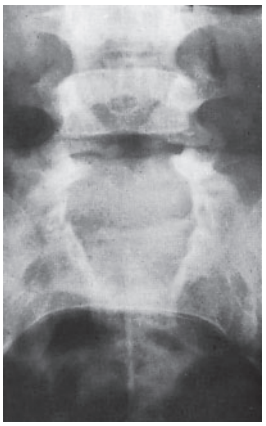

(c)

Figure 10.30 Spina bifida (a) Baby with spina bifida cystica (myelomeningocele). (b) Tuft of hair over the lumbosacral junction. X-ray in this case showed a sacral defect (c).

there is complete LMN paralysis and loss of sensation and sphincter control below the affected level; in one-third there is a complete lesion at some level but a distal segment of cord is preserved, giving a mixed neurological picture with intact segmental reflexes and spastic muscle groups; in the remaining third the cord lesion is incomplete and some movement and sensation are preserved.

Spina bifida occulta is often encountered by accident as an incidental finding on an abdominal X-ray and can usually be ignored. However, in some cases, and especially if several vertebrae are affected, there are clinical signs in the overlying skin, such as a dimple, a pit, a tuft of hair or a pigmented lesion, which may signify something more serious (Figure 10.30). Occasionally there are associated intraspinal anomalies, such as tethering of the conus medullaris below L1, splitting of the spinal cord (diastematomyelia) and cysts or lipomas of the cauda equina that might need to be identified. In the neonate, an USS can identify spina bifida and a low-lying cord but, in the older infant, MRI is most appropriate.

Children may present with mild neurological symptoms (enuresis, urinary frequency or intermittent incontinence) and neurological examination may reveal weakness and some loss of sensibility in the lower limbs.

Hydrocephalus may be present at birth; with a communicating hydrocephalus the intracranial pressure may not be elevated until leakage from the spinal lesion is arrested by surgical closure of the lesion.

Plain X-rays may show the laminar defect and any associated vertebral anomalies; a midline ridge of bone suggests bifurcation of the cord (diastematomyelia). Intraspinal anomalies are best shown by MRI.

DIAGNOSIS IN OLDER CHILDREN

The minor forms of spina bifida may present clinically at any age. The physical signs mentioned above may have been noted previously and the child (or teenager) now presents with clawing of the toes, a change in gait pattern, incontinence or abnormal sensation. This delayed presentation is often attributed to the *tethered cord syndrome*. Tethering may be secondary to the early surgical reconstruction of the major defect or to conditions such as a diastematomyelia, and with growth there is progressive damage to the cord and/or nerve roots. MRI with gadolinium enhancement is the investigation of choice, and neurosurgical release before any further neurological damage occurs is the treatment of choice.

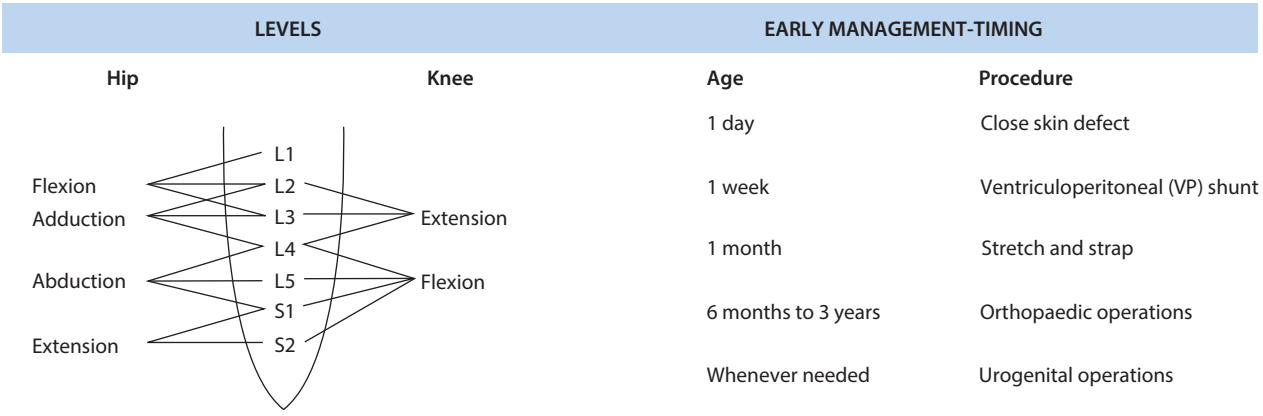
Any orthopaedic deformity secondary to the altered neurological status should be reassessed following release of the tether in case there has been a significant improvement in, for example, the muscle strength or sensation which might influence the choice of surgical procedure. Fixed contractures will need releases and muscle imbalance may need to be addressed with tendon transfers and/or corrective orthoses.

Older children with neurological lesions are liable to suffer fractures after relatively minor injuries (see Figure 10.27). These may not always be obvious clinically and the altered sensation means pain may not be a major feature, but suspicion should be raised by the appearance of swelling, warmth and redness in the limb.

Treatment

In recent years intrauterine surgery (open or fetoscopic) has been attempted: closure of the defect is possible but a reduction in neurological disability has not yet been identified.

After birth, care must be taken to dress the 'wound' and prevent infection of these vulnerable tissues. Formal neurosurgical closure of the defect should take place within 48 hours of birth in order to prevent drying and ulceration, or infection of the lesion (Figure 10.31). All neural tissue should be carefully preserved and covered with dura; the skin is then undercut widely to facilitate complete closure. Some



EARLY MANAGEMENT-TIMING

Age	Procedure
1 day	Close skin defect
1 week	Ventriculoperitoneal (VP) shunt
1 month	Stretch and strap
6 months to 3 years	Orthopaedic operations
Whenever needed	Urogenital operations

Figure 10.31 Spina bifida The diagram shows the root levels concerned with hip and knee movements. The table is a simple guide to the timing of operations.

centres avoid urgent operation if the neurological level is high (above L1), if spinal deformities are severe or if there is marked hydrocephalus.

A few weeks later, when the back has healed, the degree of *hydrocephalus* is assessed. Almost all children also have the *Arnold–Chiari* malformation with displacement of the posterior fossa structures through the foramen magnum. Thus, up to 90% of children may require active management of their real or potential hydrocephalus in the form of a ventriculoperitoneal shunt (VP shunt) to reduce the risk of further damage to their CNS. A chronically raised intracranial pressure may be associated with learning difficulties and other problems. Similarly, if a child's neurological status changes unexpectedly, shunt problems such as infection/blockage should be considered.

VP drainage can be maintained (if necessary, by changing the valve as the baby grows) for 5–6 years, by which time the tendency to hydrocephalus usually ceases.

Management of neonatal deformities will vary depending on the overall clinical picture, but physiotherapy and/or splinting will be the mainstays of early treatment. It must be remembered that the skin is likely to be insensate and pressure area care is essential.

In the more severe forms of spina bifida, there must be a multidisciplinary approach to treatment from early infancy through to adulthood. Orthopaedic management is important but so is the management of the neurological lesion in terms of urological function and bladder/bowel control. The vast majority of patients have urological problems necessitating the use of catheters or urinary diversion. Medical management of the detrusor muscle overactivity may include the use of botulinum toxin injections.

The psychosocial aspects of the condition must also be borne in mind; they can be overwhelming to the child and his or her family and require patient attention.

ORTHOPAEDIC MANAGEMENT

The orthopaedic surgeon, working as part of a team, must identify the important treatment goals while bearing in mind some basic observations:

- Except in the mildest cases, the late functional outcome cannot be predicted until the child is assessed both intellectually and in terms of neuromuscular function around the age of 3–4 years.
- Most patients with myelomeningocele will never be functionally independent.
- The maintenance and development of intellectual skills and upper-limb function are often more important for independence in the activities of daily living than walking and, for many patients, the ability to sit comfortably is more important than the ability to stand awkwardly.
- The best predictor of walking ability and function is the motor level of the paralysis. Children with lesions below L4 will have quadriceps control and active knee extension and should be encouraged to walk. Children with higher lesions may start off walking with the aid of orthotic devices but they are likely to opt for a wheelchair with time.
- Immobilization and muscle imbalance both lead to joint deformity and the risk of pathological fracture. Physiotherapists working to correct, or indeed prevent, joint deformity must understand the risk of fracture, and orthotists must take into consideration the need for lightweight appliances and beware the risk of pressure sores when using splints on insensate skin.
- *Latex allergy* is present in some children with spina bifida and a history of allergic reactions should be noted. All treatment, including surgery, must be conducted in a latex-free environment. If a positive history is identified, antihistamines and/or corticosteroids should be given.

Regional survey

Spine

Spinal deformity (scoliosis and/or kyphosis) is common in children with myelomeningocele, due to a combination of muscle weakness and imbalance, associated congenital vertebral anomalies (in about 20% of cases) and the tethered cord syndrome.

Distal tethering of the cord or other neural elements is almost inevitable after repair of a myelomeningocele; this may be harmless, but it can cause pain and progression of neurological dysfunction during phases of rapid growth, and in some cases it gives rise to a scoliosis. Diagnosis may be aided by MRI. Indications for operative release of the tethered cord are increasing pain and neurological dysfunction or progressive spinal deformity.

Kyphosis This may result in stretching and breakdown, or chronic ulceration, of the overlying skin posteriorly and compression of the abdominal and thoracic viscera anteriorly. Treatment is difficult and may require localized vertebral resection and arthrodesis. However, the cord at the affected level is often non-functioning and therefore the risks of further neurological insult influencing the outcome are small.

Neuromuscular scoliosis This appears as a long C-shaped curve which is usually progressive and makes sitting particularly difficult. It is unlikely to respond to a brace. Moulded seat inserts for the wheelchair are essential to aid sitting balance and independence and may help reduce the rate of curve deterioration. Surgery via an anterior, a posterior or a combined approach is often necessary, and fusion to the pelvis may be required, although this tends to reduce walking ability in ambulant patients – at least temporarily. The operation is always difficult and carries a high risk of complications, particularly postoperative infection and implant failure.

Hip

Patients with spina bifida present a wide spectrum of hip problems, the management of which is still being debated (Figure 10.32). In our approach the general aim is to secure hips that have enough movement (and muscle strength) to enable the child both to stand up in knee–ankle–foot orthoses (KAFOs) or reciprocating gait orthoses (RGOs) and to sit comfortably.

If the neurological level of the lesion is above L1, all muscle groups are flaccid and splintage is the only option; in the long term, the child will probably use a wheelchair. With lesions below S1, a hip flexion contracture is the most likely problem and this can be corrected by elongation of the psoas tendon combined with detachment of both heads of the rectus femoris from the ilium.

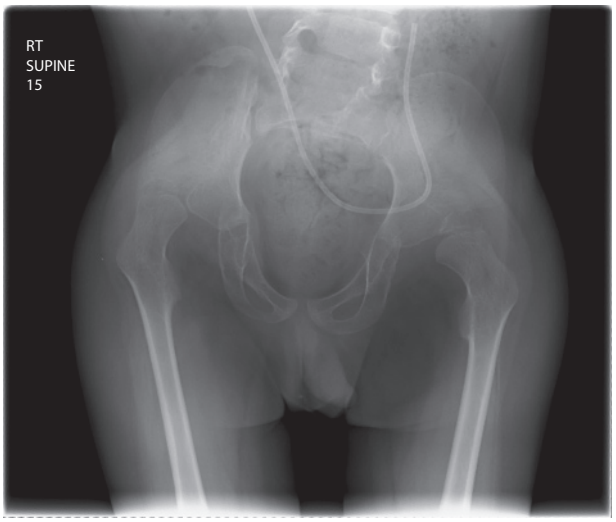


Figure 10.32 Spina bifida – pelvis AP pelvic radiograph of a young boy with spina bifida. Note the presence of the VP shunt in the abdomen, the bilateral hip dislocations and the widened pedicles in the lower lumbar vertebrae which implies that the posterior elements have not been formed well. The child is constipated: a common problem in children with neurological disabilities.

For children with 'in-between' lesions, muscle imbalance is the main problem and many hips (up to 50%) will sublux or dislocate by early childhood. The effect of hip joint subluxation or dislocation (and its associated pelvic obliquity) on spinal development is unclear, but the natural history of hip joint function in these children can be surprisingly good, perhaps because of the associated sensory loss and lack of overt spasticity. This has led to the recognition that retaining hip movement may be more useful than striving for hip reduction by multiple operations, with their attendant complications and uncertain prognosis. So, while in CP, despite a lack of evidence that surgery benefits quality of life, there is an increasing trend for aggressive surgery to maintain hip reduction, in contrast in spina bifida the lack of convincing evidence to suggest that function is improved significantly by operative hip relocation means a cautious approach is still advocated.

Knee

Unlike the hip, the knee usually presents no problem, because the aim is simple: a straight knee suitable for wearing orthoses and using gait-training devices. In older children fixed flexion may follow prolonged sitting. If physiotherapy and/or the use of orthotics fails to correct this deformity, one or more of the hamstrings may be lengthened, divided or reinserted into the femur or patella; this may have to be combined with a posterior capsular release. However, if

the likely prognosis is that the patient will be wheelchair-dependent, flexion contractures are, of course, less of a problem.

Some children are born with a hyperextension contracture and on occasion the hamstring tendons are subluxed anteriorly. Physiotherapy and sometimes serial casting are the treatments of choice initially but a V–Y quadricepsplasty and hamstring lengthening/relocation may be required in order to achieve enough passive knee flexion to facilitate sitting and standing. Following surgical release, it is difficult to maintain any degree of functional quadriceps activity and thus orthotic devices are required for standing/walking.

Walking patients often develop a valgus knee, in some cases with torsional abnormalities in the lower limb. Secondary joint instability can further exacerbate the problems of walking, with patients relying more and more on the use of forearm crutches and a swing-through gait.

Foot

Foot deformities are among the most common problems in children with spina bifida. The aim of treatment is a mobile foot, with healthy skin and soft tissues that will not break down easily, so that it can be held or braced in a plantigrade position.

A flail foot or one that has a balanced paralysis or weakness is relatively easy to treat and only requires the use of carefully made orthoses (e.g. an ankle–foot orthosis) or occasionally just well-fitting ankle boots. Appropriate padding is vital to avoid skin breakdown.

A talipes equinovarus deformity is likely to be more severe (and more resistant to treatment) than an idiopathic clubfoot deformity. The standard treatment is now to use the Ponseti technique of gentle manipulation towards progressive correction, holding the feet in well-moulded plaster casts which are changed weekly until all but the equinus has been corrected. Most cases will require a percutaneous tendo Achillis tenotomy to achieve full correction. The relapse rate is high and, depending on the child's level of function, further surgical treatment may be required with bony procedures reserved for residual or recurrent deformity in the older child in whom it is still important to obtain a plantigrade foot.

A vertical talus deformity can be treated in a similar way by a 'reverse Ponseti' regimen and transfer of the tibialis anterior tendon to the neck of the talus, but surgical correction of this deformity is often required.

Toe deformities (or indeed flail toes) sometimes cause concern because of pressure points and difficulty fitting shoes. 'Orthopaedic shoes' with a high toe box may be needed and could be more appropriate than surgical intervention.

SACRAL AGENESIS

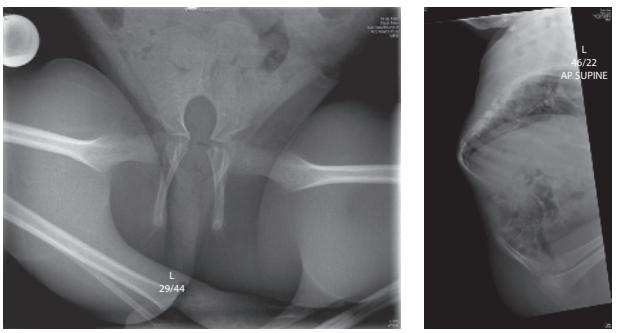
Sacral agenesis or caudal regression syndrome is a very rare congenital disorder in which there is abnormal development of the lower spine (the caudal partition of the spine) and other organ systems in the lower half of the body such as the urogenital and gastrointestinal systems (Figure 10.33). The lower-limb function depends to a large extent on the severity of the neurological developmental problem. Some patients can walk and may behave much like a low-level spina bifida but others with major anomalies may have an absent sacrum and lower lumbar spine with legs fixed in a 'Buddha position'.

POLIOMYELITIS

Poliomyelitis is an acute infectious viral disease, spread by the oropharyngeal route that passes through several distinct phases. Only around 10% of patients exhibit any symptoms at all, and involvement of the CNS occurs in less than 1% of cases with effects on the anterior horn cells of the spinal cord and brainstem leading to LMN (flaccid) paralysis of the affected muscle groups. The poliomyelitis viruses have varying virulence and in countries where vaccination is encouraged it has become a rare disease; since 1988 there has been a 99% decrease in the number of cases reported annually. However, the effects of previous infection are still with us today.

Clinical features

Poliomyelitis typically passes through several clinical phases, from an acute illness resembling meningitis to paralysis, then slow recovery or convalescence and



(a) (b)

Figure 10.33 Sacral agenesis (a) AP pelvis and (b) lateral spinal radiographs of a young girl with severe sacral agenesis. Her hips are in wide abduction and she sits in a Buddha position. The two iliac wings are fused together and there is no sacrum; in addition, the lateral view confirms there is no lumbar spine either. She has essentially no motor or sensory function below L2.

finally the long period of residual paralysis. The disease strikes at any age but most commonly in children.

The acute illness Early symptoms are fever and headache; in about one-third of cases the patient gives a history of prodromal symptoms of a sore throat, mild headache and slight pyrexia. As the symptoms increase in severity, neck stiffness appears and meningitis may be suspected. The patient lies curled up with the joints flexed; the muscles are painful and tender and passive stretching provokes painful spasms.

Paralysis Soon muscle weakness appears; it reaches a peak in the course of 2–3 days and may give rise to difficulty with breathing and swallowing. If the patient does not succumb from paralysis of the respiratory muscles, pain and pyrexia subside after 7–10 days and the patient enters the convalescent stage. However, he or she should be considered to be infective for at least 4 weeks from the onset of illness.

Recovery and convalescence A return of muscle power is most noticeable within the first 6 months, but there may be continuing improvement for up to 2 years.

Residual paralysis In some patients the illness does not progress beyond the early stage of meningeal irritation; some who develop muscle weakness do recover completely; in others recovery is incomplete and they are left with some degree of asymmetric flaccid (LMN) paralysis or unbalanced muscle weakness that in time leads to joint deformities and growth defects (Figure 10.34). Although sensation is intact, the limb often appears cold and blue.

Post-polio syndrome Although it was generally believed that the pattern of muscle weakness became firmly established by 2 years, it is now accepted that in up to 50% of cases reactivation of the virus results in progressive muscle weakness in both old and new muscle groups, giving rise to unaccustomed fatigue. If this occurs in patients with a confirmed history of poliomyelitis and a period of neurological stability of at least 15 years, then the diagnosis of post-polio syndrome (PPS) must be considered. PPS is, however, a diagnosis of exclusion and care must be taken to investigate for other medical diagnoses that might explain the new symptoms. The older the child was at the onset of disease, the more severe the disease was likely to have been and the more likely is it that the adult will develop PPS.

Early treatment

During the acute phase the patient is isolated and kept at complete rest, with symptomatic treatment for pain and muscle spasm. Active movement is avoided but gentle passive stretching helps to prevent contractures. Paralysis of the respiratory muscles and respiratory failure calls for intermittent positive pressure ventilation and sometimes a tracheotomy.

Once the acute illness settles, physiotherapy is stepped up, active movements are encouraged and every effort is made to regain maximum power. Between exercise periods, splintage may be necessary to maintain joint and limb alignment and prevent fixed deformities.

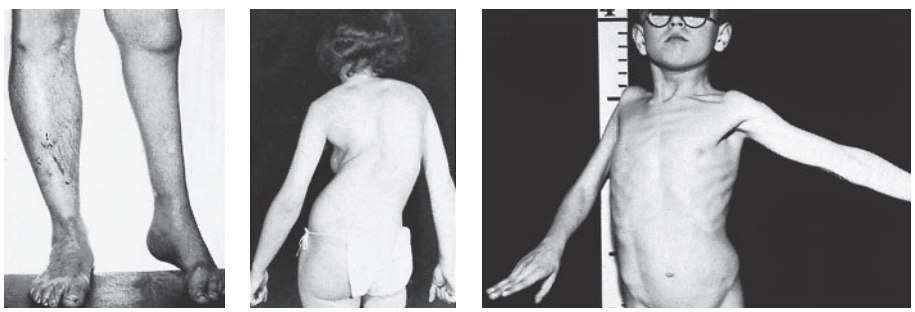
Muscle charting is carried out at regular intervals until no further recovery is detected.

Late treatment

Once the severity of residual paralysis has been established, there are a number of basic problems that need to be addressed.

Isolated muscle weakness without deformity Isolated muscle weakness, even in the absence of joint deformity, may cause instability (e.g. quadriceps paralysis which makes weight-bearing and walking impossible without some type of brace) or loss of complex function (e.g. thumb opposition, which can be treated by tendon transfer).

Passively correctible deformity Any unbalanced paralysis (i.e. muscle weakness on one aspect of a joint and greater power in the antagonists) can lead to deformity. At first this is passively correctable and can be counteracted by a suitable orthosis. However, an appropriate tendon transfer may solve the problem permanently. It is here that muscle charting is particularly important. A muscle usually loses one grade of power when it is transferred; therefore, to be useful, it should have grade 4 or 5 power prior to transfer



(a) (b) (c)

Figure 10.34 Poliomyelitis (a) Shortening and wasting of the left leg, with equinus of the ankle. (b) This long curve is typical of a paralytic scoliosis. (c) Paralysis of the right deltoid and supraspinatus makes it impossible for this boy to abduct his right arm.

and then be asked to work with gravity (rather than against gravity) in its transferred position. On the other hand, a transferred grade 3 muscle may act as a type of tenodesis and reduce the deformity caused by gravity.

Fixed deformity Fixed deformities cannot be corrected by either splintage or tendon transfer alone; it is important also to restore alignment operatively and to stabilize the joint, if necessary, by arthrodesis (rather than relying on orthoses again). This is especially applicable to fixed deformities of the ankle and foot, but the same principle applies in treating paralytic scoliosis.

Occasionally a fixed deformity is beneficial. Thus, an equinus foot may help to compensate mechanically for quadriceps weakness (by keeping the ground reaction force (GRF) forward of the knee) and/or a leg length difference; if so, it should not be corrected.

Flail joint Balanced paralysis, because it causes no deformity, may need no treatment. However, if the joint is unstable or flail, it must be stabilized, either by permanent splintage or by arthrodesis.

Shortening Normal bone growth depends on normal muscle activity; thus many children who have been affected with poliomyelitis in their early years can be expected to develop a difference in leg length. Discrepancies of up to 3–5 cm can, in theory, be compensated for with a shoe raise although this tends to make the shorter (and weaker) leg clumsier. While leg lengthening is always an option, the fact that the increase in length discrepancy with growth can be calculated fairly accurately from growth tables means it can also be mitigated by a well-timed epiphyseodesis of the distal femoral and/or the proximal tibial physis in the normal limb.

Disturbance of skeletal modelling As with all childhood paralytic disorders, the effects of muscle imbalance on the growing skeleton must be anticipated. Changes may become obvious with growth, appearing as torsional deformities or angular deformities in either the sagittal or the coronal plane. Moreover, muscle and joint contractures may aggravate the effects of any bone distortion. Any changes that interfere with function should be prevented or treated as soon as possible.

Vascular dysfunction Sensation is intact but the paralysed limb is often cold and blue. Large chilblains sometimes develop and sympathectomy can be considered.

Regional survey

Treatment is often concentrated on the lower limbs but this should not be at the expense of upper-limb function. For children who are dependent on walking aids and/or wheelchairs, obtaining and maintaining bimanual function can be very important.

Shoulder

Provided the scapular muscles are strong, abduction at the shoulder can be restored by arthrodesing the gleno-humeral joint (50 degrees abducted and 25 degrees flexed) (Figure 10.35). Contracted adductors may need release.

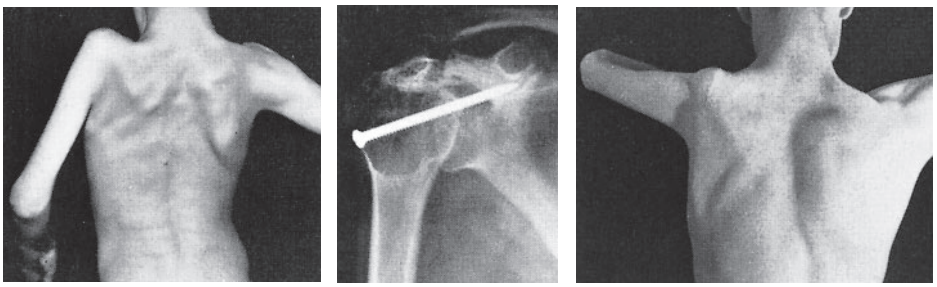
Elbow and forearm

At the elbow, *flexion* can be restored in one of two ways. If there is normal power in the anterior forearm muscles (wrist and finger flexors), the common flexor origin can be moved more proximally on the distal humerus to provide better leverage across the elbow. Alternatively, if the pectoralis major is strong, the lower half of the muscle can be detached at its origin on the rib-cage, swung down and joined to the biceps tendon.

Pronation of the forearm can be strengthened by transposing an active flexor carpi ulnaris tendon across the front of the forearm to the radial border. Loss of *supination* may be countered by transposing flexor carpi ulnaris across the back of the forearm to the distal radius.

Wrist and hand

Wrist deformity or instability can be markedly improved by arthrodesis. Any active muscles can then be used to restore finger movement.



(a) (b) (c)

Figure 10.35 Poliomyelitis – arthrodesis (a) This patient had paralysis of the left deltoid: after arthrodesis (b) he could lift his arm (c) by using his scapular muscles.

In the thumb, weakness of opposition can be overcome by a flexor superficialis transfer (Figure 10.36). The tendon (usually of the ring finger) is wound round that of flexor carpi ulnaris (which acts as a pulley), threaded across the palm and fixed to the distal end of the first metacarpal.

Spine

Unbalanced paralysis causes scoliosis, frequently a long thoracolumbar curve which may involve the lumbosacral junction, causing pelvic obliquity. Operative treatment is often needed but instrumented fusions do have a high complication rate even though patient satisfaction is generally good.

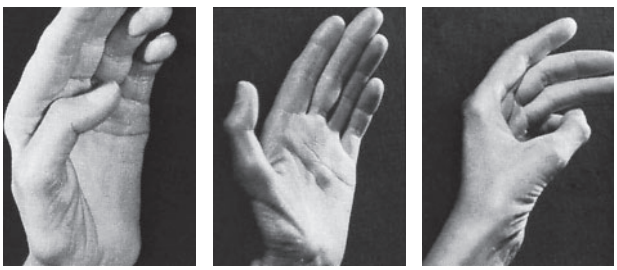
Hip

Hip deformities are usually complex and difficult to manage; the problem is often aggravated by the gradual development of subluxation or dislocation due either to muscle imbalance (abductors weaker than adductors) or pelvic obliquity associated with scoliosis.

Furthermore, since paralysis usually occurs before the age of 5 years, growth of the proximal femur is abnormal and this may result in secondary deformities such as persistent anteversion of the femoral neck, coxa valga and underdevelopment of the acetabulum, all of which will increase the tendency to instability and dislocation.

The keys to successful treatment are to:

1 reduce any scoliotic pelvic obliquity by correcting or improving the scoliosis
2 overcome or improve the muscle imbalance by suitable tendon transfer
3 correct the proximal femoral deformities by intertrochanteric or subtrochanteric osteotomy, and
4 deepen the acetabulum, if necessary, by an acetabuloplasty which will prevent posterior displacement of the femoral head.



(a) (b) (c)

Figure 10.36 Poliomyelitis – treatment Opponens paralysis (a) has been treated by superficialis tendon transfer. In (b) the tendon can be seen in action at the start of thumb opposition. (c) Full opposition achieved.

Fixed flexion can be treated by Soutter's muscle slide operation or by transferring psoas to the greater trochanter. For fixed abduction with pelvic obliquity the fascia lata and iliotibial band may need division; occasionally, for severe deformity, proximal femoral osteotomy may also be required. With this type of obliquity the 'higher' hip tends to be unstable and the 'lower' hip to have fixed abduction; if the abducted hip is corrected first, the pelvis may level and the other hip become normal.

Knee

Instability due to relative weakness of the knee extensors is a major problem. Unaided walking may still be possible provided the hip has good extensor power and the foot good plantarflexion power (or fixed equinus); with this combination the knee is stabilized by being thrust into hyperextension as body weight comes onto the leg. The patient has often learnt to help this manoeuvre by placing a hand on the front of the thigh and pushing the thigh backwards with every stance phase of gait. If the hip or ankle joints are also weak, a full-length RGO or KAFO will be required, or a supracondylar extension osteotomy of the femur must be considered.

Fixed flexion with flexors stronger than extensors is more common and must be corrected. Flexor-to-extensor transfer (e.g. hamstring muscles to the patella or the quadriceps tendon) is feasible if the flexor muscles are normal; however, quadriceps power is unlikely to be improved by more than one grade. If the flexors are not strong enough, the deformity can be corrected by supracondylar extension osteotomy.

Marked hyperextension (genu recurvatum) sometimes occurs, either as a primary deformity or secondary to fixed equinus. It can be improved by supracondylar flexion osteotomy; an alternative is to excise the patella and slot it into the upper tibia where it acts as a bone block.

Foot

Instability and *foot-drop* can be controlled by an ankle–foot orthosis or a simpler drop-foot splint. Often there is imbalance causing varus, valgus or calcaneocavus *deformity*; fusion in the corrected position should be combined with tendon re-routing to restore balance, otherwise there is risk of the deformity recurring.

For hindfoot varus or valgus, the simplest procedure is to slot bone grafts into vertical grooves on each side of the sinus tarsi (Grice); alternatively, a triple arthrodesis (Dunn) of subtalar and mid-tarsal joints is performed, relying on bone carpentry to correct deformity. With associated foot-drop, Lambrinudi's modification is valuable; a triple arthrodesis is performed but the fully plantarflexed talus is slotted into

the navicular with the forefoot in only slight equinus: foot-drop is corrected because the talus cannot plantarflex further, and slight equinus helps to stabilize the knee. With calcaneocavus deformity, Elmslie's operation is useful: a triple arthrodesis is performed in the calcaneus position but corrected at a second stage by posterior wedge excision combined with tenodesis using half of the tendo Achillis.

There is a low incidence of secondary osteoarthritis in the joints adjacent to the arthrodesed joint because of the relatively low demands placed on them by the paralytic limb.

Mobile claw toe deformities are corrected by transferring the toe flexors to the extensors; if the deformity is fixed, the interphalangeal joints should be arthrodesed in the straight position and the long extensor tendons reinserted into the metatarsal necks.

MOTOR NEURON DISORDERS

Rare degenerative disorders of the large motor neurons may cause progressive and sometimes fatal paralysis.

Motor neuron disease (MND)

This is a progressive degenerative disease of unknown aetiology. It affects both cortical (upper) motor neurons and the anterior horn cells of the cord, causing widespread UMN and LMN symptoms and signs. There are four main types of MND with amyotrophic lateral sclerosis (ALS) being the most common form. Patients usually present in middle age with dysarthria and difficulty in swallowing or, if the limbs are affected, with muscle weakness (e.g. clumsy hands or unexplained foot-drop) and wasting in the presence of exaggerated reflexes. Muscle cramps are troublesome; muscle atrophy and fasciculations may be obvious. Sensation and bladder control are normal. Some of these features are also seen in spinal cord compression, which can be excluded by MRI.

The disease is progressive and incurable. Patients usually end up in a wheelchair and have increasing difficulty with speech and eating. Cognitive function is usually spared although some patients have associated frontotemporal dementia or a pseudobulbar effect causing emotional lability. Most of them die within 5 years of symptom onset from a combination of respiratory weakness and aspiration pneumonia.

Spinal muscular atrophy

In this rare group of heritable disorders (a defect on the long arm of chromosome 5 has been identified) there is widespread degeneration of the anterior horn cells in the cord, leading to progressive LMN weakness. The commonest form (*Werdnig–Hoffman disease*) is inherited in an autosomal recessive manner and is diagnosed at birth or soon afterwards. The baby is floppy and weak, feeding is difficult and breathing is shallow. Death occurs, usually within a year.

A less severe form (*Kugelberg–Welander disease*), of either dominant or recessive inheritance, is usually seen in adolescents or young adults who present with limb weakness, proximal muscle wasting and 'paralytic' scoliosis. However, it sometimes appears in early childhood as a cause of delayed walking. Patients may live to 30–40 years of age but are usually confined to a wheelchair. Spinal braces are used to improve sitting ability; if this cannot prevent the spine from collapsing, operative instrumentation and fusion is advisable.

PERIPHERAL NEUROPATHIES

Disorders of the peripheral nerves (Box 10.3) may affect motor, sensory or autonomic functions, may be localized to a short segment or may involve the full

BOX 10.3 CAUSES OF POLYNEUROPATHY

Hereditary
- Hereditary motor and sensory neuropathy
- Friedreich's ataxia
- Hereditary sensory neuropathy

Infections
- Viral infections
- Herpes zoster
- Neuralgic amyotrophy
- Leprosy

Inflammatory
- Acute inflammatory polyneuropathy
- Guillain-Barré syndrome
- Systemic lupus erythematosus

Nutritional and metabolic
- Vitamin deficiencies
- Diabetes
- Myxoedema
- Amyloidosis

Neoplastic
- Primary carcinoma
- Myeloma

Toxic
- Alcohol
- Lead

Drugs
- Various

length of the nerve fibres including their cell bodies in the anterior horn (motor neurons), posterior root ganglia (sensory neurons) and autonomic ganglia. In some cases spinal cord tracts are involved as well. There are over 100 types of neuropathy; in this section we consider those conditions that are most likely to come within the ambit of the orthopaedic surgeon.

Classification

Classification by anatomical level and distribution is probably the simplest. Although it does not fully cover pathological causation, it does relate to clinical presentation and provides a framework for further investigations. It is well to remember that in over 40% of cases no specific cause is found!

1 *Radiculopathy* – involvement of nerve roots, most commonly by vertebral trauma, intervertebral disc herniation or bony spurs, space-occupying lesions of the spinal canal and root infections such as herpes zoster.
2 *Plexopathy* – direct trauma (e.g. brachial plexus traction injuries, lumbosacral plexopathy in pelvic trauma), compression by local tumours (Pancoast's tumour), entrapment in thoracic outlet syndrome, and viral infection such as neuralgic amyotrophy.
3 *Distal neuropathy* – involvement of neurons in distinct peripheral nerves, which is usually subdivided into:
   - *Mononeuropathy* – involvement of a single nerve, usually mixed sensorimotor (e.g. nerve injury, nerve compression, entrapment syndromes and nerve tumours).
   - *Multiple mononeuropathy* – involvement of several isolated nerves (e.g. leprosy and some cases of diabetes or vasculitis).
   - *Polyneuropathy* – widespread symmetrical dysfunction (e.g. diabetic neuropathy, alcoholic neuropathy, vitamin deficiency, Guillain–Barré syndrome and a host of less common disorders (see Box 10.3).

Abnormalities may be predominantly sensory (e.g. diabetic polyneuropathy), predominantly motor (e.g. Charcot–Marie–Tooth disease) or mixed. Chronic motor loss with *no* sensory component is usually due to anterior horn cell disease rather than more esoteric pathology like lead poisoning.

Pathology

In general terms, large nerve fibres (those over 4 μm in diameter, which includes α-motor neurons, γ-motor neurons to the muscle spindles and sensory neurons serving touch and pressure) are myelinated whereas small fibres (less than 4 μm in diameter, mainly sensory neurons serving pain sensibility and autonomic neurons effecting vasomotor control, piloerection and neuroendocrine functions) are unmyelinated.

There are three basic types of peripheral neuronal pathology: (1) acute interruption of axonal continuity; (2) axonal degeneration; and (3) demyelination. In all three, conduction is disturbed or completely blocked, with consequent loss of motor and/or sensory and/or autonomic functions.

ACUTE AXONAL INTERRUPTION

This occurs most typically after nerve division and is described in Chapter 11. Loss of motor and sensory functions is immediate and complete. The distal segments of axons that are crushed or severed will degenerate, as will the muscle fibres which are supplied by motor neurons if nerve conduction is not restored within 2 years. These changes are detectable at an early stage by nerve conduction studies and EMG. Axonal regeneration, when it occurs, is slow and often incomplete; new axons grow by about 1 mm per day.

CHRONIC AXONAL DEGENERATION

In non-traumatic neuronal neuropathies the changes are slower and progressive. Most *large-fibre disorders* affect both sensory and motor neurons causing 'stocking' and 'glove' numbness, altered postural reflexes and ataxia as well as muscle weakness and wasting, all beginning distally and progressing proximally. Symptoms tend to appear in the feet and legs before the hands and arms (stocking before glove rather than 'glove and stocking' distribution). Some disorders are predominantly either motor or sensory. Nerve conduction studies show a reduction in the size of CMAP and SNAP responses proportionate to the loss of peripheral nerve fibres, but relatively little conduction slowing (in contrast to the demyelinating neuropathies). EMG may demonstrate denervation changes in distal muscles and confirm the extent and severity of nerve loss.

Small-fibre neuropathies (as occur in diabetes) may cause orthostatic hypotension, cardiac arrhythmias, reduced peripheral limb perfusion, ischaemia and a predisposition to limb infection. Small nerve fibres also convey pain, heat and cold sensibility and when disturbed give rise to burning dysaesthesias. Neurophysiological tests are not sensitive enough to distinguish small-fibre disturbances.

DEMYELINATING NEUROPATHIES

Focal demyelination occurs most commonly in nerve entrapment syndromes and blunt soft-tissue trauma. The main effects are slowing of conduction and sometimes complete nerve block, causing sensory and/or motor dysfunction distal to the lesion. These changes are potentially reversible; recovery usually takes less than 6 weeks, and in some cases only a few days.

Demyelinating polyneuropathies are rare, with the exception of Guillain–Barré syndrome. Other conditions are the heritable motor and sensory neuropathies and some inherited metabolic disorders, but most of these show a mixture of axonal degeneration and demyelination.

Clinical features

Patients usually complain of sensory symptoms: 'pins and needles', numbness, a limb 'going to sleep', 'burning', shooting pains or restless legs. They may also notice weakness or clumsiness, or loss of balance in walking. Occasionally (in the predominantly motor neuropathies) the main complaint is of progressive deformity, such as claw hand or cavus foot (Figure 10.37). The onset may be rapid (over a few days) or gradual (over weeks or months). Sometimes there is a history of injury, a recent infective illness, a known disease such as diabetes or malignancy, alcohol abuse or nutritional deficiency.

Examination may reveal motor weakness in a particular muscle group. In the polyneuropathies the limbs are involved symmetrically, usually legs before arms and distal before proximal parts. Reflexes are usually depressed, though in small-fibre neuropathies (e.g diabetes) this occurs very late. In mononeuropathy, sensory loss follows the map of the affected nerve. In polyneuropathy, there is a symmetrical 'glove' or 'stocking' distribution. Trophic skin changes may be present. Deep sensation is also affected and some patients develop ataxia. If pain sensibility and proprioception are depressed, there may be joint instability and/or breakdown of the articular surfaces classical of a Charcot joint.

Clinical examination alone may establish the diagnosis. Further help is provided by electromyography (which may suggest the type of abnormality) and nerve conduction studies (which may show exactly where the lesion is).

The *mononeuropathies,* mainly nerve injuries and entrapment syndromes, are dealt with in Chapter 11.

The more common polyneuropathies are listed in Box 10.3 and some are described below.

Hereditary neuropathies

These rare disorders present in childhood and adolescence, usually with muscle weakness and deformity.

Hereditary sensory neuropathy

Congenital insensitivity or indifference to pain and temperature is inherited as either a dominant or a recessive trait but it is an extremely rare condition. Patients develop neuropathic joint disease and ulceration of the feet. The cycle of painless injury and progressive deformity can lead to severe disability.

Hereditary motor and sensory neuropathy (HMSN)

This is the preferred name for a group of conditions which includes *peroneal muscular atrophy* (Figure 10.38), *Charcot–Marie–Tooth disease* and some *benign forms of spinal muscular atrophy.* They are the commonest of the inherited neuropathies and they are usually autosomal dominant disorders.

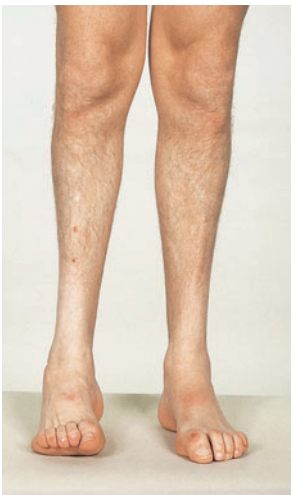
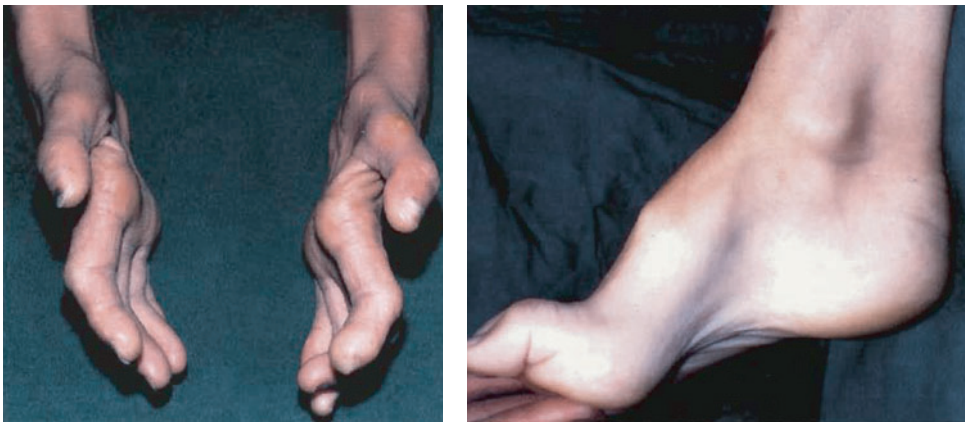


Figure 10.38 Hereditary neuropathies – peroneal muscular atrophy This patient has the typical wasting of the legs, cavus feet and claw toes associated with peroneal muscular atrophy.



(a) (b)

Figure 10.37 Peripheral neuropathy Two typical deformities in patients with peripheral neuritis: (a) ulnar claw hands and (b) pes cavus and claw toes.

HMSN type I is seen in children who have difficulty walking and develop claw toes and pes cavus or cavo-varus. There may be severe wasting of the legs and (later) the upper limbs, but often the signs are quite subtle. Spinal deformity may occur in severe cases. This is a demyelinating disorder and nerve conduction velocity is markedly slowed. The diagnosis can be confirmed by finding demyelination on sural nerve biopsy or by genetic testing of blood samples.

HMSN type II presents in adolescents and young adults and is much less disabling than type I; it affects only the lower limbs, causing milder foot deformities and peroneal muscle wasting. Nerve conduction velocity is only slightly reduced, indicating primary axonal degeneration.

Treatment In the early stages physiotherapy stretches and AFOs are helpful. If the deformities are progressive or disabling, operative correction may be indicated (see Chapter 21). Equinus deformities may need correction and a peroneus longus to brevis tenodesis is useful for flexible hindfoot varus as evaluated by the Coleman block test. The fixed deformity would require an osteotomy. The cavus deformity often needs no treatment. In children a plantar fascia release may help but in adults, if it is causing pain, a mid-tarsal osteotomy may be more appropriate or (in severe cases) a triple arthrodesis.

Claw toes (due to intrinsic muscle weakness) can be corrected by transferring the toe flexors to the extensors, with or without fusion of the interphalangeal joints. Clawing of the big toe is best corrected by the Robert Jones procedure: transfer of the extensor hallucis longus to the metatarsal neck and fusion of the interphalangeal joint.

Hereditary neuropathy with liability to pressure palsies (HNPP)

This is a relatively common, dominant disorder which often presents as multiple mixed entrapment mononeuropathies (e.g. carpal tunnel syndrome and ulnar nerve palsy), even in young patients. The general recommendation is to avoid surgery if at all possible.

Friedreich's ataxia

This rare autosomal recessive condition is the classic archetype of a large group of genetic disorders – the *spinocerebellar ataxias* – characterized by spinocerebellar dysfunction but where there may also be degeneration of the posterior root ganglia and peripheral nerves. The genetic defect is a triplet expansion localized to chromosome 9. In the UK the incidence is only 1–2/50 000 but in the USA, about 1 in 90 adults is a carrier for this condition.

The condition presents in childhood (rarely adulthood) and all patients develop progressive ataxia of the limbs and of their gait with associated extensor plantar responses but absent knee and ankle reflexes and sensory disturbances such as loss of vibration sense and two-point discrimination. Dysarthria appears within 5 years of disease onset. The neurological degeneration is seen in the spinocerebellar tracts, the corticospinal tracts, the posterior columns of the spinal cord and parts of the cerebellum itself. Nerve conduction studies demonstrate slowed motor velocities in both median and tibial nerves with absent sensory action potentials in the sural and digital nerves.

Painful muscle spasms occur in some patients and, if so, they tend to worsen with time. As with the other hereditary peripheral neuropathies, the more common orthopaedic complaints are a progressive cavo-varus foot deformity that is usually rigid, the development of clawed toes and a scoliosis. In general, the earlier the onset of the disease the greater is the risk of significant curve progression. In the more severe cases, functional and neurological deterioration may be rapid with the development of a cardiomyopathy and death in early to mid adulthood. Despite the potentially poor prognosis, surgical correction of foot and spine deformities is worthwhile.

Metabolic neuropathies

Diabetic neuropathy

Diabetes is one of the commonest causes of peripheral neuropathy. The metabolic disturbance associated with hyperglycaemia interferes with axonal and Schwann cell function, leading to mixed patterns of demyelination and axonal degeneration. Autonomic dysfunction and vascular disturbance also play a part.

The onset is insidious and the condition often goes undiagnosed until patients start complaining of numbness and paraesthesiae in the feet and lower legs. Even at that early stage there may be areflexia and diminished vibration sense. Another suspicious pattern is an increased susceptibility to nerve entrapment syndromes.

Later, muscle weakness becomes more noticeable in proximal parts of the limbs. In advanced cases trophic complications can arise: neuropathic ulcers of the feet, regional osteoporosis, insufficiency fractures of the foot bones, or Charcot joints in the ankles and feet. Another late feature is loss of balance. Autonomic dysfunction may produce postural hypotension and abnormal sphincter control and may also account for an increased susceptibility to infection.

Treatment It is vital to ensure that the underlying disorder is properly controlled. Local treatment consists of skin care, management of fractures and splintage or arthrodesis of grossly unstable or deformed joints. Management of the diabetic foot is discussed in Chapter 21.

Alcoholic neuropathy

Axonal degeneration may be due to some toxic effect of the alcohol, but the main cause is the accompanying nutritional deficiency, especially thiamine deficiency. Presenting symptoms are, typically, 'burning' paraesthesiae, numbness and muscle weakness in the feet and legs. The calf muscles are tender to pressure and reflexes are depressed or absent. Men are likely to complain of urinary difficulty and impotence.

Treatment Early cases may respond to nutritional supplementation and administration of thiamine. Patients should be protected from trauma. Of course, steps should also be taken to deal with the alcohol abuse.

Infective neuropathies

Herpes zoster (shingles)

This common disorder is caused by the varicella (chickenpox) virus. The virus, having lain dormant for many years in the dorsal root ganglia, is then reactivated and migrates down the nerve. The exact cause of the reactivation is unknown but immunocompromise, age and stress are contributory factors; thus elderly or immunosuppressed patients are particularly susceptible.

Following an injury or intercurrent illness, the patient develops severe unilateral pain in the distribution of several adjacent nerve roots. Motor roots and even the spinal cord may (rarely) be affected and involvement of the lumbar roots can closely mimic sciatica. Days or weeks later an irritating vesicular rash appears (Figure 10.39); characteristically it trails out along the dermatomes corresponding to affected nerves. The condition usually subsides spontaneously but post-herpetic neuralgia may persist for months or years.

Treatment Treatment is symptomatic and oral antiviral therapy may be justified.

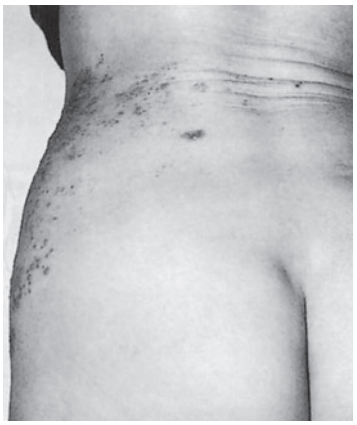


Figure 10.39 Herpes zoster This patient was treated for several weeks for 'sciatica' – then the typical rash of shingles appeared.

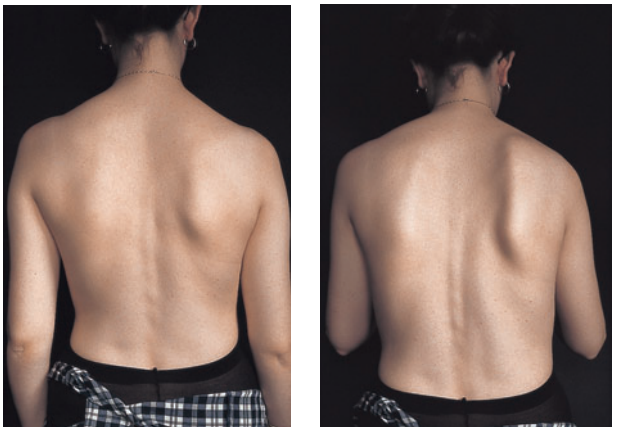
Neuralgic amyotrophy (acute brachial neuritis)

This unusual cause of severe shoulder girdle pain and weakness is of unknown aetiology but is believed to be due to a para-infectious disorder of one or more of the cervical nerve roots and the brachial plexus, sometimes producing a pseudomononeuropathic pattern (e.g. scapular winging (Figure 10.40) or wrist-drop). There is often a history of an antecedent viral infection or antiviral inoculation; sometimes a small epidemic occurs among several inmates of an institution.

The history alone often suggests the diagnosis. Pain in the shoulder and arm is typically sudden in onset, intense and unabating; the patient can often recall the exact hour when symptoms began. Pain may extend into the neck and down as far as the hand; usually it lasts for 2–3 weeks. Other symptoms are paraesthesiae in the arm or hand and weakness of the muscles of the shoulder, forearm and hand.

Winging of the scapula (due to serratus anterior weakness), wasting of the shoulder girdle muscles, and occasionally involvement of more distal arm muscles may be profound, becoming evident as the pain improves. Shoulder movement is initially limited by pain but this is superseded by weakness due to muscle atrophy. Sensory loss and paraesthesiae in one or more of the cervical dermatomes is not uncommon. Involvement of overlapping root territories of the brachial plexus is a feature that helps to distinguish neuralgic amyotrophy from an acute cervical disc herniation which is monoradicular.

There is no specific treatment; pain is controlled with analgesics and sometimes steroids in the early phase. The prognosis is variable with around 75% of



(a) (b)

Figure 10.40 Neuralgic amyotrophy A common feature of neuralgic amyotrophy is winging of the scapula due to serratus anterior weakness. Even at rest (a) the right scapula is prominent in this young woman. When she thrusts her arms forwards against the wall (b) the abnormality is more pronounced.

patients regaining prior function within 2 years, but many are left with reduced exercise tolerance and poor muscle coordination.

Guillain–Barré Syndrome (acute inflammatory demyelinating polyneuropathy – AIDP)

Guillain–Barré syndrome describes an acute demyelinating motor and sensory (though mainly motor) polyneuropathy. It can occur at any age and usually appears 2–3 weeks after an upper respiratory or gastrointestinal infection, probably as an autoimmune reaction. Occasionally surgery may act as the trigger.

The typical history is of aching and weakness in the legs, often accompanied by numbness and paraesthesiae, which steadily progresses upwards over a period of hours, a few days or a few weeks. Symptoms may stop when the thigh and pelvic muscles are reached, and then gradually retreat, or they may go on ascending to involve the upper limbs, facial muscles and diaphragm, resulting in quadriplegia and respiratory failure. In the established case there will be areflexia and loss of position sense. In severe cases patients may develop features of autonomic dysfunction. Unsurprisingly, the condition is also known as 'ascending paralysis'.

Cerebrospinal fluid analysis may show a characteristic pattern: elevated protein concentration in the presence of a normal cell count (unlike an infection, in which the cell count would also be elevated).

Nerve conduction studies may show conduction slowing or block; in severe cases there may be EMG signs of axonal damage.

Treatment Treatment consists essentially of bed rest, pain-relieving medication and supportive management to monitor, prevent and deal with complications such as respiratory failure and difficulty with swallowing. In severe cases specific treatment with intravenous immunoglobulins or plasmapheresis should be started as soon as possible. Once the acute disorder is under control, physiotherapy and splintage will help to prevent deformities and improve muscle power.

Most patients recover completely, though this may take 6 months or longer; about 30% are left with residual weakness at 3 years and about 3% are likely to die.

Leprosy

Although uncommon in Europe and North America, this is still a frequent cause of peripheral neuropathy in Africa and Asia (Figure 10.41).

Mycobacterium leprae, an acid-fast organism, causes a diffuse inflammatory disorder of the skin, mucous membranes and peripheral nerves: it is only mildly contagious. Depending on the number of bacteria present, two main types of disease are recognised: paucibacillary (PB) and multibacillary (MB). The two are distinguished by the number of poorly pigmented, anaesthetic skin patches which develop over the extensor surfaces of the limbs: 1–5 patches is PB and >5 patches is MB leprosy.

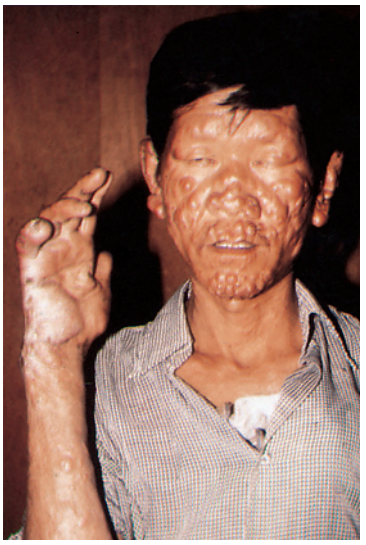


Figure 10.41 Leprosy – ulnar nerve paralysis Ulnar nerve paralysis is relatively common in long-standing leprosy. This patient has the typical ulnar claw-hand deformity.

Loss of motor function leads to weakness and deformities of the hands and feet. Thickened nerves may be felt as cords under the skin or where they cross the bones (e.g. the ulnar nerve behind the medial epicondyle of the elbow). Trophic ulcers are common and may predispose to osteomyelitis.

Leprosy is curable with a multidrug therapy regime: rifampicin and dapsone for PB and rifampicin, clofazimine and dapsone for MB.

The condition is discussed in greater detail in Chapter 2 and the peripheral nerve complications are dealt with in Chapter 11.

ARTHROGRYPOSIS

'Arthrogryposis' is a broad term used to describe a large group of congenital disorders, all of them rare, in which children are born with multiple non-progressive soft-tissue contractures and associated restriction of joint movement. The underlying pathology is very varied and may be neurological or myopathic, leading to varying phenotypes or clinical pictures.

In the most common form, *arthrogryposis multiplex congenita* (nowadays known as *amyoplasia*), all joints of the upper and lower limbs are involved including the spine. In others, restriction is more minor and predominantly affects the distal extremities.

The incidence is said to be about 1 in 3000 live births; in some cases a genetic linkage has been demonstrated. Extrinsic causes may include intrauterine lack of room for movement (for whatever reason)

during fetal development. Joint capsules are often fibrotic.

The deformities are associated with unbalanced muscle weakness which follows a neurosegmental distribution, and necropsy specimens show sparseness of anterior horn cells in the cervical and lumbar cord. Deformities and contractures develop *in utero* and remain largely unchanged throughout life. Myopathic and neuropathic features may coexist in the same muscle.

Classification

Considering arthrogryposis as a whole, the conditions can be placed in three major categories:

1 *Those with total body involvement* – typified by the condition formerly known as *arthrogryposis multiplex congenita* and now termed *amyoplasia*, but also including other congenital disorders showing widespread joint contractures.
2 *Those with predominantly hand or foot involvement* – conditions with joint features similar to those of amyoplasia but usually limited to distal joints (wrists, hands, feet) and therefore termed *distal arthrogryposis*; included also are more severe types of distal myopathy such as the Freeman–Sheldon syndrome in which there are, in addition, abnormal facial features (the 'whistling face syndrome').
3 *Pterygia syndromes* – conditions characterized by arthrogrypotic joint contractures with identifiable soft-tissue webs, usually across the flexor aspects of the knees and ankles (Figure 10.42).

Clinical features

Clinical examination is still the best way of making the diagnosis: involved limbs are often tubular and the joints 'featureless' (Figure 10.43). The normal skin creases on both sides of the joints are missing but there are often deep dimples over the joints. Muscle mass is markedly reduced. In some cases there is true muscle weakness.

In the *classic form of amyoplasia* the shoulders are adducted and internally rotated, the elbows usually extended and the wrists/hands flexed and deviated ulnar-wards. In the lower limbs, the hips are flexed and abducted, the limbs externally rotated, the knees usually extended and the feet showing equinovarus or vertical talus deformities. Secondary problems include feeding difficulties due to the stiff jaw and immobile tongue.

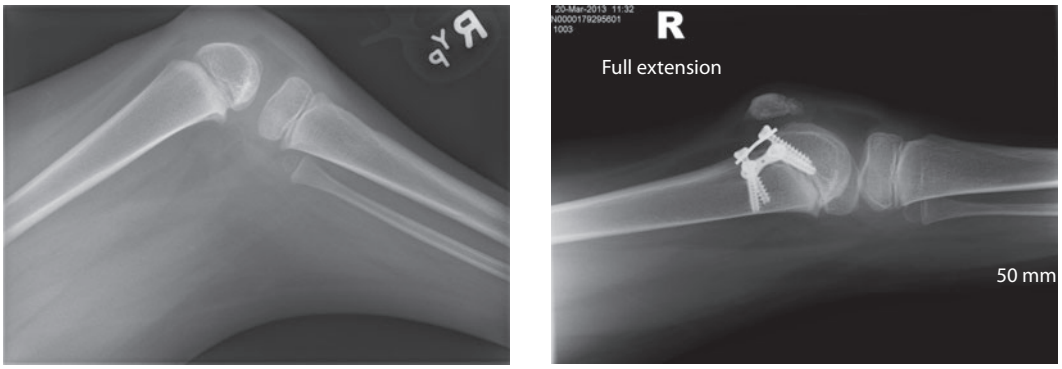
Distal arthrogryposis often manifests an autosomal dominant pattern of inheritance. Common hand deformities are ulnar deviation of the metacarpophalangeal joints, fixed flexion of the PIP joints and tightly adducted thumbs. Foot deformities are likely to be resistant forms of equinovarus or vertical talus.

Treatment

The severe forms are diagnosed at birth and many joints will gain some passive and perhaps active movement over the first few weeks of life in response to physiotherapy stretches and the fact that there is more room to move. However, in essence the condition is incurable and unlikely to improve significantly without more aggressive treatment.

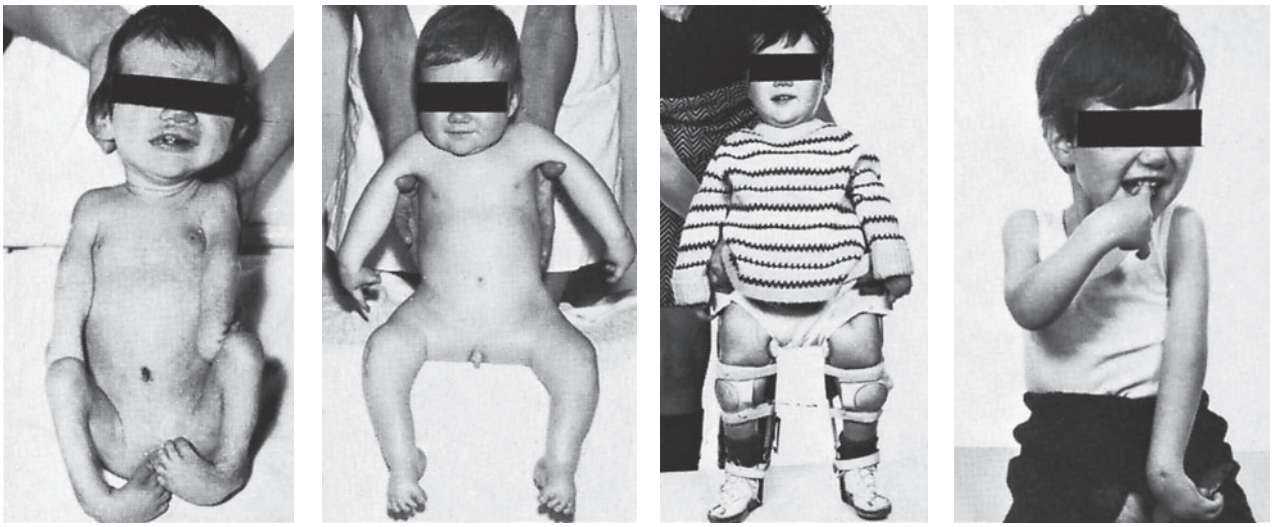
Treatment of the individual joint begins shortly after birth and follows basic principles with manipulation, stretching and splinting forming the mainstays of initial management. A few cautionary words: check for neonatal fractures before starting treatment and avoid forceful manoeuvres.

In the pterygia syndromes, physiotherapy can be tried but early release of the popliteal contractures



(a) (b)

Figure 10.42 Pterygium syndrome Lateral radiographs of the knee of a child with a pterygium syndrome. (a) The preoperative film shows the extent of the soft-tissue web behind the knee. This was released and at the same time guided growth plates were inserted anteriorly over the distal femoral physis. (b) 18 months later the femoral growth had resulted in divergence of the screws, a change in the orientation of the physis and a reduction in the deformity. The knee is now straight and the plates can be removed.



(a) (b) (c) (d)

Figure 10.43 Arthrogryposis multiplex congenita (a,b) Severe deformities are present at birth. In this case all four limbs are affected. (c,d) Operative treatment is often worthwhile. In this young boy the lower limbs were tackled first and the feet and knees are held in splints. In the upper limbs, the minimum aim is to enable a hand to reach the mouth.

should be considered. Great care is needed to avoid injury to tight neurovascular structures which may lie close to the superficial surface of the web.

In general, if progress is slow, tendon releases, tendon transfers and osteotomies may become necessary. Rigid talipes equinovarus is particularly difficult to treat and, although the Ponseti technique has been successful in correcting many feet, the relapse rate is high and operative treatment is often required. Displacement or dislocation of the hip, likewise, often defies conservative treatment and open reduction is then needed: many prefer the medial approach.

Unfortunately, recurrences of deformity are common. Before surgical intervention is considered, it should be noted that children often cope surprisingly well with their deformities and a holistic approach to the child is essential in order to ensure that the interaction of all the involved joints is understood; changing the position of one joint can have a significant adverse effect on overall function. The passive movement in one upper limb, for example, may be utilized by the active movement in a stiff contralateral limb to gain bimanual function. If both elbows are rigidly extended, function may be improved by leaving one elbow in extension and the other in partial flexion.

MUSCULAR DYSTROPHIES

The muscular dystrophies are a group of over 50 rare inherited disorders characterized by progressive muscle weakness and wasting. Pathological changes include malformation of muscle fibres, death of muscle cells and replacement of muscle by fibrous tissue and fat. Historically, they have been grouped according to their various inheritance patterns, age of onset, distribution of affected musculature and severity of the muscle weakness. The following are those most likely to be encountered in orthopaedic practice:

- *Duchenne muscular dystrophy (DMD)* – a severe, generalized sex-linked disorder affecting only boys in early childhood.
- *Becker muscular dystrophy* – is similar to DMD but less severe, starts somewhat later and progresses more slowly.
- *Limb girdle dystrophies* – a mixed group, usually of autosomal recessive inheritance, with more localized changes, affecting boys and girls in later childhood.
- *Facioscapulohumeral dystrophy* – an autosomal dominant condition of variable severity, usually appearing in early adulthood.

DUCHENNE MUSCULAR DYSTROPHY

This is a progressive disease of sex-linked inheritance with recessive transmission. It is therefore seen only in boys (or in girls with sex chromosome disorders), affecting 1 in 3500 male births. Some women are 'manifesting carriers' who have slight muscle weakness and cramps.

The DMD gene is the largest one known and a defect at locus p21 on the X chromosome results in failure to code for the dystrophin protein, which is essential for maintaining the integrity of cardiac and skeletal muscle cells. Absence of functional dystrophin leads to cell membrane leakage, muscle fibre damage and replacement by fat and fibrous tissue.

Clinical features

The condition is usually unsuspected until the child starts to walk. He has difficulty standing and climbing stairs, he cannot run properly and he falls frequently. Weakness begins in the proximal muscles of the lower limbs and progresses distally, affecting particularly the glutei, the quadriceps and the tibialis anterior, giving rise to a wide-based stance and gait with the feet in equinus, the pelvis tilted forwards, the back arched in lordosis and the neck extended. The calf muscles look bulky, but much of this is due to fat and the pseudohypertrophy belies the obvious weakness. A characteristic feature is the child's method of rising from the floor by climbing up his own legs (Gowers' sign); this is due to weakness of the gluteus maximus and thigh muscles.

Shoulder girdle weakness follows around 5 years after the clinical onset of the disease, making it difficult for the patient to use crutches. Facial muscle involvement occurs later. By the age of 10–12 years the child has usually lost the ability to walk and becomes wheelchair-dependent; from then on there is a rapid deterioration in spinal posture with the development of scoliosis and, consequently a further deterioration in lung function. Cardiopulmonary failure is the usual cause of death, generally in the fourth decade.

Investigations

The diagnosis is usually based on the clinical features and family history and by looking at serum creatinine phosphokinase levels which are 200–300 times the normal in the early stages of the disease (and also elevated, but less so, in female carriers). Confirmation is achieved by muscle biopsy and genetic testing with a DNA polymerase chain reaction.

Treatment

It is important to prolong the child's walking phase, and physiotherapy and splintage along with muscle-lengthening procedures may help achieve this.

Corticosteroids are useful in preserving muscle strength but there are significant side effects such as weight gain, osteoporosis, increased risk of fractures and cataract formation.

Research studies in which dystrophin is introduced into diseased muscle have been successful in animal models but not so far in humans. Gene therapy has also been tried but there have been difficulties with the viral vectors and associated immunological responses.

Once the scoliosis has developed, early instrumentation and spinal fusion (with the curve around 30 degrees) helps to maintain pulmonary function and improves quality of life although not necessarily lifespan. Preoperative cardiac and pulmonary function evaluation must be performed and there is always a risk of perioperative death even with minor surgical procedures.

Family counselling is important. Up to 20% of families already have a younger affected sibling by the time the proband is diagnosed.

Becker muscular dystrophy

This condition, also an X-linked recessive disease, is similar to but milder than Duchenne dystrophy. Dystrophin is decreased and/or abnormal in character. Affected boys retain the ability to walk into their teens and patients may survive until middle age. The muscles of facial expression are not affected and nor are the muscles controlling bowel or bladder function or swallowing.

Limb girdle dystrophy

This form of muscular dystrophy, characterized by weakness of the pelvic and shoulder girdle muscles, represents a heterogeneous group of conditions, many of which show an autosomal recessive inheritance pattern affecting both sexes.

Symptoms usually start in late adolescence. Pelvic girdle weakness causes a waddling gait and difficulty in rising from a low chair; pectoral girdle weakness makes it difficult to raise the arms above the head. However, the muscles of facial expression are spared. Disease progression is usually slow. The clinical features can be mistaken for those of a mild form of spinal muscular atrophy.

Treatment consists of physiotherapy and splintage to prevent contractures and operative correction when necessary.

Facioscapulohumeral dystrophy

This is an autosomal dominant condition with very variable expression. In general, males are more severely affected than females and from a younger age. Characteristically, muscle weakness is first seen in the face (inability to purse the lips or close the eyes tightly). This is followed by weakness of scapular muscles causing winging of the scapula and difficulty with shoulder abduction. Shoulder movements can sometimes be improved by fixing the scapula to the ribs posteriorly, so improving deltoid leverage. There may also be weakness of the anterior tibial muscles.

The condition is due to gene deletion on the long arm of chromosome 4; genetic testing to confirm the diagnosis is both sensitive and specific.

MYOTONIA

Myotonia, the inability to relax a muscle after the end of a voluntary contraction or effort, is a prominent feature in certain genetic disorders. The two least rare of these conditions are considered here: *dystrophia myotonica*, in which myotonia is part of a more widespread systemic disorder, and *myotonia congenita*, in which myotonia is usually the only abnormal clinical feature.

Dystrophia myotonica

Myotonic dystrophy is the most common adult-onset muscular dystrophy, with an incidence of approximately 1 in 7000. There are two types, both inherited in an autosomal dominant manner, with type 1 tending to be more common and more severe than type 2. Type 1 affects hands, lower legs, face and neck; type 2 is more proximal with shoulder, elbow and hip involvement. As the disease is passed through the generations, it starts noticeably earlier and follows a more severe course in keeping with the genetic principle of 'anticipation'. Patients usually present in adult life with distal muscle weakness and wasting. The defining feature of myotonia (perceived by the patient as 'muscle stiffness') may have been present for some years.

Myotonia (see above) can be demonstrated by asking the patient to flex and extend the fingers rapidly. Some patients are only mildly affected while others develop more widespread muscle weakness; the face and tongue may be involved, causing ptosis and difficulty with chewing. EMG changes may be diagnostic. Direct questioning may identify the family history. With time, systemic features appear – diabetes, cataracts and cardiorespiratory problems – and by middle age patients are often severely disabled.

Treatment is essentially palliative but foot deformities may benefit from physiotherapy and the use of orthotic devices to maintain a plantigrade and stable foot position.

Myotonia congenita

This is a rare condition consisting of two forms. One presents in younger children or in infancy and is inherited in an autosomal dominant manner, symptoms appear early but do not progress and are usually mild enough not to need treatment. The other, usual form of congenital myotonia is inherited in an autosomal recessive fashion. Symptoms due to 'muscle stiffness' appear in childhood and usually progress slowly. Common complaints are that walking and climbing stairs are difficult; typically this is worse after periods of inactivity and is relieved by exercise (the 'warm-up' effect). Symptoms tend also to be triggered by exposure to cold and can cause pain ('muscle cramps'). By adulthood there may be muscle weakness, though the forearms and calves are unusually bulky. There is no specific treatment for this condition although some medications have been tried. Patients are advised to avoid aggravating activities.

Other very rare subgroups have also been identified and their diagnosis can be difficult. The best advice is that children with 'atypical' features of congenital myotonia should be referred to a centre specializing in muscle disorders.

PAIN

Many, perhaps most, musculoskeletal disorders are accompanied by pain or at least discomfort. Whatever the nature of the underlying condition, pain usually requires treatment in its own right; sometimes it becomes the main focus of attention even after the initiating factors have disappeared or subsided.

Pain perception

Pain is confounding. The same receptors that appreciate discomfort also respond to tickling with feelings of pleasure. The electrical discharge in 'mild' pain is no different from that in 'severe' pain. That the degree of discomfort is related to the magnitude of the physical stimulus cannot be doubted, but ultimately both the severity of the pain and its character are experienced subjectively and cannot be measured.

Pain receptors

Nociceptors in the form of free nerve endings are found in almost all tissues. They are stimulated by mechanical distortion, by chemical, thermal or electrical irritation or by ischaemia. Musculoskeletal pain associated with trauma or inflammation is due to both tissue distortion and chemical irritation (local release of kinins, prostaglandins and serotonin). Visceral nociceptors respond to stretching and anoxia. In nerve injuries the regenerating axons may be hypersensitive to all stimuli.

Pain transmission

Pain sensation is transmitted via both myelinated axons (large-diameter fibres), which carry well-defined and well-localized sensation, and the far more numerous unmyelinated fibres, which are responsible for crude, poorly defined pain. From the dorsal horn synapses in the cord, some fibres participate in ipsilateral reflex motor and autonomic activities while others

connect with axons in the contralateral spinothalamic tracts that run to the thalamus and cortex (where pain is appreciated and localized) as well as the reticular system, which may be responsible for reflex autonomic and motor responses to pain.

Pain modulation

Pain impulses may be suppressed or inhibited by (1) simultaneous sensory impulses travelling via adjacent axons or (2) impulses descending from the brain. Thus, the gate control theory, proposed by Melzak and Wall in the 1960s, suggests that pain impulses are 'checked' in the dorsal horn of the cord; some impulses are effectively blocked and others are allowed through. This could explain why counter-stimulation sometimes reduces pain perception by encouraging impulses from fast sensory fibres. In addition, certain morphine- like compounds (endorphins and enkephalins), normally produced in the brain and spinal cord, can inhibit pain sensibility. These neurotransmitters are activated by a variety of agents, including severe pain itself, other neurological stimuli, psychological messages and placebos.

Pain threshold

The so-called 'pain threshold' is the level of stimulus needed to induce pain. There is no fixed threshold for any individual; pain perception is the result of all the factors mentioned above, operating against a complex and changing psychological background. The threshold is lowered by fear, anxiety, depression, lack of self-esteem and mental or physical fatigue; and it is elevated by relaxation, diversion, reduction of anxiety and general psychological support. The management of pain involves not only the elimination of noxious stimuli, or the administration of painkillers, but also holistic care of the person themselves.

Acute pain

Severe acute pain, as seen typically after injury, is accompanied by an autonomic 'fight or flight' reaction: increased pulse rate, peripheral vasoconstriction, sweating, rapid breathing, muscle tension and anxiety. Similar features are seen in pain associated with acute neurological syndromes or in malignant disease. Lesser degrees of pain may have negligible side effects.

Treatment is directed at:

- removing or counteracting the painful disorder
- making the patient feel comfortable and secure
- splinting/resting the painful area, if appropriate
- administering analgesics, anti-inflammatory drugs or, if necessary, narcotic preparations
- alleviating anxiety.

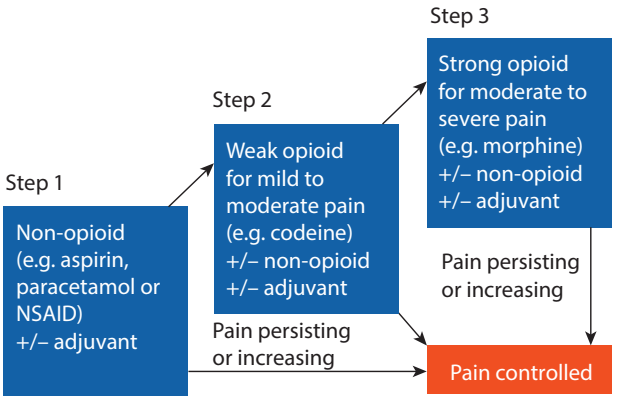


Figure 10.44 World Health Organization pain ladder The concept was devised for adults with cancer-related pain: it can be adapted usefully to the management of acute pain (or, indeed, chronic pain) in other settings.

NOTE: The concept behind the WHO pain ladder for the management of cancer-associated pain can be adapted for use in any patient with pain (Figure 10.44).

Chronic pain

Chronic pain usually occurs in degenerative and arthritic disorders or in malignant disease and is accompanied by vegetative features such as fatigue and depression. Treatment again involves alleviation of the underlying disorder if possible and general analgesic therapy, but there is an increased need for rehabilitative and psychologically supportive measures (see 'The "syndrome" of chronic pain' below).

Complex regional pain syndrome (CRPS)

This is a poorly understood condition associated with severe pain, swelling and skin changes which previously went by several different names including *Sudeck's atrophy, reflex sympathetic dystrophy, algodystrophy, shoulder–hand syndrome* and, particularly after a nerve injury, *causalgia* (see below). What they have in common is pain out of proportion (in both intensity and duration) to the precipitating cause, vasomotor instability, trophic skin changes, regional osteoporosis and functional impairment.

Precipitating causes are trauma (often trivial), operation including, classically, an arthroscopy for knee pain, a peripheral nerve lesion, myocardial infarction, stroke and hemiplegia. The incidence of post-traumatic CRPS is unknown, largely because there are no agreed criteria for diagnosing mild cases. However, the condition is more common than is generally recognized and it has been suggested that as many

as 30% of patients with fractures of the extremities develop at least some features of this condition which, fortunately, are short-lived in most cases; surgeons involved in fracture clinics therefore need to be on the lookout for symptoms 'out of proportion' to the injury and treat them promptly. Adults are the usual sufferers but the condition can occur in children.

In 1994, the International Association for the Study of Pain (IASP) came to a consensus on the diagnostic criteria for CRPS and defined two types. *CRPS Type II* (previously called causalgia) is characterized by the presence of a confirmed/defined nerve injury. The criteria were modified in 2003 (Box 10.4).

PATHOGENESIS

The pathophysiology of this condition has been argued over since it was first described 100 years ago. As many of the features involve autonomic pathways, it was usually regarded as a type of sympathetic 'overactivity', although this never explained why the abnormal activity was maintained for so long (sometimes indefinitely). It is now recognized that multiple mechanisms are involved: abnormal cytokine release, neurogenic inflammation, sympathetic-mediated enhancement of pain responses and as yet poorly understood cortical reactions to noxious stimuli. For the time being, the purely descriptive term 'complex regional pain syndrome' will have to suffice.

BOX 10.4 IASP-PROPOSED REVISED CRPS CLINICAL DIAGNOSTIC CRITERIA

A clinical diagnosis of CRPS can be made when the following criteria are met:

1. Continuing pain that is disproportionate to any inciting event
2. At least **1 symptom** reported in at least 3 of the following categories:
   - *Sensory:* Hyperesthesia or allodynia
   - *Vasomotor:* Temperature asymmetry, skin color changes/asymmetry
   - *Sudomotor/edema:* Edema, sweating changes, or sweating asymmetry
   - *Motor/trophic:* Decreased range of motion, motor dysfunction (e.g. weakness, tremor, dystonia), or trophic changes (e.g. hair, nail, skin)
3. At least **1 sign** at time of evaluation in at least 2 of the following categories:
   - *Sensory:* Evidence of hyperalgesia (to pinprick), allodynia (to light touch, temperature sensation, deep somatic pressure, or joint movement)
   - *Vasomotor:* Evidence of temperature asymmetry (>1 °C), skin color changes or asymmetry
   - *Sudomotor/edema:* Evidence of edema, sweating changes, or sweating asymmetry
   - *Motor/trophic:* Evidence of decreased range of motion, motor dysfunction (e.g. weakness, tremor, dystonia), or trophic changes (e.g. hair, nail, skin)
4. No other diagnosis better explaining the signs and symptoms

CLINICAL FEATURES

Following some precipitating event, the patient complains of burning pain and sometimes cold intolerance in the affected area, usually the hand or foot, sometimes the knee or hip and sometimes the shoulder, for example in a hemiplegia. In the mild or early case there may be no more than slight swelling, with tenderness and stiffness of the nearby joints. More suspicious signs are local redness and warmth, sometimes changing to cyanosis with a blotchy, cold and sweaty skin. X-rays are at first normal but triple-phase radionuclide scanning at this stage shows increased activity.

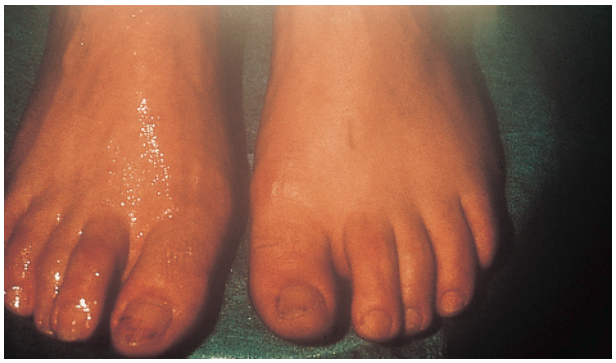
Later, or in more severe cases, trophic changes become apparent: a smooth shiny skin with scanty hair and atrophic, brittle nails (Figure 10.45). Swelling and tenderness persist and there may be marked loss of movement. X-rays now show patchy osteoporosis, which may be quite diffuse.

In the most advanced stage, there can be severe joint stiffness and fixed deformities. The acute symptoms may subside after 12–18 months, but some degree of pain often persists indefinitely. Pain is intense, often 'burning' or 'penetrating' and exacerbated by touching (allodynia), jarring or sometimes even by a loud noise. Symptoms may start distally and progress steadily up the limb to involve an entire quadrant of the body.

TREATMENT

Treatment should be started as early as possible; if the condition is allowed to persist for more than a few weeks, the changes may be irreversible. Mild cases often respond to a simple regimen of reassurance, anti-inflammatory drugs and physiotherapy. Other conservative measures include the administration of corticosteroids, calcium-regulating drugs such as calcitonin and bisphosphonates, and tricyclic antidepressants: many other treatment modalities have been tried but there is a lack of evidence to support their use. Guanethidine regional blocks were considered useful, but the effectiveness of these measures is unpredictable and somewhat doubtful.

A small percentage of patients go on complaining of pain and impaired function almost indefinitely. Psychological treatment may help them to deal with



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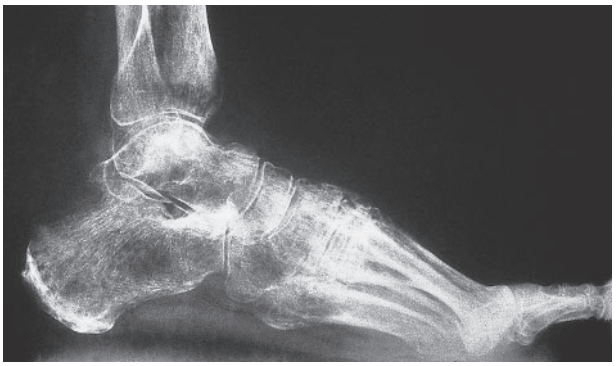


(b)

Figure 10.45 Complex regional pain syndrome
(a) A 53-year old woman suffered an undisplaced fracture of her right tibia. The fracture healed but her foot became swollen, warm to the touch and tender, the skin reddish-purple and sweaty.
(b) X-rays showed an unusual degree of osteoporosis.

the emotional distress and anxiety and to develop better coping strategies (see 'The "syndrome" of chronic pain' below).

The 'syndrome' of chronic pain

Information about the 'syndrome' of chronic pain is often vague and poorly defined but it is generally used to describe pain of 6 months or longer that has no obvious physical cause and which does not respond well to standard treatments. In a minority of patients with chronic pain there is an apparent mismatch between the bitterness of complaint and the degree of physical abnormality and it is to these patients that the word 'syndrome' is attached. The most common example is the patient with discogenic disease and prolonged, unresponsive, disabling low back pain or the young adult with ongoing disability and pain following a relatively minor injury. Labels such as 'functional overlay', 'compensitis', 'supratentorial reaction' and 'illness behaviour' are introduced and both patient and doctor may feel that the situation is 'hopeless'.

It is not sensible to keep searching for something (i.e. a significant pathological lesion that has a surgical or orthopaedic treatment) that is not there by entering a seemingly never-ending cycle of investigations and unhappy negative consultations. Nor is it helpful to over-diagnose and possibly over-treat a minor problem such as a small, inconsequential area of bone oedema on MRI. Pain is what it is perceived to be and therefore the perception needs as much respect as the pain and the appropriate management. Sometimes there are well-marked features of depression, or complaints of widespread somatic illness (pain in various parts of the body, muscular weakness, paraesthesiae, palpitations and impotence) but often the patient is simply unhappy having to cope with the chronic symptoms and the lack of explanation for them. There may of course be two elements to the pain, and the perception part may have to change before you can identify what the remaining true orthopaedic problem is and what change can be made (if any) with surgical treatment.

Management of chronic pain is always difficult. Although continuity of care and consistency in the information given are essential, these patients are ideally managed by a team that includes a specialist in pain control, a psychotherapist, a rehabilitation specialist and a social worker. Pain may be alleviated by a variety of measures: (1) analgesics and anti-inflammatory drugs; (2) local injections to painful areas; (3) local counter-irritants; (4) acupuncture; (5) transcutaneous nerve stimulation; (6) sympathetic block; and, occasionally, (7) surgical interruption of pain pathways. These methods, as well as psychosocial assessment and therapy, are best applied in a dedicated pain clinic.

FIBROMYALGIA

Fibromyalgia is not so much a diagnosis as a descriptive term for a condition in which patients complain of widespread pain and tenderness in the muscles and other soft tissues around the back of the neck and shoulders and across the lower part of the back and the upper parts of the buttocks. What sets the condition apart from other 'rheumatic' diseases (or indeed the CRP syndromes discussed above) is the complete absence of demonstrable pathological changes in the affected tissues.

Indeed, it is often difficult to give credence to the patient's complaints, an attitude which is encouraged by the fact that similar symptoms are encountered in some patients who have suffered trivial injuries in a variety of accidents; a significant number also develop psychological depression and anxiety.

The criteria for making the diagnosis were put forward by the American College of Rheumatology in 1990. These included symptoms of widespread pain in all four quadrants of the body, together with at

least nine pairs of designated 'tender points' on physical examination. In practice, however, the diagnosis is often made in patients with much more localized symptoms and signs.

The cause of fibromyalgia remains unknown; no pathology has been found in the 'tender spots'. It has been suggested that this is an abnormality of 'sensory processing', which is perhaps another way of saying that the sufferers have a 'low pain threshold'; in fact, they often do display increased sensitivity to pain (hyperalgesia) in other parts of the body. There are also suggestions that the condition is related to stress responses which can be activated by sudden accidents or traumatic life events. This does not mean that such patients will necessarily show other features of psychological dysfunction and the condition cannot be excluded merely by psychological testing.

In mild cases, treatment can be limited to keeping up muscle tone and general fitness (hence the advice to have physiotherapy and then continue with daily exercises on their own). Analgesics help with symptom control (rather than cure) and low-dose amitriptyline aids sleep. Gabapentin and pregabalin, which are used for nerve pain, can also be helpful. Patients with more persistent and more disturbing symptoms may benefit from various types of psychotherapy.

There is some confusion, and perhaps some overlap, between FMS (fibromyalgia syndrome) and *myofascial pain syndrome* where tender spots in muscles are associated with palpable 'knots or cords' or 'trigger points'. MPS is often treated by physical therapy practitioners and massage directed at the tender areas. Trigger point injections of anaesthetic solution have also been tried.

Peripheral nerve disorders

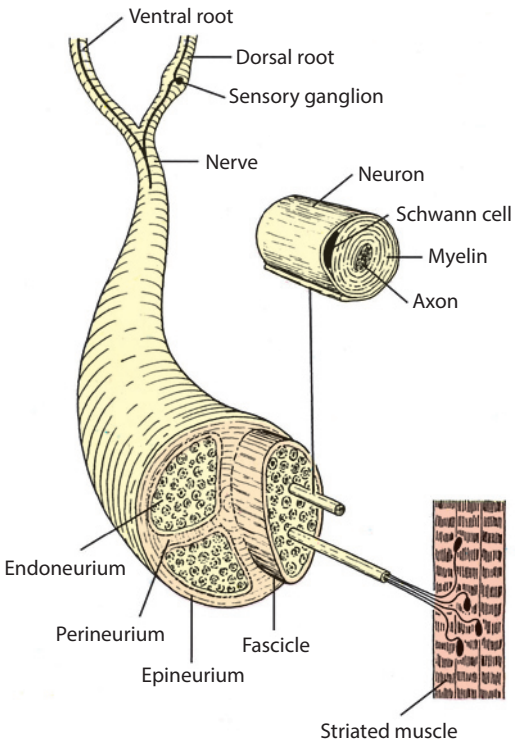
11

Michael Fox, David Warwick & H. Srinivasan

NERVE STRUCTURE AND FUNCTION

Peripheral nerves are bundles of *axons* conducting efferent (motor) impulses from cells in the anterior horn of the spinal cord to the muscles, and afferent (sensory) impulses from peripheral receptors via cells in the posterior root ganglia to the cord (Figure 11.1). They also convey sudomotor and vasomotor fibres from ganglion cells in the sympathetic chain. Some nerves are predominantly motor, some predominantly sensory; the larger trunks are mixed, with motor and sensory axons running in separate bundles.

Each axon is, in reality, an extension or elongated process of a nerve cell, or *neuron* (see Chapter 10). The cell bodies of the motor neurons supplying the peripheral muscles are clustered in the anterior horn of the spinal cord; a single motor neuron with its axon may, therefore, be more than a metre long. The cell bodies of the sensory neurons serving the trunk and



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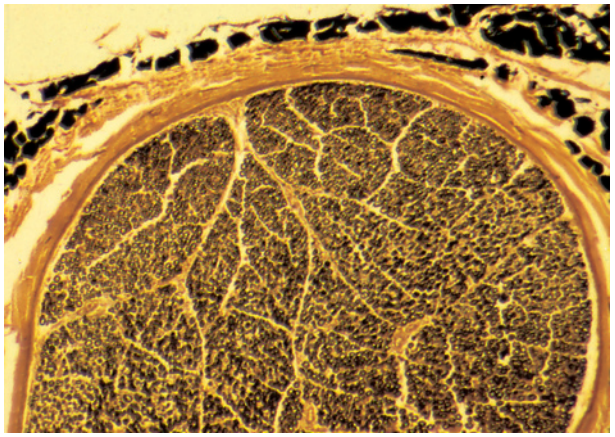

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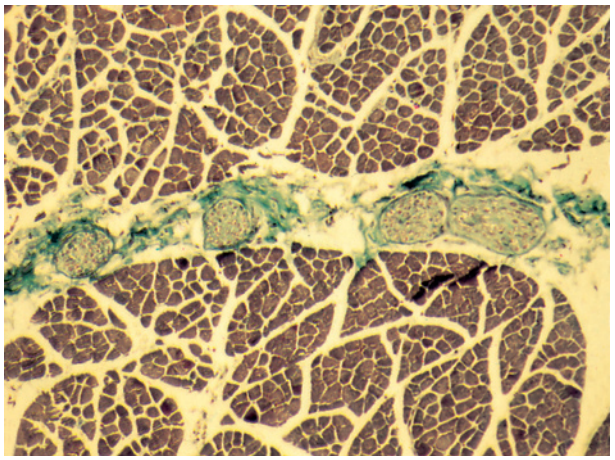


(c)

Figure 11.1 Nerve structure (a) Diagram of the structural elements of a peripheral nerve. (b) Histological section through a large nerve. (c) High-power view of the same, showing blood vessels in the perineurium.

limbs are situated in the dorsal root ganglia and each neuron has one process (axon) extending from the periphery to the cell body and another from the cell body up the spinal cord.

The peripheral ends of all the neurons are branched. A single motor neuron may supply anything from ten to several thousand muscle fibres, the ratio depending on the degree of dexterity demanded of the particular muscle (the smaller the ratio, the finer the movement). Similarly, the peripheral branches of each sensory neuron may serve anything from a single muscle spindle to a comparatively large patch of skin; here again, the fewer the end receptors served the greater the degree of discrimination.

The signal, or action potential, carried by motor neurons is transmitted to the muscle fibres by the release of a chemical transmitter, acetylcholine, at the terminal bouton of the nerve. Sensory signals are similarly conveyed to the dorsal root ganglia and from there up the ipsilateral column of the spinal cord, through the brainstem and thalamus, to the opposite (sensory) cortex. Proprioceptive impulses from the muscle spindles and joints bypass this route and are carried to the anterior horn cells as part of a local reflex arc. The economy of this system ensures that 'survival' mechanisms like balance and sense of position in space are activated with great speed.

In the peripheral nerves, all motor axons and the large sensory axons serving touch, pain and proprioception are coated with *myelin*, a multilayered lipoprotein membrane derived from the accompanying *Schwann cells*. Every few millimetres the myelin sheath is interrupted, leaving short segments of bare axon called the *nodes of Ranvier*. Nerve impulses leap from node to node at the speed of electricity, much faster than would be the case if these axons were not insulated by the myelin sheaths. Consequently, depletion of the myelin sheath causes slowing – and eventually complete blocking – of axonal conduction.

Most axons – in particular the small-diameter fibres carrying crude sensation and the efferent sympathetic fibres – are unmyelinated but wrapped in Schwann cell cytoplasm. Damage to these axons causes unpleasant or bizarre sensations and various sudomotor and vasomotor effects.

Outside the Schwann cell membrane the axon is covered by a connective tissue stocking, the *endoneurium*. The axons that make up a nerve are separated into bundles (fascicles) by fairly dense membranous tissue, the *perineurium*. In a transected nerve, these fascicles are seen pouting from the cut surface, their perineurial sheaths well defined and strong enough to be grasped by fine instruments during operations for nerve repair. The groups of fascicles that make up a nerve trunk are enclosed in an even thicker connective tissue coat, the *epineurium*. The epineurium varies in thickness and is particularly strong where the nerve is subjected to movement and traction, for example near a joint.

The nerve is richly supplied by *blood vessels* that run longitudinally in the epineurium before penetrating the various layers to become the *endoneurial capillaries*. These fine vessels may be damaged by stretching or rough handling of the nerve; however, they can withstand extensive mobilization of the nerve, making it feasible to repair or replace damaged segments by operative transposition or neurotization. The tiny blood vessels have their own *sympathetic nerve supply* coming from the parent nerve, and stimulation of these fibres (causing intraneural vasoconstriction) may be important in conditions such as reflex sympathetic dystrophy and other unusual pain syndromes.

PATHOLOGY

Nerves may be damaged by compression, stretching, laceration, thermal injury or chemical injury. The final common pathology of all these mechanisms is nerve ischaemia.

Transient ischaemia

If ischaemia is transient, such as that seen with short-term low-level compression, there are no long-term effects on the individual nerve fibres and nerve function recovers. The familiar effects of this compression are numbness with paraesthesia 'pins and needles' followed by muscle weakness after around 45 minutes. Relief of compression is followed by intense paraesthesiae lasting up to 5 minutes (the familiar 'pins and needles' after a limb 'goes to sleep'); feeling is restored within 30 seconds and full muscle power after about 10 minutes. Deep pain sensation is often preserved and prompts the release of compression.

Conduction block

In circumstances where there is an ongoing cause of compression, such as in scar tissue, axonal hypoxia may give rise to a prolonged conduction block. In situations where compression is more forceful, there may be both mechanical segmental demyelination and focal ischaemia. For example, compression of a nerve beneath a plate will result in axonotmesis.

The compression of entrapment neuropathies, such as carpal tunnel syndrome, may progress to axonal loss if sufficient compression is exerted at high force or with repeated insults to the nerve blood supply.

Seddon's classification

Seddon's description of the three different types of nerve injury (neurapraxia, axonotmesis and neurotmesis) served as a useful classification for many years. This classification is useful and readily remembered but it oversimplifies the clinical situation seen. In practice, within the commonly seen injuries to major trunk nerves there often exists a combination of neurapraxia and axonotmesis. It is whether in the main this is a degenerative or non-degenerative lesion as suggested by Birch that is of clinical relevance.

If there is persistent conduction seen in the nerve segment distal to injury, then the lesion is non-degenerative. If a Tinel's sign is observed, then there must be degeneration as this is indicative of axonal disruption. Persistent neuropathic pain suggests an ongoing ischaemic insult to the nerve.

NEURAPRAXIA

Seddon coined the term 'neurapraxia' in 1942 to describe a reversible physiological nerve conduction block in which there is loss of some types of sensation and muscle power followed by spontaneous recovery after a few days or weeks. It is due to mechanical pressure causing segmental demyelination and is seen typically in 'crutch palsy', pressure paralysis in states of drunkenness ('*Saturday night palsy*') and the milder types of tourniquet palsy.

AXONOTMESIS

This is a more severe form of nerve injury, seen typically after closed fractures and dislocations. The term means, literally, axonal interruption. There is loss of conduction but the nerve is in continuity and the neural tubes are intact. Distal to the lesion, and for a few millimetres retrograde, axons disintegrate and are resorbed by phagocytes. This *wallerian degeneration* (named after the physiologist Augustus Waller who described the process in 1851) takes only a few days and is accompanied by marked proliferation of Schwann cells and fibroblasts lining the endoneurial tubes. The denervated target organs (motor end plates and sensory receptors) gradually atrophy, and if they are not reinnervated within 2 years they will never recover.

Axonal *regeneration* starts within hours of nerve damage, probably encouraged by neurotropic factors produced by Schwann cells distal to the injury. From the proximal stumps grow numerous fine unmyelinated tendrils, many of which find their way into the cell-clogged endoneurial tubes (Figure 11.2). These axonal processes grow at a speed of 1–2 mm per day, the larger fibres slowly acquiring a new myelin coat. Eventually they join to end-organs, which enlarge and start functioning again. Some cell bodies will die after an axonotmetic insult. This is more so with a greater metabolic insult to the cell with a more proximal injury.

NEUROTMESIS

In Seddon's original classification, neurotmesis meant division of the nerve trunk, such as may occur in an open wound. It is now recognized that severe degrees of damage may be inflicted without actually dividing the nerve. If the injury is more severe, whether the nerve is in continuity or not, recovery will not occur. As in axonotmesis, there is rapid wallerian degeneration, but here the endoneurial tubes are destroyed over a variable segment and scarring thwarts any hope of regenerating axons entering the distal segment and regaining their target organs. Instead, regenerating fibres mingle with proliferating Schwann cells and fibroblasts in a jumbled knot, or 'neuroma', at the site of injury. Even after surgical repair, many new axons fail to reach the distal segment, and those that do may not find suitable Schwann tubes, or may not reach the correct end-organs in time, or may remain incompletely myelinated. Function may be adequate but is never completely normal.

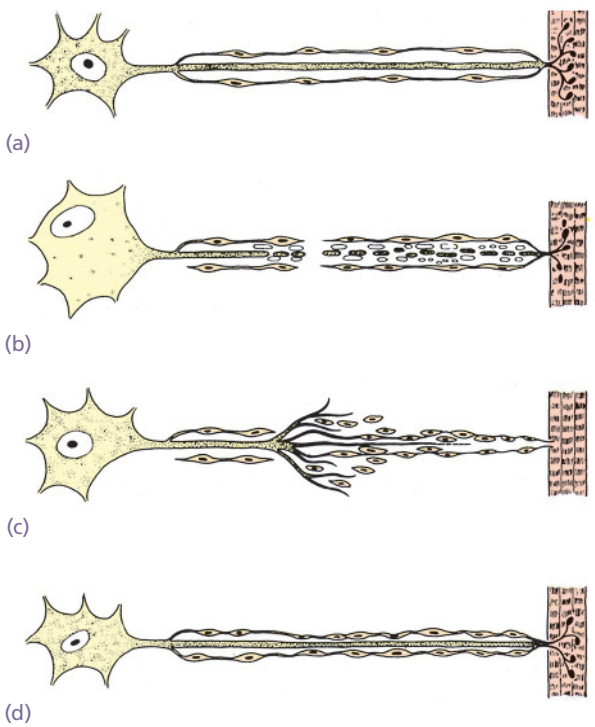


Figure 11.2 Nerve injury and repair (a) Normal axon and target organ (striated muscle). (b) Following nerve injury the distal part of the axon disintegrates and the myelin sheath breaks up. The nerve cell nucleus becomes eccentric and Nissl bodies are sparse. (c) New axonal tendrils grow into the mass of proliferating Schwann cells. One of the tendrils will find its way into the old endoneurial tube and (d) the axon will slowly regenerate.

Sunderland's classification

Sunderland's (1978) classification conversely is clinically of little use, having an anatomical basis that is only appreciated after the fact.

First degree injury This embraces transient ischaemia and neurapraxia, the effects of which are reversible.

Second degree injury This corresponds to Seddon's axonotmesis. Axonal degeneration takes place but, because the endoneurium is preserved, regeneration can lead to complete, or near-complete, recovery without the need for intervention.

Third degree injury This is worse than axonotmesis. The endoneurium is disrupted but the perineurial sheaths are intact and internal damage is limited. The chances of the axons reaching their targets are good, but fibrosis and crossed connections will limit recovery.

Fourth degree injury Only the epineurium is intact. The nerve trunk is still in continuity but internal damage is severe. Recovery is unlikely; the injured segment should be excised and the nerve repaired or grafted.

Fifth degree injury The nerve is divided and will have to be repaired.

The 'double crush' phenomenon

There is convincing evidence that proximal compression of a peripheral nerve renders it more susceptible to the effects of a second, more peripheral injury. This may explain why peripheral entrapment syndromes are often associated with cervical or lumbar spondylosis. A similar type of 'sensitization' is seen in patients with peripheral neuropathy due to diabetes or alcoholism.

CLINICAL FEATURES

Acute nerve injuries

Acute nerve injuries are easily missed, especially if associated with fractures or dislocations, the symptoms of which may overshadow those of the nerve lesion. *Always test for nerve injuries following any significant trauma*. If a nerve injury is present, it is crucial also to look for an accompanying vascular injury.

Ask the patient if there is numbness, paraesthesia or muscle weakness in the related area. Then examine the injured limb systematically for signs of abnormal posture (e.g. a wrist drop in radial nerve palsy), weakness in specific muscle groups and changes in sensibility.

Areas of altered sensation should be accurately mapped. Each spinal nerve root serves a specific dermatome (see Figure 11.3) and peripheral nerves have more or less discrete sensory territories which are illustrated in the relevant sections of this chapter. Despite the fact that there is considerable overlap in sensory boundaries, the area of altered sensibility is usually sufficiently characteristic to provide an anatomical diagnosis. Sudomotor changes may be found in the same topographic areas; the skin feels dry due to lack of sweating. If this is not obvious, the 'plastic pen test' may help. The smooth barrel of the pen is brushed across the palmar skin: normally there is a sense of slight stickiness, due to the thin layer of surface sweat, but in denervated skin the pen slips along smoothly with no sense of stickiness in the affected area.

The neurological examination must be repeated at intervals so as not to miss signs which appear hours after the original injury, or following manipulation or operation.

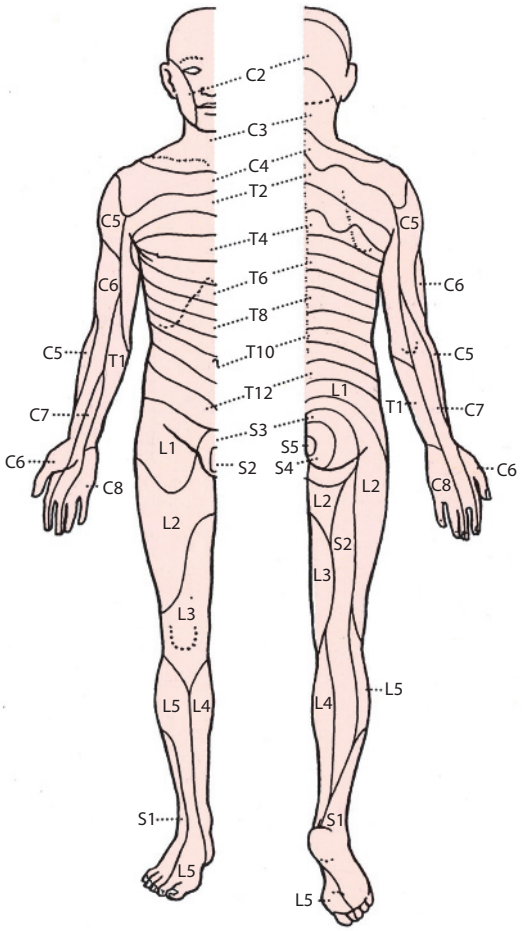


Figure 11.3 Examination Dermatomes supplied by spinal nerve roots. The sensory distribution of peripheral nerves is illustrated in the relevant sections.

Chronic nerve injuries

There are other characteristic signs in chronic injuries. The anaesthetic skin may be smooth and shiny, with evidence of diminished sensibility such as cigarette burns of the thumb in median nerve palsy or foot ulcers with sciatic nerve palsy. Muscle groups will be wasted and postural deformities may become fixed (Figure 11.4). Beware of trick movements which give the appearance of motor activity where none exists.

Assessment of nerve recovery

The presence or absence of distal nerve function can be revealed by simple clinical tests of muscle power and sensitivity to light touch and pin-prick. Remember that after nerve injury motor recovery is slower than sensory recovery. More specific assessment is required to answer two questions: How severe was the lesion? How well is the nerve functioning now?

THE DEGREE OF INJURY

The history is most helpful. A low-energy injury is likely to have caused a neurapraxia; the patient should be observed and recovery anticipated. A high-energy injury is more likely to have caused axonal and endoneurial disruption (Sunderland third and fourth degree) and so recovery is less predictable. An open injury, or a very high-energy closed injury, will probably have divided the nerve and early exploration is called for.

Tinel's sign – peripheral tingling or dysaesthesia provoked by percussing the nerve – is important. It is assessed by percussing from distal to proximal along the course of the nerve being examined. In a neurapraxia, Tinel's sign is negative. In axonotmesis, it is positive at the site of injury because of sensitivity of the regenerating axon sprouts. After a delay of a few days or weeks, the Tinel's sign will then advance at a rate of about 1 mm each day (or the clinically measureable and more useful 3 cm a month) as the regenerating axons progress along the Schwann-cell tube. *Motor activity* also should progress down the limb. Failure of Tinel's sign to advance suggests a fourth- or fifth-degree injury and the need for early exploration. If the Tinel's sign proceeds very slowly, or if muscle groups do not sequentially recover as expected, then a good recovery is unlikely and here again exploration must be considered.

Electromyography (EMG) studies can be helpful. If a muscle loses its nerve supply, the EMG will show denervation potentials by the third week. This excludes neurapraxia but of course it does not distinguish between axonotmesis and neurotmesis; this remains a clinical distinction, but if one waits too long to decide, then the target muscle may have failed irrecoverably and the answer hardly matters.

ASSESSMENT OF NERVE FUNCTION

Two-point discrimination (Figure 11.5) is a measure of innervation density. After nerve regeneration or repair, a proportion of proximal sensory axons will fail to reach their appropriate sensory end-organ; they will either have regenerated down the wrong Schwann-cell tube or will be entangled in a neuroma at the site of injury.

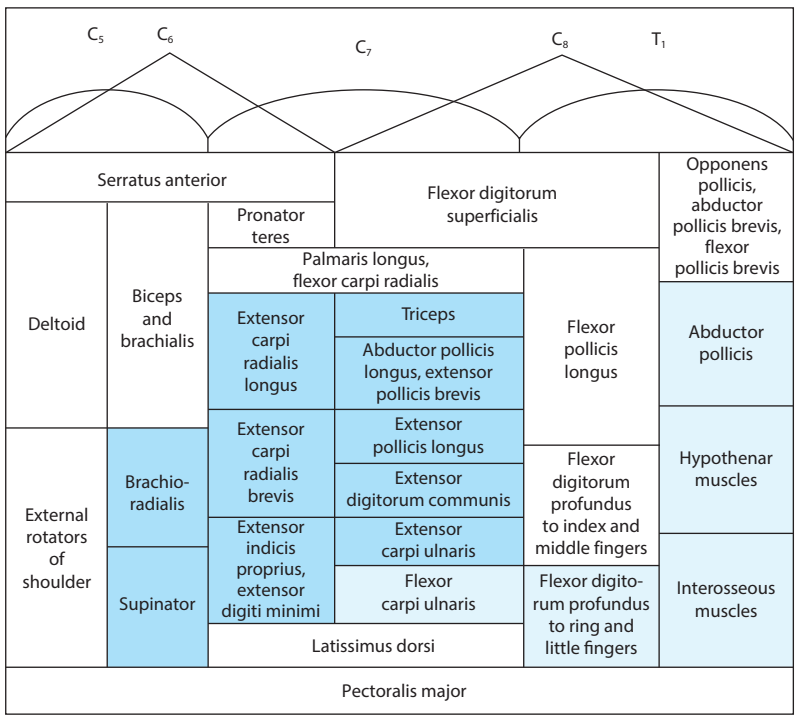


Figure 11.4 Examination Type of form used for recording muscle power in new and recovering nerve lesions (after Merle d'Aubigné). Power is recorded in individual blocks on the MRC scale 1–5.

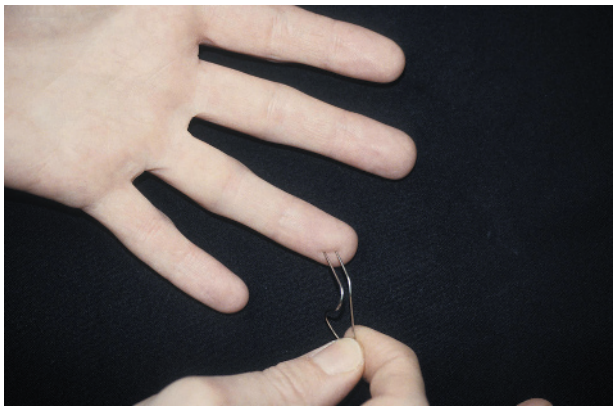


Figure 11.5 Two-point discrimination

Therefore, two-point discrimination (measured with a bent paperclip and compared with the opposite normal side) gives an indication of how completely the nerve has recovered. Static two-point discrimination measures slowly adapting sensors (Merkel cells) and moving two-point discrimination measures rapidly adapting sensors (Meissner corpuscles and pacinian corpuscles). Moving two-point discrimination is more sensitive and returns earlier. Normal static two-point discrimination is about 6 mm and moving is about 3 mm.

Threshold tests measure the threshold at which a sensory receptor is activated. They are more useful in nerve-compression syndromes, where individual receptors fail to send impulses centrally; two-point discrimination is preserved because the innervation density is not affected. Fine nylon monofilaments of varying widths are placed perpendicularly on the skin and the size of the lightest perceptible filament is recorded (Figure 11.6).

Locognosia is the ability to localize touch and can be tested with a standardized hand map.

The Moberg pick-up test measures tactile gnosis. The patient is blindfolded and instructed to pick up and identify nine objects as rapidly as possible.

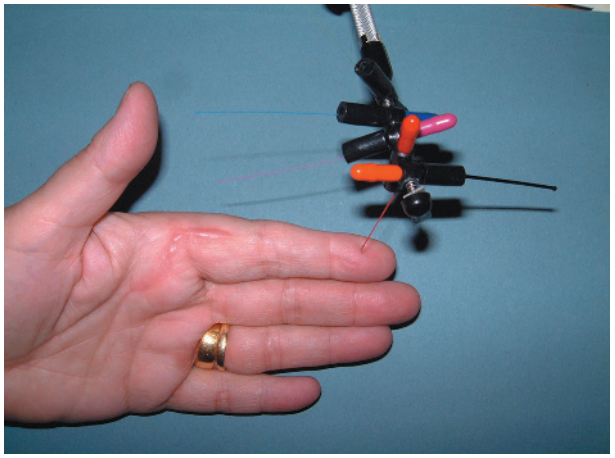


Figure 11.6 Monofilament assessment

Motor power is graded on the Medical Research Council (MRC) scale as:

0 No contraction
1 A flicker of activity
2 Muscle contraction but unable to overcome gravity
3 Contraction able to overcome gravity
4 Contraction against resistance
5 Normal power

PRINCIPLES OF TREATMENT

Nerve exploration

Closed low-energy injuries usually recover spontaneously and it is worth waiting until the most proximally supplied muscle should have regained function. Exploration is indicated: (1) if the nerve was seen to be divided and needs to be repaired; (2) if the type of injury (e.g. a knife wound or a high-energy injury) suggests that the nerve has been divided or severely damaged; (3) if recovery is inappropriately delayed and the diagnosis is in doubt.

Vascular injuries, unstable fractures, contaminated soft tissues and tendon divisions should be dealt with before the nerve lesion. The incision will be long, as the nerve must be widely exposed above and below the lesion before the lesion itself is repaired. Thought must be given to the surgical approach used. For example, in a midshaft humeral with radial nerve injury, the anterolateral approach may be employed rather than the posterior. The nerve must be handled gently with suitable instruments. Bipolar diathermy and magnification are essential. A nerve stimulator is essential if scarring makes recognition uncertain. If microsurgical equipment and expertise are not available, then the nerve lesion should be identified and the wound closed pending transfer to an appropriate facility.

Primary repair

A divided nerve is best repaired as soon as this can be done safely. Primary suture at the time of wound toilet has considerable advantages: the nerve ends have not retracted much; their relative rotation is usually undisturbed; and there is no fibrosis.

A clean cut nerve is sutured without further preparation; a ragged cut may need paring of the stumps with a sharp blade, but this must be kept to a minimum. The stumps are anatomically orientated and fine (8/0–10/0 depending on the nerve in question) sutures are inserted in the epineurium (Figure 11.7). There should be no tension on the suture line.

Sufficient relaxation of the tissues to permit tension-free repair can usually be obtained by positioning the nearby joints or by mobilizing and

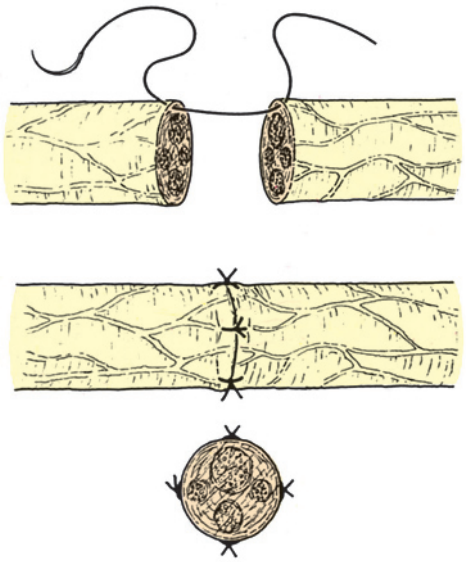


Figure 11.7 Nerve repair The stumps are correctly orientated and attached by fine sutures through the epineurium.

rerouting the nerve. If this does not solve the problem, a primary nerve graft must be considered. A traction lesion – especially of the brachial plexus – may leave a gap too wide to close. These injuries are best dealt with in specialized centres, where primary grafting or nerve transfer can be carried out.

If a tourniquet is used, it should be a pneumatic one; it must be released and bleeding stopped before the wound is closed.

The limb is splinted in a position to ensure minimal tension on the nerve; if flexion needs to be excessive, a graft is required. The splint is retained for 3 weeks and thereafter physiotherapy is encouraged.

Delayed repair

Late repair, i.e. weeks or months after the injury, may be indicated because: (1) a closed injury was left alone but shows no sign of recovery at the expected time; (2) the diagnosis was missed and the patient presents late; or (3) primary repair has failed. The options must be carefully weighed: if the patient has adapted to the functional loss, if it is a high lesion and re-innervation is unlikely within the critical 2-year period, or if there is a pure motor loss which can be treated by tendon transfers, it may be best to leave well alone. Excessive scarring and intractable joint stiffness may, likewise, make nerve repair questionable; yet in the hand it is still worthwhile simply to regain protective sensation.

The lesion is exposed, working from normal tissue above and below towards the scarred area. When the nerve is in continuity, it is difficult to know whether resection is necessary or not. If the nerve is only slightly thickened and feels soft, or if there is conduction across the lesion, resection is not advised; if the 'neuroma' is hard and there is no conduction on nerve stimulation, it should be resected, paring back the stumps until healthy fascicles are exposed.

How to deal with the gap? The nerve must be sutured without tension. The stumps may be brought together by gently mobilizing the proximal and distal segments, by flexing nearby joints to relax the soft tissues, or (in the case of the ulnar nerve) by transposing the nerve trunk to the flexor aspect of the elbow. In this way, gaps of 2 cm in the median nerve, 4–5 cm in the ulnar nerve and 6–8 cm in the sciatic nerve can usually be closed, the limb being splinted in the 'relaxing' position for 4–6 weeks after the operation. Elsewhere, gaps of more than 1–2 cm usually require grafting.

The authors' preference is for a tension free graft, but this method of achieving primary closure in segmental defects of large nerves, such as the sciatic still has validity.

Nerve guides

It is now apparent that nerve gaps of less than 2 cm can regenerate through a tube which excludes the surrounding tissue from each end. Historically, vein, silicone, metal or freeze-dried muscle conduits were all used for this purpose. More recent technologies include collagen tubes of animal origin and a variety of nano-engineered tubes which have the addition of nerve growth factors and/or Schwann cells. Human processed allograft nerve is now commercially available and shows early promise. The gold standard for mixed/motor nerve grafting remains autologous nerve graft, such as the patient's own sural nerve.

Nerve grafting

Free autogenous nerve grafts can be used to bridge gaps too large for direct suture. The sural nerve is most commonly used; up to 40 cm can be obtained from each leg. Because the nerve diameter is small, several strips may be used (cable graft). The graft should be long enough to lie without any tension, and

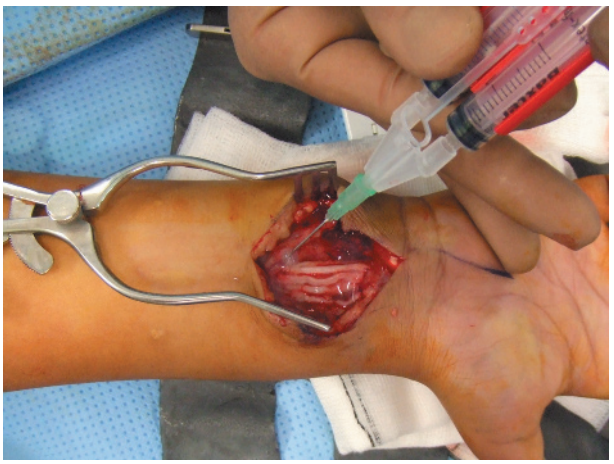


Figure 11.8 Nerve graft using fibrin polymer glue

it should be routed through a well-vascularized bed. The graft is attached at each end either by fine sutures or with fibrin glue (Figure 11.8).

It is crucial that the motor and sensory fascicles are appropriately connected by the graft. There are various techniques which can help. Careful inspection of the fascicular alignment, structure and vascular markings is often helpful. Enzyme-staining techniques can be used. Vascularized grafts are used in special situations. If the ulnar and median nerves are both damaged (e.g. in Volkmann's ischaemia), a pedicle graft from the ulnar nerve may be used to bridge the gap in the median. It is also possible to use free vascularized grafts for certain brachial plexus lesions.

Nerve transfer

The principle of nerve transfers is similar to that of tendon transfers in that a less essential function is sacrificed to reinstate a more vital one. The classic example of this is the Oberlin transfer, where the ulnar nerve is opened in the mid brachium and a fascicle destined for the flexor carpi ulnaris is detached and swung up to be attached directly to the nerve to biceps (Table 11.1). This brings motor axons close to the target organ and does not waste motor axons as the transfer is to the motor component only.

This restores antigravity biceps function in around 70% of subjects. If combined with a median fascicle to the nerve to brachialis, this is achieved in 90%. It should be noted that the outcome of nerve transfers remains less predictable than that of the classical tendon transfers, despite the former's increasing popularity.

The indication for nerve transfer is in the circumstances of very proximal nerve injury, such as root avulsion, or where the distance to the target organ, or length of graft required, precludes any chance of recovery.

Sensory nerve transfers have also been described. Here the rationale is to restore sensibility to a more vital part of the limb, for example sacrificing the lateral cutaneous nerve of the forearm to restore a degree of superficial radial nerve function.

Table 11.1 Examples of nerve transfers and function restored: spinal accessory nerve to suprascapular nerve

Procedure/ function restored	Nerve transfer
Oberlin	Ulnar to nerve to biceps
Double Oberlin	Ulnar and median fascicles to nerve to biceps and nerve to brachialis
Somsak	Radial triceps branch to axillary motor
Thumb opposition	Anterior interosseous to thenar muscles
Intrinsic function	Anterior interosseous to ulnar nerve Median to radial flexor digitorum superficialis to extensor carpi radialis brevis Flexor carpi radialis / palmaris to posterior interosseous nerve

BOX 11.1 PREREQUISITES FOR SUCCESSFUL TENDON TRANSFER

Patient factors

- A motivated patient who comprehends the rehabilitation and will attend therapy
- No progressive neurological lesion

Donor muscle/tendon unit factors

- Adequate power (one MRC grade lost in transfer)
- Adequate excursion
- Synergistic function
- Functionally expendable

Target function factors

- Supple joints and soft tissues
- Passage through a non-scarred field

Tendon transfer

Motor recovery may not occur if the axons, regenerating at about 1 mm per day, do not reach the muscle within 18–24 months of injury. This is most likely when there is a proximal injury in a nerve supplying distal muscles. In such circumstances, nerve and tendon transfers (Box 11.1) should be considered.

Recommended transfers are discussed under the individual nerve lesions.

Further reconstructive options in nerve injury

In practice, a number of staged operations may be undertaken in the willing patient to restore useful limb function following nerve injury. This may take several months or even years. Following initial nerve repair and/or nerve transfers and dependent on the outcome of those, these further procedures may include joint fusion, tendon transfer and free muscle transfer.

PROGNOSIS

Factors affecting outcome in nerve injury will include the following:

- *Delay to repair:* The best outcomes are with immediate repair.
- *Age of patient:* Children do better than adults.
- *Nature of nerve injury:* Clean cut > Crush > Traction.
- *Length of injured segment:* A graft of >10 cm is unlikely to work.

- *Distal > Proximal level of lesion:* The higher the lesion, the worse the prognosis.
- *Associated vascular injury:* Both the nerve and its target organ, be that skin or muscle, require an adequate blood supply.
- *Associated direct muscle damage:* This may prevent good re-innervation and in some circumstances (e.g. hamstring injury in sciatic nerve lesions) make nerve repair technically difficult.
- *Type of nerve:* Purely motor or purely sensory nerves recover better than mixed nerves, because there is less likelihood of axonal confusion.
- *Surgical techniques:* Skill, experience and suitable facilities are needed to treat nerve injuries. If these are lacking, it is wiser to perform the essential wound toilet and then transfer the patient to a specialized centre.

REGIONAL SURVEY OF NERVE INJURIES

BRACHIAL PLEXUS INJURIES

Pathological anatomy

The brachial plexus is formed by the confluence of nerve roots from C5 to T1; the network and its branches are shown diagrammatically in Figure 11.9.

It should be noted that the nomenclature of the cord level brachial plexus refers to the relationship of the cords to the subclavian artery in its segment immediately below the clavicle.

When the plexus is explored in its infraclavicular portion, the lateral cord is observed to lie anterior to the artery, with the medial cord lying posterior to it and sending the medial contribution to the median nerve around from posterior to anterior. The large medial cutaneous nerve of the forearm can easily be mistaken for the ulnar nerve, which lies deep to it. In addition, at this level, the posterior cord can be found lying lateral to the artery as it divides into axillary and radial branches.

The plexus, as it passes from the cervical spine between the muscles of the neck and beneath the clavicle en route to the arm, is vulnerable to injury – either a stab wound or severe traction caused by a fall on the side of the neck or the shoulder.

Traction injuries are generally classed as supraclavicular (50%), infraclavicular (40%) and combined (10%). *Supraclavicular lesions* typically occur in motorcycle accidents: as the cyclist collides with the ground or another vehicle, his neck and shoulder are wrenched apart. In the most severe injuries the arm is practically avulsed from the trunk, with rupture of the subclavian artery. *Infraclavicular lesions* are usually associated with fractures or dislocations of the shoulder; in about a quarter of cases the axillary artery also is torn. Anatomical variants such as a double contribution of lateral cord to the median nerve, or a very proximal

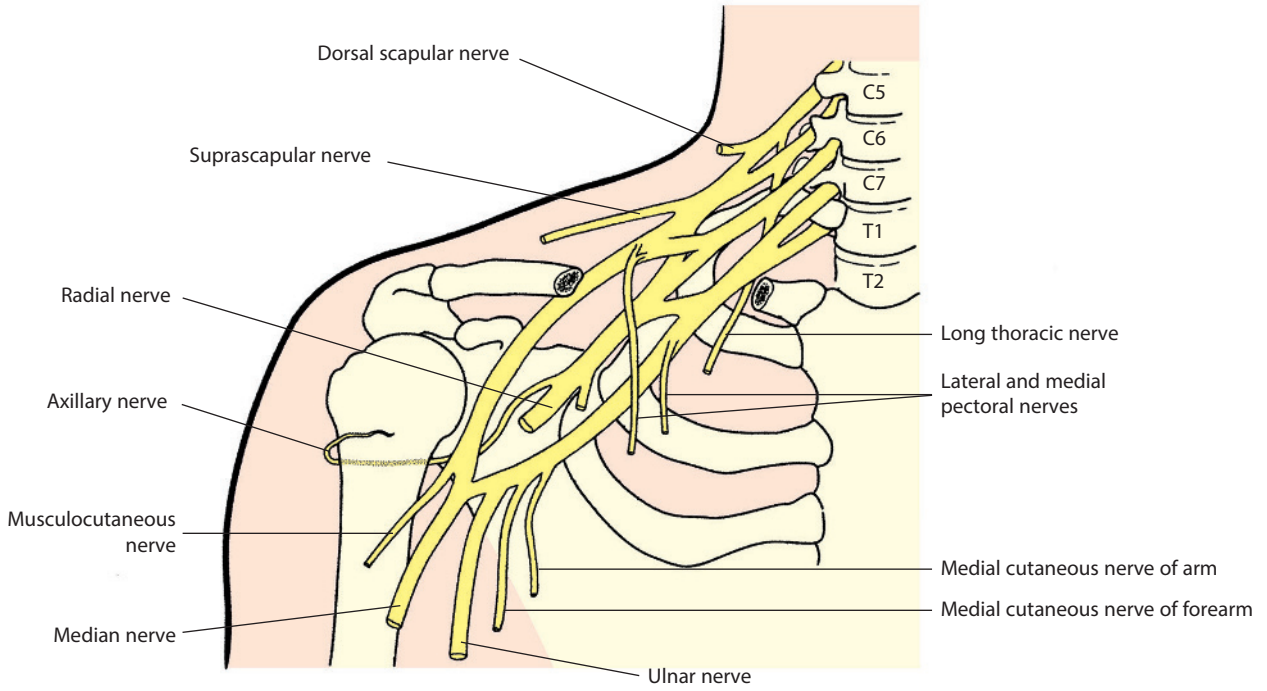


Figure 11.9 Brachial plexus Diagram of the brachial plexus and its relationship to the clavicle. (Some of the less important nerve branches and the posterior attachment of the second rib have been omitted.)

entry of the musculocutaneous nerve into the biceps, may predispose to injury by allowing less excursion of the infraclavicular plexus. *Fractures of the clavicle* rarely damage the plexus and then only if caused by a direct blow. The plexus may, however, be at risk in delayed fixation of clavicle fractures.

The injury may affect any level, or several levels within the plexus, often involving a mixture of nerve root(s), trunk(s) and nerve(s). An important distinction is made between preganglionic and postganglionic lesions. Avulsion of a nerve root from the spinal cord is a *preganglionic lesion*, i.e. disruption proximal to the dorsal root ganglion; this cannot recover and it is surgically irreparable. Rupture of a nerve root distal to the ganglion, or of a trunk or peripheral nerve, is a *postganglionic lesion*, which is surgically reparable and potentially capable of recovery. *Lesions in continuity* generally have a better prognosis than complete ruptures. *Mild lesions (neurapraxia)* are fairly common and may be caused by comparatively trivial trauma such as sudden compression by a tight harness or motor vehicle seatbelt; these recover spontaneously but mild residual symptoms may prove a nuisance for many months.

Clinical features

Brachial plexus injuries are often high-energy injuries and associated with other, life-threatening trauma which needs immediate attention. Associated injuries, such as rupture of the subclavian or axillary artery, should be sought and attended to; otherwise a poor outcome is inevitable. A major vessel injury must be stented or repaired to restore adequate blood flow for neurological recovery. Neurological dysfunction soon becomes obvious. Detailed clinical examination is directed at answering specific questions: What is the level of the lesion? Is it preganglionic or postganglionic? If postganglionic, what type of lesion is it?

THE LEVEL OF THE LESION

In upper plexus injuries (C5 and 6) the shoulder abductors and external rotators and the forearm supinators are paralysed. Sensory loss involves the outer aspect of the arm and forearm.

Pure lower plexus injuries are rare. Wrist and finger flexors are weak and the intrinsic hand muscles are paralysed. Sensation is lost in the ulnar forearm and hand.

If the entire plexus is damaged, the whole limb is paralysed and numb.

Sometimes the scapular muscles and one side of the diaphragm too are involved. By examining systematically for each component of the brachial plexus (roots, trunks, divisions, cords and branches) the exact site of the lesion may be identified. For instance, preservation of the dorsal scapular nerve (rhomboids), long thoracic nerve (serratus anterior) and suprascapular nerve (supraspinatus), but loss of musculocutaneous nerve function (biceps), radial nerve (triceps) and axillary nerve (deltoid) suggest a lateral and posterior cord injury.

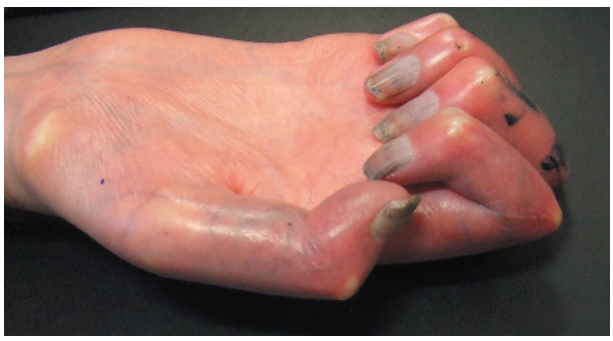


Figure 11.10 Brachial plexus injury Ischaemic insensate hand.

PRE- OR POST-GANGLIONIC?

It is crucial to establish how far from the cord the lesion is. Preganglionic lesions (root avulsions) are irreparable; postganglionic lesions may either recover (axonotmesis) or may be amenable to repair. Features suggesting root avulsion are: (1) crushing or burning pain in an anaesthetic hand; (2) paralysis of scapular muscles or diaphragm; (3) Horner's syndrome – ptosis, miosis (small pupil), enophthalmos and anhidrosis; (4) severe vascular injury; (5) associated fractures of the cervical spine; and (6) spinal cord dysfunction (e.g. hyper-reflexia in the lower limbs).

The *histamine test* is intriguing. Intradermal injection of histamine usually causes a triple response in the surrounding skin (central capillary dilatation, a wheal and a surrounding flare). If the flare reaction persists in an anaesthetic area of skin, the lesion must be proximal to the posterior root ganglion, which means that it is probably a root avulsion. With a postganglionic lesion the test will be negative because nerve continuity between the skin and the dorsal root ganglion is interrupted.

CT myelography or *MRI* may show pseudo-meningoceles produced by root avulsion (Figure 11.11). Note that during the first few days a 'positive' result is unreliable because the dura can be torn without there being root avulsion. MRI neurography is becoming increasingly sophisticated but does not substitute for careful clinical examination.

Nerve conduction studies need careful interpretation. If there is sensory conduction from an anaesthetic dermatome, this suggests a preganglionic lesion (i.e. the nerve distal to the ganglion is not interrupted). This test becomes reliable only after a few weeks, when wallerian degeneration in a postganglionic lesion will block nerve conduction.

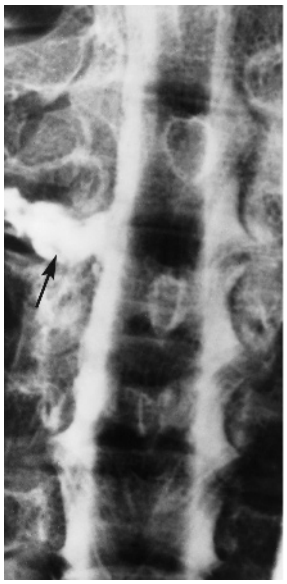


Figure 11.11 Brachial plexus The myelogram shows leakage of the contrast medium, indicating root avulsion.

THE TYPE OF LESION

Once a postganglionic lesion has been diagnosed, it becomes important to decide how severely the nerve has been damaged. The history is informative: the mechanism of injury and the impact velocity may suggest either a mild (first or second degree) or a severe (fourth or fifth degree) injury. With the former a period of observation is justified; a first or second degree lesion may show signs of recovery by 6–8 weeks. If a neurotmesis seems likely, early operative exploration is called for. Since there may be different degrees of injury within the plexus, some muscles may recover while others fail to do so.

Management

The patient is likely to be admitted to a general unit where fractures and other injuries will be given priority. Emergency surgery is required for brachial plexus lesions associated with penetrating wounds, vascular injury or severe (high-energy) soft-tissue damage whether open or closed; clean cut nerves should be repaired or grafted. This is best performed by a team specializing in this field of work.

All other closed injuries are left until detailed examination and special investigations have been completed. Patients with severe, mutilating injuries of the limb will be unsuitable for nerve surgery, at least until the prognosis for limb function becomes clear. In cases of root avulsion, limited reimplantation may be indicated. This requires the patient to have a stable cervical spine with no other unstable injuries and hence the applicable pool of suitable candidates is limited. Reimplantation is only possible within a 4-week window from time of injury.

Progress of the neurological features is carefully monitored. As long as recovery proceeds at the expected rate, watchful conservation is the byword. If recovery falters, or if special investigations show that it is more than a conduction block, then the patient should be referred to a special centre for surgical exploration of the brachial plexus and nerve repair, grafting or nerve transfer procedures. The sooner this decision is made, the better: during the early days operative exposure is easier and the response to repair more reliable. Repairs performed after 6 months are unlikely to succeed.

THE PATTERN OF INJURY

Surgical exploration reveals three typical patterns of injury:

- *C5,6(7) avulsion or rupture with C(7)8, T1 intact:* This group has the most favourable outcome as hand function is preserved and muscles innervated from the upper roots often recover after plexus repair or nerve transfer.
- *C5,6(7) rupture with avulsion of C7,8,T1:* These may recover shoulder and elbow movement after repair and grafting of the upper levels, but hand function is irretrievably lost.
- *C5–T1 avulsion:* These cases have a poor outcome. There are few donor axons available to neurotize the upper levels (shoulder and elbow function) and no recovery will take place in the hand. The implication is that all efforts for nerve repair or nerve transfer are directed towards lesions involving C5 and C6. The objectives are to regain shoulder abduction, elbow flexion, wrist extension, finger flexion, and sensibility over the lateral (radial) side of the hand.

NERVE GRAFTING AND NERVE TRANSFER

Nerve grafting is often necessary and the results for restoration of shoulder and elbow function are quite good; however, the outcome for lesions affecting the forearm and hand is disappointing.

Nerve transfer is an alternative way of providing functioning axons. If C5 and C6 are avulsed, then the spinal accessory nerve can be transferred to the suprascapular nerve; or two or three intercostal nerves can be transferred to the musculocutaneous nerve.

If one nerve root is available (e.g. C5), this should be grafted on to the lateral cord which will supply elbow flexion, finger flexion and sensation over the radial side of the hand. If two roots are available (e.g. C5, C6), these can be grafted on to the lateral and posterior cords. These procedures bypass the suprascapular nerve, which is then joined to the spinal accessory nerve.

With complete preganglionic loss, the contralateral C7 root can be extended across the chest with

autologous graft and then used as an axon source into the plexus. This is not without donor deficit, with reporting of up to 10% of significant deficit, and it should be noted that almost 90% of nerve fibres in the C7 root are afferent. The long-term outcome of this denervation is not known. Similarly, some surgeons use the phrenic nerve as an extraplexal transfer. Preservation of the one remaining useful upper limb should be considered a priority over the desire to employ any novel surgical technique.

Two or three years must pass before the final results of plexus reconstruction are apparent.

LATER RECONSTRUCTION

The best results of plexus reconstruction are obtained after very early operation. If the patient is not seen until very late after injury, or if plexus reconstruction has failed, then there are a number of options:

Tendon transfer to achieve elbow flexion Various muscles can be transferred as elbow flexors: pectoralis major (Clarke's transfer), the common flexor origin (Steindler transfer), latissimus dorsi, or triceps. The nerve supply to these muscles must remain intact, so they are suitable only for certain patterns of injury.

Free muscle transfer Gracilis, rectus femoris or the contralateral latissimus dorsi can be transferred as a free flap and innervated with two or three intercostal nerves or contralateral C7. Elbow flexion and wrist extension can be regained.

Shoulder arthrodesis Arthrodesis is usually reserved for an unstable or painful shoulder, perhaps after failure of reinnervation of the supraspinatus. The position must be tailored to the needs of the particular patient. Classically the glenohumeral joint is fused in flexion, abduction and internal rotation, all of 30 degrees. In practice, the position is determined by placing the hand to the mouth at time of operation.

Wrist arthrodesis Where posterior cord muscles have not recovered and insufficient flexor function remains to power a tendon transfer, the wrist may be fused. This may improve grip function, or free up a residual wrist flexor to power another useful function.

OBSTETRICAL BRACHIAL PLEXUS PALSY

Obstetrical palsy is caused by excessive traction on the brachial plexus during childbirth, for example by pulling the baby's head away from the shoulder or by exerting traction with the baby's arm in abduction. The incidence in the United Kingdom is around 1 in 2300 live births. Babies of diabetic mothers or unrecognized large-for-dates babies are at risk. Classically three patterns of injury were described: (1) *upper root injury (Erb's palsy)*, typically in overweight babies with shoulder dystocia at delivery; (2) *lower root injury (Klumpke's palsy)*, usually after breech delivery of smaller babies; (3) *total plexus injury.*

In practice, the injury is a spectrum of brachial plexus injury with the upper nerve roots being injured in the majority of cases. Isolated injury to the C8/T1 is very rare. Erb's palsy is now synonymous with obstetric brachial plexus injury for parents and patient groups.

Narakas classified the injury for babies delivered with a cephalic presentation:

Group 1 C5/C6
Group 2 C5/C6/C7
Group 3 Entire plexus involved
Group 4 Entire plexus with Horner's sign

Clinical features

The diagnosis is usually obvious at birth: after a difficult delivery the baby has a floppy or flail arm. Further examination a day or two later will define the type of brachial plexus injury.

Erb's palsy is caused by injury of C5, C6 and (sometimes) C7. The abductors and external rotators of the shoulder and the supinators are paralysed. The arm is held to the side, internally rotated and pronated (Figure 11.12a). There may also be loss of finger extension. Sensation cannot be tested in a baby.

Klumpke's palsy is due to injury of C8 and T1 (Figure 11.12b). The baby lies with the arm supinated and the elbow flexed; there is loss of intrinsic muscle power in the hand. Reflexes are absent and there may be a unilateral Horner's syndrome.

With a total plexus injury the baby's arm is flail and pale; all finger muscles are paralysed and there may also be vasomotor impairment and a unilateral Horner's syndrome.

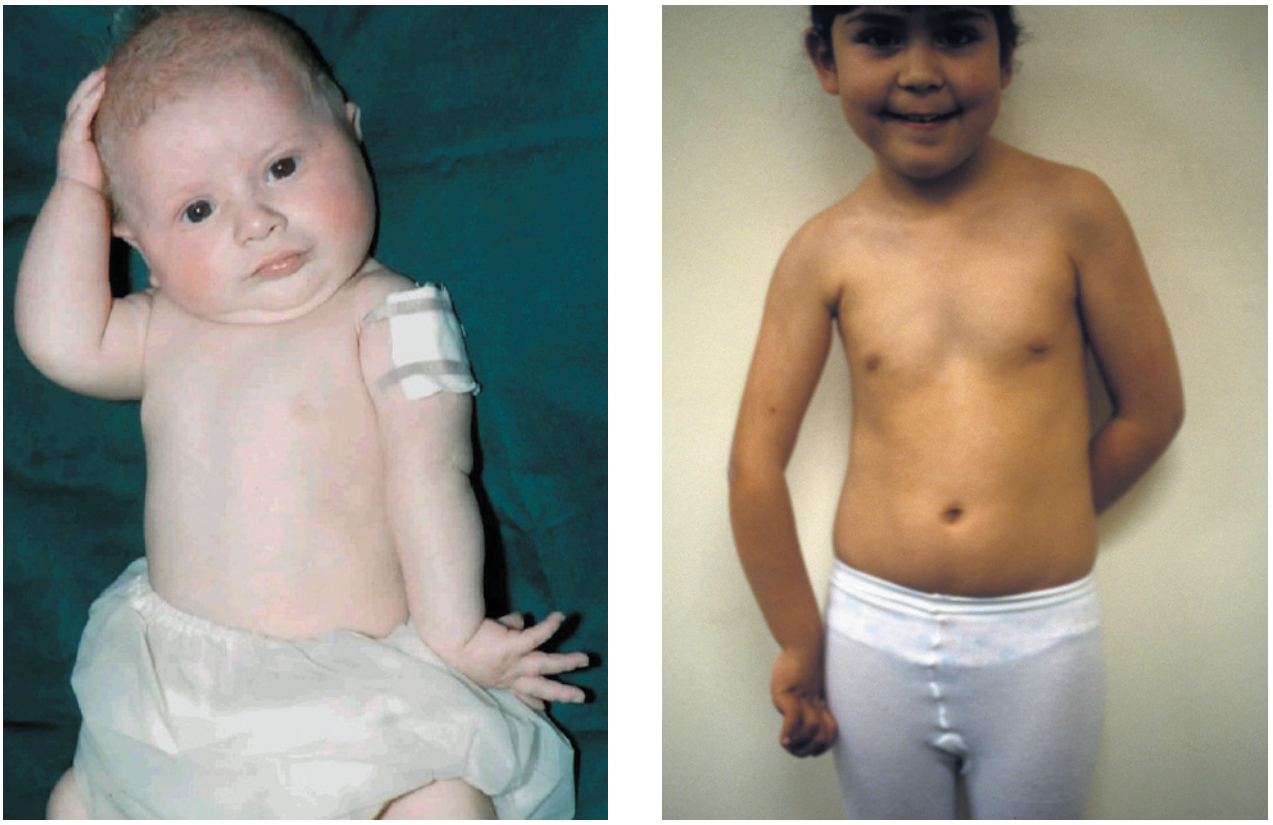
In Narakas group 1 injuries of C5, C6 the abductors and external rotators of the shoulder and the supinators are paralysed. The arm is held to the side, internally rotated and pronated. The elbow is extended.

In Narakas Group 2 the wrist and digital extensors are also paralysed, with weak or absent triceps function. The baby presents with a clenched fist.

In Group 3 injuries there is flaccid paralysis. Finger flexion is usually the first movement seen to recover.

In Group 4 injuries flaccid paralysis is accompanied by a Horner's sign. Note that the sympathetic outflow in babies also encompasses C8 as well as T1, unlike that in adults.

X-rays should be obtained to exclude fractures of the shoulder or clavicle (which are not uncommon and which can be mistaken for obstetrical palsy).



(a) (b)

Figure 11.12 Obstetrical brachial plexus palsy (a) Paralysis of the abductors and external rotators of the shoulder, as well as the forearm supinators, results in the typical posture demonstrated in this baby with Erb's palsy of the left arm. (b) Young boy with Klumpke's palsy of the right arm.

Management

OBSERVATION

Over the next few weeks one of several things may happen. While waiting for recovery, physiotherapy is applied to keep the joints mobile.

Paralysis may recover completely Around 60–70% of all cases recover spontaneously. A fairly reliable indicator is return of biceps activity by the third month. However, absence of biceps activity does not completely rule out later recovery. Residual C7 activity is a favourable prognostic indicator.

Paralysis may improve A total lesion may partially resolve, leaving the infant with a partial paralysis.

Paralysis may remain unaltered This is more likely with complete lesions, especially in the presence of a Horner's syndrome.

OPERATIVE TREATMENT

Early neurophysiological studies (at 6 weeks of age) may be a useful adjunct in decision making for these infants. These should be carried out in a specialist centre.

If there is no shoulder activity and no biceps recovery by 3 months, operative intervention should be considered. Unless the roots are avulsed, it may be possible to excise the scar and bridge the gap with free sural nerve grafts; if the roots are avulsed, nerve transfer may give a worthwhile result. This is highly demanding surgery which should be undertaken only in specialized centres.

The shoulder is prone to fixed internal rotation and adduction deformity secondary to imbalanced motor activity. This in turn may result in progressive posterior subluxation of the glenohumeral articulation. If diligent physiotherapy does not prevent this, then this is amenable to treatment by release of the subscapularis, commonly in its superior segment.

On occasion this may be supplemented by a tendon transfer. In older children, the deformity can be treated by rotation osteotomy of the humerus. Fixed flexion deformity of the elbow may be treated by splinting and serial casting. The child should be followed up through the adolescent growth spurt until the shoulder approaches adult size and is no longer at risk.

BRACHIAL NEURITIS

This is an inflammatory condition of unknown aetiology that affects the upper portion of the brachial plexus. It is characterized by severe neuralgic pain that lasts 4–10 days. Following this, muscle weakness is revealed. Recovery may take 18–24 months. The pain of brachial neuritis prevents sleep in the acute phase. It is synonymous with Parsonage Turner syndrome and neuralgic amyotrophy.

It may be associated with a preceding viral infection, or unaccustomed physical activity or stress. The pain is out of all proportion to the preceding history of activity. It is poorly recognized by orthopaedic surgeons.

The most commonly affected nerves are those of smaller diameter, such as the suprascapular, long thoracic and axillary nerves, although the muscle weakness may be more widespread and patchy.

Hereditary neuralgic amyotrophy is caused by a mutation in the *SEPT9* gene on the long arm of chromosome 17. It presents in the second or third decade of life and is characterized by recurrent episodes of brachial neuritis.

Electromyography is usually diagnostic and helpful to exclude primary muscle pathologies such as muscular dystrophy.

LONG THORACIC NERVE

The long thoracic nerve of Bell (C5, 6, 7) may be damaged in shoulder or neck injuries (usually an axonotmesis) or during operations such as first rib resection, transaxillary sympathectomy or radical mastectomy. It is often affected in cases of brachial neuritis (neuralgic amyotrophy) where an initial 4–10-day history of intense pain is followed by winging. The patient may relate the dysfunction to an apparently innocuous activity which seems inconsistent with the injury.

Clinical features

Paralysis of serratus anterior is the commonest cause of winging of the scapula. The patient may complain of aching and weakness on lifting the arm. Examination shows little abnormality until the arm is elevated in flexion or abduction. The classic test for winging is to have the patient pushing forwards against the wall or thrusting the shoulder forwards against resistance (Figure 11.13). Abduction will reveal an opening of the inferior scapulohumeral angle as viewed from behind. The fulcrum for supra and infraspinatus is unstable and activation of supraspinatus elevates both the humerus and the scapula. The inferior pole of the scapula rotates superiorly due to the unopposed pull of the trapezius muscle and the rhomboids.

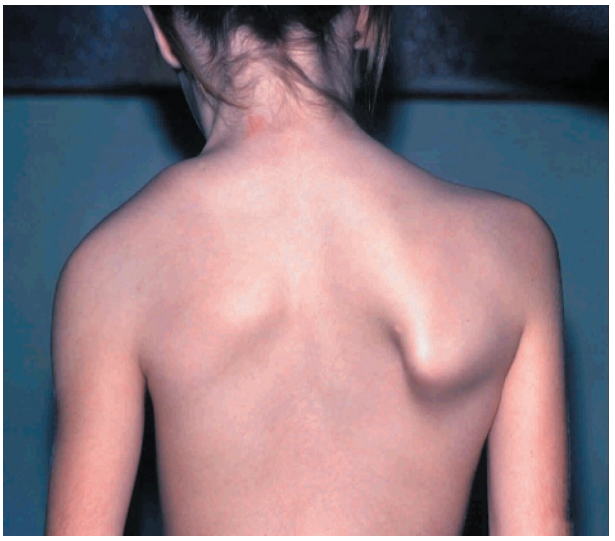


Figure 11.13 Long thoracic nerve palsy Winging of the scapula is demonstrated by the patient pushing forwards against the wall. If the serratus anterior is paralysed, the scapula cannot be held firmly against the rib cage.

Treatment

After an episode of brachial neuritis the nerve usually recovers spontaneously, though this may take 12–18 months. Persistent winging of the scapula may be amenable to operative stabilization by transferring pectoralis minor or major to the lower part of the scapula. In recalcitrant cases, scapulothoracic fusion may be considered.

SPINAL ACCESSORY NERVE

The spinal accessory nerve (C2–6) supplies the sternomastoid muscle and then runs obliquely across the posterior triangle of the neck to innervate the upper half of the trapezius (Figure 11.14). Because of its superficial course, it is easily injured in stab wounds and operations in the posterior triangle of the neck. The most common cause of injury to this nerve is lymph node biopsy. It is occasionally damaged in whiplash injuries. As the trapezius muscle is a large postural muscle, the majority of its nerve fibres are afferent, although it has no cutaneous supply. The pain of injury to the nerve is that of deep-seated, poorly localizing aching pain, reflective of deafferentation of the muscle sense organs.

Clinical features

Following an open wound or operation, the patient complains of severe deep-seated and poorly localizing pain with a restriction of shoulder movement. They may complain of clothing sliding off the affected shoulder.

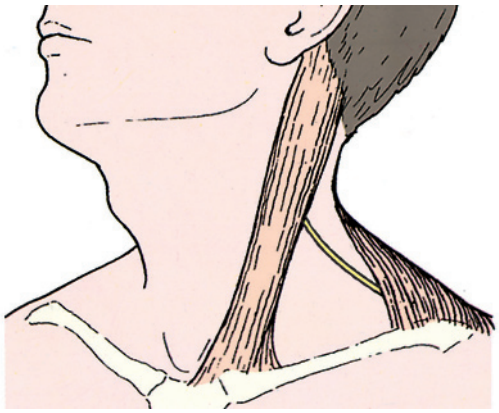


Figure 11.14 Accessory nerve The accessory nerve is embedded in the fascia which covers the posterior triangle and is easily damaged during lymph node biopsy or excision (and in stab wounds).

Examination reveals drooping of the shoulder, with weak abduction, typically limited to an abduction range of 90 degrees or less.

There is winging of the scapula on abduction, with a reduction of the inferior scapulohumeral angle. The winging differs from that seen in serratus anterior palsy in that the medial border of the scapula lifts from the posterior thorax uniformly and so may seem less pronounced. The contour of the shoulder may appear to be maintained by a preserved levator scapulae muscle, although on examination of the muscle bulk wasting of the trapezius will be evident within the first 2 months. The reduced mass of trapezius when grasped between forefinger and thumb at the border of the posterior triangle is readily appreciated.

Presentation of these injuries is often late (at an average of around 6 months after injury).

Treatment

Stab injuries and surgical injuries should be explored immediately and the nerve repaired (Figure 11.15). If the exact cause of injury is uncertain, it is prudent to wait for about 8 weeks for signs of recovery. If this does not occur, the nerve should be explored, to confirm the diagnosis and if necessary to repair the lesion by direct suture or grafting. While waiting for recovery, the arm is held in a sling to prevent dragging on the neck muscles. The results of early nerve repair are good.

Late repair or missed injury are difficult to treat. Transfer of the levator scapulae and the rhomboids to the scapular spine in the manner of Eden Lange may improve the winging and shoulder range, although results are variable.

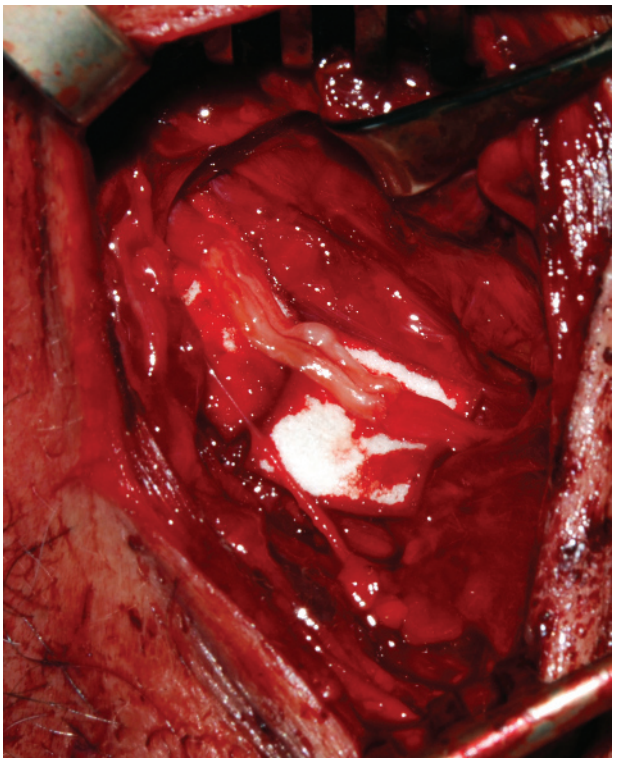


Figure 11.15 Accessory nerve Example of graft repair of spinal accessory nerve utilizing supraclavicular donor nerve.

SUPRASCAPULAR NERVE

The suprascapular nerve, which arises from the upper trunk of the brachial plexus (C5, 6), runs through the suprascapular notch to supply the supra- and infraspinatus muscles. It may be injured in fractures of the scapula, dislocation of the shoulder, by a direct blow or sudden traction, or simply by carrying a heavy load over the shoulder.

Clinical features

There may be a history of injury, but patients sometimes present with unexplained pain in the suprascapular region and weakness of shoulder abduction – symptoms readily mistaken for a rotator cuff syndrome. There is usually wasting of the supraspinatus and infraspinatus, with diminished power of abduction and external rotation. Electromyography may help to establish the diagnosis.

Treatment

This is usually an axonotmesis which clears up spontaneously after 3 months. If no recovery is seen at this stage, the nerve should be explored. In the absence of trauma one might suspect a nerve entrapment syndrome,

and decompression by division of the suprascapular ligament often brings improvement. The operative approach is through a posterior incision above and parallel to the spine of the scapula. A distal transfer of spinal accessory nerve to the suprascapular nerve may be employed if the nerve lesion is not a repairable one.

AXILLARY NERVE

The axillary nerve (C5, 6) arises from the posterior cord of the brachial plexus and runs along subscapularis and across the axilla just inferior to the shoulder joint. It emerges behind the humerus, deep to the deltoid; after supplying the teres minor, it divides into a medial branch which supplies the posterior part of the deltoid and a patch of skin over the muscle and an anterior branch that curls round the surgical neck of the humerus to innervate the anterior two-thirds of the deltoid. The landmark for this important branch is 5 cm below the tip of the acromion (Figure 11.16).

The nerve is sometimes ruptured in a brachial plexus injury. More often it is injured during shoulder dislocation or fractures of the humeral neck. Iatrogenic injuries occur in transaxillary operations on the shoulder and with lateral deltoid-splitting incisions. It is sometimes injured at the same time as the suprascapular nerve in shoulder dislocation where it is most commonly injured at its take-off from the posterior cord, just medial to the coracoid process.

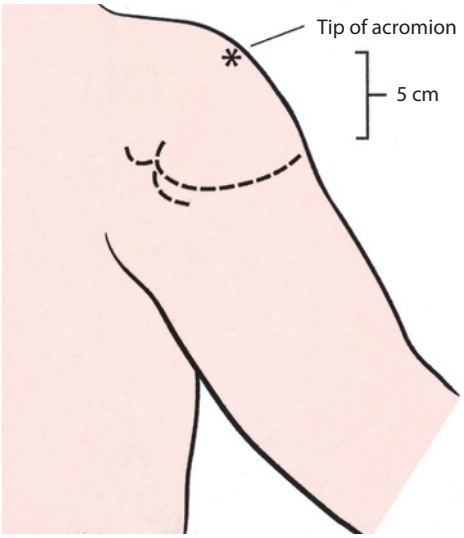
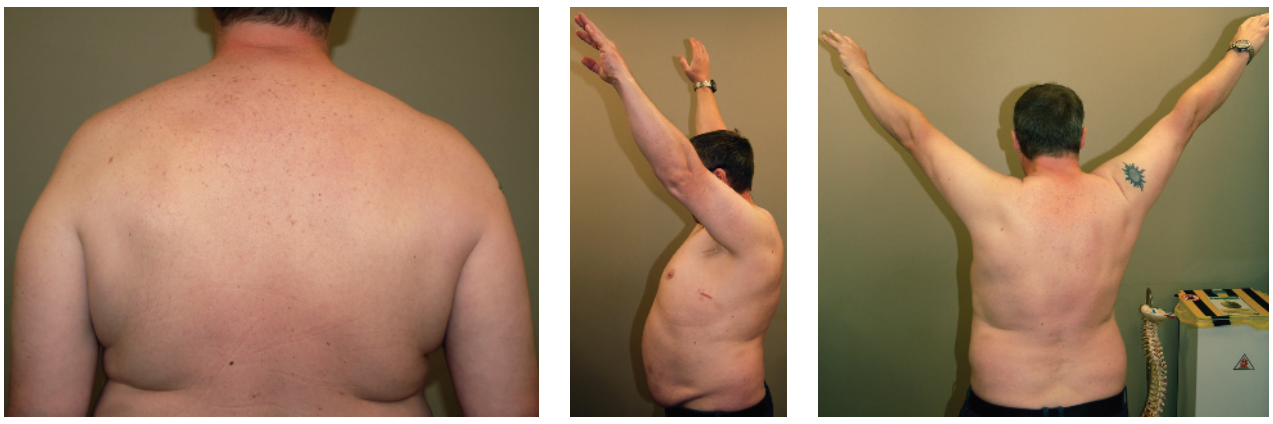


Figure 11.16 Axillary nerve Surface marking of the axillary nerve.

Clinical features

The patient complains of shoulder 'weakness', and the deltoid is wasted (Figure 11.17a). Although abduction can be initiated (by supraspinatus), it cannot be maintained. Retropulsion (extension of the shoulder with the arm abducted to 90 degrees) is impossible. Careful testing will reveal a small area of numbness over the deltoid (the 'sergeant's patch').

Abduction of the arm is possible, mediated by the supraspinatus muscle. This is commonly misunderstood by orthopaedic surgeons. Supraspinatus alone is entirely capable of elevating the upper limb above head height (Figure 11.17b,c). Deltoid confers power and stability with the elevated arm. Its force vector is such that, until 30 degrees or more of abduction has been achieved, the deltoid action is predominantly that of a vertical shearing force. Thus we see in complete rotator cuff tears that the action of deltoid results in a shrug or hitching movement. Beyond 30 degrees of abduction, the predominant force vector of deltoid is horizontal and it can now act as an abductor (Figure 11.18). This is critically important in understanding the significance of lack of abduction where there is nerve injury with shoulder dislocation. *With complete axillary nerve*



(a) (b) (c)

Figure 11.17 Axillary nerve (a) Deltoid wasting in complete axillary nerve lesion. (b,c) Elevation is achievable by intact supraspinatus alone. Note that the elevation is in the scapular plane compared to the normal side.

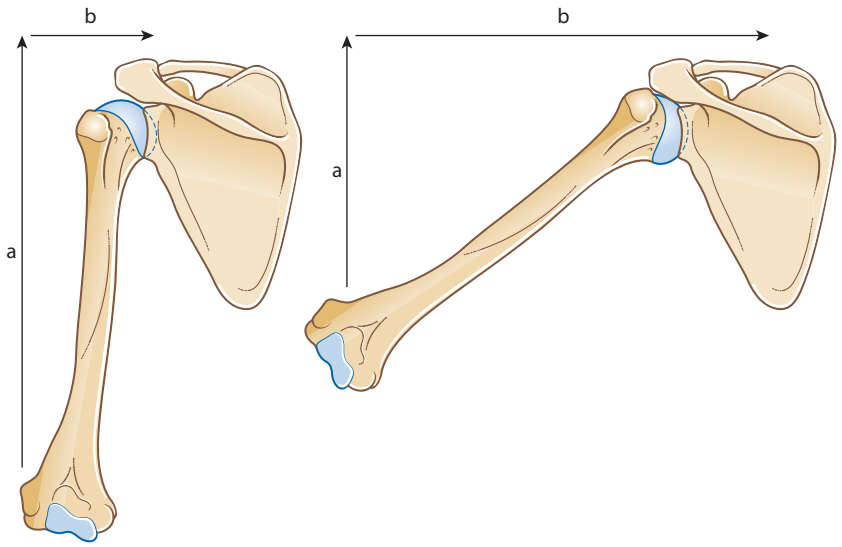


Figure 11.18 Force vector of deltoid With the adducted arm, initiation of abduction will result in a vertical shear force effect on the glenohumeral joint by the deltoid. With an incompetent supraspinatus, the shoulder will hitch rather than abduct.

palsy following shoulder dislocation, lack of abduction indicates injury to the rotator cuff, or the suprascapular nerve, until proven otherwise. Unrecognized rotator cuff injury is associated with a poor outcome in the presence of axillary nerve injury.

Treatment

Nerve injury associated with fractures or dislocations recovers spontaneously in about 80% of cases. If the deltoid shows no sign of recovery by 8 weeks, EMG should be performed; if the tests suggest denervation, then the nerve should be explored through a combined deltopectoral and posterior (quadrilateral space) approach. Excision of the nerve ends and grafting are usually necessary; a good result can be expected if the nerve is explored within 3 months of injury. The Somsak nerve transfer of a triceps branch to the anterior branch of the axillary nerve may be useful, particularly in the late-presenting lesion.

RADIAL NERVE

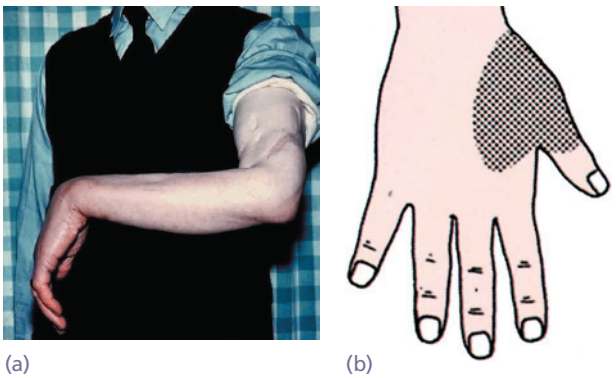
The radial nerve may be injured at the elbow, in the upper arm or in the axilla.

Clinical features

Low lesions are usually due to fractures or dislocations at the elbow, or to a local wound. Iatrogenic lesions of the posterior interosseous nerve where it winds through the supinator muscle are sometimes seen after operations on the proximal end of the radius. The patient complains of clumsiness and, on testing, cannot extend the metacarpophalangeal joints of the hand. In the thumb there is also weakness of extension and retroposition. Wrist extension is preserved because the branch to the extensor carpi radialis longus arises proximal to the elbow. The wrist is seen to extend into radial deviation without the balance of the extensor carpi radialis brevis.

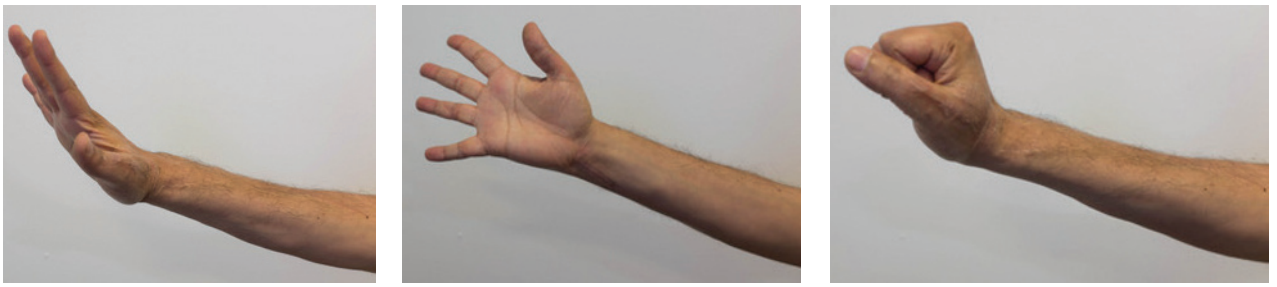
High lesions occur with fractures of the humerus or after prolonged tourniquet pressure. There is an obvious wrist drop, due to weakness of the radial extensors of the wrist, as well as inability to extend the metacarpophalangeal joints or elevate the thumb (Figure 11.19). Sensory loss is limited to a small patch on the dorsum around the anatomical snuffbox.

Very high lesions may be caused by trauma or operations around the shoulder. More often, though, they



(a) (b)

Figure 11.19 Radial nerve palsy (a) This man developed a complete drop-wrist palsy following a severe open fracture of the humerus and division of the radial nerve. (b) The typical area of sensory loss.



(a) (b) (c)

Figure 11.20 Radial nerve Function with Tendon transfers for radial nerve palsy (a) FCU to EDC and EPL; (b) Palmaris Longus to APL (c) Pronator Teres to ECRB

are due to chronic compression in the axilla; this is seen in drink and drug addicts who fall into a stupor with the arm dangling over the back of a chair ('Saturday night palsy') or in thin elderly patients using crutches ('crutch palsy'). In addition to weakness of the wrist and hand, the triceps is paralysed and the triceps reflex is absent.

Treatment

Open injuries should be explored and the nerve repaired or grafted as soon as possible.

Closed injuries are usually neurapraxia or conduction block lesions, and function eventually returns. In patients with fractures of the humerus it is important to examine for a radial nerve injury on admission, before treatment and again after manipulation or internal fixation. If the palsy is present on admission, one can afford to wait for 12 weeks to see if it starts to recover. If it does not, then EMG should be performed; if this shows denervation potentials and no active potentials, a neurapraxia is excluded and the nerve should be explored. The results, even with delayed surgery and quite long grafts, can be gratifying as the radial nerve has a straightforward motor function.

If it is certain that there was no nerve injury on admission, and the signs appear only after manipulation or internal fixation, then the chances of an iatropathic injury are high and the nerve should be explored and – if necessary – repaired or grafted without delay.

While recovery is awaited, the small joints of the hand must be put through a full range of passive movements. The wrist is splinted in extension. 'Lively' hand splints are avoided as they tend to hold the metacarpophalangeal joints in extension with the proximal interphalangeal (IP) joints flexed and this will lead to fixed contractures. If recovery does not occur, the disability can be largely overcome by tendon transfers: pronator teres to the short radial extensor of the wrist, flexor carpi radialis or ulnaris to the long finger extensors and palmaris longus (where present) to the long thumb abductor (Figure 11.20).

ULNAR NERVE

Injuries of the ulnar nerve are usually either near the wrist or near the elbow, although open wounds may damage it at any level.

Clinical features

Low lesions are often caused by cuts on shattered glass. There is numbness of the ulnar one and a half fingers. The hand assumes a typical posture in repose – the claw-hand deformity – with hyperextension of the metacarpophalangeal joints of the ring and little fingers, due to weakness of the intrinsic muscles. Hypothenar and interosseous wasting may be obvious by comparison with the normal hand. Finger abduction is weak and this, together with the loss of thumb adduction, makes pinch difficult. The patient is asked to grip a sheet of paper forcefully between thumbs and index fingers while the examiner tries to pull it away; powerful flexion of the thumb interphalangeal joint signals weakness of adductor pollicis and first dorsal interosseous with overcompensation by the flexor pollicis longus (*Froment's sign*) (Figure 11.21).

Entrapment of the ulnar nerve in the pisohamate tunnel (Guyon's canal) is often seen in long-distance cyclists who lean with the pisiform pressing on the handlebars. Unexplained lesions of the distal (motor) branch of the nerve may be due to compression by a deep carpal ganglion or ulnar artery aneurysm.

High lesions occur with elbow fractures or dislocations. The hand is not markedly deformed because the ulnar half of flexor digitorum profundus is paralysed and the fingers are therefore less 'clawed' (the '*high ulnar paradox*'). Otherwise, motor loss and sensory loss are the same as in low lesions.

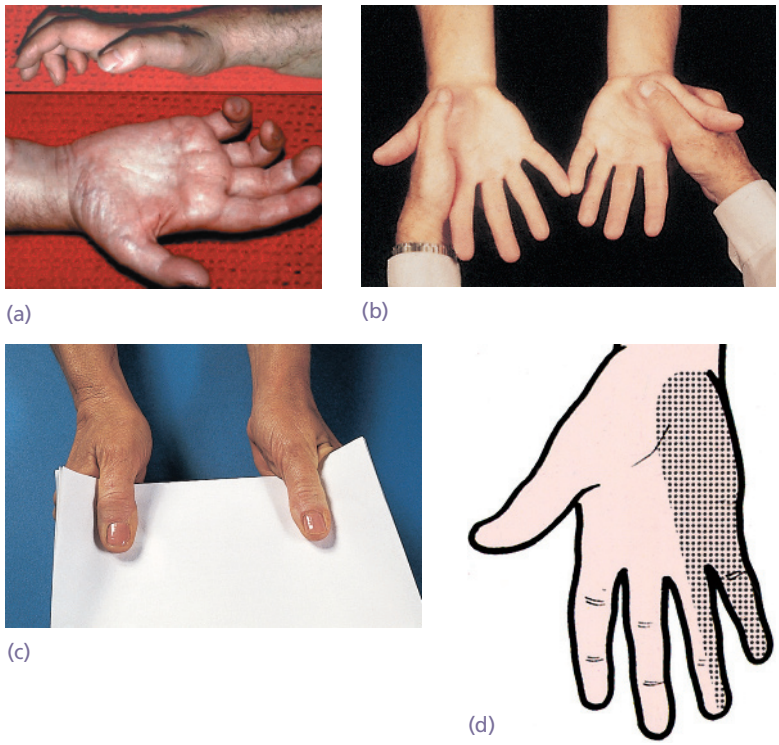


Figure 11.21 Ulnar nerve palsy
(a) Clawing of the ring and little fingers and wasting of the intrinsic muscles. (b) A good test for interosseous muscle weakness. Ask the patient to spread his fingers (abduct) as strongly as possible and then force his hands together with the little fingers apposed; the weaker side will collapse (the left hand in this case). (c) Froment's sign: the patient is asked to grip a card firmly between thumbs and index fingers; normally this is done using the thumb adductors while the interphalangeal joint is held extended. In the right hand, because the adductor pollicis is weak, the patient grips the card only by acutely flexing the interphalangeal joint of the thumb (flexor pollicis longus is supplied by the median nerve). (d) Typical area of sensory loss.

'*Ulnar neuritis*' may be caused by compression or entrapment of the nerve in the medial epicondylar (cubital) tunnel, especially where there is severe valgus deformity of the elbow or prolonged pressure on the elbows in anaesthetized or bedridden patients. It is important to be aware of this condition in patients who start complaining of ulnar nerve symptoms some weeks after an upper limb injury; one can easily be misled into thinking that the nerve lesion is due to the original injury!

Treatment

Exploration and suture of a divided nerve are well worthwhile, and anterior transposition at the elbow permits closure of gaps up to 5 cm. While recovery is awaited, the skin should be protected from burns. Hand physiotherapy keeps the hand supple and useful.

If there is no recovery after nerve division, hand function is significantly impaired. Grip strength is diminished because the primary metacarpophalangeal flexors are lost, and pinch is poor because of the weakened thumb adduction and index finger abduction. Fine, coordinated finger movements are also affected.

Metacarpophalangeal flexion can be improved by extensor carpi radialis longus to intrinsic tendon transfers (Brand), or by looping a slip of flexor digitorum superficialis around the opening of the flexor sheath (Zancolli procedure). Index abduction is improved by transferring extensor pollicis brevis or extensor indicis to the interosseous insertion on the radial side of the finger.

Distal nerve transfer from the anterior interosseous nerve may be considered in high lesions of the ulnar nerve.

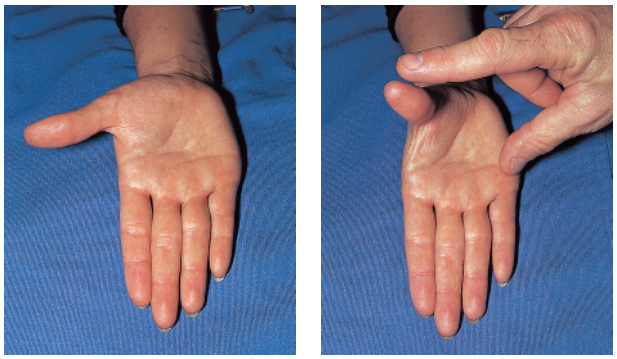
MEDIAN NERVE

The median nerve is most commonly injured near the wrist or high up in the forearm.

Clinical features

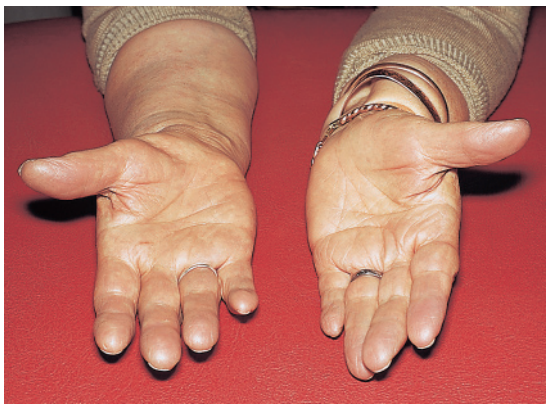
Low lesions may be caused by cuts in front of the wrist or by carpal dislocations. The patient is unable to abduct the thumb, and sensation is lost over the radial three and a half digits (Figure 11.22). In long-standing cases the thenar eminence is wasted and trophic changes may be seen.

High lesions are generally due to forearm fractures or elbow dislocation, but stabs and gunshot wounds may damage the nerve at any level. The signs are the same as those of low lesions but, in addition, the long flexors to the thumb, index and middle fingers, the radial wrist flexors and the forearm pronator muscles are all paralysed. Typically the hand is held with the ulnar fingers flexed and the index straight (the 'pointing index sign') (Figure 11.23). Also, because the thumb and index flexors are deficient, there is a characteristic pinch defect: instead of pinching with the thumb and index fingertips flexed, the patient pinches with the distal joints in full extension.



(a) (b)

Figure 11.22 Median nerve – testing for abductor power (a) The hand must remain flat, palm upwards. (b) The patient is told to point the thumb towards the ceiling against the examiner's resistance.



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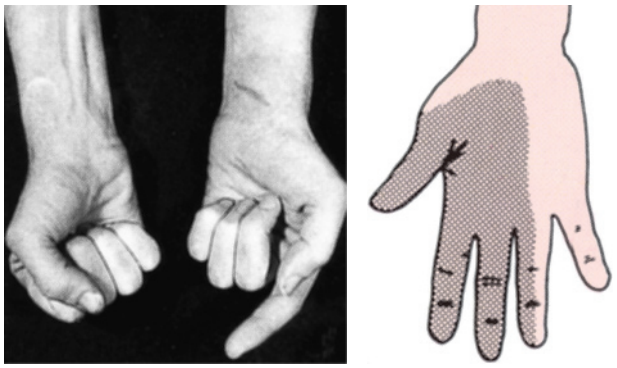


(b) (c)

Figure 11.23 Median nerve lesions (a) Wasting of the thenar eminence on the right side. (b) In high median nerve lesions, the long flexors to the thumb and index fingers are also paralysed and the patient shows the 'pointing index sign'. (c) Typical area of sensory loss.

Isolated anterior interosseous nerve lesions are extremely rare. The signs are similar to those of a high median nerve injury, but without any sensory loss. The usual cause is brachial neuritis (Parsonage–Turner syndrome) which is associated with shoulder girdle pain after immunization or a viral illness.

Treatment

If the nerve is divided, suture or nerve grafting should always be attempted. Postoperatively the wrist is splinted in flexion to avoid tension; when movements are commenced, wrist extension should be prevented.

Late lesions are sometimes seen. If there has been no recovery, the disability is severe because of sensory loss and deficient opposition. If sensation recovers but opposition does not, extensor indicis proprius or, less suitably, abductor digiti minimi can be rerouted to the insertion of abductor pollicis brevis. Extensor carpi radialis longus is available as a transfer for flexor digitorum profundus, brachioradialis for flexor pollicis longus and extensor indicis for abductor pollicis brevis.

Nerve transfers from the radial nerve in its branches to extensor carpi radialis brevis and the supinator have been proposed to restore pronation and anterior interosseous nerve function in high median nerve injuries.

UPPER LIMB NERVE INJURIES IN CHILDREN

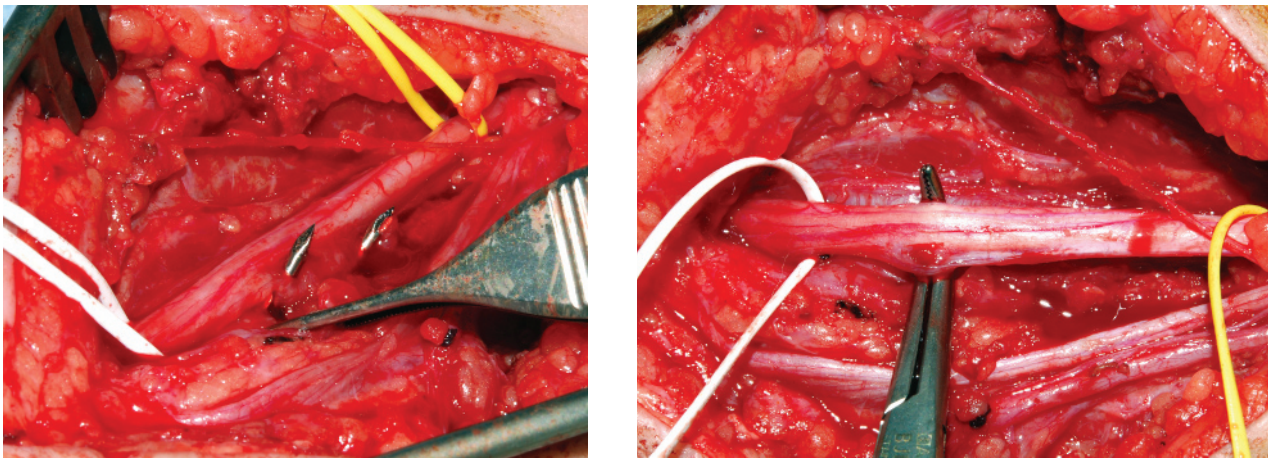
Any or all of the ulnar, median and radial nerves may be injured in the child presenting with a supracondylar fracture of the humerus. This fracture occurs in the active 4–8 year old and is technically difficult to reduce and hold, particularly where there is discontinuity of posterior periosteum in the Gartland 3 extension type.

Nerve injury may occur at time of injury or at time of reduction. It is vitally important to examine and document neurological function in these children prior to any intervention.

A nerve injury presenting following a procedure should be followed closely; a non-progressing Tinel should prompt intervention, as should light touch allodynia. The majority of iatrogenic injuries to the nerve occur as a result of nerve tether within the fracture, rather than by k-wire penetration (Figures 11.24).

LUMBOSACRAL PLEXUS

The plexus may be injured by massive pelvic trauma. These lesions are usually incomplete and often missed; the patient may complain of no more than patchy muscle weakness and some difficulty with micturition. Sensation is diminished in the perineum or in one or more of the lower-limb dermatomes. Some patients, however, have significant problems with incontinence, impotence and neurogenic pain. *Plexus injuries should always be sought in patients with fractures of the pelvis.* Surgery is rarely undertaken.



(a) (b)

Figure 11.24 (a) K-wire penetration of the median nerve in fixation of a supracondylar fracture. (b) The extent of the fascicular injury following k-wire removal.

FEMORAL NERVE

The femoral nerve may be injured by a gunshot wound, by pressure or traction during an operation or by bleeding into the thigh.

Clinical features

Quadriceps action is lacking and the patient is unable to extend the knee actively. There is numbness of the anterior thigh and medial aspect of the leg. The knee reflex is depressed. Severe neurogenic pain is common.

Treatment

This is a disabling lesion and, where possible, countermeasures should be undertaken. A thigh haematoma may need to be evacuated. A clean cut of the nerve may be treated successfully by suturing or grafting, but results are disappointing. The alternative would be a caliper to stabilize the knee, or the anterior transfer of the hamstrings to the extensor apparatus.

SCIATIC NERVE

Division of the main sciatic nerve is rare except in gunshot wounds and stab injuries to the proximal thigh or buttock. Traction lesions may occur with traumatic hip dislocations and with pelvic fractures. Intraneural haemorrhage in patients receiving anticoagulants is a rare cause of intense pain and partial loss of function. Caution should be exercised in prescribing treatment doses of anticoagulants rather than prophylactic doses in total hip replacement.

Iatropathic lesions are sometimes discovered after total hip replacement – due to inadvertent division, compression by bone levers or possibly thermal injury from extruded acrylic cement; in most cases, though, no specific cause can be found and injury is assumed to be due to traction (see below).

Clinical features

In a complete lesion the hamstrings and all muscles below the knee are paralysed; the ankle jerk is absent. Sensation is lost below the knee, except on the medial side of the leg which is supplied by the saphenous branch of the femoral nerve. The patient walks with a drop foot and a high-stepping gait to avoid dragging the insensitive foot on the ground.

Sometimes only the deep part of the nerve is affected, producing what is essentially a common peroneal (lateral popliteal) nerve lesion (see below and Figure 11.25). This is the usual presentation in patients suffering foot-drop after hip replacement; however, careful examination will often reveal minor abnormalities also in the tibial (medial popliteal) division. Electrodiagnostic studies will help to establish the level of the injury.

If sensory loss extends into the thigh and the gluteal muscles are weak, suspect an associated lumbosacral plexus injury.

In late cases the limb is wasted, with fixed deformities of the foot and trophic ulcers on the sole.

Treatment

If the nerve is known to be divided, suture or nerve grafting should be attempted even though it may take more than a year for leg muscles to be reinnervated.

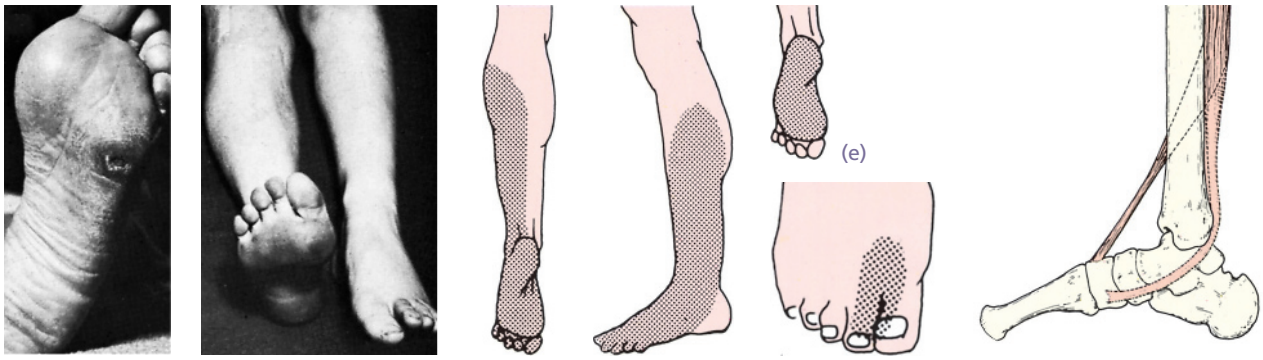


Figure 11.25 Two problems in sciatic nerve lesions (a) Trophic ulcers, which may develop because of sensory loss, and (b) foot drop. Sensory loss following division of (c) complete sciatic nerve, (d) common peroneal nerve, (e) posterior tibial nerve and (f) anterior tibial nerve. (g) Drop foot can be treated by rerouting tibialis posterior so that it acts as a dorsiflexor.

While recovery is awaited, a below-knee drop-foot splint is fitted. Great care is taken to avoid damaging the insensitive skin and to prevent trophic ulcers (Figure 11.25a).

The chances of recovery are generally poor and, at best, will be long delayed and incomplete. Partial lesions, in which there is protective sensation of the sole, can sometimes be managed by transferring tibialis posterior to the front in order to counteract the drop foot. This transfer has the advantage of correcting the inversion deformity as well as the drop foot. Tibialis posterior is best transferred to the extensor hallucis longus and the extensor digitorum longus as this prevents the claw-toe deformity which impinges on footwear and prevents its easy donning. The deformities should be corrected if they threaten to cause pressure sores. If there is no recovery whatever, amputation may be preferable to a flail, deformed, insensitive limb.

Sciatic palsy after total hip replacement

The incidence of overt sciatic nerve dysfunction is reported as 0.5–3% following primary hip replacement and about twice as high after revision. However, subclinical EMG changes are quite common. The vast majority of these resolve fairly quickly and do not manifest as postoperative nerve lesions. The less fortunate patients present soon after operation with weakness of ankle dorsiflexion, or a foot-drop, and abnormal sensibility in the distribution of the common peroneal nerve – a combination which is readily mistaken for a peroneal nerve lesion (wishful thinking in almost every case!). The reason for this is that the 'peroneal' portion of the sciatic nerve lies closest to the acetabulum and is most easily damaged. Careful examination will often show minor abnormalities also in the tibial nerve. If there is any doubt about the level of the lesion, EMG and nerve conduction tests will help.

X-rays may show a bone fragment or extruded cement (with the possibility of thermal damage) in the soft tissues; MRI may be needed to establish its proximity to the sciatic nerve. However, in most cases no cause is identified and one is left guessing whether the nerve was inadvertently injured by a scalpel point, haemostat, electrocautery, suture knot or traction levers. Delayed onset palsy may be due to a haematoma.

In about half the cases the lesion proves to be a neurapraxia or conduction block; some of these recover within weeks, others take months and may not recover completely. Unless a definite cause is known or strongly suspected, it is usually worth waiting for 6 weeks to see if the condition improves. During this time the patient is fitted with a drop-foot splint and physiotherapy is begun.

There is no agreement about the indications for immediate operation. Those who argue against it say they are unlikely to find any specific pathology and anyway, if they do discover evidence of nerve damage, the chances of functional recovery after nerve repair are probably no better than those of waiting for spontaneous improvement. Our own indications for early operation are: (1) total sciatic palsy; (2) a partial lesion associated with severe burning pain; and (3) strong evidence of a local, and possibly reversible, cause such as a bone fragment, acrylic cement or haematoma near the nerve. If the exploratory operation reveals a local cause, it should be corrected. If the nerve is divided or shows full thickness damage, repair or grafting may be worthwhile. At best, recovery will take several years and will be incomplete. Partial lesions are better left alone and the resulting disability managed by splintage and/or tendon transfers.

PERONEAL NERVES

Injuries may affect either the common peroneal (lateral popliteal) nerve or one of its branches, the deep or superficial peroneal nerves.

Clinical features

The *common peroneal nerve* is often damaged at the level of the fibular neck by severe traction when the knee is forced into varus (e.g. in lateral ligament injuries and fractures around the knee, or during operative correction of gross valgus deformities), or by pressure from a splint or a plaster cast, from lying with the leg externally rotated, by skin traction or by wounds. It is commonly injured in knee dislocations, where the posterolateral corner tears and traction is expended upon the nerve by the embracing fascial sling of biceps femoris as it attempts to violently resist the varus and rotational force. In these injuries the nerve may even be avulsed. The prognosis for recovery is poor. A ganglion from the superior tibiofibular joint can also present with this palsy when ganglion fluid tracks into the nerve via the articular branch. This is rare. The patient has a drop foot and can neither dorsiflex nor evert the foot. He or she walks with a high-stepping gait to avoid catching the toes. Sensation is lost over the front and outer half of the leg and the dorsum of the foot. Pain may be significant.

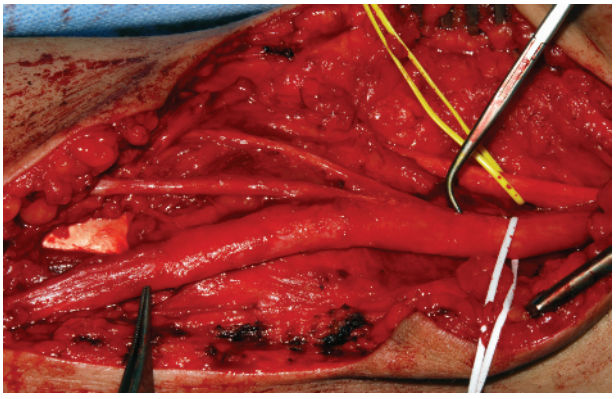
The *deep peroneal nerve* runs between the muscles of the anterior compartment of the leg and emerges at the lower border of the extensor retinaculum of the ankle. It may be threatened in an anterior compartment syndrome, causing pain and weakness of dorsiflexion and sensory loss in a small area of skin between the first and second toes. Sometimes the distal portion is cut during operations on the ankle, resulting in paraesthesia and numbness on the dorsum around the first web space.

The *superficial peroneal nerve* descends along the fibula, innervating the peroneal muscles and emerging through the deep fascia 5–10 cm above the ankle to supply the skin over the dorsum of the foot and the medial four toes. The muscular portion may be involved in a lateral compartment syndrome. The patient complains of pain in the lateral part of the leg and numbness or paraesthesia of the foot; there may be weakness of eversion and sensory loss on the dorsum of the foot. The cutaneous branches alone may be trapped where the nerve emerges from the deep fascia, or stretched by a severe inversion injury of the ankle, causing pain and sensory symptoms without muscle weakness.

Treatment

Direct injuries of the common peroneal nerve and its branches should be explored and repaired or grafted wherever possible (Figure 11.26). As usual, the earlier the repair, the better the result. While recovery is awaited, a splint may be worn to control ankle weakness. Pain may be relieved and drop foot is improved in almost 50% of patients, especially those who are operated on early. If there is no recovery, the disability can be minimized by tibialis posterior tendon transfer or by hindfoot stabilization; the alternative is a permanent splint.

Traction injuries from a knee dislocation may damage the nerve over a large length, needing a graft so long that recovery is hopeless. Splintage and tendon transfers are required.



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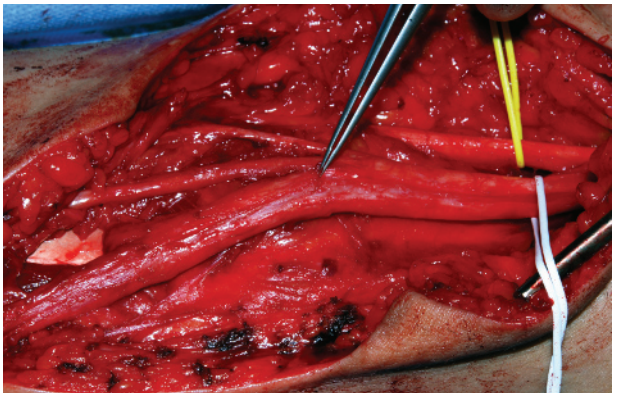


(b)

Figure 11.26 Peroneal nerve (a) The extent of long traction lesion to the common peroneal nerve. Note the normal size tibial nerve on the yellow sling for comparison. (b) Thickened epineurium opened along the neuromatous section.

TIBIAL NERVES

The tibial (medial popliteal) nerve is rarely injured except in open wounds. The distal part (posterior tibial nerve) is sometimes involved in injuries around the ankle.

Clinical features

The *tibial nerve* supplies the flexors of the ankle and toes. With division of the nerve, the patient is unable to plantarflex the ankle or flex the toes; sensation is absent over the sole and part of the calf. Because both the long flexors and the intrinsic muscles are involved, there is not much clawing. With time the calf and foot become atrophic and pressure ulcers may appear on the sole.

The *posterior tibial nerve* runs behind the medial malleolus under the flexor retinaculum, gives off a small calcaneal branch and then divides into *medial* and *lateral plantar nerves,* which supply the intrinsic muscles and the skin of the sole. Fractures and dislocations around the ankle may injure any of these branches and the resultant picture depends on the level of the lesion. Thus, posterior tibial nerve lesions cause wide sensory loss and clawing of the toes due to paralysis of the intrinsics with active long flexors; but injury to one of the smaller branches causes only limited sensory loss and less noticeable motor weakness. A compartment syndrome of the foot (e.g. following metatarsal fractures) is easily missed if one fails to test specifically for plantar nerve function.

Treatment

A complete nerve division should be sutured as soon as possible. A peculiarity of the tibial nerve is that injury or repair (especially delayed repair) may be followed by causalgia.

While recovery is awaited, a suitable orthosis is worn (to prevent excessive dorsiflexion) and the sole is protected against pressure ulceration. In suitable cases, weakness of plantarflexion can be treated by hindfoot fusion or transfer of the tibialis anterior to the back of the foot.

NERVE COMPRESSION (ENTRAPMENT) SYNDROMES

Pathophysiology

Wherever peripheral nerves traverse fibro-osseous tunnels, they are at risk of entrapment and compression, especially if the soft tissues increase in bulk (as they may in pregnancy, myxoedema or rheumatoid arthritis) or if there is a local obstruction (e.g. a ganglion or osteophytic spur).

Nerve compression impairs epineural blood flow and axonal conduction, giving rise to symptoms such as numbness, paraesthesia and muscle weakness; the relief of ischaemia explains the sudden improvement in symptoms after decompressive surgery. Prolonged or severe compression leads to segmental demyelination, target muscle atrophy and nerve fibrosis; symptoms are then less likely to resolve after decompression.

Peripheral neuropathy associated with generalized disorders such as diabetes or alcoholism may render a nerve more sensitive to the effects of compression. There is evidence, too, that proximal compression (e.g. discogenic root compression) impairs the synthesis and transport of neural substances, so predisposing the nerve to the effects of distal entrapment – the so-called '*double-crush syndrome*'.

Common sites for nerve entrapment are the *carpal tunnel* (median nerve) and the *cubital tunnel* (ulnar nerve); less common sites are the *tarsal tunnel* (posterior tibial nerve), the *inguinal ligament* (lateral cutaneous nerve of the thigh), the *suprascapular notch* (suprascapular nerve), the *neck of the fibula* (common peroneal nerve) and the *fascial tunnel of the superficial peroneal nerve.* A special case is the *thoracic outlet*, where the subclavian vessels and roots of the brachial plexus cross the first rib between the scalenus anterior and medius muscles. In these cases there may be vascular as well as neurological signs.

Clinical features

The patient complains of unpleasant tingling or pain or numbness. Symptoms are usually intermittent and sometimes related to specific postures which compromise the nerve. Thus, in the *carpal tunnel syndrome* they occur at night when the wrist is held still in flexion, and relief is obtained by moving the hand 'to get the circulation going'. In *ulnar neuropathy*, symptoms recur whenever the elbow is held in acute flexion for long periods. In the *thoracic outlet syndrome*, paraesthesia in the distribution of C8 and T1 may be provoked by holding the arms in abduction, extension and external rotation.

Areas of altered sensation and motor weakness are mapped out. In long-standing cases there may be obvious muscle wasting. The likely site of compression should be carefully examined for any local cause. Electromyography and nerve conduction tests help to confirm the diagnosis, establish the level of compression and estimate the degree of nerve damage. Conduction is slowed across the compressed segment and EMG may show abnormal action potentials in muscles that are not obviously weak or wasted, or fibrillation in cases with severe nerve damage.

Treatment

In early cases splintage may help (e.g. holding the wrist or elbow in extension) and steroid injection into the entrapment area can reduce local tissue swelling. If symptoms persist, operative decompression will usually be successful. However, in long-standing cases with muscle atrophy there may be endoneurial fibrosis, axonal degeneration and end-organ decay; tunnel decompression may then fail to give complete relief.

MEDIAN NERVE COMPRESSION

Three separate syndromes are recognized: (1) carpal tunnel syndrome (far and away the most common); (2) proximal median nerve compression (the 'pronator syndrome'); and (3) anterior interosseous nerve compression.

Carpal tunnel syndrome

This is the best known of all the entrapment syndromes. In the normal carpal tunnel there is barely room for all the tendons and the median nerve; consequently, any swelling is likely to result in compression and ischaemia of the nerve. Usually the cause eludes detection; the syndrome is, however, common at the menopause, and in one-quarter of new presentations of rheumatoid arthritis, pregnancy and hypothyroidism.

Clinical features

The history is most helpful in making the diagnosis. Pain and paraesthesia occur in the distribution of the median nerve in the hand. Night after night the patient is woken with burning pain, tingling and numbness. Hanging the arm over the side of the bed, or shaking the arm, may relieve the symptoms. In advanced cases there may be clumsiness and weakness, particularly with tasks requiring fine manipulation such as fastening buttons.

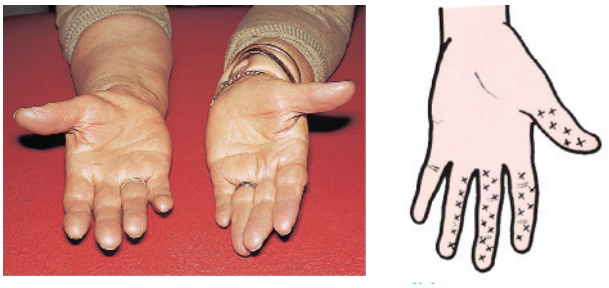
The condition is far more common in women than in men. The usual age group is 40–50 years; in younger patients it is not uncommon to find related factors such as pregnancy, rheumatoid disease, chronic renal failure or gout.

Sensory symptoms can often be reproduced by percussing over the median nerve (*Tinel's sign*) or by holding the wrist fully flexed for less than 60 seconds (*Phalen's test*). In late cases there is wasting of the thenar muscles, weakness of thumb abduction and sensory dulling in the median nerve territory (Figure 11.27).

Electrodiagnostic tests, which show slowing of nerve conduction across the wrist, are reserved for those with atypical symptoms. Radicular symptoms of cervical spondylosis may confuse the diagnosis and may coincide with carpal tunnel syndrome.

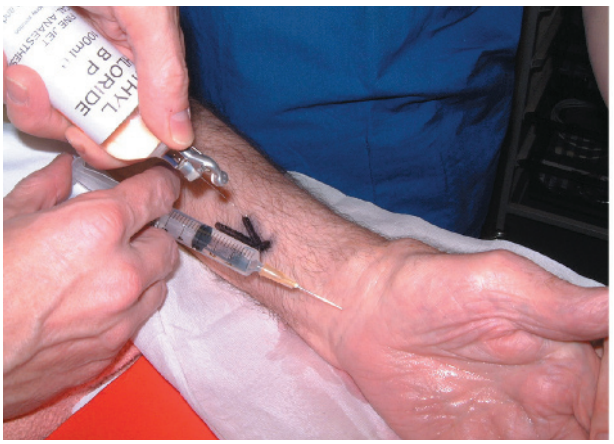
Treatment

Splints prevent wrist flexion and are especially helpful with night pain or with pregnancy-related symptoms. *Steroid injection* into the carpal canal will usually provide temporary relief and is a useful diagnostic tool (Figure 11.28a). For some it is durable, for others just temporary. It is also useful in late pregnancy when symptoms usually resolve after birth and so a temporary respite is welcomed.



(a) (b)

Figure 11.27 Median nerve compression (a) Thenar wasting in the right hand; (b) sensory loss.



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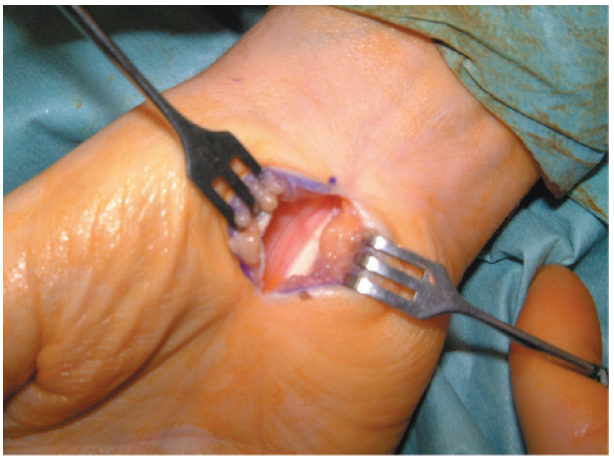


(b)

Figure 11.28 Median nerve compression treatment (a) Carpal tunnel injection; (b) open carpal tunnel release.

Surgical division of the transverse carpal ligament usually provides a quick and simple cure (Figure 11.28b). The incision should be kept to the ulnar side of the thenar crease so as to avoid accidental injury to the palmar cutaneous (sensory) and thenar motor branches of the median nerve. Internal neurolysis is not recommended. Endoscopic carpal tunnel release offers an alternative with slightly quicker postoperative rehabilitation; however, the complication rate is higher.

Proximal median nerve compression

PRONATOR SYNDROME

The median nerve can be (very rarely) compressed beneath one of several structures around the elbow including the ligament of Struthers (a connection between the medial epicondyle and the humerus), the bicipital aponeurosis or the arch-like origins of either pronator teres or flexor digitorum superficialis. This variability is not well conveyed by the more common term '*pronator syndrome*'. Symptoms are similar to those of carpal tunnel syndrome, although night pain is unusual and forearm pain is more common. Phalen's test will obviously be negative; instead, symptoms can be provoked by resisted elbow flexion with the forearm supinated (tightening the bicipital aponeurosis), by resisted forearm pronation with the elbow extended (pronator tension) or by resisted flexion of the middle finger proximal interphalangeal joint (tightening the superficialis arch). Pain may be felt in the forearm and there may be altered sensation in the territory of the palmar cutaneous branch of the median nerve (which originates proximal to the carpal tunnel). Tinel's sign may be positive over the nerve proximally but not at the carpal tunnel. Nerve conduction studies may localize the level of the compression but are often negative, particularly in postural compression. X-ray examination may show a bony spur at the attachment of Struthers' ligament (a very rare association).

Surgical decompression involves division of the bicipital aponeurosis and any other restraining structure (pronator teres, arch of flexor digitorum superficialis); great care is needed in the dissection.

ANTERIOR INTEROSSEOUS NERVE SYNDROME

The anterior interosseous nerve can be selectively compressed at the same sites as the proximal median nerve. However, spontaneous (and usually temporary) physiological failure (Parsonage–Turner syndrome) is a more likely cause. There is motor weakness without sensory symptoms. The patient is unable to make the 'OK sign' – pinching with the thumb and index finger joints flexed, like a ring – because of weakness of the flexor pollicis longus and flexor digitorum profundus. Isolated loss of flexor pollicis longus can occur. Pressure over the belly of this muscle in the forearm will flex the thumb-tip, thus excluding tendon rupture. The condition usually settles spontaneously within a few months. If it does not, surgical exploration and release or tendon transfer may be considered.

ULNAR NERVE COMPRESSION

This occurs most commonly at the elbow and less commonly at the wrist.

Cubital tunnel syndrome

The ulnar nerve is easily felt behind the medial epicondyle of the humerus (the 'funny bone'). It can be trapped or compressed within the cubital tunnel (by bone abnormalities, ganglia or hypertrophied synovium) (Figure 11.29), proximal to the cubital tunnel (by the fascial arcade of Struthers) or distal to the cubital tunnel as it passes through the two heads of flexor carpi ulnaris to enter the forearm (Osbourne's canal). Sometimes it is 'stretched' by a cubitus valgus deformity or simply by holding the elbow flexed for long periods.

Clinical features

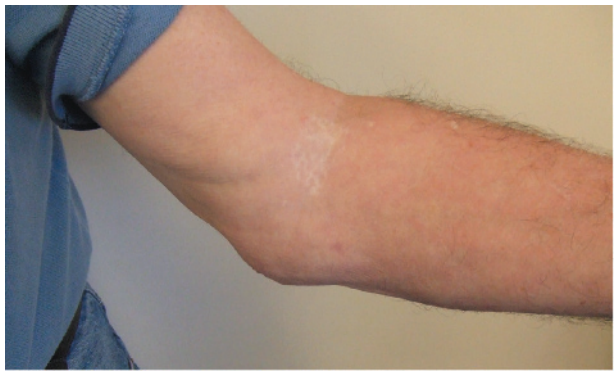
The patient complains of numbness and tingling in the little and the ulnar half of the ring finger; symptoms may be intermittent and related to specific elbow postures (e.g. they may appear only while the patient is lying down with the elbows flexed, or while reading or using a mobile phone – again with the elbows flexed). Initially there is little to see, but in late cases there may be weakness of grip, slight clawing, intrinsic muscle wasting and diminished sensibility in the ulnar nerve territory. Froment's sign and weakness of abductor digiti minimi can often be demonstrated.

Bone or soft-tissue abnormalities may be obvious. Tinel's percussion test, tenderness over the nerve behind the medial epicondyle, reproduction of the symptoms with flexion of the elbow, and weakness of flexor carpi ulnaris and the flexor digitorum profundus to the little finger all suggest compression at the elbow rather than at the wrist.

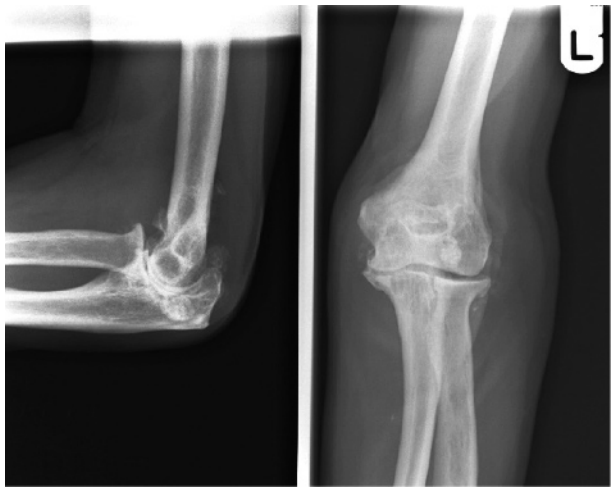
The diagnosis may be confirmed by nerve conduction tests; however, since the symptoms are often postural or activity related, a negative test does not exclude the diagnosis.

Treatment

Conservative measures such as modification of posture and splintage of the elbow in mid-extension at night should be tried.



(a)



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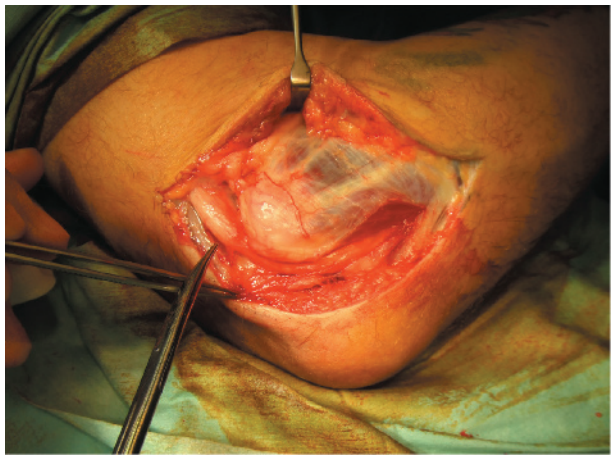


(c)

Figure 11.29 Ulnar nerve compression at the elbow
The ulnar nerve may be compressed in the cubital tunnel by (a) tension in a valgus elbow or (b) osteoarthritic spurs. (c) Surgical release in situ.

If symptoms persist, and particularly if there is intrinsic wasting, operative decompression is indicated. Options include simple release of the roof of the cubital tunnel, anterior transposition of the nerve into a subcutaneous or submuscular plane, or medial epicondylectomy. Simple release is preferable as it avoids the potential denervation associated with transposition or the persisting epicondylar pain associated with epicondylectomy. Transposition also usually affects the segmental blood supply of an already compromised nerve. During the surgical approach, great care is taken to avoid damaging the posterior branch of the medial cutaneous nerve of the forearm; otherwise troublesome numbness, if not neurogenic pain or even complex regional pain syndrome, may result.

A branch of the medial cutaneous nerve is always found within 5 cm distal to the medial epicondyle.

Compression in Guyon's canal

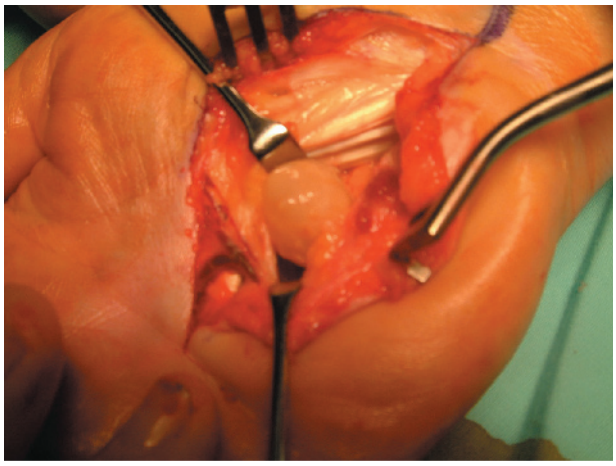
The ulnar nerve can be compressed as it passes through Guyon's canal at the ulnar border of the wrist. The symptoms can be pure motor, pure sensory or mixed, depending on the precise location of entrapment. A ganglion from the triquetrohamate joint is the most common cause; a fractured hook of hamate and ulnar artery aneurysm (seen with overuse of a hammer) are much rarer causes. Preservation of sensation in the dorsal branch of the ulnar nerve (which leaves the nerve proximal to Guyon's canal) suggests entrapment at the wrist rather than elbow; similarly, power to flexor carpi ulnaris and flexor digitorum profundus to the little finger will be maintained.

After electrophysiological localization of the lesion to the wrist, further investigations should be considered: MRI may demonstrate a ganglion, CT a carpal fracture and Doppler studies an ulnar artery aneurysm. Depending on the results of these investigations, surgery can be planned (Figure 11.30).

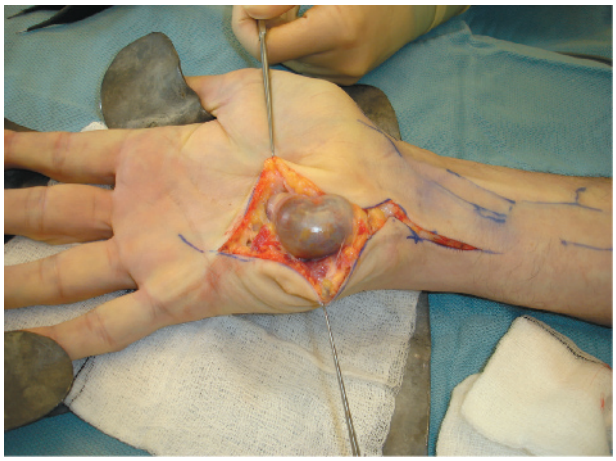
RADIAL (POSTERIOR INTEROSSEOUS) NERVE COMPRESSION

The radial nerve itself is rarely the source of 'entrapment' symptoms. Just above the elbow, it divides into a superficial branch (sensory to the skin over the anatomical snuffbox) and the posterior interosseous nerve, which dives between the two heads of the supinator muscle before supplying motor branches to extensor carpi ulnaris and the metacarpophalangeal extensors (branches to extensor carpi radialis longus and brevis arise above the elbow).

Posterior interosseous nerve compression may occur at five sites, represented by the mnemonic *FREAS* (*F*ibrous bands around radiocapitellar joint; *R*ecurrent arterial branches; *E*xtensor carpi radialis brevis, *A*rcade of Frohse (a thickening at the proximal edge of supinator); distal edge of *S*upinator). It may also be caused by a space-occupying lesion pushing



(a)



(b)

Figure 11.30 Ulnar nerve compression in Guyon's canal (a) Schwannoma pushing on the ulnar nerve. (b) Ulnar artery aneurysm.

on the nerve – a ganglion, a lipoma or severe radiocapitellar synovitis.

Two clinical patterns are encountered: the *posterior interosseous syndrome* and the *radial tunnel syndrome*.

Posterior interosseous syndrome

Clinical features

This is a pure motor disorder and there are no sensory symptoms. Gradually emerging weakness of metacarpophalangeal extension affects first one or two and then all the digits. Wrist extension is preserved (the nerves to extensor carpi radialis longus arise proximal to the supinator) but the wrist veers into radial deviation because of the weak extensor carpi ulnaris (Figure 11.31). This feature helps to distinguish posterior interosseous nerve entrapment from conditions such as neuralgic amyotrophy, in which the more proximally supplied muscles are often affected.

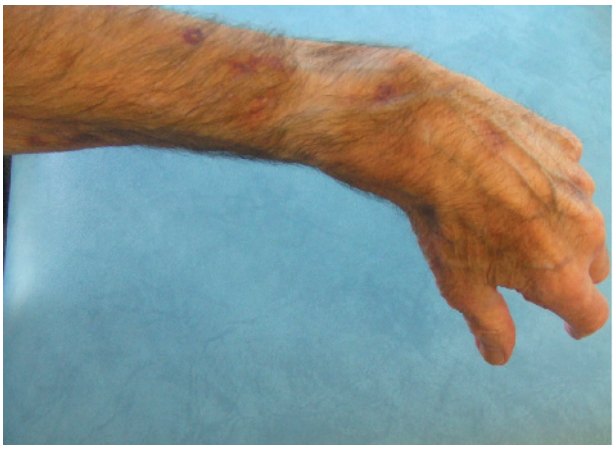


Figure 11.31 Posterior interosseous nerve compression Wrist in radial deviation; fingers dropped.

Compression usually occurs within the tunnel (FREAS) but it may also be caused by swellings (a lipoma, a ganglion or synovial proliferation) in or around the radial tunnel. MRI may help to pinpoint the diagnosis.

Treatment

Surgical exploration is warranted if the condition does not resolve spontaneously within 3 months or earlier if MRI shows a swelling. Recovery after surgery is slow; if there is no improvement by the end of a year, and if muscle weakness is disabling, tendon transfer is needed.

Radial tunnel syndrome

This syndrome is controversial; the symptoms resemble those of 'tennis elbow' and the condition is sometimes labelled '*resistant tennis elbow*'. However, a careful history and examination should distinguish between the two.

Although a motor nerve is involved, the patient presents with pain, often work-related or at night, just distal to the lateral aspect of the elbow. Resisted wrist extension may precipitate the pain. Unlike posterior interosseous syndrome, there is no weakness and there is not an association with a mass lesion. Electrodiagnostic tests are not helpful.

If the symptoms do not resolve with prolonged non-operative measures (modification of activities and splintage), surgery is considered. The nerve is freed beneath the extensor carpi radialis brevis and supinator muscle. However, the patient should be warned that surgery often fails to relieve the symptoms.

SUPRASCAPULAR NERVE COMPRESSION

Chronic or repetitive compression of the suprascapular nerve and its branches is much more common than is generally recognized. The peculiar anatomy of the nerve makes it unusually vulnerable to both traction and compression. However, the symptoms of this condition closely mimic those of rotator cuff lesions and cervical radiculopathy; unless the diagnosis is kept in mind in all such cases, it is likely to be missed.

The suprascapular nerve arises from the upper trunk of the brachial plexus in the posterior triangle of the neck and then courses through the suprascapular notch beneath the superior transverse scapular ligament to supply the supraspinatus and infraspinatus muscles. It also sends sensory branches to the posterior part of the glenohumeral joint, the acromioclavicular joint, the subacromial bursa, the ligaments around the shoulder and (in a small proportion of people) the skin on the outer, upper aspect of the arm.

Compression or entrapment occurs at two sites: (a) the suprascapular notch and (b) a fibro-osseous tunnel where the infraspinatus branch curves around the edge of the scapular spine. Causes are continuous pressure or intermittent impact on the supraclavicular muscles (e.g. by carrying loads on the shoulder) or repetitive traction due to forceful shoulder movements (e.g. in games which involve pitching and throwing). In some cases nerve compression may be produced by a soft-tissue mass such as a large 'ganglion' at the back of the shoulder joint.

Clinical features

There may be a history of injury to the pectoral girdle; more often patients present with unexplained pain in the suprascapular region or at the back of the shoulder, and weakness of shoulder and upper arm movements – symptoms readily mistaken for cervical radiculopathy or a rotator cuff disorder. There is usually wasting of the supraspinatus muscle and diminished power of abduction and external rotation. Tensing the nerve by forceful adduction (pulling the arm across the front of the chest) causes increased pain.

Special investigations

Electromyography and measurement of nerve conduction velocity may help to establish the diagnosis. Ultrasonography and MRI are useful in excluding a soft-tissue mass.

Treatment

The first step is to stop any type of activity which might stress the suprascapular nerve; after a few weeks, graded muscle-strengthening exercises can be introduced. If the condition is likely to settle, it will do so within 3–6 months.

If there is no improvement, or if imaging studies reveal a soft-tissue mass, operative decompression is justified. The nerve is approached through a posterior incision above and parallel to the spine of the scapula. Provided the diagnosis was correct, there is a good chance that symptoms will be improved; however, some muscle wasting will probably remain.

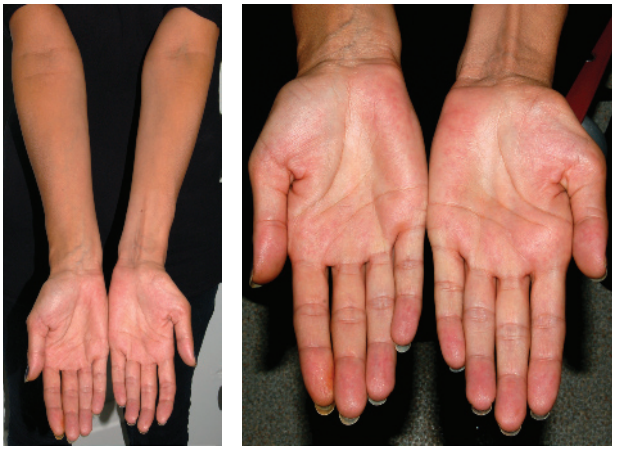
THORACIC OUTLET SYNDROME

Neurological and vascular symptoms and signs in the upper limbs may be produced by compression of the lower trunk of the brachial plexus (C8 and T1) and subclavian vessels between the clavicle and the first rib.

The subclavian artery and lower brachial trunk pass through a triangle based on the first rib and bordered by scalenus anterior and medius. These neurovascular structures are made taut when the shoulders are braced back and the arms held tightly to the sides; an extra rib (or its fibrous equivalent extending from a large costal process), or an anomalous scalene muscle, exaggerates this effect by forcing the vessel and nerve upwards.

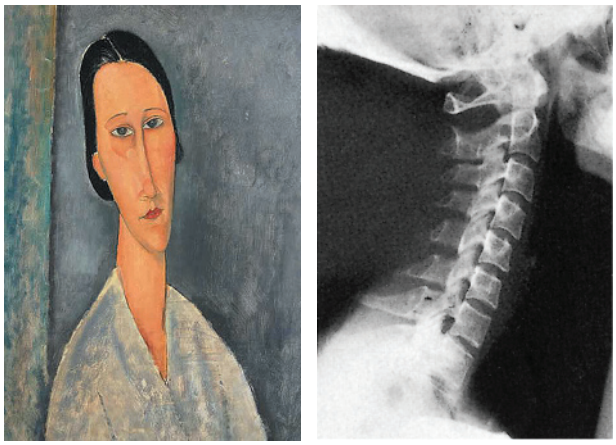
These anomalies are all congenital, yet symptoms are rare before the age of 30. This is probably because the blood vessels supplying the trunk level of the plexus become less compliant in the third decade.

Stretching or compression of the lower nerve trunk produces sensory changes along the ulnar side of the forearm and hand, and weakness of the intrinsic hand muscles. In true *Gilliatt–Sumner type neurogenic thoracic outlet*, there is characteristic wasting of the hypothenar eminence and forearm musculature (Figure 11.32). The subclavian artery is rarely



(a) (b)

Figure 11.32 Gilliatt–Sumner type neurogenic thoracic outlet syndrome (a) Characteristic wasting of the forearm and hand in the long-standing condition. (b) Hand wasting.



(a) (b)

Figure 11.33 Thoracic outlet syndrome (a) Amadeo Modigliani's painting of Madame Zoborowska. (Courtesy of Tate Gallery London.) (b) X-ray of a long-necked woman. All the vertebrae down to T1 are above the clavicle.

compressed but the lumen may contract due to irritation of its sympathetic supply, or else its wall may be damaged leading to the formation of small emboli. Even more unusual are signs of venous compression – oedema, cyanosis or thrombosis.

Clinical features

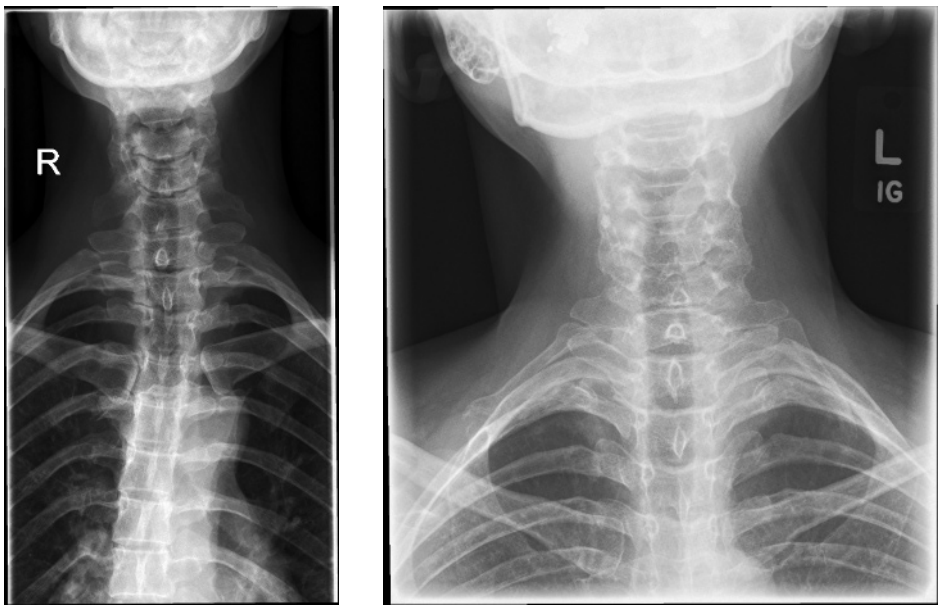
The patient, typically a woman in her thirties, complains of pain and paraesthesia extending from the shoulder, down the ulnar aspect of the arm and into the medial two fingers. Symptoms tend to be worse at night and are aggravated by bracing the shoulders (wearing a back-pack) or working with the arms above shoulder height. Examination may show mild clawing of the ulnar two fingers with wasting and weakness of the intrinsic muscles. If a female, the patient is often long-necked with sloping shoulders (like a Modigliani painting – Figure 11.33).

Vascular signs are uncommon, but there may be cyanosis, coldness of the fingers and increased sweating. *Unilateral Raynaud's phenomenon should make one think 'thoracic outlet'.*

Symptoms and signs may be reproduced by various provocative manoeuvres. In *Adson's test* the patient's neck is extended and turned towards the affected side while he or she breathes in deeply; this compresses the interscalene space and may cause paraesthesia and obliteration of the radial pulse. In *Wright's test* the arms are abducted and externally rotated; again the symptoms recur and the pulse disappears on the abnormal side. The examination is continued by asking the patient to hold his or her arms high above their head and then open and close the fingers rapidly; this may cause cramping pain on the affected side (*Roos test*). Unfortunately, these tests are neither sensitive nor specific enough to clinch the diagnosis.

Investigations

X-rays of the neck occasionally demonstrate a cervical rib or an abnormally long C7 cervical process (Figure 11.34). X-rays should also be obtained of the lungs (is there an apical tumour?) and the shoulders (to exclude any painful local lesion). *MRI* scanning of the cervical spine and MR of the brachial plexus with angiographic sequences with elevated arms may be helpful (Figure 11.35).



(a) (b)

Figure 11.34 Cervical ribs (a) Elongated transverse process of C7 associated with cervical band and (b) bilateral cervical ribs.

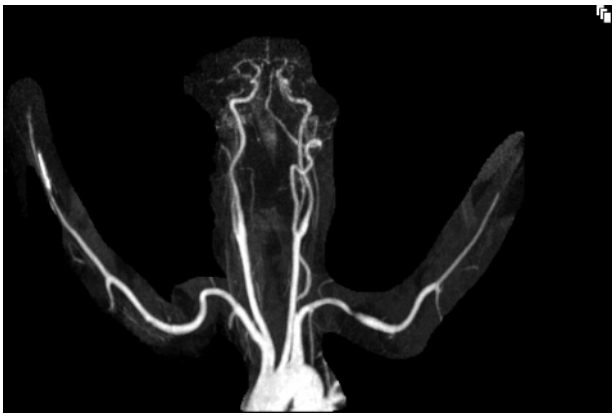


Figure 11.35 Cervical ribs MR Angiogram showing left subclavian stenosis with arm elevation.

Electrodiagnostic tests are helpful mainly to exclude peripheral nerve lesions such as ulnar or median nerve compression which may confuse the diagnosis.

Diagnosis

In the absence of clear motor signs (which are rare!) the diagnosis of thoracic outlet syndrome is not easy. Some of the symptoms occur as transient phenomena in *normal individuals*, and 'cervical ribs' (Figure 11.36) are sometimes discovered as incidental findings in patients who are X-rayed for other reasons. Postural obliteration of the radial pulse, likewise, may be quite normal in up to a third of individuals; the provocative tests should be interpreted as positive only if they affect the pulse *and* reproduce the sensory symptoms.

The early symptoms and signs can be mistaken for those of *ulnar nerve compression*. In fact, ulnar neuropathy may accompany thoracic outlet compression as a manifestation of the double-crush syndrome. There is pain and numbness over the medial side of the forearm and hand. In severe cases there will be wasting of all the intrinsic muscles (T1) and weakness of the long flexors (C8).

Cervical spondylosis is sometimes discovered on X-ray. This disorder seldom involves the T1 nerve root.

Pancoast's syndrome, due to apical carcinoma of the bronchus with infiltration of the structures at the root of the neck, includes pain, numbness and weakness of the hand. A hard mass may be palpable in the neck, and X-ray of the chest shows a characteristic opacity.

Rotator cuff lesions sometimes cause pain radiating down the arm, but there are no neurological symptoms and shoulder movement is likely to be abnormal.

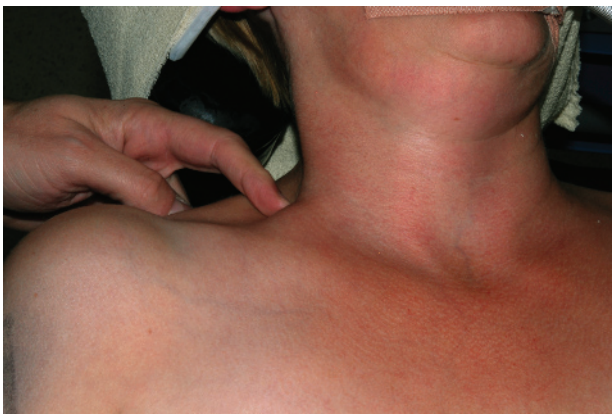
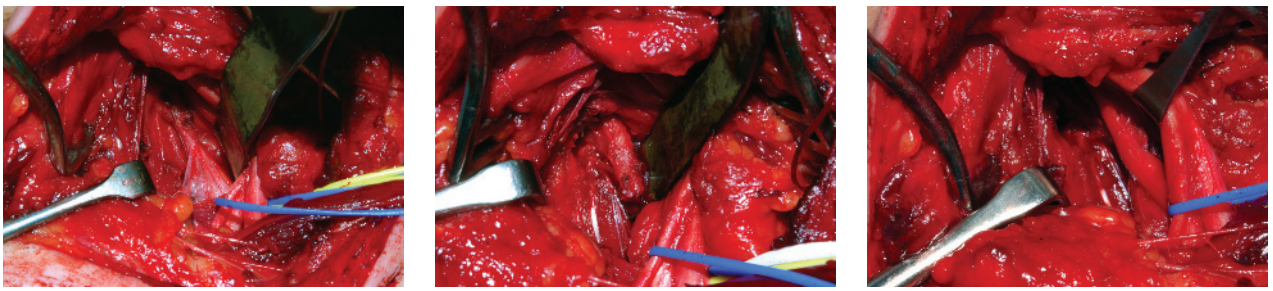


Figure 11.36 Palpation of a large cervical rib

Treatment

Most patients can be managed by *conservative treatment*: exercises to strengthen the shoulder girdle muscles, postural training and instruction in work practices and ways of preventing shoulder droop and muscle fatigue. Analgesics may be needed for pain.

Operative treatment is indicated if pain is severe, if muscle wasting is obvious or if there are vascular disturbances. The thoracic outlet is decompressed by removing the first rib (or the cervical rib) (Figure 11.37). This is accomplished by either a supraclavicular approach or a transaxillary approach; in the latter, care must be taken to prevent injury to the brachial plexus and subclavian vessels, or perforation of the pleura. Patients with arterial obstruction, distal embolism or a local aneurysm will need vascular reconstruction as



(a) (b) (c)

Figure 11.37 Thoracic outlet syndrome (a) Short cervical rib with cervical band. (b) Following excision of cervical band. (c) Following excision of cervical rib.

well as decompression. The transaxillary approach is not recommended where a cervical band is suspected. Our much preferred approach is the supraclavicular. Intraoperative nerve monitoring may be useful where available. Nerve stimulation should be used.

LOWER LIMB COMPRESSION SYNDROMES

Compression of the lateral cutaneous nerve of the thigh

The lateral cutaneous nerve can be compressed as it runs through the inguinal ligament just medial to the anterior superior iliac spine.

The patient complains of numbness, tingling or burning discomfort over the anterolateral aspect of the thigh (*meralgia paraesthetica*). Testing for sensibility to pinprick will reveal a patch of numbness over the upper outer thigh.

If the symptoms are troublesome, the nerve can be released.

Tarsal tunnel syndrome

Pain and sensory disturbance over the plantar surface of the foot may be due to compression of the posterior tibial nerve behind and below the medial malleolus. The pain may be precipitated by prolonged weight-bearing. It is often worse at night and the patient may seek relief by walking around or stamping his or her foot. Paraesthesia and numbness should follow the characteristic sensory distribution, but these symptoms are not as well defined as in other entrapment syndromes. Tinel's percussion test may be positive behind the medial malleolus. The diagnosis is difficult to establish but nerve conduction studies may show slowing of motor or sensory conduction.

Treatment

Tarsal tunnel entrapment may be relieved by fitting a medial arch support that holds the foot in slight varus. If this fails, surgical decompression is indicated. The nerve is exposed behind the medial malleolus and followed into the sole; sometimes it is trapped by the belly of abductor hallucis, arising more proximally than usual. Unfortunately, symptoms are not consistently relieved by this procedure.

Digital nerve compression in the foot

Compression neuropathy of the digital nerve (Morton's metatarsalgia) is dealt with in Chapter 21.

OTHER PERIPHERAL NERVE DISORDERS

Compartment syndromes

Capillary perfusion of a nerve may be markedly reduced by swelling within an osteofascial compartment. Direct trauma, prolonged compression or arterial injury may result in muscle swelling and a critical rise in compartment pressure; if unrelieved, this causes further impedance of blood flow, more prolonged ischaemia and so on into a vicious circle of events ending in necrosis of nerve and muscle. This may occur after proximal arterial injury, soft-tissue bleeding from fractures or operations, circular compression by tight dressings or plasters, and even direct pressure in a comatose person lying on a hard surface. Lesser, self-relieving effects are sometimes produced by muscle swelling due to strenuous exercise. Common sites are the forearm and leg; less common are the foot, upper arm and thigh.

ACUTE COMPARTMENT SYNDROME

Acute compartment syndrome and its late effects (Volkmann's contracture) are described in Chapter 23.

CHRONIC COMPARTMENT SYNDROME

Long-distance runners sometimes develop pain along the anterolateral aspect of the calf, brought on by muscular exertion. Swelling of the anterior calf muscles contained within the inexpansile deep fascia causes ischaemia of the deep peroneal nerve as it traverses the compartment. The condition is diagnosed from the history and can be confirmed by measuring the compartment pressure before and after exercise. Release of the fascia is curative. The same syndrome is very rarely seen in the forearm muscles (known as 'arm pump' in motorcyclists).

IATROPATHIC INJURIES

Positioning the patient for diagnostic or operative procedures needs careful attention so as to avoid compression or traction on nerves at vulnerable sites. The brachial plexus, radial nerve, ulnar nerve and common peroneal nerve are particularly at risk. Recovery may take anything from a few minutes to several months; permanent loss of function is unusual.

During operation an important nerve may be injured by accidental scalpel or diathermy wounds, excessive traction, compression by instruments, snaring by sutures or heating and compression by extruded acrylic cement. Nerves most frequently involved are the spinal accessory or the trunks of the brachial plexus (during

operations in the posterior triangle of the neck), the axillary and musculocutaneous nerves (during operations for recurrent dislocation of the shoulder), the posterior interosseous branch of the radial nerve (during approaches to the proximal end of the radius), the median nerve at the wrist (in tendon surgery), the palmar cutaneous branch of the median nerve (in carpal tunnel release), the cutaneous branch of the radial nerve (when operating for de Quervain's disease), the digital nerves (in operations for Dupuytren's contracture), the sciatic nerve (in hip arthroplasty), the common peroneal nerve (in operations around the knee) and the sural nerve (in operations on the calcaneum).

Tourniquet pressure is an important cause of nerve injury in orthopaedic operations. Damage is due to direct pressure rather than prolonged ischaemia; injury is therefore more likely with very high cuff pressure (it need never be more than 75 mmHg above systolic pressure), a non-pneumatic tourniquet or a very narrow cuff. However, ischaemic damage may occur at 'acceptable' pressures if the tourniquet is left on for more than 2 hours.

Manipulative pressure or traction – for example, during reduction of a fracture or dislocation – may injure a nerve coursing close to the bone or across the joint. Shoulder abduction and varus angulation of the knee under anaesthesia are particularly dangerous. Even moderate pressure or traction can be harmful in patients with peripheral neuropathy; this is always a risk in alcoholics and diabetics.

Injections are occasionally misdirected and delivered into a nerve (usually the radial or sciatic during intramuscular injection, the median nerve during nonoperative treatment of carpal tunnel syndrome or the brachial plexus during axillary blockade). Direct nerve injury at anaesthetic blockade is rare following the advent of routine nerve stimulator use or ultrasound guidance. One should be wary of attributing a postoperative palsy to the anaesthetic block.

Irradiation may cause irreparable nerve damage, a mishap not always avoidable when treating cancer. The effects may not appear until a year or two after exposure.

Diagnosis

Following operations in 'high-risk' areas of the body, local nerve function should always be tested as soon as the patient is awake. Even then it may be difficult to distinguish true weakness or sensory change from the 'normal' postoperative discomfort and unwillingness to move.

Initially it may be impossible to tell whether the lesion is a neurapraxia, axonotmesis or neurotmesis. With closed procedures it is more likely to be a lesser injury, with open ones a greater. If there is no recovery after a few weeks, EMG may be helpful. The demonstration of denervation potentials suggests either axonotmesis or neurotmesis. Surgical exploration at this early stage gives the best chance of a favourable outcome.

Prevention and treatment

Awareness is all. Knowing the situations in which there is a real risk of nerve injury is the best way to prevent the calamity. The operative exposure should be safe and well-rehearsed; important nerves should be given a wide berth or otherwise kept under vision and out of harm's way; retraction should be gentle and intermittent; hidden branches (such as the posterior interosseous nerve in the supinator muscle) should be retracted with their muscular covering. It goes without saying that self-retaining retractors should never be used to retract nerves.

If a nerve is seen to be divided during surgery, it should be repaired immediately; if this cannot be done, the wound can be closed, help can be summoned and the nerve can be re-explored as soon as possible. If the injury is discovered only after the operation, it is best to re-operate as soon as possible, referring the patient to a specialist centre if needed.

If nerve division is thought to be unlikely, then it is wiser to wait for signs that might clarify the diagnosis. If there is marked loss of function and no flicker of recovery by 6 weeks, the nerve should be explored. Even then, fibrosis may make diagnosis difficult; nerve stimulation will show whether there is conduction across the injured segment. Partial lesions or injuries that cause only minor disability are probably best left alone. More serious lesions may need excision and repair or grafting.

LEPROSY

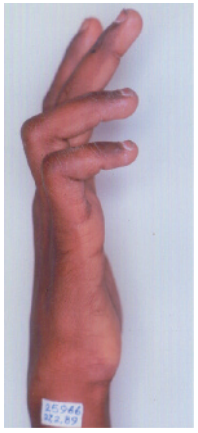
Long-term disabilities in patients with leprosy are due mainly to peripheral nerve abnormalities which result in *loss of sensibility* and *muscle weakness* affecting the hands and feet (see Chapter 2). The former may result in poor wound healing, ulceration and scarring – mainly affecting the hands. The latter may result in deformity and joint instability.

The hand in leprosy

The ulnar nerve is most often affected; combined ulnar and median nerve paralysis is less common, and triple nerve (ulnar, median and radial) paralysis is rare. Any other kind of paralysis is extremely rare. The clinical features associated with these conditions are summarized in Table 11.2 and typical deformities are shown in the accompanying figures (Figures 11.38–11.41).

Table 11.2 Clinical features of paralytic hand deformities in leprosy

Pattern of paralysis	Frequency	Deformity	Consequence	Disability
Isolated (high or low) ulnar nerve paralysis	Most common	Partial claw hand Ulnar palsy thumb Z deformity	Intrinsic muscle deficiency Froment's sign	Poor precision handling Weak grip
Combined ulnar and low median nerve paralysis	Less common	Total claw hand Claw fingers and claw thumb	Intrinsic zero hand All intrinsic muscles paralysed Fingers and thumb activated By long muscles only	Only thumb–index (lateral pinch or 'key grip') and hook grips possible. Power grip and precision handling become difficult or impossible
Ulnar, low median and radial nerve paralysis	Rare	Drop-wrist and dropped digits	'Long-flexor driven' hand All intrinsic muscles and long flexors paralysed	Severe loss of function Cannot grip or hold objects
Combined ulnar and high median nerve paralysis	Very rare	Mild clawing	'Extensor driven' hand All intrinsic muscles and long flexors paralysed	Very severe functional loss Cannot grip
Ulnar, high median and radial nerve paralysis	Very rare	Drop-wrist	Denervated hand All muscles below elbow paralysed	Total loss of function



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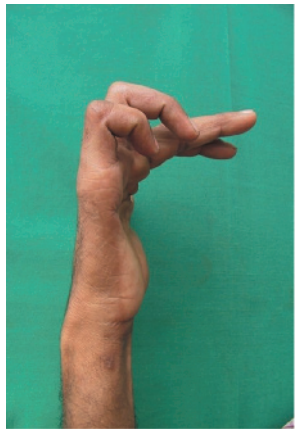
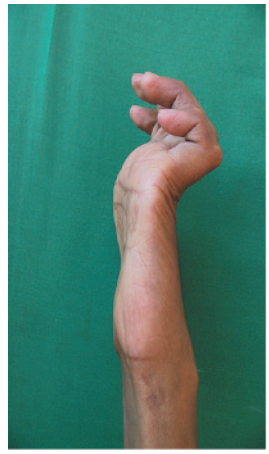


(b)

Figure 11.38 Partial claw hand (a) Partial claw-hand deformity in ulnar nerve paralysis: ring and little fingers are clawed more severely than index and middle fingers. The virtually straight terminal phalanges of the clawed ring and little fingers indicate that flexor digitorum profundus going to these two fingers is paralysed, so this must be a case of 'high' ulnar paralysis. (Courtesy of Dr G. N. Malaviya.) (b) 'Intrinsic minus' disability: isolated PIP extension. Keeping the metacarpophalangeal joints in flexion is not possible. (Courtesy of Dr Santosh Rath.)



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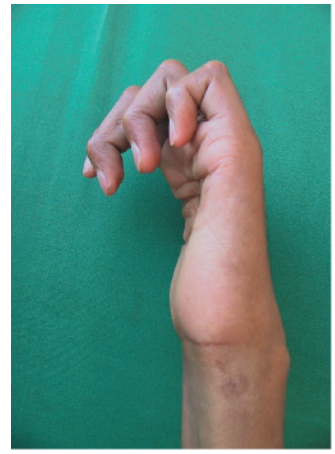


(b)

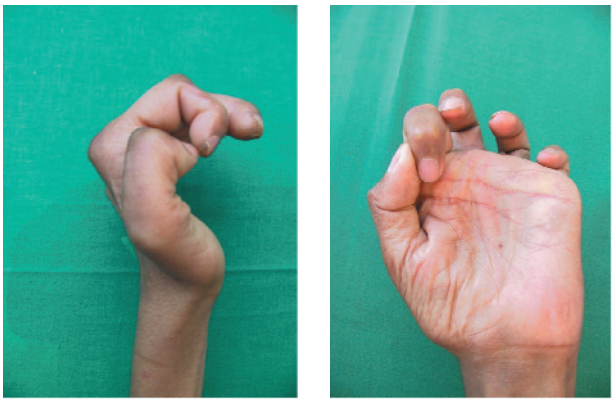
Figure 11.39 Total claw hand (a) Total claw-hand deformity in combined paralysis of ulnar and median nerves. The flexed terminal phalanges of the ring and little fingers indicate that this is a case of 'low' ulnar paralysis, i.e. distal to the elbow beyond the points of origin of the motor branches of flexor digitorum profundus. (b) With intrinsic minus disability, isolated metacarpophalangeal flexion is not possible. (Courtesy of Dr Santosh Rath.)

Correction of claw-finger

This deformity is improved, and the movements lost due to intrinsic muscle paralysis are restored, by rebalancing muscle pull at the metacarpophalangeal (MCP) or proximal interphalangeal (PIP) joint or at both joints. A number of operations have been employed to achieve this end (Table 11.3). The operation currently favoured by most surgeons is the 'lasso operation' of Zancolli in which one tendon of flexor digitorum superficialis (FDS) is split into four slips and one slip each is looped around the A1 pulley of each affected finger so as to provide an independent flexor to MCP joints.

The thumb in ulnar palsy

The severely unstable thumb due to flexor pollicis brevis (FPB) paresis can be corrected by augmenting



(a) (b)

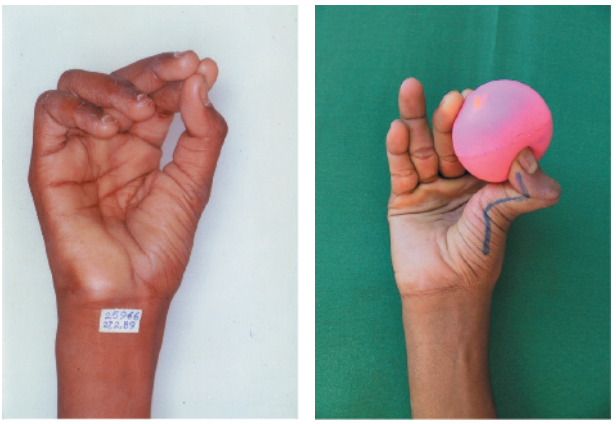
Figure 11.40 Claw thumb (a) 'Claw thumb' (hyperextended at the basal and flexed at the middle and distal joints) in combined ulnar and median nerve paralysis. Note wasting of the thenar eminence. (b) Illustrating pinch in thenar paralysis. Only the lateral or 'key-pinch' is possible for these hands. (Courtesy of Dr Santosh Rath.)

Table 11.3 Strategies and tactics for correction of claw finger

Strategy	Tactic	Procedure
Restore balance at MCP joint	Reduce extending force Increase flexing forces Increase flexor moment arm	Extensor diversion graft operation Capsulodesis Tenodesis Dermodesis Pulley advancement
Restore balance at PIP joint	Reduce flexing force Increase extending force	FDS transfer of Bunnell
Restore balance at both joints	Tendon transfer operations in which the transfer is routed volar to MCP and dorsal to PIP joints	ECRL/ECRB transfers of Brand Palmaris longus transfer of Antia Fowler's digital extensor transfer and other similar procedures

ECRB, extensor carpi radialis brevis; ECRL, extensor carpi radialis longus; FDS, flexor digitorum superficialis; MCP, metacarpophalangeal; PIP, proximal interphalangeal.

flexion at the MCP joint or extension at the interphalangeal joint or both. In one procedure, the radial half of flexor pollicis longus (FPL) tendon is 'dorsalized' by bringing it over the proximal phalanx distal to the MCP joint and fixing it to the extensor pollicis longus tendon, turning FPL into an MCP flexor. Alternatively, FPB can be substituted by transferring the radial half of the index flexor superficialis tendon.



(a) (b)

Figure 11.41 Thumb in ulnar palsy with paralysis of flexor pollicis brevis (a) While at rest the proximal phalanx is de-rotated and lies in line with the metacarpal instead of being flexed by about 25 degrees, and the distal phalanx is flexed by about 15 degrees. (Courtesy of Dr G. N. Malaviya.) (b) Acting against resistance, the thumb collapses into hyperextension at the metacarpophalangeal joint and hyperflexion at the interphalangeal joint (Z deformity). (Courtesy of Dr Santosh Rath.)

The thumb in combined ulnar and median nerve paralysis

Complete paralysis of all thenar muscles (the '*intrinsic-zero*' thumb) results in loss of effective power and precision grip. Correction requires stabilization of the carpometacarpal joint in the 'opponens position' (abduction, flexion and internal rotation) by opponensplasty using flexor superficialis of the middle or ring finger or extensor indicis proprius.

Triple paralysis

Combined loss of ulnar, median and radial nerve function causes very severe disability. The patient has a 'flexor-driven' hand as only the long flexors of the fingers and the wrist flexors are active. Multiple tendon transfers to stabilize the wrist, fingers and thumb in extension are needed; the resulting functionally 'intrinsic-zero' hand is then corrected.

The foot in leprosy

Feet are involved less often than hands but the consequences are more serious. Problems include drop foot, claw toes, plantar ulceration and tarsal disorganization.

Drop foot

'Drop foot' occurs in 1–2% of leprosy patients, because of paralysis of muscles in the anterior and

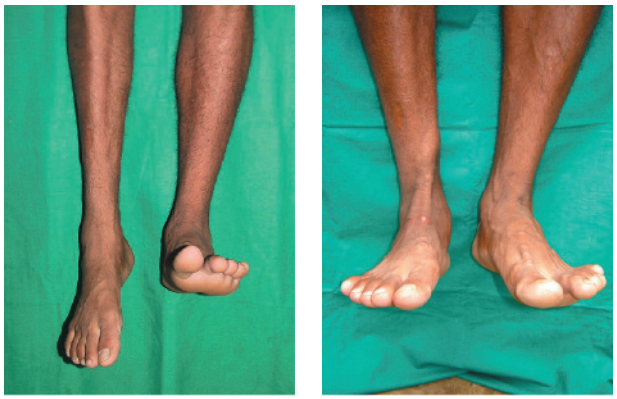
lateral compartments of the leg consequent to damage to the common peroneal nerve. Sometimes only the dorsiflexors or the evertors of the foot are paralysed.

In *dorsiflexor paralysis* the patient has to lift the leg higher than usual during walking for clearing the ground (high-stepping gait). If the condition is neglected, the foot becomes stiff in equinus with intractable forefoot ulceration.

In *evertor paralysis* the foot remains inverted when striking the ground and during the push-off stage of walking. In the course of time, the foot becomes stiff in varus, the weight-bearing lateral part of the foot gets damaged and ulcers develop here. In neglected cases the outer part of the foot is destroyed by repeated ulceration.

A suitable drop-foot orthosis offers a temporary solution, only until corrective surgery is available. The choice of operation depends on whether the deformity is mobile or fixed.

Mobile drop-foot This is corrected by transfer of tibialis posterior tendon, which is almost never paralysed in leprosy. The tendon is rerouted to run in front of the ankle and is fixed in the foot so that the muscle now acts as a dorsiflexor (Figure 11.42b). Skeletal fixation of the transferred tendon is not advised as that might precipitate tarsal disorganization. Circumtibial tibialis posterior tendon transfer to extensor hallucis and extensor digitorum longus tendons over the dorsum of the foot is most commonly done; it is usually combined with tendo calcaneus lengthening. When only the anterior compartment muscles are paralysed, a similar transfer of peroneus longus is done.



(a) (b)

Figure 11.42 Right drop foot (a) Preoperative deformity. The patient is attempting to lift both feet but can do so only on the left side. (b) The same patient 1 year after surgical correction by two-tailed circumtibial transfer of tibialis posterior to extensor hallucis longus and extensor digitorum longus tendons over the dorsum of the foot. (Courtesy of Dr Santosh Rath.)

Fixed drop-foot deformity Fixed equinus or equinovarus usually requires triple arthrodesis of the hindfoot (Lambinudi's operation), which should provide the patient with a plantigrade foot.

Claw toes

This condition, due to plantar intrinsic muscle paralysis, is more common than drop foot. It increases the risk of plantar ulceration greatly. Treatment depends on the severity of the deformity.

First-degree (mild) claw toes There is no joint stiffness but the tips of the toes become ulcerated. The deformity is corrected by transfer of the long flexor to the extensor expansion of each toe.

Second-degree (moderate) claw toes The interphalangeal joints have fixed flexion but the metatarsophalangeal joints remain mobile. PIP arthrodesis, with or without excision of the distal interphalangeal (DIP) joint, is needed.

Third-degree (severe) claw-toes Fixed flexion of the IP joints is associated with dorsal migration of the toes and fixed hyperextension of the metatarsophalangeal (MTP) joints; the metatarsal heads are pushed down towards the sole of the forefoot (plunger effect). Trans-metatarsal amputation is probably the treatment of choice, but patients usually reject this option. A more conservative solution requires open reduction of the MTP joints, proximalization of the long extensor tendons and PIP arthrodesis. Surgical syndactyly helps to fix a 'floating toe'.

Plantar ulceration (trophic ulcers)

Painless chronic ulcers that occur 'spontaneously' are commonly seen in the soles of neurologically compromised feet. They heal with difficulty and recur easily. Loss of sensibility is the main predisposing cause, and the risk of ulceration increases greatly when plantar intrinsic muscles are paralysed or when there is some deformity. Plantar ulcers are colonized by 'street bacteria'; they remain chronic because they are not treated properly.

About 80% of the ulcers are located in the ball of the foot (the majority under the first MTP joint), about 8% in the cubometatarsal joint region, about 10% in the heel, and about 2% over the tips of the toes.

PATHOGENESIS

During walking the body load shifts from the heel to the forefoot and from the lateral to the medial side of the forefoot. In this process the subcutaneous tissues suffer significant compression, shear and stretch, which is normally countered by the intrinsic muscles. These stresses are increased momentarily with each

step when the intrinsic muscles are paralysed. Even slightly increased stresses, if repetitive, eventually lead to tissue damage. A *necrosis blister* develops and that breaks down to form an ulcer.

Injuries occurring in insensitive feet are often neglected because the patient does not experience pain. Wounds fester and develop into ulcers. Even in the absence of any injury, the lack of sweating in the denervated sole predisposes to the development of cracks and fissures and they easily become infected.

NATURAL HISTORY

The natural history of plantar ulcers is a dismal cycle of: ulceration–infection–tissue loss–healing–breakdown of scar–recurrent ulceration–spread of infection with further tissue loss–healing with deformity–more frequent recurrences, and so on until the forefoot is destroyed, tarsal sepsis supervenes and the foot is lost or removed. Sometimes lethal complications (gas gangrene, septicaemia or malignancy) supervene.

MANAGEMENT

A *necrosis blister* should be treated promptly by compression bandaging, rest and elevation for 3 days, followed by a below-knee walking plaster of Paris cast for 3 weeks. If the blister is likely to burst, it is opened under aseptic conditions and dressed before applying the cast.

Simple ulcers present as chronic, shallow lesions. They remain unhealed because they are subjected to the repetitive trauma of walking. A below-knee walking cast, which eliminates the forefoot stage of the walking cycle, is applied and kept on for 6 weeks. Split-thickness skin grafting hastens healing of large simple ulcers. Walking is resumed gradually and only with protective footwear.

Acute infected ulcers require bed rest, elevation of the foot, frequent wet dressings and local irrigation. Systemic antibiotics are used if there are symptoms and signs of general infection. Surgery is limited to drainage procedures.

Complicated ulcers are chronic ulcers associated with additional factors such as infection of deeper structures or deformity. The principles of management are ulcer debridement (which may have to be repeated many times) and protected weight-bearing; deformity correction and stabilizing operations (like arthrodesis) are performed, if needed, after sound healing has been obtained. Sometimes chronic ulcers present as 'cauliflower growths' which commonly turn out to be pseudo-epitheliomas or less commonly epitheliomas of low-grade malignancy. Deep local excision is adequate as treatment and essential for histological confirmation.

Recurrent plantar ulcers occur because the original causes (anaesthesia, muscle paralysis and walking) persist. Additional factors are: poor quality skin, excessive loading of the scar, deep-seated infection and poor blood supply. The risk can be minimized by constant vigilance and attention to hydration of the sole, the use of protective footwear, restricted walking and correction of stress-inducing deformities.

Excessive pressures due to prominent metatarsal heads on the sole of the foot can be treated by: (a) plantar condylectomy and transfer of the long extensor tendons to the metatarsal necks; (b) dorsal displacement metatarsal osteotomies; or (c) excision of an entire ray in the foot.

Intractable ulceration along the lateral border of the foot, due to equinovarus deformity, will need an appropriate triple arthrodesis or a more complicated joint-sparing procedure to render the foot plantigrade.

Heel scars may require plastic surgical flaps combined with 'bumpectomy' to remove bony prominences. Deformities of the calcaneum which produce high-pressure areas should be treated by re-establishing the posterior pillar of the arch of the foot, by doing an appropriately designed calcaneal osteotomy. Sometimes subtotal resection of the calcaneum is needed to get rid of persistent infection; after this type of surgery, the inside of the shoe heel will need to be padded.

OTHER OPERATIONS

In suitably selected cases, posterior tibial neurovascular decompression behind and above the ankle improves the blood supply to the sole and helps heal a recurring or non-healing ulcer.

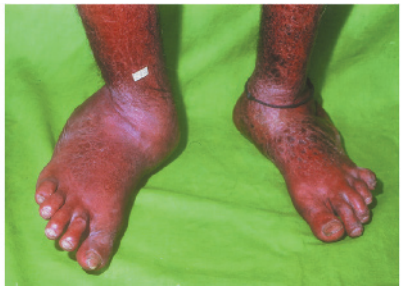
'Flail foot' after loss of the talus is corrected by tibiocalcaneal fusion.

Neuropathic tarsal disorganization

ASEPTIC DISORGANIZATION

Aseptic tarsal disorganization is uncommon. It may follow an inadequately treated fracture of a tarsal bone. In the early stages the patient may have mild pain during walking and on examination there is local swelling, warmth and tenderness. X-rays show the typical features of neuropathic bone necrosis and disorganization (Figure 11.43).

Treatment consists of complete avoidance of all weightbearing and movement, enforced bed rest and application of a total-contact cast that is renewed periodically until the soft-tissue swelling disappears (usually 8–12 weeks), and then for another 4 weeks. If the foot is then found to be stable, a walking cast is applied for a further 4–6 weeks, to be followed by the use of an appropriate orthosis. If the foot is unstable, operative stabilization will be needed.



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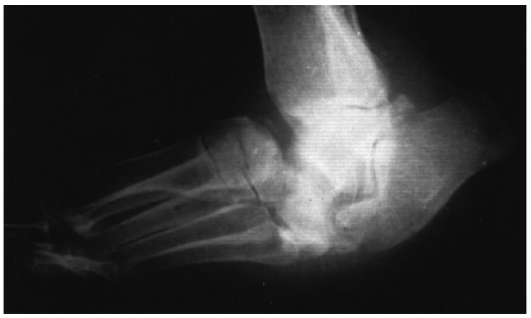


(b)

Figure 11.43 The neuropathic foot (a) Neuropathic tarsal disorganization (right foot). (b) Radiograph of the same foot. There is disruption at the mid-tarsal level with separation of the forefoot from the talus and calcaneum. The talocalcaneal articulation is intact, the talus is plantarflexed and the calcaneum is in equinus. The head of the plantarflexed talus has ploughed through the midfoot and has become directly weight-bearing, as may be seen from the clinical photograph. Because he could feel no pain in the foot, this patient was able to walk on the foot despite the severe damage. (Courtesy of Dr G. N. Malaviya.)

SEPTIC TARSAL DISORGANIZATION

Infection may spread from a plantar ulcer to underlying tarsal bones and joints and destroy these structures. Once the infection is controlled, the foot is immobilized in a below-knee cast; the involved bones fuse together and a stable, rigid foot results. An unstable foot will need surgical stabilization after clearing the infection.

Amputations

Occasionally amputation is necessary to keep the patient ambulatory. However, this step should not be taken without careful consideration; amputation merely shifts the problem to a more proximal level where it will be even more difficult to manage because the stump is often insensitive in these patients. Moreover, facilities for prostheses are scarce in many of the areas where leprosy is endemic, and even where they are available, hand deformities or poor vision in affected persons make their use difficult. The guiding principles are: amputate only if you must, amputate conservatively and try to provide an end-bearing stump where possible.

FURTHER READING

MRC Memorandum *Aids to the Examination of the Peripheral Nervous System*, MRC Memorandum 45. London: HM Stationery Office, 1976.

Birch R. *Surgical Disorders of the Peripheral Nerves*, 2nd edn. New York: Springer, 2011.

Brandsma W, Schwarz R (eds). *Surgical Reconstruction & Rehabilitation in Leprosy and Other Neuropathies.* Kathmandu (Nepal): Ekta Books, 2004.

Orthopaedic operations

12

Michael Whitehouse, David Warwick & Ashley Blom

The art and skill of orthopaedic surgery is directed not simply to reshaping or constructing a particular arrangement of parts but to restoring function to the whole.

In this chapter, principles applying to orthopaedic operations will be discussed and fundamental techniques of soft-tissue and bone repair will be described. For detailed descriptions of the various operative procedures the reader is referred to standard textbooks on operative orthopaedic surgery and monographs dealing with specific regional subjects.

PREPARATION

Planning

Operations upon the musculoskeletal system must be carefully planned in advance, when accurate measurements can be made, bones can be compared for symmetry with those on the other side of the axis or with those of the opposite limb. This stage allows the surgeon to plan for appropriate equipment, expertise and support to be available. X-rays, ultrasound, magnetic resonance imaging (MRI) and computed tomography (CT) (if necessary with three-dimensional reformatting) can be helpful; transparent templates or digital templating software may be needed to help size and select the most appropriate implant.

Corrective osteotomies and implant positioning can be simulated on X-ray, paper cut-outs or templating software before the operation is undertaken (Figure 12.1). Before new or complex reconstructive operations are undertaken they should, ideally, be rehearsed using artificial bones and joints at a workbench. Where available, 3D printing of models and mock implants can aid the surgeon preoperatively and in theatre.

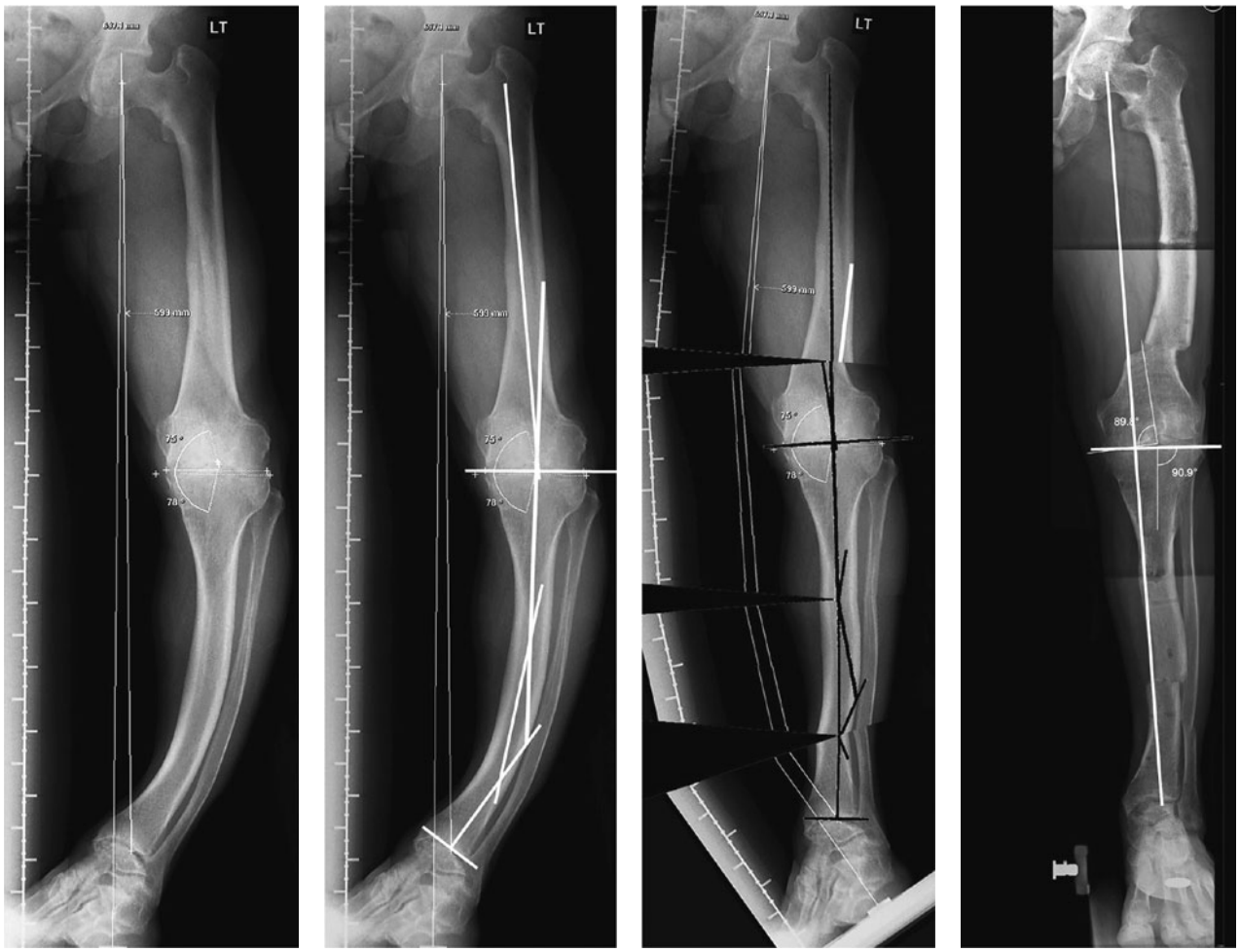
Equipment

The minimum requirements for orthopaedic operations are drills (for boring holes), osteotomes (for cutting cancellous bone), saws (for cutting cortical bone), chisels (for shaping bone), gouges (for removing bone) and plates, screws and screwdrivers (for fixing bone).

Many operations such as joint replacement, spinal fusion and the various types of internal fixation require more specialized implants and instruments to ensure that the bone and implants are correctly aligned and fixed. Surgeons should be familiar with the techniques and implants they plan to use, their advantages and disadvantages and the pitfalls encountered in their use. Most importantly, the surgeon is responsible for ensuring that the necessary instruments and implants (in appropriate sizes!) are available before the anaesthetic commences.

INTRAOPERATIVE RADIOGRAPHY

Intraoperative radiography is often helpful and sometimes essential. Fracture reduction, osteotomy alignments and the positioning of implants and fixation devices can be checked before completing the procedure. Fluoroscopy, where available, is quick and easy to use. Although the resolution is not as high as standard radiographs, sufficient detail is normally visible for intraoperative decisions. Smaller low-emission surgeon-operated devices are particularly useful in extremity surgery. Formal radiographs or CT scans may still be required postoperatively to confirm adequacy of reduction or implant positioning. Where these are not available radiograph cassettes may be used but must be wrapped in sterile drapes and introduced to the operative field. Angiography may be needed to diagnose a vascular injury or demonstrate the success of a vascular repair.



(a) (b) (c) (d)

Figure 12.1 Preoperative planning on digitized X-ray images The computer software allows the deformity to be analysed (a,b) and the correction simulated (c). The end result then mimics the simulation (d).

Imaging guidance systems

By using a navigation system based on techniques such as optical tracking, implanted markers and intraoperative radiography with suitable computer software, surgeons may be able to improve their accuracy and consistency in placing implants correctly. Examples are insertion of screws into vertebral pedicles and positioning of joint replacement components.

Robotically assisted surgery

In an attempt to improve the accuracy and reproducibility of certain surgical procedures, such as joint replacement, robotically assisted surgery can be considered. There are a variety of different ways in which surgeons can interact with robots including haptic feedback, force constraint and telemanipulation. The use of robotics in orthopaedics remains relatively rare and is limited by the associated costs.

Radiation exposure

Intraoperative radiography involves the risks to both patient and members of the theatre team of radiation exposure. The recommended dose limit for the general public is 1 mSv per year, which is the equivalent of 1000 chest X-rays. Each chest X-ray in turn produces the same radiation dose as is endured during a 4-hour airline flight. Fluoroscopic images acquired during operations are usually pulsed exposures rather than continuous screening, thus reducing total exposure and amounting to a negligible additional risk of developing cancer. However, for the surgeon the risk is far greater because of the repeated use of fluoroscopy.

Total exposure varies with the type of procedure performed (operations on limb extremities produce the least, hip operations, midline and spine operations the most), the number of procedures needing X-ray assistance and the protective measures used. The latter influence the cumulative exposure significantly and lead aprons are therefore compulsory;

further attenuation of radiation exposure is gained through the use of thyroid shields and, if practical, eye goggles. Using a hip procedure as an example, lead aprons will reduce the effective dose received by a factor of 16 for anteroposterior projections and by a factor of 4–10 for lateral projections. Using a thyroid shield decreases the dose by another 2.5 times.

Magnification

Magnification is an integral part of peripheral nerve and hand surgery. The improved view minimizes the risk of inadvertent injury to structures and allows more accurate apposition of tissues during reconstruction.

Operating loupes range in power from 2× to 6× magnification. As the magnification increases, the field of view decreases and the interruption by unwanted head movements becomes more apparent. Most surgeons, therefore, choose between 2.5× and 3.5× magnification.

The operating microscope allows much greater magnification with a stable field of view. It is particularly important when very accurate apposition of tissue is required, for example when aligning nerve fascicles during nerve repair or nerve grafting, when anastomosing small vessels or when operating in a narrow corridor of safety such as in microdiscectomy of the spine.

THE 'BLOODLESS FIELD'

Many operations on limbs (and particularly the hand) can be done more rapidly and accurately if bleeding is prevented and a bloodless field is created. This is usually achieved by the application of a tourniquet.

Wide awake local anaesthetic no tourniquet (WALANT)

A bloodless field can be achieved by slowly infiltrating large volumes of very dilute lidocaine and adrenaline (epinephrine). Traditional concerns about the risk of using adrenaline in the extremities have been overestimated so long as the dose is low enough. The advantage of WALANT is that there is no paralysis from the tourniquet. This allows the surgeon to assess the effect of a tenolysis, the tension of a tendon transfer, the rotation of a fixed finger fracture throughout range of movement, and the alignment and offset of a small joint replacement.

Tourniquet cuff

Only a pneumatic cuff should be used and it should be at least as wide as the diameter of the limb. Wide cuffs reduce the pressure needed for vascular occlusion. Tied rubber bandages are potentially dangerous and should not be used; the pressure beneath the bandage cannot be controlled and there is a real risk of damage to the underlying nerves and muscle. A protective layer should be applied to the skin prior to placement of the tourniquet. Specifically manufactured devices are now available, but a layer of wool bandage can be used to distribute the tourniquet pressure and cushion underlying skin. During skin preparation, it is essential that the sterilizing fluid does not leak beneath the cuff as this can cause a chemical burn. Isolating the tourniquet with a plastic drape can help prevent this complication.

Exsanguination

Elevation of the lower limb at 60 degrees for 30 seconds will reduce the blood volume by 45%; increasing the elevation time does not alter the percentage significantly. This simple manoeuvre will therefore suffice to 'drain' the tissues if a truly bloodless field is not essential, or when surgery is being undertaken for tumour or infection and forceful exsanguination might squeeze pathological tissue into the proximal part of the limb. The 'squeeze' method, in which pressure on the palm or foot is followed by sequential squeezing of the limb in a proximal direction, is also effective. If a clearer field is required, exsanguination can be achieved by pressure using a rubber tubular exsanguinator prior to skin preparation, or if tourniquet time is to be kept to a minimum, a sterile Esmarch or gauze bandage wrapped from distal to proximal. These methods reduce blood volume by an additional 20%.

Tourniquet pressure

A tourniquet pressure of 150 mmHg above systolic is recommended for the lower limb and 80–100 mmHg above systolic for the upper limb. This may need to be increased in hypertensive, obese or very muscular patients. Higher pressures are unnecessary and will increase the risk of damage to underlying muscles and nerves.

Tourniquet time

Tourniquet time is ischaemia time and thus an absolute maximum tourniquet time of 2.5 hours is allowed, although it is safer (and more advisable) to keep this under 2 hours; transient nerve-related symptoms may occur with 3-hour tourniquet times, but full recovery is usual by the fifth day. Time can be saved by ensuring that the limb is shaved, prepared, draped and marked before inflating the cuff. The time of application of the tourniquet should be recorded

and the surgeon should be informed of the elapsed time at regular intervals, particularly as the 2-hour period is approached.

Deflating and re-inflating the tourniquet

This has serious local and systemic effects. Locally deflation is followed by a hyperaemic response that reduces by half in 5, 12 and 25 minutes, respectively after ischaemic times of 1, 2 and 3 hours. This information is useful to the surgeon trying to obtain haemostasis after tourniquet release. There is also a variable amount of swelling, unrelated to the length of the ischaemic period; it is therefore wise not to use a tourniquet when it is not required to perform the procedure safely and for those limbs where significant swelling is already evident. At the systemic level, tourniquet deflation induces a free radical-mediated reperfusion syndrome, which adds to any muscle damage already produced by the ischaemic period. 'Breathing periods' (deflation followed after a pause by re-inflation), which were once popular to enable extended tourniquet times, are no longer recommended as the reperfusion effects are cumulative even though the local limb anoxia is relieved at each tourniquet deflation. If a prolonged tourniquet time is required and unavoidable, it is wise to warn the patient of the possibility of transient nerve-related symptoms and to obtain consent to use the absolute maximum period of 3 hours. WALANT should be considered when suitable.

Finger tourniquet

This is suitable for relatively minor hand operations. A sterile rubber glove-finger makes a good cuff; the tip is cut and the margin is then rolled back proximally. This has the combined effect of exsanguinating the finger and acting as a tourniquet. A rubber glove can be obscured by blood and be left on inadvertently; brightly coloured devices with a long tag are available commercially and indeed in some health services a glove is no longer allowed. A stretched rubber catheter must not be used as this may damage the underlying structures. *Always check that the finger tourniquet has been removed at the end of the operation.*

Complications

Complications of tourniquet usage usually relate to nerve injury (more often due to compression than duration of ischaemia), skin burns from leakage of alcoholic antiseptic solutions beneath the tourniquet cuff and a failure to diagnose peripheral vascular disease before surgery. The risk of these can be minimized by always using a wide cuff, sealing the cuff against seeping fluids and avoiding excessive tourniquet pressures. A wise precaution is to not employ a bloodless field at all in patients with impaired peripheral circulation or those with arterial prostheses or stents that may not expand sufficiently after tourniquet deflation to re-establish an adequate blood flow.

MEASURES TO REDUCE RISK OF INFECTION

Skin preparation and draping

Hair removal

Shaving the limb is more likely to be harmful than helpful. Shaving before surgery causes superficial skin damage and leads to local bacterial proliferation. Depilatory creams and hair clippers do not lead to this and are safe to use. If depilatory creams are used, they are typically applied the day before surgery.

Skin cleaning

A 'social wash' with a soap solution removes particulate matter and grease. This is particularly useful in visibly contaminated cases, managing open fractures and in cases where the limb has been wrapped in a cast or splint for some time. Skin preparation prior to surgery should be carried out with an alcohol-based preparation where safe; alcohol is not to be applied over open wounds, exposed joints or nerve tissue. Iodine or chlorhexidine preparations are available, but there is evidence that chlorhexidine is more effective after a single application, having longer residual activity and maintaining efficacy in the presence of blood and serum. A staining fluid will help to ensure that the limb is fully covered. However, use of a red staining fluid should be avoided if a tourniquet is used since it may make it difficult to determine whether blood flow has returned after releasing the tourniquet.

Drapes

Drapes function to isolate the surgical field from the rest of the patient and reduce contamination from outside. Disposable drapes have been shown to be superior at preventing passage of bacteria and strike through of fluids. They should have the following qualities:

- barrier effectiveness throughout the length of the procedure
- configurability to cover the appropriate areas of body or limb
- tear-resistance
- non-allergenicity
- reasonable cost.

Plastic adhesive coverings, some impregnated with iodine, function primarily to secure the drapes,

especially if the limb is moved during surgery. Iodine-impregnated adhesive drapes decrease bacterial counts at the incision site but there is no strong evidence they decrease infection rates. Modern drapes tend to come in packs designed for single use and designed to expose certain areas of the body or for specific operations.

Surgical attire

Gowns

Gowns need to share the requisite qualities of drapes but should also be comfortable to wear. An anterior panel with increased resistance to fluid penetration is an advantage.

Gloves

Gloves are available in latex and non-latex varieties. The latter are needed if either the surgeon or the patient has a latex hypersensitivity. This could apply to patients who are constantly exposed to latex devices such as urinary catheters. Latex allergy is second only to muscle relaxants for inducing anaphylaxis during surgery. Double gloving, with a coloured inner glove (so-called 'indicator glove') reduces the number of inner-glove perforations and allows outer-glove perforations to be picked up more quickly. Reassuringly, studies have shown that very small penetrations to inner gloves do not lead to significant passage of bacteria.

Face mask

This hallmark of the surgeon in theatre has been questioned in its ability to reduce surgical site infections. Certainly the mouth is a potent source of bacterial contamination of theatre air. As studies provide conflicting views as to whether masks reduce infection rates rather than just decrease bacterial counts in the theatre air, for the time being at least, face masks should continue to be used if only for protection of the surgical staff. Modern face masks incorporate visors (eye shields), which substantially reduce the risk of contact with blood.

Glasses and eye protection

For procedures in which blood and bone may be splattered, or if the patient carries a blood-borne virus, protective eye wear should be worn.

Vaccination

There is a risk of transmission of blood-borne infections to orthopaedic surgeons, not least because of the nature of surgery but also due to frequent handling of instruments and bone fragments with sharp edges (exposure prone procedures). Transfer of infectious agents through blood occurs mainly by contact (percutaneous or mucocutaneous) and through aerosols. The face and neck may become contaminated and this may go unnoticed until after the procedure; splashes and aerosol sprays often happen during the use of power tools and irrigation fluids. Exposure is more likely if the operation continues for over 3 hours or when blood loss is greater than 300 mL. A barrier created by a surgeon's attire must be coupled to the correct etiquette for handling and passing instruments between staff. This reduces the likelihood of accidental needle-stick injury but is augmented by prophylaxis through vaccination. Sharp tools should not be passed from hand to hand, but via a receptacle.

Hepatitis B

Transmission may occur through inoculation or even from contact with a contaminated surface (the virus is able to survive for a week in dried blood). There is a 30% risk of transmission from a single inoculation of an unvaccinated person. Vaccination is safe and effective, and immunity, for those who respond after a course of injections, is indefinite. Those who do not respond to immunization and are exposed will need post-exposure prophylaxis using a combination of hepatitis B immunoglobulin and the vaccine.

Hepatitis C

The risk of accidental transmission is lower for hepatitis C than for hepatitis B (less than 7%). Unfortunately, no effective vaccine or post-exposure protection is available.

Human immunodeficiency virus

The risk of human immunodeficiency virus (HIV) infection after accidental injury is very low (less than 0.5%) although this may vary between individuals. Vaccination is not available but post-exposure treatment with antivirals is essential and effective.

THROMBOPROPHYLAXIS

Venous thromboembolism (VTE) is one of the commonest complications of lower limb surgery. It comprises three associated disorders: *deep vein thrombosis (DVT)*, *pulmonary embolism (PE)* and the later complication of *chronic venous insufficiency* in some cases. Approximately one in 30–40 patients operated on for hip fractures or hip and knee replacements will develop a symptomatic thromboembolic complication despite the use of prophylaxis during their hospital stay. The most important risk factors are increasing

age, obesity, prolonged immobility, malignancy and in particular a personal or family history of previous thrombosis.

Pathophysiology

According to Virchow, thrombosis results from an interaction between vessel wall damage, alterations in blood components and venous stasis. All of these occur in major orthopaedic operations. The surgery is highly thrombogenic. Soft-tissue exposure, bone cutting and reaming induce a systemic hypercoagulable state and fibrinolytic inhibition. Blood flow in the femoral vein is obstructed by the torsion needed to expose the femoral canal and the acetabulum in hip replacements; this damages the endothelium, both in the proximal femoral vein (by torsion) and in the distal veins (by distension). Furthermore, venous obstruction allows a concentration of clotting factors. In knee replacement, the anterior subluxation of the tibia and vibration from the saw may cause local endothelial damage. In addition, the relative immobility that follows lower-limb operations induces some degree of venous stasis. DVT occurs most frequently in the veins of the calf and less often in the proximal veins of the thigh and pelvis. It is from the larger and more proximal thrombi that fragments sometimes get carried to the lungs, where they may give rise to symptomatic pulmonary embolism (PE) and, in a small percentage of cases, fatal pulmonary embolism (FPE).

Clinical features and diagnosis

Thromboembolic events can be represented as a pyramid (Figure 12.2); most of these events are asymptomatic but a proportion cause symptoms. Hence DVT is, in the main, an occult disease and considerably more common than the symptoms and signs suggest.

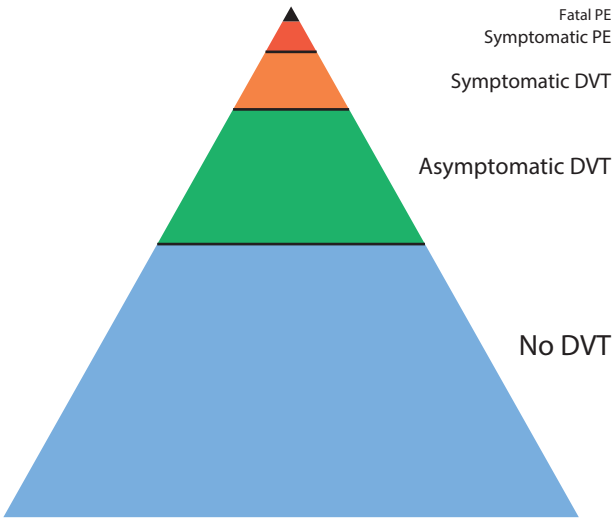


Figure 12.2 The thromboembolism pyramid

Deep venous thrombosis

DVT is usually asymptomatic, although some patients present with pain in the calf or thigh. An increase in temperature and pulse rate may develop. There are usually no signs but there may be calf swelling and tenderness. Homans' test (increased pain on passive dorsiflexion of the foot), although still frequently employed, is now regarded as unreliable. The signs of DVT may be confused with, or obscured by, the normal swelling and tenderness that can affect the calf after a knee or shin procedure.

Pulmonary embolism (PE)

Patients may develop pleuritic pain in the chest and shortness of breath, but other conditions, such as myocardial infarction or fulminant pneumonia, can be mistaken for PE. In most cases PE is asymptomatic and fatal PE usually presents as a sudden collapse without any prior symptoms in the legs or chest; in such cases the diagnosis is confirmed by post-mortem examination (Figure 12.3).

Imaging studies help to confirm the diagnosis in patients who have a moderate or high clinical probability of thromboembolism. Ultrasound or venography is important for demonstrating DVT and computerized tomographic pulmonary angiography or ventilation–perfusion (VQ) scans are helpful in the diagnosis of PE.

Post-thrombotic syndrome

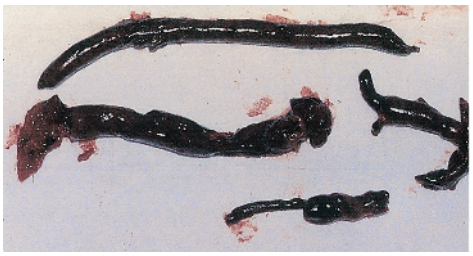
Post-thrombotic syndrome (PTS) presents with leg discomfort, swelling, skin changes and even ulceration. This is a debilitating condition that directly influences quality of life. Approximately one-third of patients with *symptomatic* DVT will go on to develop features of PTS within 2 years but it is not yet established whether the much more frequent *asymptomatic* DVT after joint replacement predisposes to this long-term outcome.

Chronic pulmonary hypertension

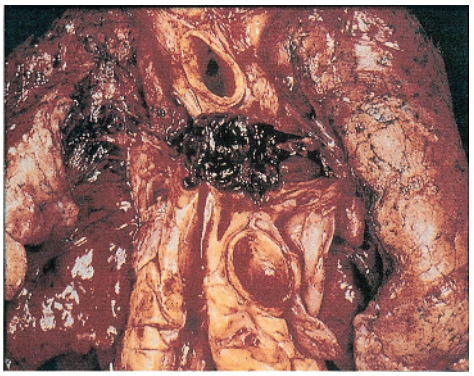
This is a potential sequel for those who survive a symptomatic PE but the incidence is unknown.

Incidence of thromboembolic events

The incidence of symptomatic post-surgical thromboembolism and fatal PE is decreasing with time, due to more efficient surgery as well as earlier mobilization and the widespread use of prophylaxis and regional anaesthesia. In particular, the risk of death from pulmonary embolus following common procedures such as hip and knee replacement is now very low.



(a)



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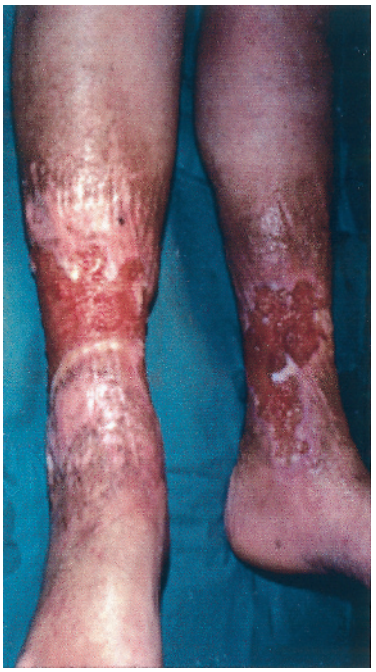

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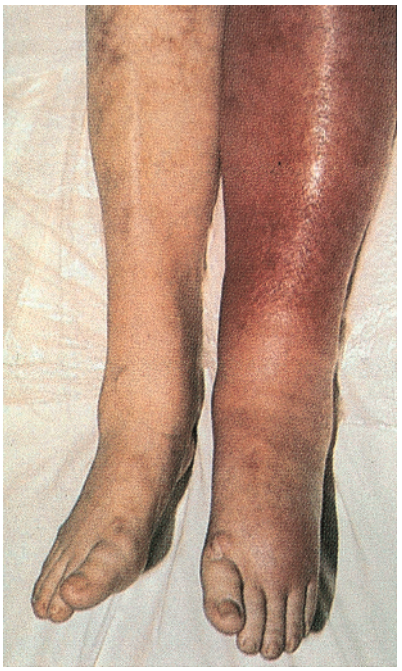


(d)

Figure 12.3 Venous thromboembolism (a) Venous thrombosis – embolism from the deep veins of the leg, extracted from the lung at post mortem. (b) Fatal pulmonary embolism at post mortem. (c) Chronic venous insufficiency. (d) Acute thrombophlebitis.

Much of the information used to calculate risk reduction with prophylaxis is derived from screening studies using ultrasound or venographic assessment. Venograms and to a lesser extent ultrasound scans are sensitive and specific in identifying venous thrombi but the relationship between 'screened DVT' and symptomatic events has not been fully defined. Many asymptomatic venographic thrombi resolve without untoward effects. However, it is reasonable to assume (and there is some evidence) that a reduction in venographic DVT does produce a proportionate reduction in symptomatic DVT or even fatal PE, thus shrinking the pyramid of risk (Table 12.1).

Prevention

The overall risk of DVT and PE can be reduced by prophylaxis. Patients admitted for surgery, whether electively or in emergency, need an individualized risk assessment, which can be simplified by including an active reminder or checklist prior to surgery. This ensures that safe, effective prophylaxis is routinely given according to a protocol that has been accepted by the surgeons and anaesthetists.

Table 12.1 Risk of venous thromboembolism (VTE)

Procedure or condition	Fatal PE	Symptomatic VTE	Asymptomatic DVT
Hip fracture	1%	4%	60%
Hip replacement	0.2–0.4%	3–4%	55%
Knee replacement	0.2%	3–4%	60%
Isolated lower limb trauma	Unknown	0.4–2%	10–35%
Spinal surgery	Unknown	0.4–2%	10–35%
Knee arthroscopy	Unknown	0.2%	7%
Major trauma	Unknown	Unknown	58%
Spinal cord injury	Unknown	13%	35%
Upper limb surgery	Unknown	Very rare	Very rare
Minor lower limb surgery	Very rare	Very rare	Very rare

DVT, deep vein thrombosis; PE, pulmonary embolism; VTE, venous thromboembolism. (Derived from the International Consensus Statement 2013 and ACCP Guidelines 2015.)

General measures

- *Neuraxial anaesthesia* – Spinal or epidural anaesthesia reduces mortality, enhances peri-operative analgesia and reduces the risk of VTE by about 50%. It is wise to avoid giving neuraxial anaesthesia and chemical prophylaxis too close together to avoid a spinal haematoma. Local guidelines should be followed.
- *Surgical technique* – Rough surgical technique will potentiate thromboplastin release. Prolonged torsion of a major vein, when maintaining a dislocated hip for purposes of replacement or during aggressive dorsal retraction of the tibia during knee replacement, inhibits venous return and damages the endothelium.
- *Tourniquet* – A tourniquet probably does not change the risk; clotting factors that accumulate while the tourniquet is inflated are flushed out by the hyperaemia on tourniquet deflation.
- *Early mobilization* – This is a simple physiological means of improving venous flow.

Physical methods

- *Graduated compression stockings* can halve the incidence of DVT when they are correctly sized and fitted compared to no prophylaxis; there is a suggestion that below-knee stockings may be just as effective as above-knee types, as long as the stockings are properly woven and well-fitted.
- *Intermittent plantar venous compression* takes advantage of the fact that blood from the sole of the foot is normally expressed during weight-bearing by intermittent pressure on the venous plexus around the lateral plantar arteries; this, in turn, increases venous blood flow in the leg. A mechanical foot pump can reproduce this physiological mechanism in patients who are confined to bed. It should not be used in combination with compression stockings as these impair refill of the venous plexus after emptying by the foot pump. There is some evidence that this technique provides effective thromboprophylaxis in hip fracture, hip arthroplasty and knee arthroplasty, especially if combined with a chemical method.
- *Intermittent pneumatic compression of the leg* has also been shown to reduce the risk of 'radiological DVT' after hip replacements and in trauma. It is, however, impractical for patients undergoing operations at or below the knee. Portable intermittent pneumatic compression devices are relatively expensive but have been popularized in North America as part of rapid discharge protocols.
- *Inferior vena cava filters* resemble an umbrella and are percutaneously passed through the femoral vein and lodged in the inferior vena cava. They merely catch an embolus to prevent it from reaching the lungs. They have a specific role in the occasional case where the risk of embolism is high yet anticoagulation is contra-indicated, for example in a patient with a pelvic fracture who has already developed a DVT but needs major surgical reconstruction. The complication rate, which includes death from proximal coagulation, should restrict use of these devices.
- *Electrical stimulation* with a small device over the peroneal nerve at the knee may enhance blood flow.

Chemical methods

These are generally safe, effective, easy to administer (tablet or injection) and can be used for extended periods. They are relatively inexpensive compared with the overall cost of surgery. However, all chemical methods incur a risk of bleeding, which is a natural concern for both the orthopaedic surgeon and the anaesthetist. Methods include the following:

- *Aspirin* – Some recent guidelines (American Academy of Orthopaedic Surgeons, Australian Orthopaedic Association, American College of Chest Physicians) recommend aspirin in conjunction with mechanical methods for joint surgery with average risk of VTE. Other guidelines (e.g. NICE in the United Kingdom and the International Consensus Statement) advise against its use.
- *Unfractionated heparin* – This carries a risk of increased bleeding after operation and is contraindicated in elderly people.
- *Low molecular weight heparin (LMWH)* – This class of drug has haematological and pharmacokinetic advantages over unfractionated heparin including ready bio-availability and a wide window of safety; therefore monitoring is not required. It is safe if used properly (with an adequate time between administration and surgery or regional anaesthesia, and a reduced dose for those with impaired renal function). LMWH is more effective than placebo or unfractionated heparin and at least as effective as warfarin, compression devices and foot pumps. Randomized studies have shown that it effectively reduces the prevalence of venographic DVT in hip and knee replacement surgery, and the effect is probably amplified when coupled with physical methods.
- *Pentasaccharide* – This synthetic injectable antithrombotic drug (fondaparinux) precisely inhibits activated Factor X. It is at least as effective as LMWH but must not be given too close to surgery (it is best given 6–8 hours after surgery) or bleeding may become a significant problem. The drug is excreted by the kidneys rather than metabolized by the liver and so must be used carefully or avoided in those with poor renal function.
- *Direct anti-Xa inhibitors* (e.g. rivaroxaban) *and direct thrombin inhibitors* (e.g. dabigatran) – These drugs are given orally and have a broad therapeutic

and safety window (so that no monitoring is required). They are given after surgery and should be continued for as long as the patient is at risk of VTE. They have efficacy equivalent with LMWH in hip and knee replacement surgery. They provide a pragmatic solution for after-hospital prophylaxis, requiring neither injections nor complex monitoring. Drug activity is difficult to reverse.

- *Warfarin* – Warfarin has been used fairly widely, particularly in North America. It reduces the prevalence of DVT after hip and knee replacement and FPE is extremely rare. Drawbacks are the difficulty in establishing appropriate dosage levels and the need for constant monitoring. If it is used at all, it must be maintained at an international normalized ratio (INR) level of 2–3.

Timing and duration of prophylaxis

Risk factors for thromboembolism are most pronounced during surgery, but in some patients (particularly those with hip or major long-bone fractures of the lower limb), immobility and a hypercoagulable state may begin before the operation. In general, prophylaxis is given on admission to hospital in this group, particularly if surgery will be delayed beyond 24 hours. Chemical prophylaxis should not be given too close to surgery because there is a risk of provoking a bleeding complication. If it is given too long before surgery, metabolism or excretion may reduce its potency; if given too long after surgery, the thrombogenic process will be established and the drug is now therapeutic instead of prophylactic.

The ideal duration of thromboprophylaxis is not known and depends on many factors, including individual patient factors, which are difficult to quantify. Without prophylaxis, VTE would often, if not usually, occur beyond hospital discharge; robust studies have shown that the risk of after-discharge symptomatic DVT can be reduced by two-thirds by prolonging thromboprophylaxis. Traditional recommendations suggesting that it should be continued until the patient is fully mobile have been superseded by recommendations that prophylaxis is continued for between 14 and 35 days following knee replacement and 35 days following hip replacement.

The duration of risk in other orthopaedic conditions is not known. While injectible chemical methods may be appropriate, oral agents that do not require monitoring (e.g. aspirin, anti-Xa and anti-thrombin inhibitors) facilitate effective and practical extended duration prophylaxis.

Multimodal prophylaxis

Risk assessment of patients may determine that a combination of physical and chemical prophylaxis is needed. This form of *multimodal prophylaxis* is gaining popularity and some studies point to increased efficacy. For patients at particularly high risk of bleeding, the mechanical method should be used until the bleeding risk has diminished such that the chemical agent can be introduced and which is continued for as long as there is a risk of thrombosis. For patients with a particularly high risk of thrombosis, the mechanical device is started preoperatively or immediately after surgery and continued for as long as tolerated; the chemical is started as close to surgery as is safe (e.g. 6 hours postoperatively) and continued for as long as the risk of VTE persists.

OPERATIONS ON BONES

OSTEOTOMY

Osteotomy may be used to correct deformity, to change the shape of the bone, or to redirect load trajectories in a limb so as to influence joint function. Preoperative planning is essential.

Knowledge of the limb axes and their relation to the joints is the foundation for analysing skeletal deformity. 'Corrective' surgery is an exercise in balancing the extent of operative interventions needed to produce *anatomical 'normality'* with the *anticipated gain in function*. 'Anatomical' correction, while desirable in most cases, is not always necessary. An appropriate example is a skeletal deformity due to a neuromuscular disorder where correction to achieve maximal *functional* gain has to be considered to be a greater priority than *anatomical* accuracy.

Modern deformity analysis recognizes the three-dimensional basis of most deformities, whether the origin of the problem is within a bone or a joint or a combination of both. Deformity of bone exists as a deviation in the coronal or sagittal plane (or any plane in between) where it can be measured in degrees of angulation or millimetres of translation, or in the axial plane, where it exists as degrees of rotation or millimetres of length abnormality.

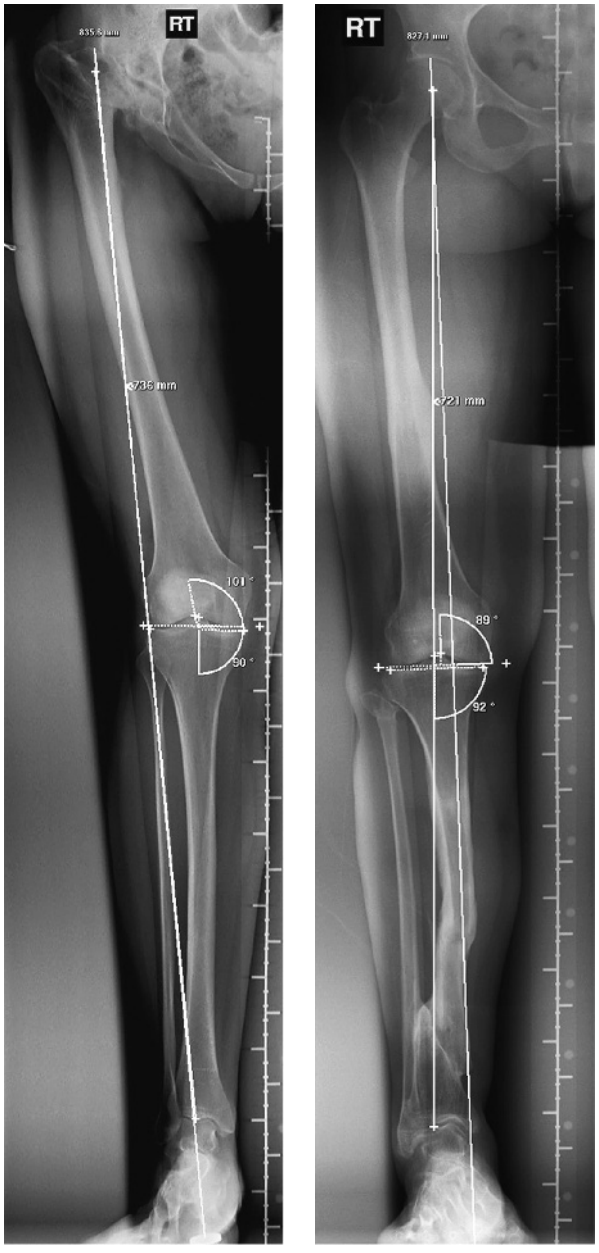
The use of 3D printing from CT data now allows very accurate planning, especially if scans of the normal limb are reversed to match the deformed limb as a template. 3D printing of customized fixation plates will enhance accuracy even further.

In the section below, the lower limb is used to illustrate the principles as applied to the coronal plane.

Limb axes and reference angles

The mechanical axis of a limb is defined by an imaginary line connecting the centre of the most proximal major joint to the centre of the most distal, for instance in the lower limb from the centre of the hip to the centre of the ankle. In most individuals this line passes close to the centre of the knee joint, usually

8(±7) mm medial to it. If a deformity is present, the line may be displaced away from its usual position (Figure 12.4a). Interestingly, if a deformity exists at two or more levels in the limb, the resulting displacements may cancel each other out, so that the overall mechanical limb axis ends up in the 'normal' position (Figure 12.4b). It follows that the observed position of the mechanical axis of the lower limb in relation to the knee joint is a 'screening' assessment and does not rule out the presence of deformity. A further step would be to compare reference angles subtended by the mechanical axes of the individual bone segments to joints. It is usual to compare these angles with those of the contralateral 'normal' side but in the event the other is also affected some 'normal' reference ranges are available (Figure 12.5):



(a) (b)

Figure 12.4 Deformity in the lower limb (a) It may be sufficient to alter the mechanical axis of the limb – here it is shifted laterally due to changes at the hip joint. (b) If there are two deformities, the mechanical axis may be normal if the effect of each is to shift the axis in equal and opposite directions – a compensated deformity.

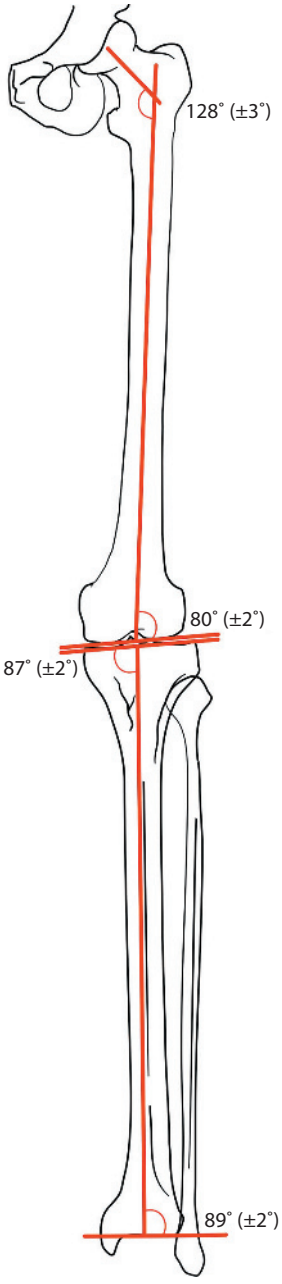


Figure 12.5 Analysis of coronal plane deformity This can be based on a contralateral normal limb or use of reference angles in relation to the anatomical (or mechanical) axes.

1 *At the hip* – the angle between the anatomical axis of the femur and the axis of the femoral neck is approximately 128 degrees (±3 degrees).
2 *At the knee* – the angle between the anatomical axis of the femur and a tangent to the joint line of the knee is, on the lateral aspect, approximately 80 degrees (±2 degrees).
3 *At the knee* – the angle between the anatomical axis of the tibia and a tangent to the joint line of the knee is, on the medial aspect, approximately 87 degrees (±2 degrees).
4 *At the ankle* – the angle between the anatomical axis of the tibia and a tangent to the tibial plafond is, on the lateral aspect, approximately 89 degrees (±2 degrees).

If an abnormal value is encountered, it suggests a deformity is present within that bone. However, when drawing out these reference angles and seeking to identify a source of deformity, it is easy to be carried away by abnormal values that differ from the reference ranges by a few degrees. The clinical significance of these 'abnormalities' must be taken in context; an intrinsic (naturally present) varus angulation of a few degrees at the distal femur matters little if the main source of deformity is a larger varus malunion of a tibial fracture further distally – in which case the correction should be in the tibia.

Rules for osteotomy

Most surgeons are familiar with the simple method of drawing the anatomical axes of the bone segments proximal and distal to a deformity and measuring the extent of the deformity (in degrees) at the intersection of these axes. In contemporary deformity analysis this intersection of anatomical axes is referred to as a *centre of rotation of angulation (CORA)* and can also be determined by noting the intersection of the mechanical axes of the segments proximal and distal to the deformity (Figure 12.6).

The CORA is important for the following reasons:

1 It indicates where an axis of rotation, named angulation correction axis (ACA), should be placed about which the two intersecting axes of the CORA can be brought in line and hence the deformity corrected. This axis of rotation, which enables appropriate realignment of the intersecting axes, should be positioned on either side of the CORA but along a line termed 'the bisector'. This is, as is implied in its name, the line that bisects the angle described by the deformity (Figure 12.7a). The effect of placing the axis of rotation on the convex side of the deformity is to envisage an opening wedge correction; conversely, if it is placed on the concave side, it is a closing wedge correction. Moving the rotation axis further along the bisector increases or decreases the size of the opening, i.e. it achieves simultaneous lengthening or shortening with the angular correction (Figure 12.7b,c). If the rotation axis is not placed on the bisector, a translation deformity will ensue despite satisfactory correction of angulation.
2 It reveals the presence of translation as well as angulation as components of the deformity and can also indicate the presence of multi-apical deformities.
   a When the CORA is identified and is found to lie within the boundaries of the bone involved as well as coinciding in level with the apex of the deformity, this indicates only an angular component to the deformity. The rotation axis to correct the deformity can be sited on the bisector and the osteotomy performed at the same level – this is equivalent to classic correction through opening or closing wedge methods (Figure 12.7b).
   b When the CORA lies within the boundaries of the bone involved but is at a different level to that of the apex of deformity, it indicates the presence of translation and angulation within the deformity (Figure 12.8a). The rotation axis to enable correction should be maintained on the bisector of the CORA but the osteotomy can be sited at either of the two levels (coincident with the apex of deformity or at the CORA): (1) when positioned on the former, correction of both translation and angulation is simultaneously accomplished at the site of

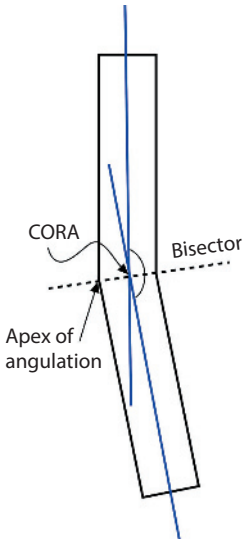


Figure 12.6 Location of the CORA It is found at the intersection of the anatomical (or mechanical) axes of the segments proximal and distal to the deformity. The bisector is the line that divides the supplement to the angle of deformity. While the apex of angulation and CORA coincide in this example, that is not always so.

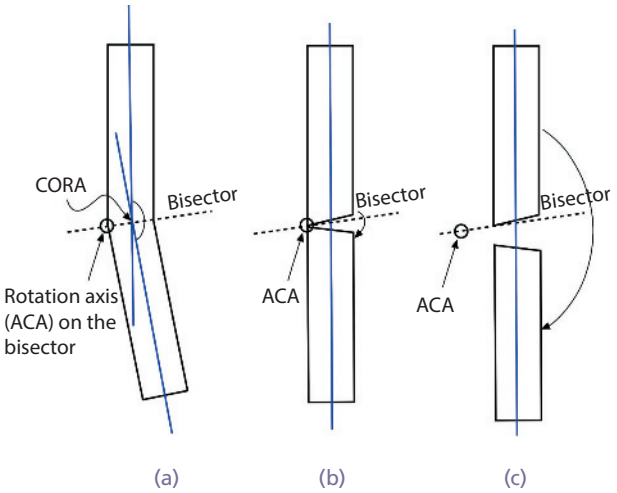


Figure 12.7 Axis of rotation It is placed along the bisector of the CORA on the convex side. This achieves an open wedge correction (a,b). If the rotation axis is moved further along the bisector, lengthening – in addition to the open wedge realignment – is obtained (c).

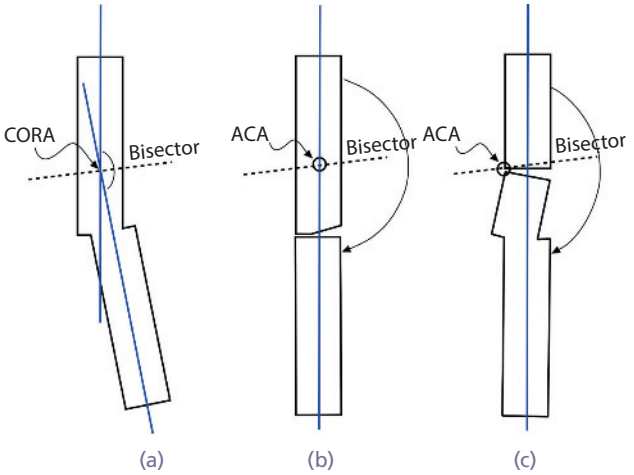


Figure 12.8 CORA If the CORA is found to be proximal or distal to the apex of angulation, but within the boundaries of the bone, this suggests the presence of translation as an additional component to the deformity (a). Simultaneous correction can be achieved by placing the axis of rotation on the CORA or its bisector; the osteotomy can be either at the apex of angulation (b) or at the same level as the CORA (c).

original deformity (Figure 12.8b); (2) when sited on the latter, a new deformity is created which correctly 'balances' the malalignment produced from the original site (Figure 12.8c).

c When the CORA lies outside the boundaries of the involved bone, a multi-apical deformity is likely to be present (and the deformity more akin to a curve). The deformity would need to be resolved through multiple osteotomies.

These features of the CORA are, in essence, the rules of osteotomy as described by Paley. It explains why it is permissible to perform osteotomies away from the apex of the deformity as long as the correction is achieved through a rotation axis placed on the CORA or on its bisector. Many examples in orthopaedics illustrate this principle, including performing an intertrochanteric or subtrochanteric osteotomy to correct malalignment of the femoral neck in a child with a slipped capital femoral epiphysis, or inducing translation in correcting a genu valgum arising from the femoral joint line – both of which are examples of the 2(b) scenario above.

Complications of osteotomy and deformity correction

General As with all bone operations, thromboembolism and infection are calculated risks.

Undercorrection and overcorrection Under- and overcorrection of the deformity can be avoided by careful preoperative planning. In difficult cases, intra-operative radiographs or fluoroscopy are essential. If the fault is recognized while the patient is still under the anaesthetic, it should be corrected straightaway. If discovered on a postoperative radiographic check, the impact of the mistake will need to be gauged and, if significant, it may still be advisable to revise the procedure.

Nerve tension Correction of severe deformities may put excessive tension on a nearby nerve. The commonest example is peroneal nerve palsy after corrective osteotomy for a marked valgus deformity of the knee. In general, acute long-bone corrections greater than 20 degrees should be avoided and, if there is a known risk of nerve injury, it should be limited to 10 degrees. If greater correction is needed, it can be done gradually in an appropriate external fixator (see the Ilizarov method, discussed below).

Compartment syndrome Osteotomy of the tibia or forearm bones is at risk of this rare but potentially limb-threatening complication. The limb should be checked repeatedly for signs and prompt action taken if danger signals appear (see Chapter 23).

Non-union Non-union may occur if fixation is inadequate or if the soft tissues are damaged by excessive stripping during surgical exposure. Gentle handling of tissues and respect for the blood supply to bone together with sound fixation techniques will minimize the risk.

BONE FIXATION

Stabilizing two or more segments or fragments of bone is usually by internal or external fixation methods. In internal fixation, this may involve screws, wires, plates or intramedullary rods. External fixators come in a variety of types. There are basic rules for choosing and using either method.

Internal fixation by screws

Simple screw

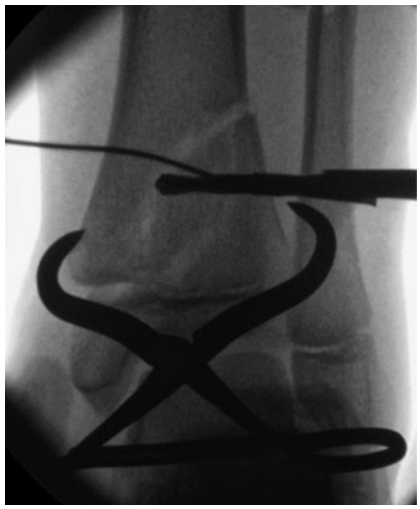
Screws are devices that convert rotational movement into longitudinal movement. Turning of the head causes the screw to advance. They can be used to hold two fragments of bone in close proximity to allow healing or to fix an implant such as a plate to bone.

Lag screw

Screws are also used to compress two fragments together through what is called the 'lag principle' (Figure 12.9). By overdrilling the near fragment, the threads of the screw only engage the far fragment; when the screw is tightened, the head of the screw pushes the near fragment towards the far fragment and causes compression between the two.

The lag screw works best if passed at right angles to the plane between the bone fragments. If there is a long fracture line, several screws can be inserted at different levels with each screw at right angles to the fracture plane at their respective sites. A similar lag effect is achieved if the screw is threaded only near its tip – a partially threaded screw. The pull-out strength of a screw fixed in bone depends on factors involving both the screw and the bone: it increases (1) with the diameter of screw thread and the length of screw embedded; (2) with the thickness and density of the bone in which it is embedded; (3) if both cortices are engaged by the screw.

Most screws are inserted after drilling a pilot-hole and tapping, although self-drilling and self-tapping screws are available. In cancellous bone, and particularly if it is osteoporotic, it may be preferable not to tap after pre-drilling; tapping removes additional bone that may help anchor the screw.



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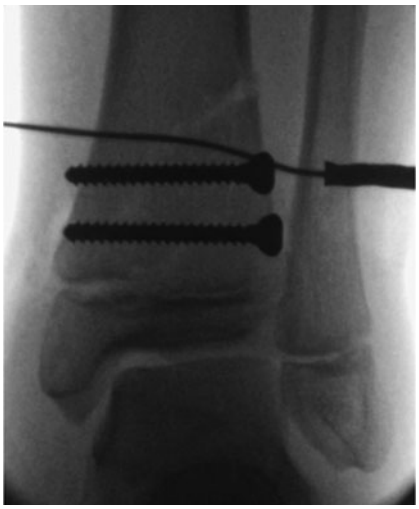

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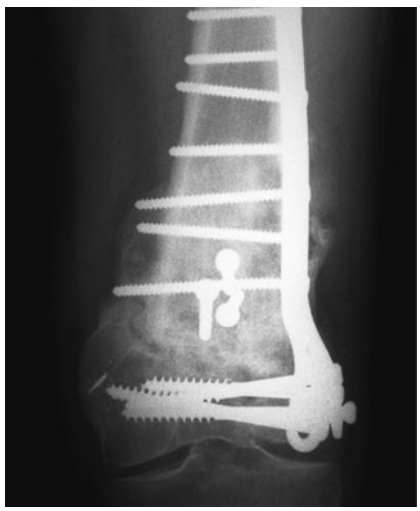

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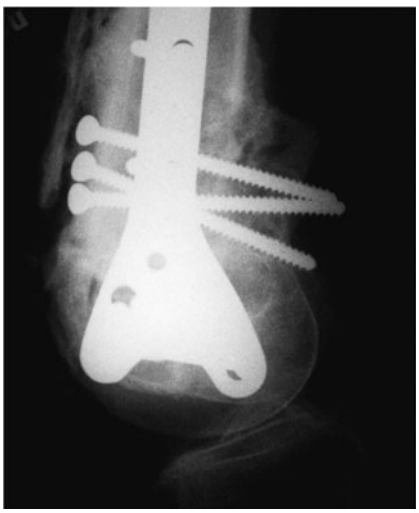

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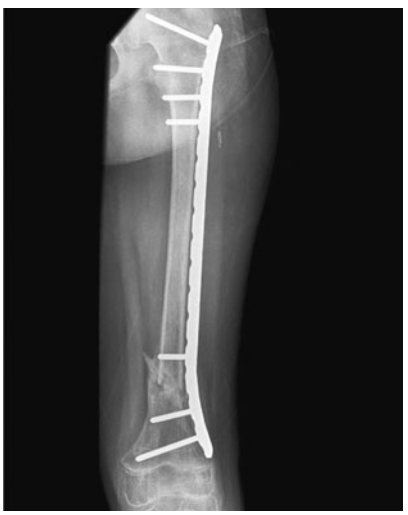


(e)

Figure 12.9 Lag screw fixation
This is accomplished through design of the screw (being unthreaded for part of the shank) or through overdrilling the near fragment (a,b). Lag screws are thus used individually or in conjunction with a plate (c,d). Plates can be applied to control twisting forces (here they are used in conjunction with lag screws) or simply as long internal splints, as in indirect submuscular plating of fractures (e).

Cannulated variable pitch screws

A pragmatic alternative to lag screw fixation involves screws with a variable pitch along the length. As the screw is advanced the ends are compressed together. The provisional passage of k-wires to confirm the position under fluoroscopy then threading a cannulated variable pitch screw is particularly useful. The devices were first used in the scaphoid (Herbert screw) but are invaluable for other carpal fractures, capitellum fractures and metacarpal head fractures.

Internal fixation by plates and screws

Plates have various designs and purposes:

1 Simple *straight compression plates* allow compression along the axis of the plate.
2 *Contoured plates* fit specific bones.
3 *Low-profile plates* reduce the 'footprint' on the bone so as to preserve local vascularity.
4 *Locked plates* enable the screw to engage the plate by a secure mechanism so as to create a rigid three-dimensional construct which prevents toggling of the screw in the hole.
5 *Bridge plates* span a comminuted defect without disturbing the fracture haematoma or periosteum and do not apply compression.
6 *Neutralization plates* do not apply compression but add some extra stability to a fracture that has been primarily compressed and stabilized by a lag screw.

The plate may be applied subperiosteally by a formal exposure of the fracture or osteotomy, or extraperiosteally in the submuscular plane so as to span the site. These are internal splints that should not be used as load-bearing devices. The ability to control loads across the bone will depend on the degree of contact between the bone ends; it is important that this should be accomplished, usually by compression across the bone ends by a lag screw or through compression with the plate itself (Figure 12.9).

In addition to improving contact between the bone ends, compression through the plate can be utilized as part of the tension-band concept. Curved long bones have a compression side and tension side when axially loaded; plate application on the tension side will convert the loading forces that attempt to separate the fracture ends into compressive ones and thereby maintain bone contact.

Internal fixation by intramedullary devices

Two major design types are used: those with and those without interlocking capabilities.

Interlocking nails have become a standard fixation method for most shaft fractures of the tibia and femur in adults. Stability from these nails is due to a combination of an interference (frictional) fit within the medullary canal and the capture of bone to nail by means of the interlocking screws, which act as bolts. Interlocked intramedullary nails offer better control of length and torsion than the unlocked varieties of this device. Older nail designs had an open cross-section but these are being replaced by closed-section devices, which provide greater torsional stiffness.

The medullary canals of the femur and tibia are not simple cylinders and there are variations between individuals. None of the present-day nail designs are anatomically contoured; therefore intramedullary reaming to a diameter greater than the nail to be used allows unimpeded insertion of the device. Insufficient reaming potentially risks the bone splitting during nail insertion as a result of hoop stresses (expansile forces) generated.

Unlocked intramedullary nails are increasingly used in the treatment of long-bone shaft fractures in children. These flexible rods are inserted so as not to damage the physes at either end of the long bone and they function as internal splints by achieving three-point fixation until callus formation takes over (Figure 12.10). *Intra-medullary wires* have a role in metacarpal fractures.

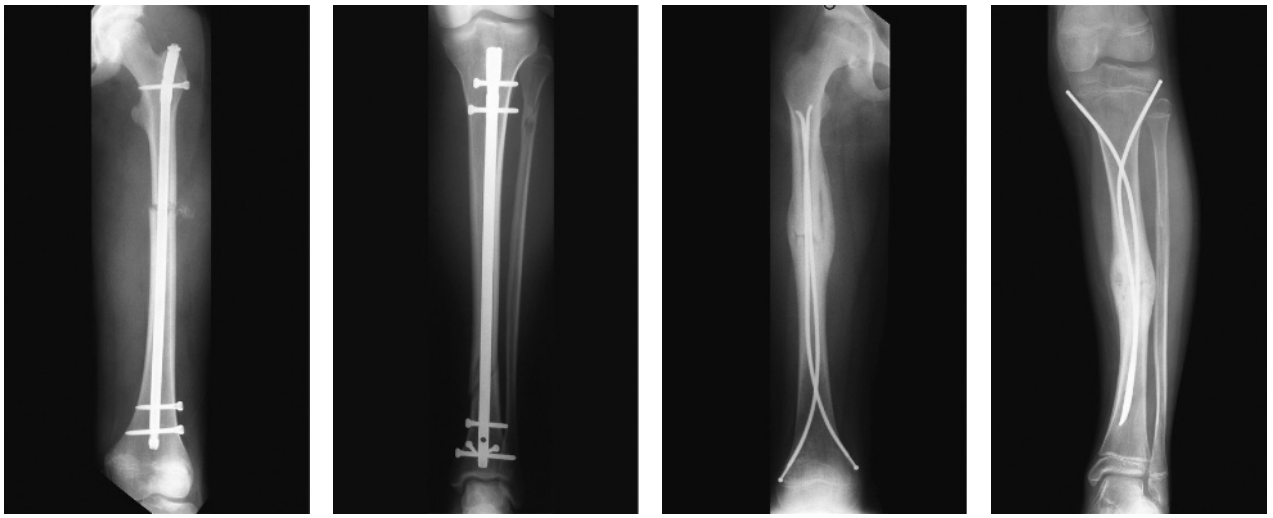
External fixation

Static external fixators

External fixators are useful for temporary stabilization of fractures with severe soft-tissue injuries, for open fractures and for reconstruction of limbs using methods such as the Ilizarov method. They may also be used during emergency stabilization of multiple long-bone fractures in the polytraumatized patient (Figure 12.11).

The fixator functions as an exoskeleton through which the patient's own skeleton can be supported and adjusted. The basic components are wires or pins inserted into bone to which rods or rings are attached and interconnected. Pin- or wire-related problems have limited more widespread adoption of this method; newer pin designs, and some with hydroxyapatite coating, have reduced the frequency of problems. The mechanics of pin-hold in bone is governed by similar factors to that of screws.

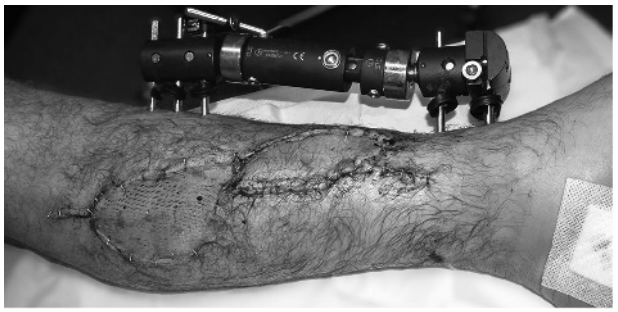
External fixators are mainly of the *unilateral-planar* or *circular* types; there are also designs that combine aspects of both types (*hybrid*). Each possesses specific biomechanical properties with regard to control of movement at the fracture or osteotomy site, especially when the patient loads the limb on walking. The choice of a fixator type will depend on many factors including the intended purpose of its use and the surgeon's familiarity with the device.



(a) (b) (c) (d)

Figure 12.10 Intramedullary nails These are excellent for stabilizing shaft fractures of the major long bones: (a) femur; (b) tibia. Locked nails have the added benefit of controlling length and torsion. Flexible and elastic nails work by three-point fixation and are suitable for paediatric fractures where damage to the physis can be avoided (c,d).

(a)



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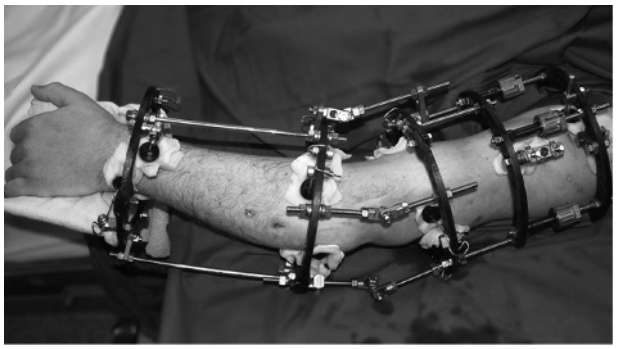


(c)

Figure 12.11 External fixators (a) These are useful for provisional fracture control, as in severe open fractures. Fixators are also used for definitive fracture treatment (b) and for Ilizarov limb reconstruction surgery (c).

Static external joint fixation

While an external fixator should provide rigid (but not too rigid) fixation for the shaft of a long bone, it can also distract a joint. This is particularly useful for comminuted intra-articular fractures when the distraction pulls on the ligament attachments to indirectly reduce the joint (ligamentotaxis). Supplementary percutaneous k-wires can hold some fragments. The distraction must not be too strong, especially in the wrist, as stiffness of the fingers may result.

Dynamic external fixation

This is especially useful for the two joints that do not tolerate stiffness – the proximal interphalangeal joint (PIPJ) and the elbow. The stiffness may be provoked by the combination of a comminuted intra-articular fracture and prolonged immobilization. Various devices are available, from complex hinged fixators for the elbow to simple home-made wire frames for the PIPJ. The essential technical point is that the fixator bends exactly at the axis of rotation of the joint.

BONE GRAFTS AND SUBSTITUTES

Bone grafts are both *osteoinductive* and *osteoconductive*: (1) they are able to stimulate osteogenesis through the differentiation of mesenchymal cells into osteoprogenitor cells; (2) they provide linkage across defects and a scaffold upon which new bone can form. Osteogenesis is brought about partly by the activity of cells surviving on the surface of the graft but mainly by the action of osteoprogenitor cells in the host bed.

Three basic requirements for osteogenesis are the presence of osteoprogenitor cells, a bone matrix and growth factors.

Autografts (autogenous grafts)

Bone is transferred from one site to another in the same individual. These are the most commonly used grafts and are satisfactory provided that sufficient bone of the sort required is available and that, at the recipient site, there is a clean vascular bed.

Cancellous autografts

Cancellous bone can be obtained from sites such as the ilium, greater trochanter, proximal metaphysis of the tibia, calcaneum, lower radius, olecranon, or from an excised femoral head. Cortical autografts can be harvested from any convenient long bone or from the iliac crest; they usually need to be fixed with screws, sometimes reinforced by a plate and can be placed on the host bone, or inlaid, or slid along the long axis of the bone. Cancellous grafts are more rapidly incorporated into host bone than cortical grafts, but sometimes the greater strength of cortical bone is needed to provide structural integrity.

The autografts undergo necrosis, though a few surface cells remain viable. The graft stimulates an inflammatory response with the formation of a fibrovascular stroma; through this, blood vessels and osteoprogenitor cells can pass from the recipient bone into the graft. Apart from providing a stimulus for bone growth (osteoinduction), the graft also provides a passive scaffold for new bone growth (osteoconduction). Cancellous grafts become incorporated more quickly and more completely than cortical grafts (Figure 12.12).

Vascularized grafts

This is theoretically the ideal graft; bone is transferred complete with its blood supply, which is anastomosed to vessels at the recipient site. The technique is difficult and time-consuming and requires microsurgical skills. Available donor sites include the iliac crest (complete with one of the circumflex arteries), the fibula (with the peroneal artery) and the radial shaft. Vascularized grafts remain completely viable and become incorporated by a process analogous to fracture healing.

Bone marrow aspirates

Bone marrow contains stem cells and osteoprogenitor cells, which are able to transform into osteoblasts in the appropriate environment and with stimulation. The number of these mesenchymal cells in aspirates from the iliac crest decreases with age and more so in females. In addition, the aspiration technique from the iliac crest can influence the number

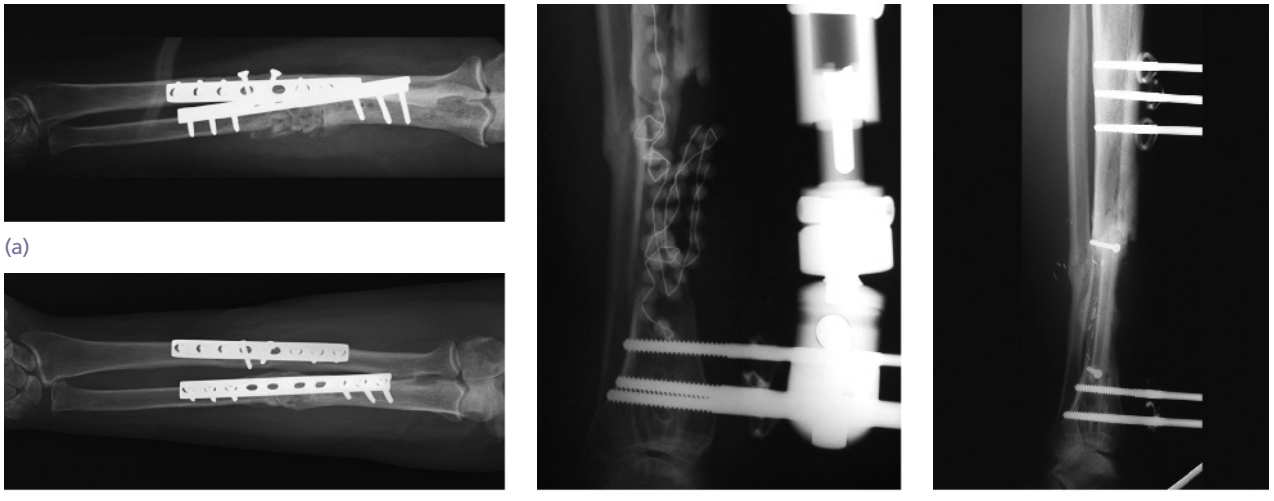


Figure 12.12 Autogenous cancellous bone grafts (a) Here autogenous grafts are used to fill a defect of the ulna and they unite with the host bone in 4 months (b). Free vascularized bone transfer (in this case a portion of fibula) is also helpful when larger defects need to be filled (c,d).

of osteoblast progenitors obtained; this may account for the variable results reported in the small clinical series published. The recommended procedure is to take multiple small-volume aspirates (four 1 mL aspirates from separate site punctures). Centrifugation of the aspirate, in order to concentrate the cellular contents, has provided encouraging results in animal experiments; early evidence suggests this also may be the optimal method for using bone marrow aspirates in humans.

Platelet-derived activators

'Activators' are now available through centrifugation of venous blood. These factors activate repair of tissues (not just bone) and may augment healing processes *in vivo*. Strong clinical evidence of their efficacy is not yet available.

Allografts (homografts)

Allografts consist of bone transferred from one individual (alive or dead) to another of the same species. They can be stored in a bone bank and, as supplies can be plentiful, are particularly useful when large defects have to be filled. However, sterility must be ensured. The potential for transfer of infection is from either diseases present in the donor or contamination at the time of harvesting. The graft must be harvested under sterile conditions and the donor must be screened for malignancy and blood-borne viruses; this requires prolonged (several months') testing of the donor before the graft is used. Sterilization of the donor material can be done by exposure to ethylene oxide or by ionizing radiation, but the physical properties and potential for osteoinduction are considerably altered by these processes.

Fresh allografts, though dead, are not immunologically acceptable. They induce an inflammatory response in the host and this may lead to rejection. However, antigenicity can be reduced by freezing (at –70 °C), freeze-drying or by ionizing radiation. Demineralization is another way of reducing antigenicity and it may also enhance the osteoinductive properties of the graft. Acid extraction of allograft bone yields *demineralized bone matrix*, which contains collagen and growth factors. It is available in a variety of forms (putty, powder, granules) and is sometimes combined with other types of bone substitutes. The osteoinductive capability of demineralized bone matrix is variable; most human studies have not shown the impressive osteoinductive capacity found in animal experiments. One way to supplement the properties of demineralized bone matrix is to use it as an autologous bone graft expander.

Allografts are most often used in reconstructive surgery where pieces are inserted for structural support; an example is revision hip arthroplasty where bone loss from prosthesis loosening is replaced. The process of incorporation of allografts (when it occurs) is similar to that with autografts but slower and less complete.

Bone morphogenetic proteins (BMPs)

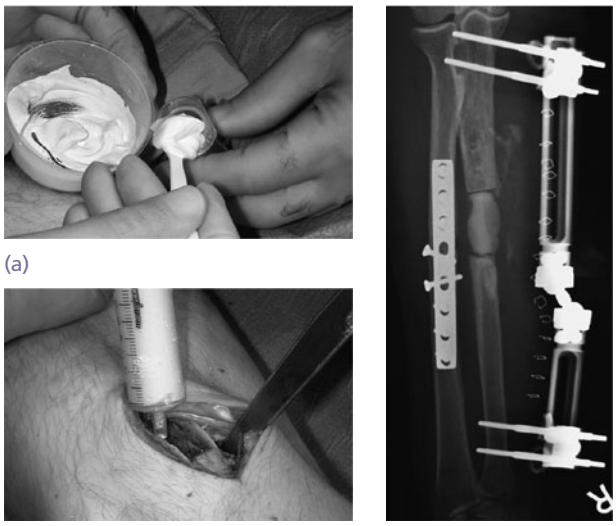
These substances were originally extracted from allograft bone but were too difficult to produce in commercially suitable quantities. BMP-2 and BMP-7 are now manufactured using recombinant techniques and are available commercially.

BMPs are osteoinductive. There is evidence to support their use in the treatment of non-union and open tibial fractures where the success rate is equivalent to that of autogenous bone grafts. They are used with a carrier, which may be allograft, demineralized bone matrix, collagen or bioactive bone cement. Currently, the cost of purchase is a barrier to widespread adoption.

Calcium-based synthetic substitutes

Calcium phosphate, hydroxyapatite (a crystalline calcium phosphate) and calcium sulphate are primarily osteoconductive and need a pore size of around 400 μm for osteoprogenitor cells to lay down bone.

The calcium phosphate and hydroxyapatite varieties are usually used to fill metaphyseal defects in fracture surgery (e.g. tibial plateau, distal radius and calcaneal fractures); in this context, several studies have reported good results. Various forms of the material are available, including granules, chips and paste. Despite claims by manufacturers to the contrary, these synthetic substitutes do not usually possess sufficient compressive strength to withstand high loads and they should be used in stabilized fractures and not as a means of contribution to stability. Compression strength can be increased by altering the ratio of calcium phosphate to hydroxyapatite, by increasing the scintering temperature and by decreasing porosity. Calcium phosphate materials are usually absorbed completely by 6–9 months, but hydroxyapatite substitutes are still visible on X-ray after several years. This slow resorption has prompted hydroxyapatite and calcium phosphate mixtures to be made available, in the hope that the faster resorption of the latter will enable more rapid bone replacement. Calcium phosphate has also been successfully mixed with autologous bone marrow and bovine collagen to produce results equivalent to those of autogenous



(a) (b) (c)

Figure 12.13 Synthetic bone substitutes These are used primarily as osteoconductive agents or as a delivery medium for antibiotics. Several forms are available, including putties and injectable pastes (a,b). They are used to fill small defects or can act as antibiotic-eluting spacers after bone resection in chronic osteomyelitis (c).

bone graft. It has also been used to deliver autologous expanded mesenchymal stem cells. Early trials of this composite biomaterial are extremely promising in treating diaphyseal non-unions.

In contrast, calcium sulphate materials are usually resorbed within 6–9 weeks and are useful, in combination with gentamicin or tobramycin, as a means of local antibiotic delivery in the treatment of cavities or 'dead space' after surgery in chronic osteomyelitis (Figure 12.13).

DISTRACTION OSTEOGENESIS AND LIMB RECONSTRUCTION – ILIZAROV METHOD

Distraction osteogenesis is a form of tissue engineering founded on the principle of *tension-stress*, which is the generation of new bone in response to gradual increases in tension. Discovered in the 1950s by Gavril Ilizarov in Russia, the application of this principle to orthopaedic conditions represents a significant advance; it has opened opportunities for treatment in conditions that hitherto were poorly treated or even untreatable. The term *'Ilizarov method'* embraces the various applications of this principle, emphasizing minimally invasive surgery (many of the techniques are performed percutaneously) and an early return of function.

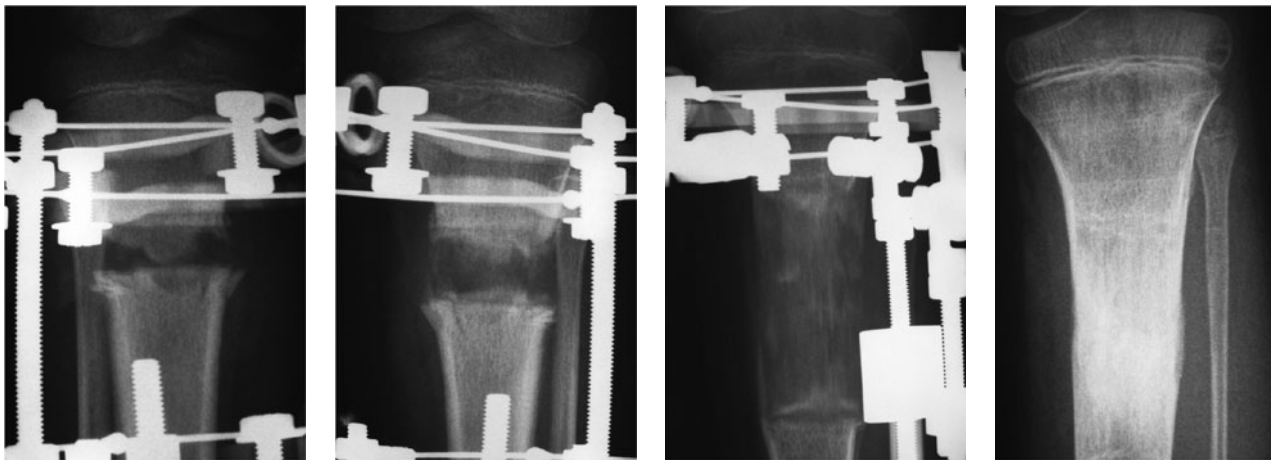
Distraction osteogenesis

Callotasis

Callus distraction, or *callotasis*, is perhaps the single most important application of the tension-stress principle. It is used for limb lengthening or filling of large segmental defects in bone, either through bone transport or other strategies. The basis of the technique is to produce a careful fracture of bone, followed by a short wait before the young callus is gradually distracted via a circular or unilateral external fixator. It is worth noting that all tissue types are created during the distraction process and the term *distraction histogenesis* is perhaps more appropriate.

The external fixator is applied using transfixing wires, pins or screws proximal and distal to the proposed osteogenesis site. The surgical fracture to allow distraction osteogenesis to commence is done by several low energy methods. In a *corticotomy*, the bony cortex is partially divided with a sharp osteotome through a small skin incision and the break completed by osteoclasis, leaving the medullary blood supply and endosteum largely intact. Alternatively, the periosteum can be incised and elevated and the bone then drilled several times before using an osteotome to complete the division; the periosteum is then repaired. Both techniques are exacting – simply dividing the bone with a power saw results in nothing being formed in the gap. After an initial wait of 5–10 days, distraction is begun and proceeds at 1 mm a day, with small (usually 0.25 mm) increments spaced out evenly throughout the day. The first callus is usually seen on X-ray after 3–4 weeks; in optimum conditions it appears on X-ray as an even column of partially radio-opaque material in the gap between the bone fragments. This is called the *regenerate*. If the distraction rate is too fast, or the osteotomy performed poorly, the regenerate may be thin with an hourglass appearance; conversely, if distraction is too slow, it may appear bulbous or worse still may consolidate prematurely, thereby preventing any further lengthening.

When the desired length is reached, a second wait follows, which allows the regenerate column to consolidate and harden. Weight-bearing is permitted throughout this period and it assists the consolidation process. The regenerate column is first seen on X-ray to be divided by an irregular line (the *fibrous interzone*), which gradually disappears when the column of bone completely ossifies. Regular radiographs allow the surgeon to check on the quality of regenerate (Figure 12.14). When cortices of even thickness are seen in the regenerate and weight-bearing is not painful, the fixator is ready to be removed. Throughout treatment, physiotherapy is important to preserve joint movement and avoid contractures.



(a) (b) (c) (d)

Figure 12.14 Distraction osteogenesis Early on there is little activity in the distracted gap (a). A little later, columns of bone are seen reaching for the centre of the distracted zone, leaving a clear space in between – the fibrous interzone (b). When the columns bridge the gap, the regenerate bone matures and, finally, a medullary cavity is re-established (c,d).

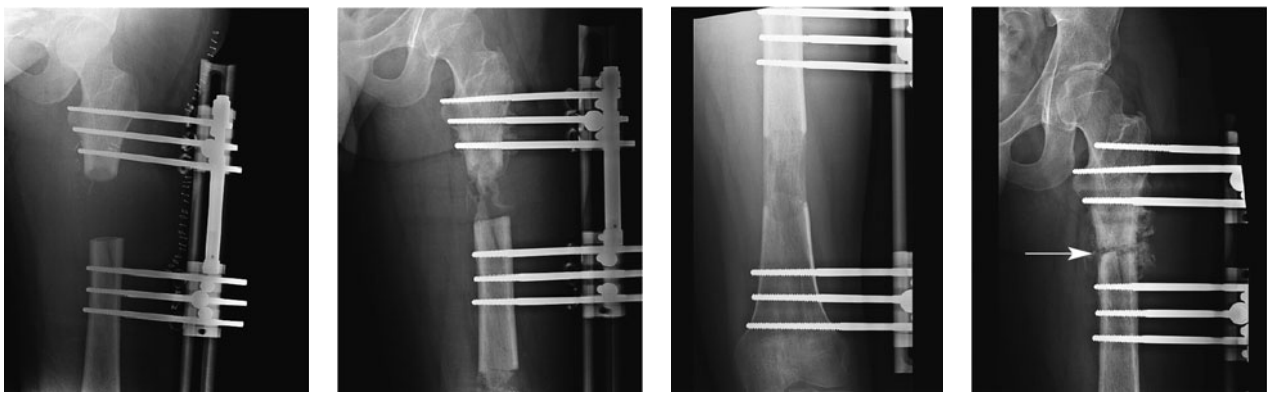
Chondrodiatasis

Bone lengthening can also be achieved by distracting the growth plate (*chondrodiatasis*). No osteotomy is needed but the distraction rate is slower, usually 0.25 mm twice daily. Although a wide, even column of regenerate is usually seen, the fate of the physis is sealed – the growth plate frequently closes after the process. This technique is best reserved for children close to the end of growth.

Bone transport

Distraction osteogenesis is used not only for limb lengthening but also as a means of filling segmental defects in bone. In *bone transport*, the defect (or gap) is filled gradually by creating a 'floating' segment of bone through a corticotomy either proximal or distal to the defect, and this segment is moved slowly across the defect. An external fixator provides stability and the ability to control this segment during the process. As the segment is transported from the corticotomy site to the new docking site, new bone is created in its wake, which effectively fills the defect (Figure 12.15).

A variant of the bone transport technique is *bifocal compression-distraction*. With this method, the defect is closed by instantly bringing the bone ends together; a corticotomy is then performed at a different level and length is restored by callotasis. In this case the limb is shortened temporarily, whereas in bone transport overall limb length remains unchanged.



(a) (b) (c) (d)

Figure 12.15 Bone transport A segment of bone 'travels' across a defect. The limb length is, therefore, unchanged. (a,b,c) The segment is created by osteotomy and gradual distraction produces new bone. The docking site (arrow) often needs attending to in order to heal (d).

Correcting bone deformities and joint contractures

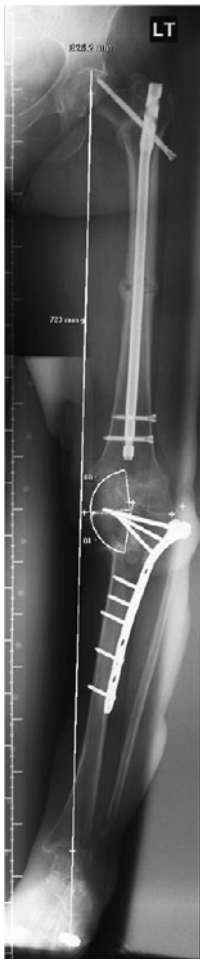
Angular deformities are corrected by carefully planned osteotomies. However, the amount of correction needed may induce, if undertaken acutely, an unwanted sudden tension on soft tissues, particularly nerves. With the Ilizarov method, it is now possible to undertake large corrections with a much lower risk. The correction is performed gradually with the aid of an external fixator; length, rotation and translation deformities can be dealt with simultaneously (Figure 12.16).

The principle of tension stress can also be applied to correcting soft-tissue contractures. For example, a resistant club-foot deformity is dealt with by applying gradual tension to the contracted soft-tissue structures through an external fixator and slowly altering the position of the ankle, subtalar and midtarsal joints until a normal position is achieved. The assembly of the external fixator to accomplish this technique is complex, but the results are often gratifying.

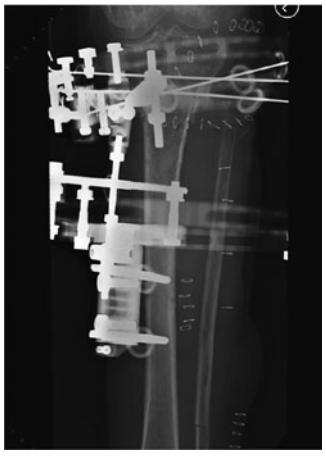
LEG LENGTH EQUALIZATION

Inequality of leg length may result from many causes, including congenital anomalies, malunited fractures, epiphyseal and physeal injuries, infections, paralysis and surgery. Marked inequality leads to inefficient walking and a noticeable limp. The longer leg has to be lifted higher to clear the ground during swing-through and the pelvis and shoulders dip noticeably during the stance phase on the shorter side; both of these adjustments increase energy consumption. Pelvic tilt and the associated compensatory scoliosis tend to cause backache, and there is a higher reported incidence of osteoarthritis of the hip on the longer side – possibly because of the 'uncovering' of the femoral head due to pelvic obliquity.

Inequality greater than 2.5 cm may need treatment, which may amount to no more than a shoe-raise, or it may involve an operation to either the shorter or the longer leg.



(a)



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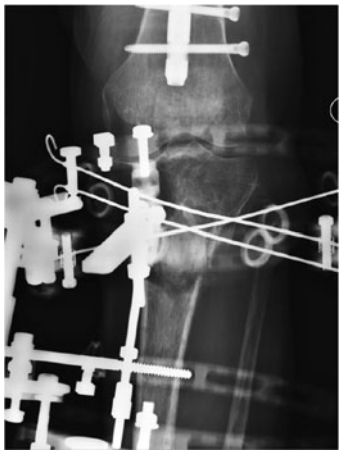

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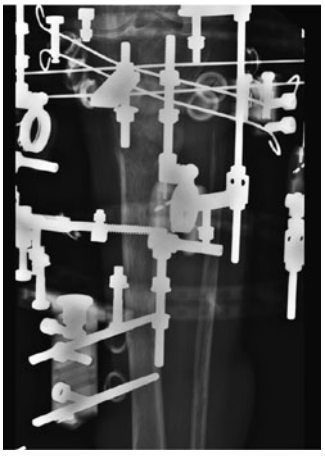

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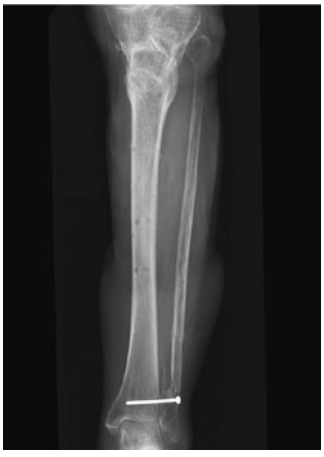

(e)

Figure 12.16 Correction of deformity (a) Varus malunion in a fracture of the proximal tibia was corrected by osteotomy and gradual realignment in a circular external fixator (b–d). A cuneiform-shaped mass of bone formed after the axes were aligned (e).

Techniques for correcting leg length

There are four choices:

- shortening the longer leg
- slowing growth in the longer leg
- lengthening the shorter leg
- speeding up growth in the shorter leg.

The problem of leg length inequality often presents in childhood. Several questions need to be answered before a technique appropriate for the particular child is determined:

- What is the expected adult height of the child?
- What will the discrepancy be when the child is mature?
- When will the child reach skeletal maturity?
- Is there an angular, translational or rotational deformity associated with the leg length discrepancy?

Leg length difference at maturity is estimated through charts and tables and by plotting the rate of change in discrepancy over a period. Expected adult height is calculated through formulae – the Tanner Whitehouse (TW3) method is one – and the time of skeletal maturity is obtained by reading the bone age from a radiograph of the non-dominant hand.

Operations on the longer leg

Physeal arrest

In children, physeal arrest is an effective method of slowing the rate of growth of the longer leg; it can be temporary, using removable staples fixed across the growth plate, or permanent, by drilling across the physis and curetting out the growth plate. Another method is to excise a rectangular block of bone across the physis, rotate the block through 90 degrees and then reinsert it into the original bed. When the physis fuses (epiphysiodesis), longitudinal growth at that site ceases and the overall gain in length of the limb is retarded. In due course, if done at the correct time, the difference in lengths should be reduced.

The timing and technique of epiphysiodesis is important. If it is inaccurately timed, a difference in leg lengths will remain, and if improperly done, deformity may occur. Physeal arrest will create a loss of 10 mm of length a year from the distal femur and 6 mm a year from the proximal tibia. As the physes close naturally at 16 years of age in boys and 14 years in girls, a predicted length discrepancy at maturity of 45 mm can, for example, be addressed by both a distal femoral and proximal tibial physeal arrest performed about 3 years before skeletal maturity.

Epiphysiodesis produces approximate length equalization, often to within 10 mm of estimated length, if performed in a timely fashion. Other methods of predicting the timing of epiphysiodesis are based on charts or use a multiplier method.

Bone shortening

Epiphysiodesis is feasible only in a growing child. In adults, it is necessary to excise a segment of bone, preferably from the femur, since tibial shortening is more complicated and is cosmetically unattractive; up to 7.5 cm of femoral shortening can be achieved without permanent impairment of function. The safest technique is to excise a segment from between the lesser trochanter and the femoral isthmus, to approximate the cut ends, and to fix them together with a locking intramedullary nail or plate. Open excision of bone segments from the long leg has several disadvantages, among which are scarring and poor muscle tone. The scarring results from a longitudinal incision being suddenly subjected to a concertina effect, which causes the wound to gape widely. Shorter segments can be removed by 'closed' intramedullary techniques, which rely on an intramedullary saw and bone splitter, and thereby avoid the problem with scars. In general, shortening of the long leg is reserved for situations where the patient is too old for an epiphysiodesis or where lengthening the short leg is deemed too risky, for instance in the presence of unstable joints or infection.

Shortening should, of course, be applied only if the patient's residual height will still be acceptable. It should also be remembered that the longer leg is usually the normal one and, if a serious complication such as non-union ensues, the patient may be worse off than not having an operation in the first place.

Lengthening the shorter leg

Lengthening the short leg is most easily accomplished by wearing a raised shoe, but this is often inadequate or unacceptable – a shoe raise of more than 5 cm can risk injury to the ankle!

Stimulation of the growth plate can be achieved by the technique of periosteal division. A circumferential 5 mm strip is excised from around the distal femoral or proximal tibial physis. The physis responds with an accelerated growth rate that may last for up to 2 years. However, like epiphysiodesis, poor technique may produce deformity; the method is probably best reserved for young children (younger than 6 years) as the effects on older children are unpredictable.

Limb lengthening by the Ilizarov method is a suitable method for predicted length discrepancies of greater than 5 cm. Distraction osteogenesis has become much safer since it was appreciated that distraction has to be slow if neural or vascular damage is to be avoided (see earlier). Major length corrections can be tackled by staging the treatment process over

several years, or by attempting to lengthen at two levels within the same bone (*bifocal lengthening*). The latter method, although attractive, has a higher rate of complications largely from the soft tissues being distracted too quickly.

OPERATIONS TO INCREASE STATURE

Bilateral leg lengthening is a feasible procedure for people with achondroplasia and other individuals of short stature, but detailed consultation is an essential preliminary process. The prospective patient must understand that treatment is painful, prolonged, and may be associated with a substantial number of complications such as pin-site sepsis, deformity or fracture. Moreover, gain in height is not the same as 'normality'. Nevertheless, successful treatment is so rewarding ('People no longer look at me in the street; I can now get things off a shelf without having to climb up') that it should not be withheld if the patient is otherwise normal and is psychologically prepared. Referral to a specialist centre is wise.

The techniques of lengthening are as described earlier and two bones can be dealt with simultaneously. It is more usual to lengthen both tibiae at one procedure and both femora at another. Gains in height averaging 20–25 cm have been achieved by combining the bone lengthening with soft-tissue releases.

OPERATIONS ON JOINTS

ARTHROTOMY

Arthrotomy (opening a joint) may be indicated to: (1) inspect the interior or perform a synovial biopsy; (2) drain a haematoma or an abscess; (3) remove a loose body or damaged structure (e.g. a torn meniscus); (4) to excise inflamed synovium. The intra-articular tissues should be handled with great care, and if postoperative bleeding is expected (e.g. after synovectomy), a drain should be inserted – postoperative haemarthrosis predisposes to infection. Following the operation the joint should be rested for a few days, but thereafter movement must be encouraged.

ARTHRODESIS

Arthrodesis is a reliable operation for a painful or unstable joint; where stiffness does not seriously affect function, this is may be the treatment of choice. Examples are the spine, tarsus, ankle, wrist, thumb metacarpophalangeal joint and the interphalangeal joints. Arthrodesis is also useful for a knee that is already stiff (provided the other knee has good movement) and for a flail shoulder. More controversial is arthrodesis of the hip. Though it is a reasonable alternative to arthroplasty or osteotomy for joint disease in young patients, there is an understandable resistance to sacrificing all movement in such an important joint. It is difficult to convey to the patient that a fused hip can still 'move' by virtue of pelvic tilting and rotation; the best approach is to introduce the patient to someone who has had a successful arthrodesis.

The principles of arthrodesis are straightforward and involve four stages:

1 *Exposure* – both joint surfaces need to be well visualized and often this means an extensile incision, but some smaller joints are now accessible by arthroscopic means.
2 *Preparation* – both articular surfaces are denuded of cartilage and sometimes the subchondral bone is 'feathered' to increase the contact area.
3 *Coaptation* – the prepared surfaces are apposed in the optimum position, ensuring good contact.
4 *Fixation* – the surfaces are held rigidly by internal or external fixation. Sometimes bone grafts are added in the larger joints to promote osseous bridging (Figure 12.17).

The main *complication* is non-union with the formation of a pseudoarthrosis. Rigid fixation lessens this

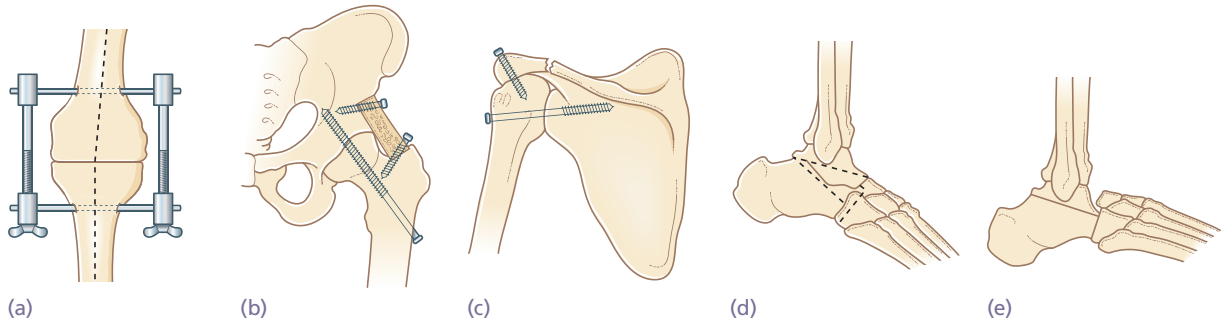

Figure 12.17 Arthrodesis (a) Compression arthrodesis; (b) screw plus bone graft; (c) similar technique using the acromion. (d,e) Subtalar mid-tarsal fusion.

risk; where feasible (e.g. the knee and ankle), the bony parts are squeezed together by compression-fixation devices.

ARTHROPLASTY

Arthroplasty, the surgical refashioning of a joint, aims to relieve pain and to retain or restore movement. The main varieties are: (1) *excision arthroscopy*, (2) *partial replacement* and (3) *total joint replacement* (Figure 12.18):

Excision arthroplasty Sufficient bone is excised from the articulating parts of the joint to create a gap at which movement can occur (e.g. Girdlestone's excision arthroplasty, trapeziectomy, proximal row carpectomy). This movement is limited and occurs through intervening fibrous tissue, which forms in the gap.

Partial replacement One articulating part only is replaced (e.g. a femoral prosthesis or hemiarthroplasty for an intracapsular hip fracture, without an acetabular component); or one compartment of a joint is replaced (e.g. the medial or lateral half of the tibiofemoral joint). The prosthesis is kept in position either by acrylic cement or by initial press-fit stability between implant and bone and later osseointegration, i.e. bonding at a molecular level between the implant and the bone.

Total joint replacement Both the articulating parts of the joint are replaced by prosthetic implants; for biomechanical reasons, the convex component is usually metal and the concave high-density polyethylene. Bearings may be hard-on-soft (e.g. metal-on-polyethylene) or hard-on-hard (e.g. ceramic-on-ceramic). All joint replacements have articulations (parts that move against each other) and all articulations produce wear. Usually the harder material causes wear of the softer material. Wear can be accelerated by roughening of the surfaces, such as by scratching, or by interposition of third bodies, such as cement fragments becoming trapped in the joint. Metal-on-metal bearings became popular in hip replacement with over one million such devices implanted worldwide, but they have been associated with higher incidences of failure, due to production of cobalt and chrome debris, and their use has been almost entirely abandoned. Irrespective of type, these components are fixed to the host bone, either with acrylic cement or by a cementless press-fit technique.

Using hip replacement as an example, the rationale, indications and complications of total joint replacement are discussed in detail in Chapter 19.

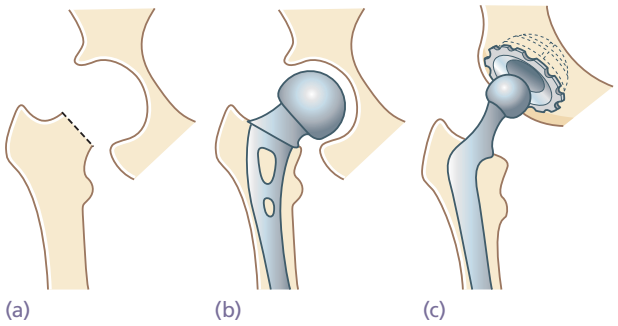

Figure 12.18 Arthroplasty The main varieties as applied to the hip joint: (a) excision arthroplasty (Girdlestone's); (b) partial replacement – an Austin Moore prosthesis has been inserted after removing the femoral head; (c) total replacement – both articular surfaces are replaced.

MICROSURGERY, LIMB REPLANTATION AND LIMB TRANSPLANTATION

Microsurgical techniques are used in repairing nerves and vessels, transplanting bone or soft tissue with a vascular pedicle, transferring a less essential digit (e.g. a toe) to replace a lost essential one (e.g. a thumb) and – occasionally – for reattaching a severed limb or digit. Essential prerequisites are an operating microscope, special instruments, microsutures, a chair with arm supports and – not least – a surgeon well practised in microsurgical techniques.

Replantation and transplantation surgery is time-consuming, expensive and often unsuccessful. It should be carried out only in centres specially equipped and by teams specially trained for this work.

Replantation

In *replantation*, the severed part should be kept cool during transport. The more muscle in the amputated part, the shorter the period it will last; warm ischaemic periods of greater than 6 hours are likely to result in permanent muscle damage and may even produce severe systemic upset in the patient when reperfusion of the muscle occurs.

Two teams dissect, identify and mark each artery, nerve and vein of the stump and the limb. Following careful debridement the bones are shortened to reduce tension and are stabilized internally. Next the vessels are sutured – veins first and (if possible) two veins for each artery. Nerves and tendons are sutured next. Only healthy ends of approximately equal diameter should be joined; tension, kinking and torsion must be prevented. Decompression of skin and fascia, as well as thrombectomy, may be needed in the postoperative period (Figure 12.19).

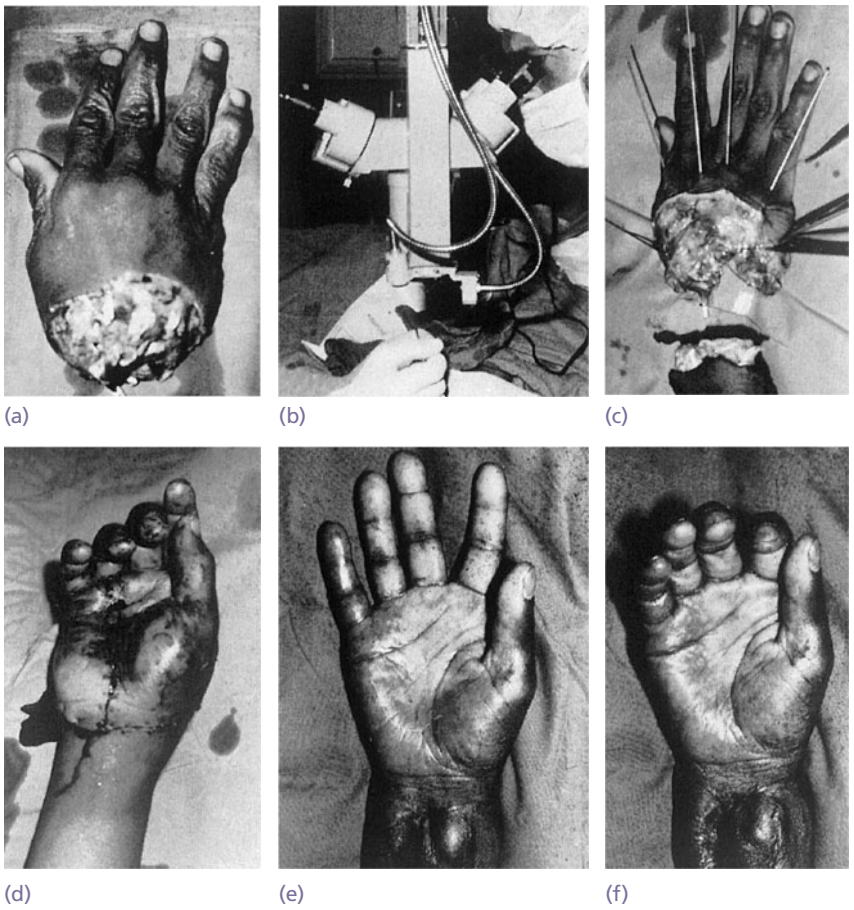

Figure 12.19 Microsurgery and limb replantation (a) The problem – a severed hand. (b) The solution – replantation with microsurgical techniques. (c) The bones of the severed hand have been fixed with k-wires as a preliminary to suturing vessels and nerves. (d) The appearance at the end of the operation. (e,f) The limb 1 year later; the fingers extend fully and bend about halfway. But the hand survived, has moderate sensation and the patient was able to return to work – as a guillotine operator in a paper works!

Transplantation

The indications, technology and immunotherapy for hand transplantation are still being developed. The cadaver hand can provide a sensate functional limb and thus transform the recipient's life. However, issues with the availability of donors, the psychological acceptance of the limb and side effects of intensive long-term immunotherapy have inhibited its adoption.

AMPUTATIONS

Indications

Alan Apley, in characteristic style, encapsulated the indications for amputation in the never-to-be forgotten 'three Ds': (1) *Dead*, (2) *Dangerous* and (3) *Damned nuisance*.

Dead (or dying) Peripheral vascular disease accounts for almost 90% of all amputations. Other causes of limb death are *severe trauma*, *burns* and *frostbite*.

Dangerous 'Dangerous' disorders are *malignant tumours*, *potentially lethal sepsis* and *crush injury*. In crush injury, releasing the compression may result in renal failure (the crush syndrome).

Damned nuisance In some cases retaining the limb may be worse than having no limb at all. This may be because of: (1) pain; (2) gross malformation; (3) recurrent sepsis or (4) severe loss of function. The combination of deformity and loss of sensation is particularly trying, and in the lower limb it is likely to result in pressure ulceration.

Varieties

A *provisional amputation* may be necessary because primary healing is unlikely. The limb is amputated as distal as the causal conditions will allow. Skin flaps sufficient to cover the deep tissues are cut and sutured loosely over a pack. Reamputation is performed when the stump condition is favourable.

A *definitive end-bearing amputation* is performed when pressure or weight is to be borne through the end of a stump. Therefore the scar must not be terminal, and the bone end must be solid, not hollow, which means it must be cut through or near a joint. Examples are through-knee and Syme's amputations.

A *definitive non-end-bearing amputation* is the commonest variety. All upper-limb and most lower-limb amputations come into this category. Because weight is not to be taken at the end of the stump, the scar can be terminal.

AMPUTATIONS AT SITES OF ELECTION

Most lower-limb amputations are for ischaemic disease and are performed through the site of election below the most distal palpable pulse. The selection of amputation level can be aided by Doppler indices; if the ankle/brachial index is greater than 0.5, or if the occlusion pressure at the calf and thigh are greater than 65 mmHg and 50 mmHg respectively, then there is a greater likelihood the below-knee amputation will succeed. An alternative means is by using transcutaneous oxygen tension as a guide, but the level that assures wound healing and avoids unnecessary above-knee amputations has not been confidently determined. The knee joint should be preserved if clinical examination and investigations suggest this is at all feasible – energy expenditure for a transtibial amputee is 10–30% greater as compared to a 40–67% increase in transfemoral cases.

The sites of election are determined also by the demands of prosthetic design and local function. Too short a stump may tend to slip out of the prosthesis. Too long a stump may have inadequate circulation and can become painful, or ulcerate; moreover, it complicates the incorporation of a joint in the prosthesis (Figure 12.20).

Discussion with the prosthetist before amputation will ensure an optimal level especially if a specialized prosthesis is available (e.g. a cosmetic fingertip or a myoelectric forearm).

Principles of technique

A tourniquet is used unless there is arterial insufficiency. Skin flaps are cut so that their combined length equals 1.5 times the width of the limb at the site of amputation. As a rule, anterior and posterior flaps of equal length are used for the upper limb and for transfemoral (above-knee) amputations; below the knee a long posterior flap is usual.

Muscles are divided distal to the proposed site of bone section; subsequently, opposing groups are sutured over the bone end to each other and to the periosteum, thus providing better muscle control as well as better circulation. It is also helpful to pass the sutures that anchor the opposing muscle groups through drill-holes in the bone end, creating an *osteomyodesis*. Nerves are divided under tension proximal to the bone with a sharp blade cut to ensure a cut nerve end will not bear weight.

The bone is sawn across at the proposed level. In transtibial amputations the front of the tibia is usually bevelled and filed to create a smoothly rounded contour; the fibula is cut 3 cm shorter.

The main vessels are tied, the tourniquet is removed and every bleeding point meticulously ligated. The skin is sutured carefully without tension. Suction drainage is usually advised and the stump covered without constricting passes of bandage; figure-of-eight passes are better suited and prevent the creation of a venous tourniquet proximal to the stump.

Aftercare

If a haematoma forms, it is evacuated as soon as possible. After satisfactory wound healing, gradual compression stump socks are used to help shrink the stump and produce a conical limb-end. The muscles must be exercised, the joints kept mobile and the patient taught to use his or her prosthesis.

AMPUTATIONS OTHER THAN AT SITES OF ELECTION

Interscapulo-thoracic (forequarter) amputation

This mutilating operation should be done only for traumatic avulsion of the upper limb (a rare event), when it offers the hope of eradicating a malignant

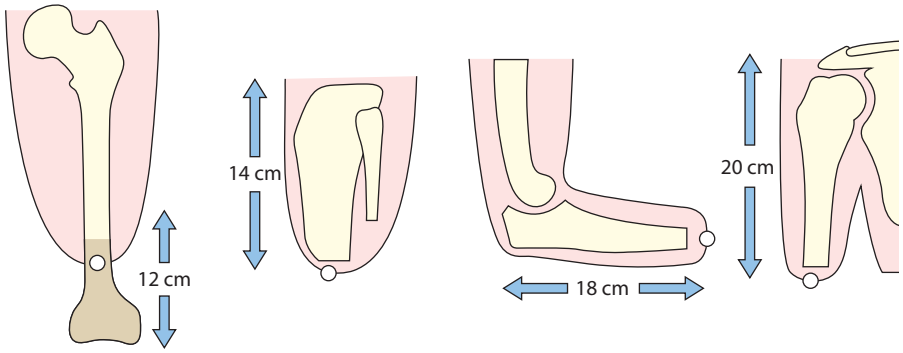

Figure 12.20 Amputations
The traditional sites of election; the scar is made terminal because these are not end-bearing stumps.

tumour, or as palliation for otherwise intractable sepsis or pain.

Disarticulation at the shoulder This is rarely indicated, and if the head of the humerus can be left, the appearance is much better. If 2.5 cm of humerus can be left below the anterior axillary fold, it is possible to hold the stump in a prosthesis.

Amputation in the forearm The shortest forearm stump that will stay in a prosthesis is 2.5 cm, measured from the front of the flexed elbow. However, if a shorter stump is required because of the injury or pathology, it still may be useful as a hook to hang things from.

Amputations in the hand These are discussed in Chapter 16.

Hemipelvectomy (hindquarter amputation) This operation is performed rarely. The indications include malignancy and intractable sepsis.

Disarticulation through the hip This is rarely indicated and prosthetic fitting is difficult. If the femoral head, neck and trochanters can be left, it is possible to fit a tilting-table prosthesis in which the upper femur sits flexed; if, however, a good prosthetic service is available, a disarticulation and moulding of the torso is preferable.

Transfemoral amputations A longer stump offers the patient better control of the prosthesis and it is usual to leave at least 12 cm below the stump for the knee mechanism. However, recent gait studies suggest some latitude is present as long as the amputated femur is at least 57% of the length of the contralateral femur.

Around the knee The Gritti–Stokes operation (in which the trimmed patella is apposed to the trimmed femoral condyle) is rarely performed because the bone may not unite securely; the end-bearing stump is rarely satisfactory and there is no room for a sophisticated knee mechanism.

Amputation through the knee is used at times but is often associated with poorer functional and psychological outcomes to above-knee amputees. Fitting a modern knee mechanism is troublesome and the sitting position reveals the knees to be grossly unequal in level. The main indication for this procedure is in children because the lower femoral physis is preserved, effectively permitting a stump length equivalent to an above-knee amputation to be reached when the child is mature.

Transtibial (below-knee) amputations Healthy below-knee stumps can be fitted with excellent prostheses allowing good function and nearly normal gait. Even a 5–6 cm stump may be fitted with a prosthesis in a thin patient; greater length makes fitting easier, but there is no advantage in prolonging the stump beyond the conventional 14 cm.

Above the ankle Syme's amputation This is sometimes very satisfactory, provided the circulation of the limb is good. It gives excellent function in children, and shares the same advantage as a through-knee amputation in that the distal physis is preserved. In adults it is well accepted by men, but women find it cosmetically undesirable. The indications are few and the operation is difficult to do well. Because the stump is designed to be end-bearing, the scar is brought away from the end by cutting a long posterior flap. The flap must contain not only the skin of the heel but the fibrofatty heel pad so as to provide a good surface for weight-bearing. The bones are divided just above the malleoli to provide a broad area of cancellous bone, to which the flap should stick firmly; otherwise the soft tissues tend to wobble about.

Pirogoff's amputation This amputation is similar in principle to Syme's but it is rarely performed. The back of the os calcis is fixed onto the cut end of the tibia and fibula.

Partial foot amputation The problem here is that the tendo Achillis tends to pull the foot into equinus; this can be prevented by splintage, tenotomy or tendon transfers. The foot may be amputated at any convenient level; for example, through the mid-tarsal joints (Chopart), through the tarsometatarsal joints (Lisfranc), through the metatarsal bones or through the metatarsophalangeal joints. It is best to disregard the classic descriptions and to leave as long a foot as possible provided it is plantigrade and that an adequate flap of plantar skin can be obtained. The only prosthesis needed is a specially moulded slipper worn inside a normal shoe.

In the foot Where feasible, it is better to amputate through the base of the proximal phalanx than through the metatarsophalangeal joint. With diabetic gangrene, septic arthritis of the joint is not uncommon; the entire ray (toe plus metatarsal bone) should be amputated.

PROSTHESES

All prostheses must fit comfortably, should function well and should look presentable. The patient accepts and uses a prosthesis much better if it is fitted soon after operation; delay is unjustifiable now that modular components are available and only the socket need be made individually.

In the *upper limb*, the distal portion of the prosthesis is detachable and can be replaced by a 'dress hand'

or by a variety of useful terminal devices. Electrically powered limbs are available for both children and adults. These are activated by detection of impulses in the stump muscles. The absence of sensory feedback limits their use, as the right amount of pressure for each object cannot be properly judged. In the hand, remarkably lifelike cosmetic prostheses can transform a patient's confidence although, again, the absence of sensory feedback limits their functional benefit.

In the *lower limb,* weight can be transmitted through the ischial tuberosity, patellar tendon, upper tibia or soft tissues. Combinations are permissible; recent developments in silicon and gel materials provide improved comfort in total-contact self-suspending sockets.

COMPLICATIONS OF AMPUTATION STUMPS

Early complications

In addition to the complications of any operation (especially secondary haemorrhage), there are two special hazards: breakdown of skin flaps and gas gangrene.

Breakdown of skin flaps This may be due to ischaemia, suturing under excess tension or (in below-knee amputations) an unduly long tibia pressing against the flap.

Gas gangrene Clostridia and spores from the perineum may infect a high above-knee amputation (or reamputation), especially if performed through ischaemic tissue.

Late complications

Skin *Eczema* is common, and tender purulent lumps may develop in the groin. A rest from the prosthesis is indicated.

Ulceration is usually due to poor circulation, and reamputation at a higher level is then necessary. If, however, the circulation is satisfactory and the skin around an ulcer is healthy, it may be sufficient to excise 2.5 cm of bone and resuture.

Muscle If too much muscle is left at the end of the stump, the resulting unstable 'cushion' induces a feeling of insecurity that may prevent proper use of a prosthesis; if so, the excess soft tissue must be excised.

Blood supply Poor circulation gives a cold, blue stump that is liable to ulcerate. This problem chiefly arises with below-knee amputations and often reamputation is necessary.

Nerve A cut nerve always forms a neuroma and occasionally this is painful and tender. Excising 3 cm of the nerve above the neuroma sometimes succeeds. Alternatively, the epineural sleeve of the nerve stump is freed from nerve fascicles for 5 mm and then sealed with a synthetic tissue adhesive or buried within muscle or bone away from pressure points.

'Phantom limb' This term is used to describe the feeling that the amputated limb is still present. In contrast, residual limb pain exists in the area of the stump. Both features are prevalent in amputees to a varying extent, and they appear to have greater significance in those who also have features of depressive symptoms. The patient should be warned of the possibility; eventually the feeling recedes or disappears but, in some, long-term medication may be needed. A painful phantom limb is very difficult to treat.

Joint The joint above an amputation may be stiff or deformed. A common deformity is fixed flexion and fixed abduction at the hip in above-knee stumps (because the adductors and hamstring muscles have been divided). It should be prevented by exercises. If it becomes established, subtrochanteric osteotomy may be necessary. Fixed flexion at the knee makes it difficult to walk properly and should also be prevented.

Bone A spur often forms at the end of the bone, but is usually painless. If there has been infection, however, the spur may be large and painful and it may be necessary to excise the end of the bone with the spur.

If the bone is transmitting little weight, it becomes osteoporotic and liable to fracture. Such fractures are best treated by internal fixation.

> **NOTE:** In addition to the physical complications of amputation stumps, amputation has significant *psychological effects* on the patient. These should never be underestimated. Careful counselling, meeting others with amputations and involvement in organized activities for amputees (the Paralympics being the ultimate example) will help.

IMPLANT MATERIALS

Metal

Metal used in implants (screws, plates, intramedullary nails and joint replacement prostheses) should be tough, strong, non-corrosive, biologically inert and easy to sterilize. Those commonly used are *stainless*

steel, *cobalt–chromium alloys* and *titanium alloys*. No one material is ideal for all purposes.

Stainless steel Because of its relative plasticity, stainless steel can be cold-worked. This is a process in which the metal is reshaped or resized, usually at room temperature, which increases its hardness and strength. The form of stainless steel used in orthopaedic surgery is 316L; in addition to iron, it contains chromium (which forms an oxide layer providing resistance to corrosion), carbon (which adds strength but needs to be in low concentrations – hence the L suffix – or else it offsets corrosion resistance), nickel and molybdenum as the main elements used in the alloy. The tensile plasticity (ductility) of stainless steel makes it possible to bend plates to required shapes during an operation without seriously disturbing their strength.

Cobalt–chromium-based alloys These alloys are widely used in joint prosthesis manufacture. Chromium is added to cobalt for *passivation*; an adherent oxide layer formed by the chromium provides corrosion resistance, as it does in stainless steel. Other elements are sometimes added, such as tungsten and molybdenum, to improve strength and machining ability. These alloys have a long track record of biocompatibility in human tissue and have also, through forging and cold-working, high strength.

Titanium alloys These are used in fracture fixation devices and joint prostheses. They usually contain aluminium and vanadium in low concentrations for strength; passivation (and thus corrosion resistance) is obtained by creating a titanium oxide layer. The elastic modulus of the metal is close to that of bone and this reduces the stress concentrations that can occur when stainless steel or cobalt chromium alloys are used. Additionally, the corrosion resistance (which is superior to that of the other two alloys) augments this metal's biocompatibility. A disadvantage of titanium alloy is notch sensitivity; this is when a scratch or sharp angle created in the metal, either at manufacture or during insertion of the implant, can significantly reduce its fatigue life.

Implant failure

Metal implants may fail for a variety of reasons: (1) defects during manufacture; (2) incorrect implant selection for the intended purpose; (3) exposure to repeated high stresses from incorrect seating of the implant or from exceeding the fatigue life as when there is delay in a fracture union (Figure 12.21).

Corrosion

Corrosion is inevitable unless the implanted metal is treated, for example by passivation, which creates a protective passive layer; this is usually an oxide layer formed from chemical treatment. In stainless steel and cobalt–chromium, it is the chromium component that helps in creating an oxide layer; in titanium, the element itself forms it. With passivated metal alloys used in orthopaedic surgery, corrosion is rarely a problem except when damage to the passive layer occurs; it may be initiated by abrasive damage or minute surface cracks due to fatigue failure. Even in the absence of these faults, failure can occur through crevice corrosion (where the process is heightened by low oxygen concentrations in crevices, e.g. beneath the heads of screws and plates) or stress corrosion (where repeated low stresses in a corrosive environment cause failure before the fatigue life of the implant is reached). The products of corrosion, metal ions and debris, cause a local inflammatory response that accelerates loosening. Recently there has been a renewed interest in wear of the trunnion (the join of the neck and modular head) in hip arthroplasty.

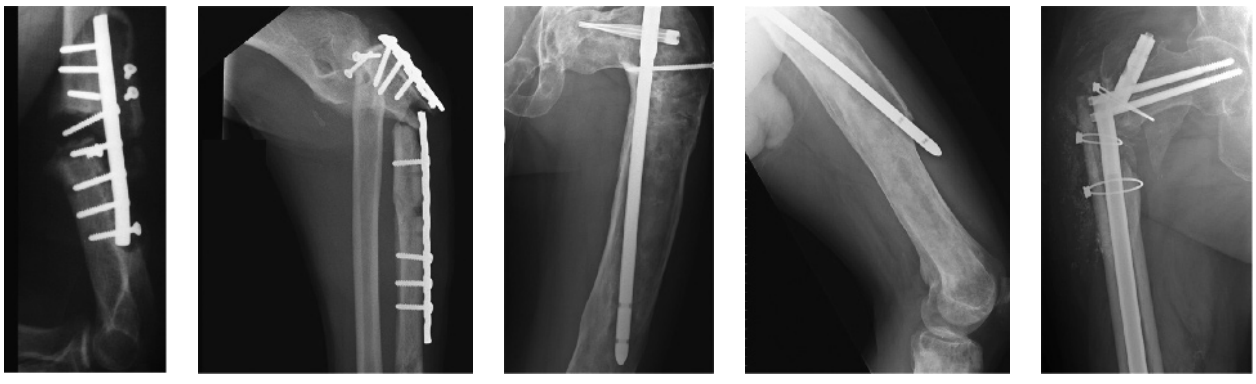

Figure 12.21 Fatigue failure of implants Fatigue failure can be due to (a,b) incorrect implant selection (too small or too weak) or (c,d) incorrect positioning. Other factors are also involved: infection may delay union and lead to eventual implant fracture (e).

Dissimilar metals

Dissimilar metals immersed in solution in contact with one another may set up galvanic corrosion with accelerated destruction of the more reactive (or 'base') metal. In the early days of implant surgery, when highly corrodible metals were used, the same thing happened in the body. However, the passive alloys now used for implants tend to resist this phenomenon (titanium being particularly resistant to chemical attack), although the application of loads at the interface between different materials (such as the trunnion in total hip arthroplasty) may potentiate this process.

Friction and wear

These mechanical concepts are relevant to understanding joint function and prosthesis design. Friction between two sliding surfaces will not be affected by the area of contact or the speed of movement but will depend on the applied load. Therefore, any two surfaces can have a coefficient of friction derived to represent this interaction – it is the ratio of the force needed to start a sliding movement to the normal compression force between the surfaces.

Normal human joints possess coefficients of friction that are about ten times lower than those of various combinations of prosthesis-bearing materials. Metal on ultra-high molecular weight polyethylene (UHMWPE – see below) produces a better (lower) coefficient of friction, and this is improved further if the metal is replaced by a ceramic, such as alumina or zirconium.

An important modulator of friction characteristics in joints is lubrication. Synovial fluid reduces the coefficient of friction by forming either a layer of fluid that is greater in thickness than the surface irregularities on normal articular cartilage (fluid film lubrication) or, in the absence of this interposed fluid layer, a molecular-width coating that resists abrasion (boundary lubrication). Both methods may be involved under different joint loading conditions.

Friction and joint lubrication are related to wear, which is the loss of surface material due to sliding motion under load. Wear is proportional to the load and distance of movement between the two surfaces. Wear between surfaces can be the result of abrasion (a harder surface eroding the surface of the softer material), adhesion (where the two surfaces bond more tightly than particles within one of the surfaces), or from debris that becomes trapped between articulating surfaces and causes abrasion (third-body wear). Metal wear particles may cause local inflammation and scarring and, occasionally, a toxic or allergic reaction; most importantly, however, they may cause implant loosening following their uptake by macrophages and subsequent activation of osteoclastic bone resorption. Metal wear particles may provoke a lymphocyte-dominated vasculitis-associated reaction locally and their presence has also been demonstrated in lymph nodes and other organs far distant from the implant; the significance of this finding is uncertain. Wear of articular cartilage is offset partly by an ability to repair, although this capacity diminishes with age; this mechanism is obviously not possessed by prostheses.

Infection

Metal does not cause infection. Titanium alloys have been shown to be less susceptible to the development of infection when exposed to the same inoculums of bacteria (as compared to stainless steel), but the mechanism of this difference is uncertain. Once infection is established, several mechanisms come into play which encourage its persistence: (1) the metal implant acts as a foreign body that is devoid of blood supply and thereby inaccessible to immune processes; (2) it promotes the formation of biofilms that encase microcolonies of the bacteria and render them immune to defence mechanisms and antibiotics; and (3) it impedes drainage.

Malignancy

A few cases of malignancy at the site of metal implants have been reported, but the number is so small in comparison with the number of implants that the risk can probably be discounted. The risk of malignancy following the use of metal-on-metal bearings in the long term remains a subject of ongoing study.

ULTRA-HIGH MOLECULAR WEIGHT POLYETHYLENE

Ultra-high molecular weight polyethylene (UHMWPE) is an inert thermoplastic polymer. Its density is close to that of the low-density polyethylenes but the very high molecular weight provides increased strength and wear resistance over other types of polyethylene. The material is most commonly used in orthopaedics for hip (acetabular cup) and knee (tibial tray) prostheses and is sterilized by gamma irradiation. The latter process was noted to cause oxidation of the material and detrimentally alter its physical and chemical properties to the extent that a 'shelf life' for the component was created. Consequently, current techniques of sterilization involve gamma irradiation in an oxygen-free, inert environment (e.g. in nitrogen). Although ethylene oxide sterilization is an alternative, irradiation of UHMWPE has the advantage of promoting cross-linking of the polymer, which also improves wear rates.

When in contact with a polished metal as part of a bearing, UHMWPE has a low coefficient of friction and it therefore seemed ideal for joint replacement. This has proved to be true in hip reconstruction with a simple ball-and-socket articulation. However, UHMWPE has *disadvantages*: (1) the cross-linked form may have improved wear properties but poorer yield strength, which may influence crack development and propagation; (2) being a viscoelastic material, it is susceptible to deformity (stretching) and creep; (3) UHMWPE is also easily abraded, a reflection of poor hardness, and chips of bone or acrylic cement trapped on its surface cause accelerated wear.

SILICON COMPOUNDS

There is a wide variety of silicon polymers, of which silicone rubber (Silastic) is particularly useful. It is firm, tough, flexible and inert, and it was used to make hinges for replacing finger and toe joints. However, long-term results are tainted by the material's susceptibility to fracture if the implant surface is nicked or torn by a sharp instrument or piece of bone. The presence of silicon particles in the body may induce a giant-cell synovitis; sometimes bone erosion and 'cyst' formation are seen at some distance from the actual implant. For these reasons the main use for Silastic is as temporary spacers to lie within tendon pulleys prior to tendon transplants.

CARBON

Carbon fibre This is extensively used for the manufacture of external fixation devices, such as connecting rods and even circular rings, as the combination of lightweight, rigidity and radiolucency is attractive. Carbon fibre is also sometimes used to replace ligaments; it induces the formation of longitudinally aligned fibrous tissue, which substitutes for the natural ligament. However, the carbon fibres tend to break up and, if particles find their way into the synovial cavity, they induce a synovitis.

Pyrocarbon For small joint components in the hand and foot, a graphite core is moulded and then coated with a carbon deposit in a very high temperature chamber. These implants are very smooth with negligible wear against another implant and probably induce less wear if articulating directly with joint cartilage. Pyrocarbon is not suitable for larger implants due to manufacturing issues and also its brittleness. It does not formally osseo-integrate and over time the implants do tend to erode and loosen.

ACRYLIC CEMENT

In joint replacement, prostheses are often fixed to the bone with acrylic cement (polymethylmethacrylate – PMMA), which acts as a form of grouting material. It is usually presented as a liquid (the PMMA monomer) and powder (the PMMA polymer plus copolymers or other additives), which is mixed to set off an exothermic polymerization reaction. Before the mixture cures, it is applied to the bone in which the prosthesis is embedded. The application of pressure causes interdigitation into the bony interstices and, when fully polymerized, the now hard compound prevents all movement between prosthesis and bone. It can withstand large compressive loads but is easily broken by tensile stress.

Cement mixing and cement introduction techniques have been shown to influence the tensile strength. An almost 50% increase in tensile strength can be obtained by vacuum mixing or centrifugation of the mixture prior to application; this reduces the number of voids within the mixture. Additionally, cleaning of the bone and pressurization of the cement within the bone cavity, prior to introduction of the implant, improves interdigitation of the cement into interstices of the bone surface. When the partially polymerized cement is forced into the bone there is often a drop in blood pressure; this is attributed to the uptake of residual monomer, which can cause peripheral vasodilatation, and fat embolization from the bone marrow (bone cement implantation syndrome – BCIS). This is seldom a problem in fit patients with osteoarthritis, but in elderly patients undergoing treatment for hip fracture, monomer and marrow fat may enter the circulation very rapidly when the cement is compressed, and the fall in blood pressure can be alarming (and occasionally fatal), particularly in a dehydrated under-resuscitated patient.

If the initial application of the cement is not perfect, a fibrous layer can form at the cement/bone interface, its thickness depending on the degree of cement penetration into the bone crevices. In this flimsy membrane fine granulation tissue and foreign body giant cells can be seen. This relatively quiescent tissue remains unchanged under a wide range of biological and mechanical conditions, but if there is excessive movement at the cement/bone interface, or if polyethylene or metallic wear products track down into the cement/bone interface, an aggressive reaction ensues that produces bone resorption and disintegration of the interlocking surface; occasionally this is severe enough to justify the term 'aggressive granulomatosis' or 'aggressive osteolysis'. Bone resorption and cement loosening may also be associated with low-grade infection, which can manifest for the first time

many years after the operation; whether the infection in these cases precedes the loosening or vice versa is still not known.

HYDROXYAPATITE

The mineral phase of bone exists largely in the form of crystalline hydroxyapatite (HA). It is not surprising, therefore, that this material has been used to reproduce the osteoinductive and osteoconductive properties of bone grafts. Porous hydroxyapatite obtained from coral exoskeleton is rapidly incorporated in living bone and synthetic implants consisting of hydroxyapatite and tricalcium phosphate are commercially available as bone graft substitutes (see earlier). HA can also be plasma sprayed onto implants; the HA coating is a highly acceptable substrate for bone cells and promotes rapid osseointegration. This technique has found a place in the use of uncemented joint replacement prostheses and with external fixator pins.

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Section 2

Regional Orthopaedics

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The shoulder and pectoral girdle

13

Andrew Cole

CLINICAL ASSESSMENT

Symptoms

Pain is the commonest symptom. However, 'pain in the shoulder' is not necessarily 'shoulder pain'! If the patient points to the top of the shoulder, think of the acromioclavicular joint, or referred pain from the neck. Pain from the shoulder joint and the rotator cuff is felt, typically, over the front and outer aspect of the joint, often as far down as the middle of the arm. The relationship to posture may be significant: pain which appears when the arm is in the 'window-cleaning' position is characteristic of rotator cuff impingement; pain which comes on suddenly when the arm is held high overhead suggests instability.

Beware the trap of *referred pain*. Mediastinal disorders, including cardiac ischaemia, can present with aching in either shoulder.

Weakness may appear as a true loss of power, suggesting a neurological disorder, or as a sudden and surprising inability to abduct the shoulder – perhaps due to a tendon rupture. Between these extremes there is weakness in performing only certain movements and weakness associated with pain.

Instability symptoms may be gross and alarming ('my shoulder jumps out of its socket when I raise my arm'); more often they are quite subtle: a click or jerk when the arm is held overhead, or the 'dead arm' sensation that overtakes the tennis player as he or she prepares to serve.

Stiffness may be progressive and severe – so much so as to merit the term 'frozen shoulder'.

Swelling may be in the joint, the muscle or the bone; the patient will not know the difference.

Deformity may consist of muscle wasting, prominence of the acromioclavicular joint, winging of the scapula or an abnormal position of the arm.

Loss of function is usually expressed as difficulty with dressing and grooming, or inability to lift objects or work with the arm above shoulder height.

Signs

The patient should always be examined from in front and from behind. Both upper limbs, the neck, the outline of the scapula and the upper chest must be visible.

Look

Skin Scars or sinuses are noted; do not forget the axilla!

Shape The two sides should be compared. Asymmetry of the shoulders, winging of the scapula, wasting of the deltoid, supraspinatus and infraspinatus muscles and acromioclavicular dislocation are best seen from behind; swelling of the acromioclavicular or sternoclavicular joint or wasting of the pectoral muscles is more obvious from the front. A joint effusion causes swelling anteriorly and occasionally 'points' in the axilla. Wasting of the deltoid suggests a nerve

BOX 13.1 THE PAINFUL SHOULDER

Referred pain syndromes
- Cervical spondylosis
- Mediastinal pathology
- Cardiac ischaemia

Joint disorders
- Glenohumeral arthritis
- Acromioclavicular arthritis

Bone lesions
- Infection
- Tumours

Rotator cuff disorders
- Tendinitis
- Rupture
- Frozen shoulder

Instability
- Dislocation
- Subluxation

Nerve injury
- Suprascapular nerve entrapment

lesion whereas wasting of the supraspinatus may be due to either a full-thickness tear or a suprascapular nerve lesion. The typical 'Popeye' bulge of a ruptured biceps is more easily seen if the elbow is flexed.

Position If the arm is held internally rotated, think of posterior dislocation of the shoulder.

Feel

Skin Because the joint is well covered, inflammation rarely influences skin temperature.

Bony points and soft tissues The deeper structures are carefully palpated, following a mental picture of the anatomy. Start with the sternoclavicular joint, then follow the clavicle laterally to the acromioclavicular joint, and so onto the anterior edge of the acromion and around the acromion. The anterior and posterior margins for the glenoid should be palpated. With the shoulder held in extension, the supraspinatus tendon can be pinpointed just under the anterior edge of the acromion; below this, the bony prominence bounding the bicipital groove is easily felt, especially if the arm is gently rotated so that the hard ridge slips medially and laterally under the palpating fingers. Crepitus over the supraspinatus tendon during movement suggests tendinitis or a tear.

Move

Active movements Movements are observed first from in front and then from behind, with the patient either standing or sitting (Figure 13.1). Sideways elevation of the arms normally occurs in the plane

Figure 13.1 Examination Active movements are best examined from behind the patient, paying careful attention to symmetry and the coordination between scapulothoracic and glenohumeral movements. (a) Abduction; (b) limit of glenohumeral abduction; (c) full abduction and elevation, a combination of scapulothoracic and glenohumeral movement. (d) The range of true glenohumeral movement can be assessed by blocking scapular movement with a hand placed firmly on the top edge of the scapula. (e) External rotation. (f,g) Complex movements involving abduction, rotation and flexion or extension of the shoulder. (h) Testing for serratus anterior weakness. (i) Feeling for supraspinatus tenderness.

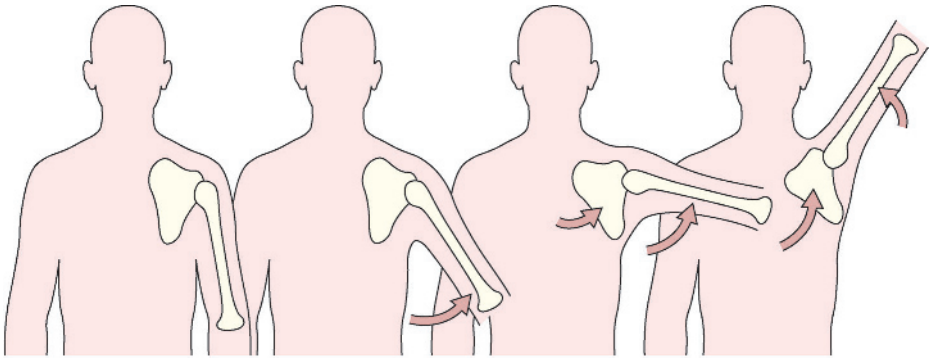


Figure 13.2 Scapulohumeral rhythm (a–c) During the early phase of abduction, most of the movement takes place at the glenohumeral joint. As the arm rises, the scapula begins to rotate on the thorax (c). In the last phase of abduction, movement is almost entirely scapulothoracic (d).

of the scapula, i.e. about 20 degrees anterior to the coronal plane, with the arm rising through an arc of 180 degrees. However, by convention, abduction is performed in the coronal plane and flexion–extension in the sagittal plane.

Abduction starts at 0 degrees; the early phase of movement takes place almost entirely at the glenohumeral joint, but as the arm rises the scapula begins to rotate on the thorax and in the last 60 degrees of movement is almost entirely scapulothoracic (hence sideways movement beyond 90 degrees is sometimes called 'elevation' rather than 'abduction') (Figure 13.2). The rhythmic transition from glenohumeral to scapulothoracic movement is disturbed by disorders in the joint or by dysfunction of the stabilizing tendons around the joint. Thus, abduction may be (1) difficult to initiate, (2) diminished in range or (3) altered in rhythm, the scapula moving too early and creating a shrugging effect. If movement is painful, the arc of pain must be noted; pain in the midrange of abduction suggests a minor rotator cuff tear or supraspinatus tendinitis; pain at the end of abduction is often due to acromioclavicular arthritis.

Flexion and extension are examined by asking the patient to raise the arms forwards and then backwards. The normal range is 180 degrees of flexion and 40 degrees of extension.

Rotation is tested in two ways: the arms are held close to the body with the elbows flexed to 90 degrees; the hands are then separated as widely as possible (external rotation) and brought together again across the body (internal rotation). This is a rather unnatural movement and one learns more by simply asking the patient to clasp his (or her) fingers behind his neck (external rotation in abduction) and then to reach up his back with his fingers (internal rotation in adduction); the two sides are compared. See Figure 13.3.

Passive movements To test the range of glenohumeral movement (as distinct from combined glenohumeral and scapular movement) the scapula must first be anchored; this is done by the examiner pressing firmly down on the top of the shoulder with one hand while the other hand moves the patient's arm.

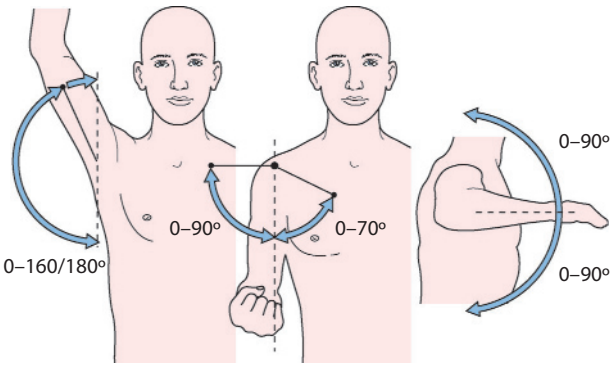


Figure 13.3 Normal range of movement (a) Abduction is from 0° to 160° (or even 180°), but only 90° of this takes place at the glenohumeral joint (in the plane of the scapula, 20° anterior to the coronal place); the remainder is scapular movement. (b) External rotation is usually about 80° but internal is rather less because the trunk gets in the way. (c) With the arm abducted to a right angle, internal rotation can be assessed without the trunk getting in the way.

Grasping the angle of the scapula as a method of anchorage is less satisfactory.

Power The deltoid is examined for bulk and tautness while the patient abducts against resistance. To test serratus anterior (long thoracic nerve, C5, 6, 7) the patient is asked to push forcefully against a wall with both hands; if the muscle is weak, the scapula is not stabilized on the thorax and stands out prominently (winged scapula). Pectoralis major is tested by having the patient thrust both hands firmly into the waist. Rotator power is tested by asking the patient to stand with his arms tucked into his side and the elbows flexed, then to externally rotate against resistance. Weakness may be associated with a rotator cuff lesion, instability or a neurological disorder.

Other systems Clinical assessment is completed by examining the cervical spine (as a common source of referred pain), testing for generalized joint laxity (a frequent accompaniment of shoulder instability) and performing a focused neurological examination.

Special clinical tests

Special clinical tests have been developed for localizing more precisely the site of pain and tenderness, the source of muscle weakness and the presence of instability. These are described in the relevant sections that follow.

Examination after local anaesthetic injection

It is sometimes possible to localize the source of shoulder pain by injecting local anaesthetic into the target site (for example, the supraspinatus tendon or the acromioclavicular joint) and thus to see whether there is a temporary reduction in pain on movement. Injection into the subacromial space may help to distinguish loss of movement due to pain from that due to a rotator cuff tear.

Diagnostic focus

Important as it is to adopt a systematic approach in the clinical examination, the practical exercise of working towards a diagnosis requires a sensible balance in the focus of attention. A young athletic person who develops pain and weakness on abduction and external rotation of the shoulder is more likely to be suffering from a rotator cuff disorder or instability than an inflammatory arthritis of the shoulder and therefore the full panoply of special tests for localization of pain and weakness would be justified, whereas some of these tests would be quite inappropriate in an elderly person with the long-standing pain and swelling of an arthritic condition.

Imaging

X-rays

At least two X-ray views should be obtained: an anteroposterior in the plane of the shoulder and an axillary projection with the arm in abduction to show the relationship of the humeral head to the glenoid (Figure 13.4a–c). Look for evidence of subluxation or dislocation, joint space narrowing, bone erosion and calcification in the soft tissues. The acromioclavicular joint is best shown by an anteroposterior projection with the tube tilted upwards 20 degrees (the cephalic tilt view). The subacromial space is viewed by tilting the tube downwards 30 degrees (the caudal tilt view).

Arthrography

This is useful for detecting rotator cuff tears and some larger Bankart lesions found with anterior instability. It is now usually combined with computed tomography (CT) or magnetic resonance imaging (MRI).

Computed tomography

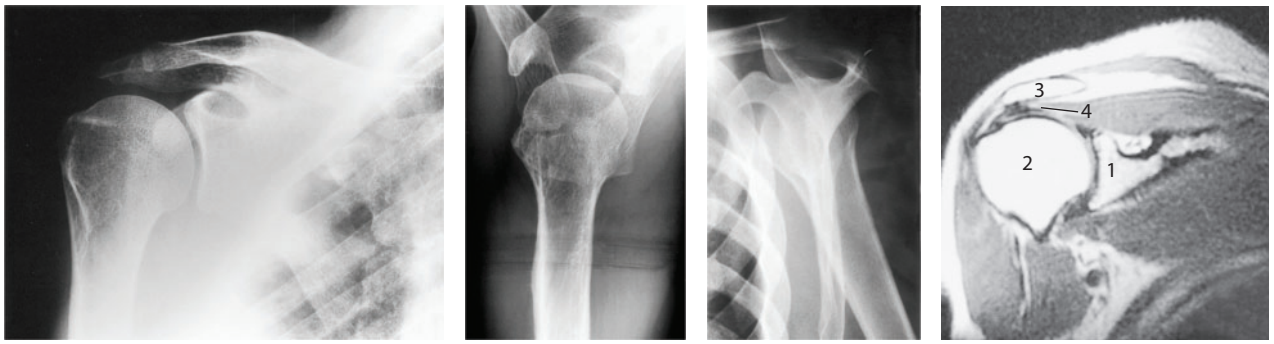
CT scans are particularly helpful for surgical planning, especially for shoulder replacement or fracture surgery. They can identify intra-articular pathology such as rotator cuff tears and labral detachments when enhanced with arthrography, but they have largely been replaced by magnetic resonance arthrography (MRA) for this purpose.

Ultrasound

In experienced hands, ultrasound provides a reliable and simple means of identifying rotator cuff tears, calcific tendinitis and biceps problems. It can also be useful to identify areas of hypervascularity and perform ultrasound-guided injections and barbotage (the practice of inserting a needle into a calcific deposit and aspirating or fragmenting the material).

Magnetic resonance imaging

The information which is provided by MRI depends on the quality of the equipment and the imaging



(a) (b) (c) (d)

Figure 13.4 Imaging (a) Anteroposterior X-ray. (b) Axillary view showing the humeral head opposite the shallow glenoid fossa, and the coracoid process anteriorly. The acromion process shadow overlaps that of the humeral head. (c) Lateral view; the head of the humerus should lie where the coracoid process, the spine of the scapula and the blade of the scapula meet. (d) MRI. Key: 1 glenoid; 2 head of the humerus; 3 acromion process; 4 supraspinatus (with degeneration of the tendon).

sequences which are chosen (Figure 13.4d). For patients with suspected rotator cuff pathology, MRI gives information on the site and size of a tear, the degree of fatty infiltration as well as the anatomy of the coracoacromial arch and acromioclavicular joint. For patients with symptoms and signs suggesting instability, it can demonstrate associated anomalies of the capsule, labrum, glenoid and humeral head, particularly when used with arthrography. MRI is also useful in detecting osteonecrosis of the head of the humerus and in the diagnosis and staging of tumours.

Magnetic resonance arthrography

MR arthrography (MRA) has a sensitivity and specificity of over 90% in the detection of pathological labral conditions. For identifying rotator cuff partial undersurface tears, MRA is more sensitive and specific than MRI alone.

Arthroscopy

Arthroscopy can be useful to diagnose (and treat) intra-articular lesions, detachment of the labrum or capsule and impingement or tears of the rotator cuff. Arthroscopy is said to be the best means by which superior labrum, anterior and posterior (SLAP) tears may be diagnosed.

DISORDERS OF THE ROTATOR CUFF

Overview

The rotator cuff comprises the lateral portions of the infraspinatus, teres minor, supraspinatus and subscapularis muscles and their conjoint tendon which is inserted into the greater and lesser tuberosity of the humerus. The teres minor inserts into the lower facet of the greater tuberosity, the infraspinatus to the middle and the supraspinatus to the superior facet. The subscapularis inserts into the lesser tuberosity. The musculotendinous cuff passes beneath the coracoacromial arch, (formed by the undersurface of the acromium and the coracoacromial ligament) from which it is separated by the subacromial bursa; during abduction of the arm the cuff slides outwards under the arch. The deep surface of the cuff is intimately related to the joint capsule and the tendon of the long head of the biceps (Figure 13.5).

Although contraction of the individual muscles that make up the rotator cuff exerts a rotational pull on the proximal end of the humerus, the other function of it is to draw the head of the humerus firmly into the glenoid socket and stabilize it there when the deltoid muscle contracts and abducts the arm. This is particularly performed by the supraspinatus and infraspinatus. The subscapularis acts as a restraint to anterior translation. Consequently, patients with rotator cuff tendinitis experience pain and weakness on active abduction and those with a severe tear of the cuff are unable to initiate abduction but may be able to hold the arm abducted once it has been raised aloft by the examiner.

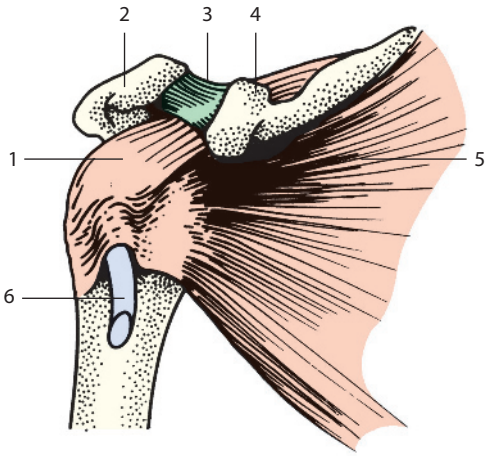


Figure 13.5 Anatomy The tough coracoacromial ligament stretches from the coracoid to the underside of the anterior third of the acromion process; the humeral head moves beneath this arch during abduction and the rotator cuff may be irritated or damaged as it glides in this confined space.
Key: 1 Rotator cuff; 2 acromion process; 3 coracoacromial ligament; 4 coracoid process; 5 subscapularis; 6 long head of biceps (LHB).

The commonest cause of pain around the shoulder is a disorder of the rotator cuff. This is sometimes referred to rather loosely as '*rotator cuff syndrome*', which comprises at least four conditions with distinct clinical features and natural history:

- supraspinatus impingement and tendinitis
- tears of the rotator cuff
- acute calcific tendinitis
- biceps tendinitis and/or rupture.

In all these conditions the patient is likely to complain of pain and/or weakness during certain movements of the shoulder. Pain may have started recently, sometimes quite suddenly, after a particular type of exertion; the patient may know precisely which movements now reignite the pain and which to avoid, providing a valuable clue to its origin. 'Rotator cuff' pain typically appears over the front and lateral aspect of the shoulder during activities with the arm abducted and medially rotated, but it may be present even with the arm at rest. Tenderness is felt at the anterior edge of the acromion.

Pain and tenderness directly in front along the delto-pectoral boundary could be associated with the biceps tendon. Localized pain over the top of the shoulder is more likely to be due to acromioclavicular pathology, and pain at the back along the scapular border may come from the cervical spine. All these sites should be inspected for muscle wasting, carefully palpated for local tenderness and constantly compared with the opposite shoulder.

If there is weakness with some movements but not with others, then one must rule out a partial or complete tendon rupture; here again, as with pain, localization to a specific site is the key to diagnosis. In both cases clinical examination should include a number of provocative tests to determine the source of the patient's symptoms. These are described in the relevant sections below.

IMPINGEMENT SYNDROME, SUPRASPINATUS TENDINITIS AND CUFF DISRUPTION

Pathology

Rotator cuff impingement is a painful disorder which is thought to arise from repetitive compression or rubbing of the tendons (mainly supraspinatus) under the coracoacromial arch (Figure 13.6). Normally, when the arm is abducted, the conjoint tendon slides under the coracoacromial arch. As abduction approaches 90 degrees, there is a natural tendency to externally rotate the arm, thus allowing the rotator cuff to occupy the widest part of the subacromial space. If the arm is held persistently in abduction and then moved to and fro in internal and external rotation (as in cleaning a window, painting a wall or polishing a flat surface), the rotator cuff may be compressed and irritated as it comes in contact with the anterior edge of the acromion process and the taut coracoacromial ligament. This attitude (abduction, slight flexion and internal rotation) has been called the 'impingement position'. Perhaps significantly, the site of impingement is also the 'critical area' of diminished vascularity in the supraspinatus tendon about 1 cm proximal to its insertion into the greater tuberosity.

The development of impingement is thought to be due to intrinsic and extrinsic factors. Intrinsic factors include degeneration of the tendon, changes in the presence of highly sulphated glycosaminoglycans and changes in the collagen composition with loading. Tendon degeneration may be age-related and a cell-mediated response. Changes in vascularity may also contribute. It is thought that these intrinsic changes result in rotator cuff dysfunction resulting in upward displacement of the humeral head and the subsequent development of extrinsic compression. It can also occur in inflammatory conditions such as gout or rheumatoid arthritis.

Extrinsic factors that may cause impingement include spurs growing down the coracoacromial

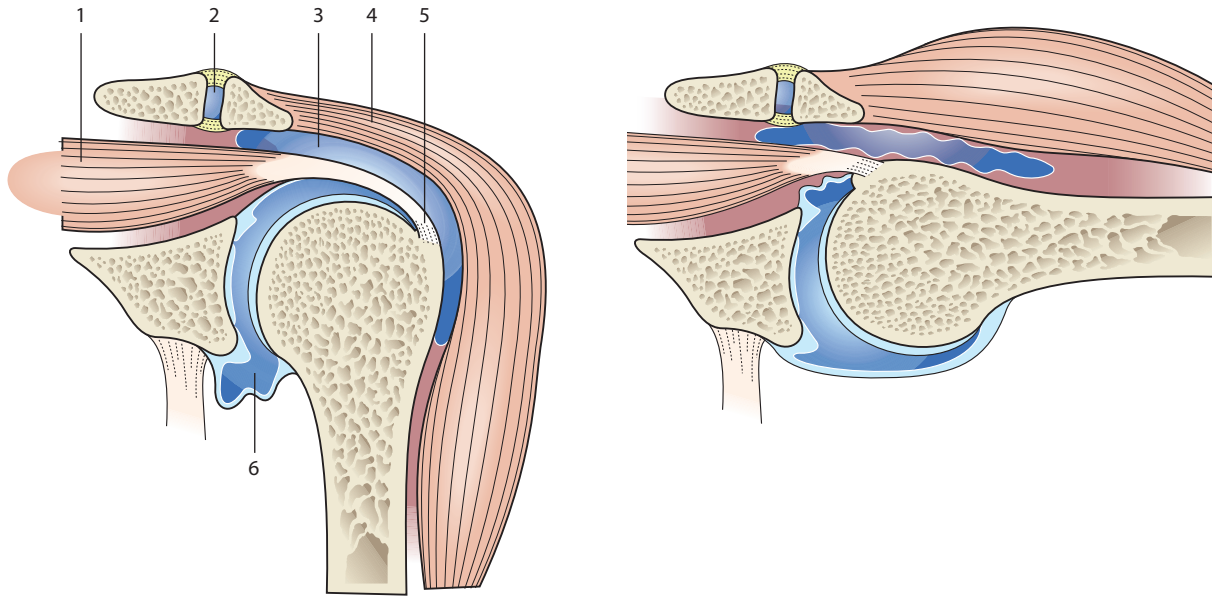


Figure 13.6 Rotator cuff impingement Coronal sections through the shoulder to illustrate show how the subdeltoid bursa and supraspinatus tendon can be irritated by repeated impingement under the coracoacromial arch or a rough acromioclavicular joint during abduction. (a) Joint at rest. (b) In abduction. Key: 1 Supraspinatus muscle; 2 acromioclavicular joint; 3 subdeltoid bursa; 4 deltoid muscle; 5 supraspinatus tendon; 6 synovial joint.

ligament, a laterally sloping acromium, and osteoarthritic thickening of the acromioclavicular joint. In 1986, Bigliani and Morrison described three variations of acromial morphology:

- *Type I* – flat
- *Type II* – curved
- *Type III* – hooked.

They suggested that the type III variety was most frequently associated with impingement and rotator cuff tears.

The mildest injury is a type of friction, which may give rise to localized oedema and swelling ('tendinitis'). This is usually self-limiting, but with prolonged or repetitive impingement – and especially in older people – minute tears can develop and these may be followed by scarring, fibrocartilaginous metaplasia or calcification in the tendon. Healing is accompanied by a vascular reaction and local congestion (in itself painful) which may contribute to further impingement in the constricted space under the coracoacromial arch whenever the arm is elevated.

Sometimes – perhaps where healing is slow or following a sudden strain – the microscopic disruption extends, becoming a partial or full-thickness tear of the cuff; shoulder function is then more seriously compromised and active abduction may be impossible.

The tendon of the long head of biceps (LHB), lying adjacent to the supraspinatus, may also be involved and is often torn.

Secondary arthropathy Large tears of the cuff may eventually lead to serious disturbance of shoulder mechanics. The humeral head migrates upwards, abutting against the acromion process, and passive abduction is severely restricted. Abnormal movement predisposes to osteoarthritis of the glenohumeral joint. Occasionally this progresses to a rapidly destructive arthropathy – the so-called 'Milwaukee shoulder' (named after the city where it was first described by McCarty).

Clinical features

Early clinical features are typically those of a 'rotator cuff syndrome', as described above. Subsequent progress depends on the stage of the disorder, the age of the patient and the vigour of the healing response. Three patterns are encountered:

1 *Subacute tendinitis* – the 'painful arc syndrome', due to vascular congestion, microscopic haemorrhage and oedema
2 *Chronic tendinitis* – recurrent shoulder pain due to tendinitis and fibrosis
3 *Cuff disruption* – recurrent pain, weakness and loss of movement due to tears in the rotator cuff.

SUBACUTE TENDINITIS (PAINFUL ARC SYNDROME)

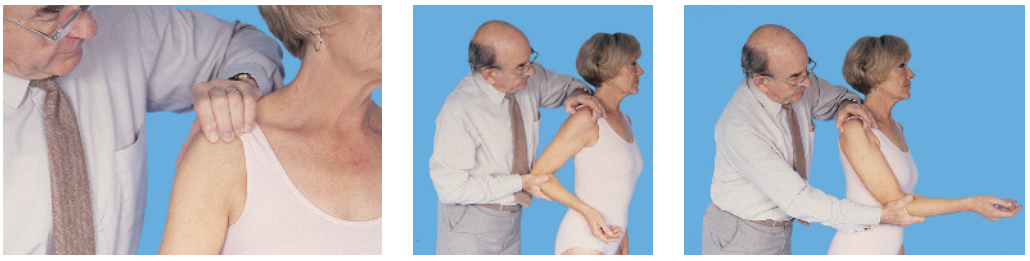
The patient develops anterior shoulder pain after vigorous or unaccustomed activity, such as competitive swimming or a weekend of house decorating. The shoulder looks normal but may be tender along the anterior edge of the acromion. Point tenderness is most easily elicited by palpating this spot with the arm held in extension, thus placing the supraspinatus tendon in an exposed position anterior to the acromion process; with the arm held in flexion the tenderness disappears (Figure 13.7).

Subacute tendinitis is often reversible, settling down gradually once the initiating activity is avoided.

CHRONIC TENDINITIS

The patient, usually aged between 40 and 50, gives a history of recurrent attacks of subacute tendinitis, the pain settling down with rest or anti-inflammatory treatment, only to recur when more demanding activities are resumed.

Characteristically pain is worse at night; the patient cannot lie on the affected side and often finds it more comfortable to sit up out of bed. Pain and slight stiffness of the shoulder may restrict even simple activities such as hair grooming or dressing. The physical signs described above should be elicited. In addition, there may be signs of bicipital tendinitis: tenderness along



(a) (b) (c)

Figure 13.7 Supraspinatus tenderness (a) The tender spot is at the anterior edge of the acromion process. When the shoulder is extended (b), tenderness is more marked; with the shoulder slightly flexed (c) the painful tendon disappears under the acromion process and tenderness disappears.

the bicipital groove and crepitus on moving the biceps tendon.

A disturbing feature is coarse crepitation or palpable snapping over the rotator cuff when the shoulder is passively rotated; this may signify a partial tear or marked fibrosis of the cuff. Small, unsuspected tears are quite often found during arthroscopy or operation.

CUFF DISRUPTION

The most advanced stage of the disorder is progressive fibrosis and disruption of the cuff, resulting in either a partial or full-thickness tear. The patient is usually aged over 45 and gives a history of refractory shoulder pain with increasing stiffness and weakness.

Partial tears may occur within the substance or on the deep surface of the cuff and are not always easily detected, even on direct inspection of the cuff. They are also deceptive in that continuity of the remaining cuff fibres permits active abduction with a painful arc, making it difficult to tell whether chronic tendinitis is complicated by a partial tear.

A *full-thickness tear* may follow a long period of chronic tendinitis, but occasionally it occurs spontaneously after a sprain or jerking injury of the shoulder. There is sudden pain and the patient is unable to abduct the arm. Passive abduction also may, in the early stages, be limited or prevented by pain. If the diagnosis is in doubt, pain can be eliminated by injecting a local anaesthetic into the subacromial space. If active abduction is now possible, the tear must be only partial. If active abduction remains impossible, then a complete tear is likely.

If some weeks have elapsed since the injury, the two types are more easily differentiated. With a complete tear, pain has by then subsided and the clinical picture is unmistakable: active abduction is impossible and attempting it produces a characteristic shrug; but passive abduction is full and, once the arm has been lifted above a right angle, the patient can keep it up by using his deltoid (the 'abduction paradox'); when the patient lowers it sideways it suddenly drops (the 'drop arm sign').

With time there may be some recovery of active abduction, though power in both abduction and external rotation is weaker than normal. There is usually wasting of the supraspinatus and infraspinatus, and on testing the biceps there may be an old tear of the long head tendon. There is often tenderness of the acromioclavicular joint. In long-standing cases of partial or complete rupture, secondary osteoarthritis of the shoulder may supervene and movements are then severely restricted (Figure 13.8).

Tests for cuff impingement pain

The painful arc On active abduction, scapula humeral rhythm is disturbed and pain is aggravated as the arm traverses an arc between 60 and 120 degrees. Repeating the movement with the arm in full external rotation may be much easier for the patient and relatively painless (Figure 13.9).

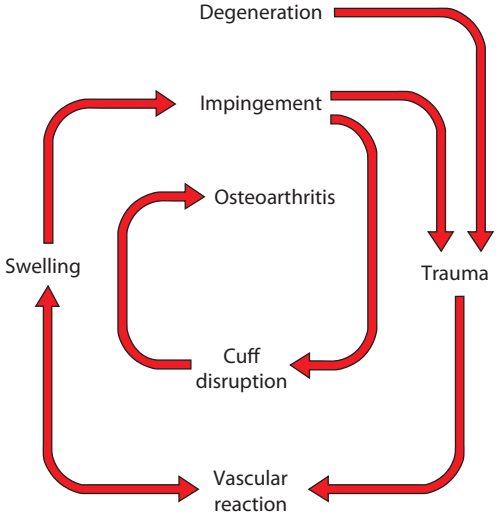
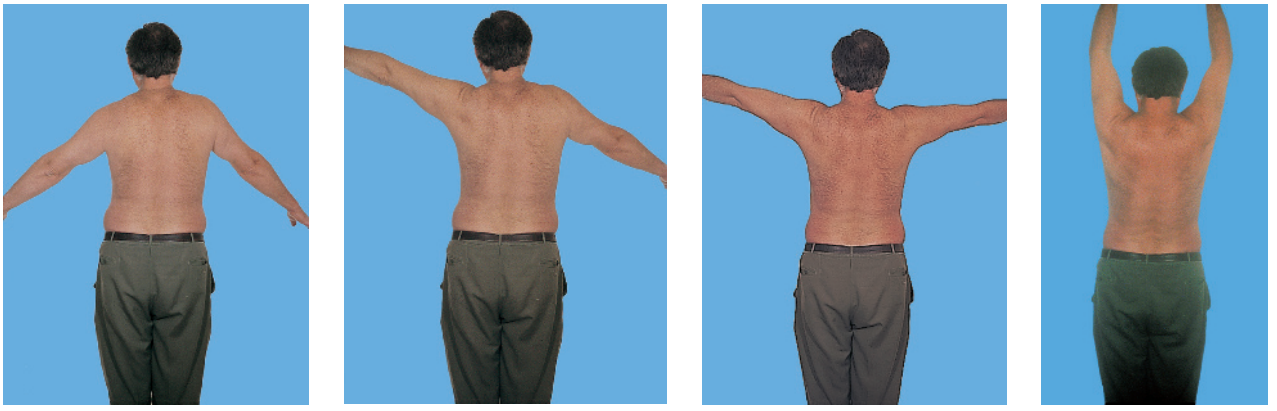


Figure 13.8 The vicious spiral of rotator cuff lesions



(a) (b) (c) (d)

Figure 13.9 The painful arc (a,b) In abduction, scapulohumeral rhythm is disturbed on the right and the patient starts to experience pain at about 60°. (c,d) As the arm passes beyond 120° the pain eases and the patient is able to abduct and elevate up to the full 180°.

Neer's impingement sign The scapula is stabilized with one hand while with the other hand the examiner raises the affected arm to the full extent in passive flexion, abduction and internal rotation, thus bringing the greater tuberosity directly under the coracoacromial arch. The test is positive when pain, located to the subacromial space or anterior edge of acromion, is elicited by this manoeuvre. The test is over 80% sensitive for subacromial impingement or a rotator cuff tear but it has poor specificity and may be positive also in patients with acromioclavicular osteoarthritis, glenohumeral instability and SLAP lesions.

Neer's test for impingement If the previous manoeuvre is positive, it may be repeated after injecting 10 mL of 1% lignocaine into the subacromial space; if the pain is abolished (or significantly reduced), this will help to confirm the diagnosis (Figure 13.10).

Hawkins–Kennedy test The patient's arm is placed in 90 degrees of elevation in the scapular plane with the elbow also flexed to 90 degrees. The examiner then stabilizes the upper arm with one hand while using the other hand to internally rotate the arm fully. Pain around the anterolateral aspect of the shoulder is noted as a positive test. As with the Neer sign, this test is highly sensitive but weakly specific.

Jobe's test The arm is elevated in the scapular plane. The elbow is extended and the thumb points to the floor in full internal rotation. The patient is then asked to hold this position with downward pressure by the examiner. Pain indicates irritation of the supraspinatus tendon (Figure 13.11a,b).

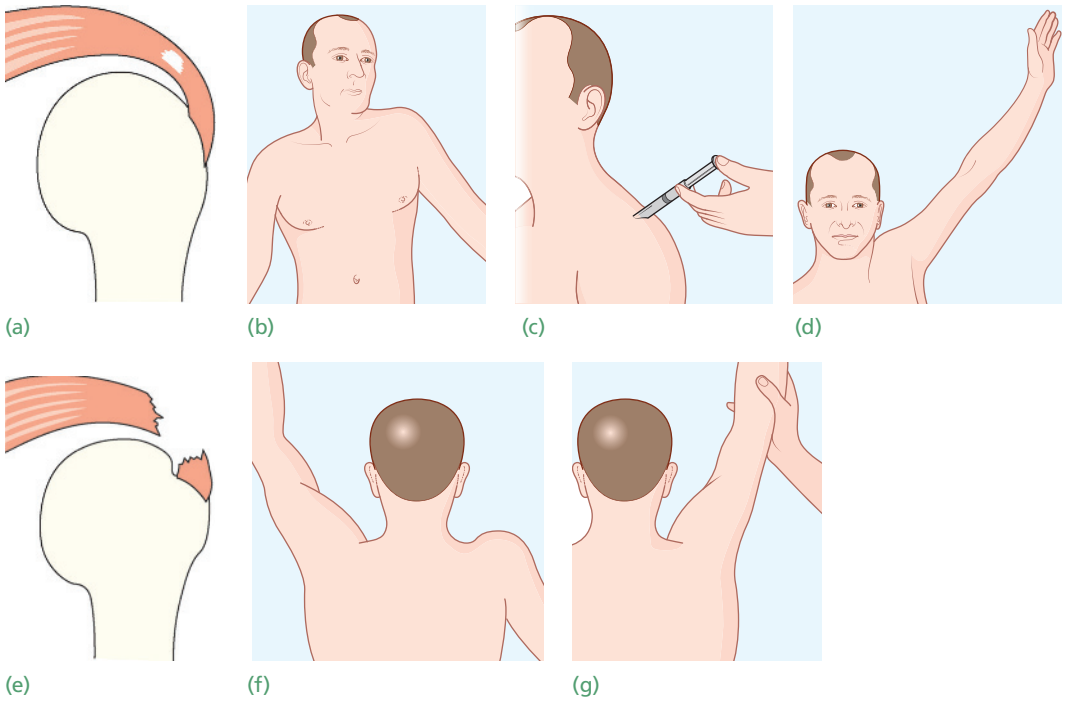


Figure 13.10 Torn supraspinatus (a–d) Partial tear of left supraspinatus: the patient can abduct actively once pain has been abolished with local anaesthetic. (e–g) Complete tear of right supraspinatus: active abduction is impossible even when pain subsides (f), or has been abolished by injection; but once the arm is passively abducted, the patient can hold it up with her deltoid muscle (g).

Figure 13.11 Tests for cuff weakness (a) Position for Jobe's test for supraspinatus power. (b) Jobe's test – right side weaker than left. (c) Test for infraspinatus – right side weaker than left. (d) Normal 'lift-off' test for subscapularis.

Tests for isolated weakness

The 'abduction paradox' and 'drop arm sign' are helpful in the diagnosis of a complete rupture of the cuff. For partial tears of the cuff, more subtle tests are used to identify weakness in isolated components of the cuff.

Supraspinatus – the 'empty can' test (Jobe) Supraspinatus strength can be tested in isolation as follows. The patient (seated or standing) is asked to raise his or her arms to a position of 90 degrees abduction, 30 degrees of forward flexion and internal rotation (thumbs pointing to the floor, as if emptying an imaginary can). The examiner stands behind the patient and applies downward pressure on both arms, with the patient resisting this force. The result is positive when the affected side is weaker than the unaffected side, suggesting a tear of the supraspinatus tendon.

Infraspinatus – resisted external rotation The patient stands holding his or her arms close to the body and the elbows flexed to 90 degrees. He or she is instructed to externally rotate both arms while the examiner applies resistance; lack of power on one side signifies weakness of infraspinatus. The test can be repeated, this time with the arm in 90 degrees of forward elevation in the plane of the scapula. The patient is asked to laterally rotate the arm against resistance; the ability to do so despite feeling pain can indicate tendinitis while an inability to resist at all suggests a tear of infraspinatus.

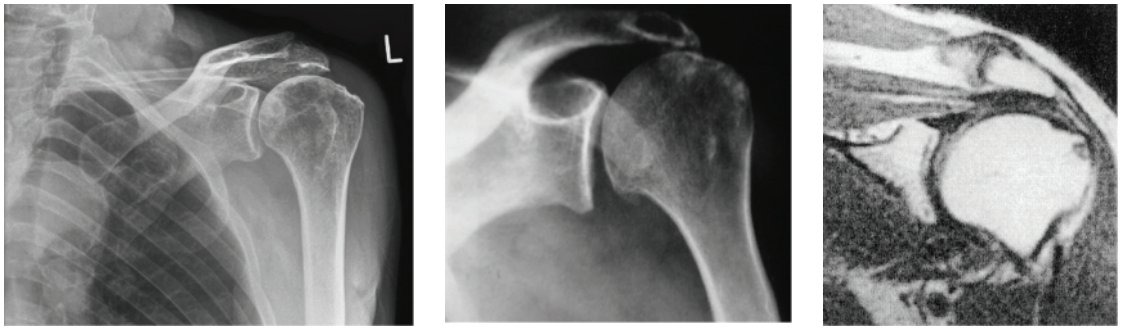
Subscapularis – belly press test The patient is asked to place their hand on their abdomen and bring the elbow forward. The patient is then asked to press their hand into their abdomen. If the subscapularis is torn, the patient will be unable to do this and the elbow will move backwards.

Infraspinatus and posterior cuff – the 'lag sign' and the 'drop sign' For the external rotation lag sign the patient's arms are lifted slightly away from the body and placed in maximum external rotation; a positive test is signalled when the patient cannot maintain that position on one side and allows the arm to drift into a more neutral position. This suggests a tear of infraspinatus or supraspinatus. The drop sign is similar: here the examiner lifts and places the arm in 90 degrees of abduction, the elbow at a right angle and the arm maximally externally rotated; when the examiner lets go, the patient would normally hold that position, but if the arm 'drops', it signals a positive test. This is seen in patients with tears of the infraspinatus and posterior part of the rotator cuff.

Subscapularis – 'the lift-off' test The patient is asked to stand and place one arm behind his or her back with the dorsum of the hand resting against the mid-lumbar spine. The examiner then lifts the patient's hand off the back and the patient is told to hold it there. Inability to do this signifies subscapularis weakness, possibly due to rupture. A drawback is that the test calls for full passive internal rotation, so it cannot be used if internal rotation is painful or restricted (Figure 13.11d).

Imaging for rotator cuff disorders

X-rays In the early stages of the cuff dysfunction, X-rays are usually normal but with chronic tendinitis there may be erosion, sclerosis or cyst formation at the site of cuff insertion on the greater tuberosity (Figure 13.12). In chronic cases the caudal tilt view may show roughening or overgrowth of the anterior edge of the acromion, thinning of the acromion process and upward displacement of the humeral head. Osteoarthritis of the acromioclavicular joint is



(a) (b) (c)

Figure 13.12 Supraspinatus tendinitis – X-rays (a) X-ray of the shoulder showing a typical thin band of sclerosis at the insertion of supraspinatus and narrowing of the subacromial space. The rest of the joint looks normal. (b) X-ray at a later stage showing upward displacement of the humeral head due to a large cuff rupture. There is almost complete loss of the subacromial space, and osteoarthritis of the glenohumeral joint. (c) MRI showing thickening of the supraspinatus and erosion at its insertion; the acromioclavicular joint is swollen and clearly abnormal.

common in older patients and in late cases the glenohumeral joint also may show features of osteoarthritis. Sometimes there is calcification of the supraspinatus, but this is usually coincidental and not the cause of pain.

Magnetic resonance imaging MRI effectively demonstrates the structures around the shoulder and gives valuable ancillary information (regarding lesions of the glenoid labrum, joint capsule or surrounding muscle or bone) (Figure 13.13). However, it should be remembered that up to a third of asymptomatic individuals have abnormalities of the rotator cuff on MRI. Changes on MRI need to be correlated with the clinical examination.

Ultrasonography The accuracy of ultrasound is comparable with MRI for identifying and measuring the size of full-thickness and partial-thickness rotator cuff tears. It has the disadvantage that it cannot identify the quality of the remaining muscle as well as MRI and cannot always be accurate in predicting the reparability of the tendons.

Conservative treatment of cuff disorders

Uncomplicated impingement syndrome (or tendinitis) is often self-limiting and symptoms settle down once the aggravating activity is eliminated. Patients should be taught ways of avoiding the 'impingement position'. Physiotherapy, including rotator cuff exercises and scapular setting, may tide the patient over the painful healing phase. A short course of non-steroidal anti-inflammatory tablets sometimes brings relief.

If all these methods fail, and before disability becomes marked, the patient should be given one or two injections of depot corticosteroid into the subacromial space. In most cases this will relieve the pain, and it is then important to persevere with protective modifications of shoulder activity for at least 6 months. Healing is slow, and a hasty return to full activity may often precipitate further attacks of tendinitis.

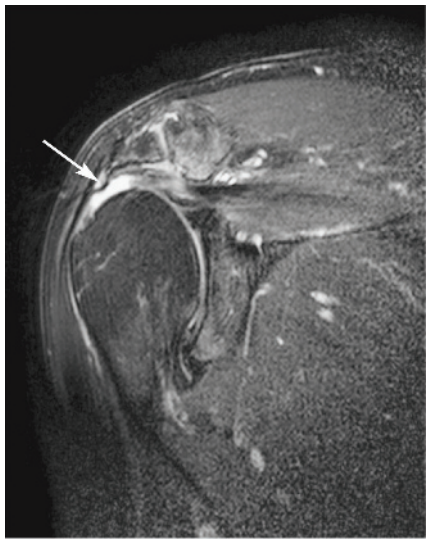


Figure 13.13 Rotator cuff tear – MRI High signal on MRI, indicating a full-thickness tear of the rotator cuff.

Surgical treatment of cuff disorders

The indications for surgical treatment are essentially clinical; the presence of a cuff tear does not necessarily call for an operation. Provided the patient has a useful range of movement, adequate strength and well-controlled pain, non-operative measures are adequate. If symptoms do not subside after 3 months of conservative treatment, or if they recur persistently after each period of treatment, an operation should be considered. Certainly this is preferable to prolonged and repeated treatment with anti-inflammatory drugs and local corticosteroids. The indication is more pressing if there are signs of a partial rotator cuff tear and in particular if there is good clinical evidence of a full-thickness tear in a younger patient. The object is to decompress the rotator cuff by excising the coracoacromial ligament, undercutting the anterior part of the acromion process and, if necessary, reducing any bony excrescences at the acromioclavicular joint (Figure 13.14). This can be achieved by open surgery or arthroscopically. The latter is technically more demanding but it can produce results equivalent to those of open surgery.

OPEN ACROMIOPLASTY

Through an anterior incision the deltoid muscle is split and the part arising from the anterior edge of the acromion is dissected free, exposing the coracoacromial ligament, the acromion and the acromioclavicular joint. The coracoacromial ligament is excised and the antero inferior portion of the acromion is removed by an undercutting osteotomy. The cuff is then inspected: if there is a defect, it is repaired. Excrescences on the undersurface of the

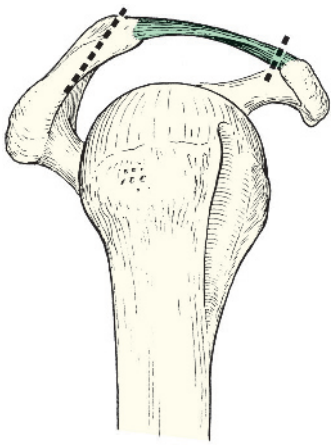


Figure 13.14 Impingement syndrome – surgical treatment The coracoacromial ligament and underside of the anterior third of the acromion are removed to enlarge the space for the rotator cuff. This can be performed by open surgery or arthroscopically.

acromioclavicular joint are pared down. If the joint is hypertrophic, the outer 1 cm of clavicle is removed; this last step exposes even more of the cuff and permits reconstruction of larger defects. An important step is careful reattachment of the deltoid to the acromion, if necessary by suturing through drill holes in the acromion; failure to obtain secure attachment may lead to postoperative pain and weakness. After the operation, shoulder movements are commenced as soon as pain subsides. This technique is now largely superceded by arthroscopic techniques.

ARTHROSCOPIC ACROMIOPLASTY

Arthroscopic acromioplasty should achieve the same basic objectives as open acromioplasty. The underside of the acromion (and, if necessary, the acromioclavicular joint) must be trimmed and the coracoacromial ligament divided or removed. If a cuff tear is encountered, then it may be possible to repair it; otherwise the edges can be debrided or an open repair undertaken.

This procedure has now become the preferred treatment and allows earlier rehabilitation than open acromioplasty because detachment of the deltoid is not performed. Arthroscopy allows good visualization inside the glenohumeral joint and therefore the detection of other abnormalities which may cause pain (present in up to 30% of patients). It allows good visualization of both sides of the rotator cuff and the identification of partial and full-thickness tears.

OPEN REPAIR OF THE ROTATOR CUFF

The indications for open repair of the rotator cuff are chronic pain, weakness of the shoulder and significant loss of function. The younger and more active the patient, the greater is the justification for surgery. The operation usually includes an acromioplasty as described above. The cuff is mobilized, if necessary by releasing the coracohumeral ligament and the glenoid attachment of the capsule; this dissection should not stray more than 2 cm medial to the glenoid rim lest the suprascapular nerve is damaged.

It may be possible to approximate the ends of the cuff defect. Larger tears can be dealt with by suturing the cuff tendon directly to a roughened area on the greater tuberosity using drill holes or soft-tissue anchors.

Postoperatively, movements are restricted for 4–6 weeks and then graded exercises are introduced. The results of open cuff repair are reasonably good, with satisfactory pain relief in about 80% of patients. This alone usually improves function, even if strength and range of movement are still restricted. Massive full-thickness tears that cannot be reconstructed are treated by subacromial decompression and debridement of degenerate cuff tissue; the relief of pain may allow reasonable abduction of the shoulder by the remaining muscles. Other methods to reconstruct irreparable tears in the younger patient include supraspinatus advancement, latissimus dorsi transfer, rotator cuff transposition, fascia lata autograft and synthetic tendon graft.

Acute rupture of the rotator cuff in patients over 70 years often becomes painless; although movement is restricted, operative repair in this age group is less reliable.

ARTHROSCOPIC ROTATOR CUFF REPAIR

Since the 1990s the repair of full-thickness tears has undergone a transition from open techniques to arthroscopically assisted (mini open) repairs and now full arthroscopic techniques. The arthroscopic instruments, suture anchors and knot tying techniques have quickly evolved to allow full arthroscopic repairs although most authors describe a steep learning curve. Advantages of the techniques include less soft-tissue damage, faster rehabilitation and a better cosmetic appearance. Double-row arthroscopic repair is now producing similar outcomes and results to open repairs.

CALCIFICATION OF THE ROTATOR CUFF

Acute calcific tendinitis

Acute shoulder pain may follow deposition of calcium hydroxyapatite crystals, usually in the 'critical zone' of the supraspinatus tendon slightly medial to its insertion, occasionally elsewhere in the rotator cuff. The condition is not unique to the shoulder, and similar lesions are seen in tendons and ligaments around the ankle, knee, hip and elbow.

The cause is unknown but it is thought that local ischaemia leads to fibrocartilaginous metaplasia and deposition of crystals by the chondrocytes. Calcification alone is probably not painful; symptoms, when they occur, are due to the florid vascular reaction which produces swelling and tension in the tendon. Resorption of the calcific material is rapid and it may soften or disappear entirely within a few weeks. Its peak incidence is in the fifth decade.

Clinical features

The condition affects 30–50 year-olds. Aching, sometimes following overuse, develops and increases in severity within hours, rising to an agonizing climax and night pain. After a few days, pain subsides and the shoulder gradually returns to normal. In some patients the process is less dramatic and recovery slower. During the acute stage the arm is held immobile; the joint is usually too tender to permit

palpation or movement. The clinical picture may resemble acute infection or gout and these conditions must be excluded.

X-rays

Calcification just above the greater tuberosity is always present (Figure 13.15). An initially well-demarcated deposit becomes more 'woolly' and then disappears.

Treatment

NON-OPERATIVE TREATMENT

Conservative treatment is successful in up to 90% of patients. The main methods are non-steroidal anti-inflammatory drugs, subacromial injection of corticosteroids, physiotherapy, extracorporeal shockwave therapy, needle aspiration and irrigation.

Non-steroidal anti-inflammatory drugs NSAIDs are the mainstay of non-operative treatment. Although corticosteroid injections are commonly used in the treatment of calcifying tendinitis, there is no conclusive evidence that they promote resorption of the calcium deposit. The efficacy of physiotherapy in the form of therapeutic ultrasound remains uncertain.

Extracorporeal shockwave therapy This therapy employs acoustic waves to induce fragmentation of the mechanically hard crystals. Its use as an alternative treatment for calcifying tendinitis has gained increasing popularity in the last few years. However, overall it seems less effective than needle barbotage or surgery.

Needle aspiration and irrigation (barbotage) Barbotage aims to drain a substantial portion of the calcium deposit, thereby stimulating cell-mediated progressive resorption. Needle aspiration can be readily carried out under local anaesthesia in the outpatient setting with ultrasound guidance. A combination of local anaesthetic and corticosteroid is used. The best results are obtained in patients with an acutely painful shoulder, typically during the resorption stage in which the calcium is of toothpaste-like consistency. There is some concern that, while the use of steroids reduces vascular and macrophage activity and reduces pain, the resorption phase may be prolonged.

OPERATIVE TREATMENT

Surgery is indicated for patients with severe disabling symptoms which have persisted for more than 6 months and are resistant to conservative treatment. The procedure involves a glenohumeral arthroscopy with special attention to the 'critical zone' of the rotator cuff. Once the calcium deposit is identified, the capsule is carefully incised from the bursal side with a knife in line with fibre orientation of the tendon; a curette is then used to milk out the toothpaste-like deposit. A subacromial decompression is also usually performed. This has been shown to be successful in around 90% in many outcome studies.

Chronic calcification

Asymptomatic calcification of the rotator cuff is common and often appears as an incidental finding in shoulder X-rays. When it is seen in association with the impingement syndrome, it is tempting to attribute the symptoms to the only obvious abnormality – supraspinatus calcification. However, the connection is spurious and treatment should be directed at the impingement lesion rather than the calcification.

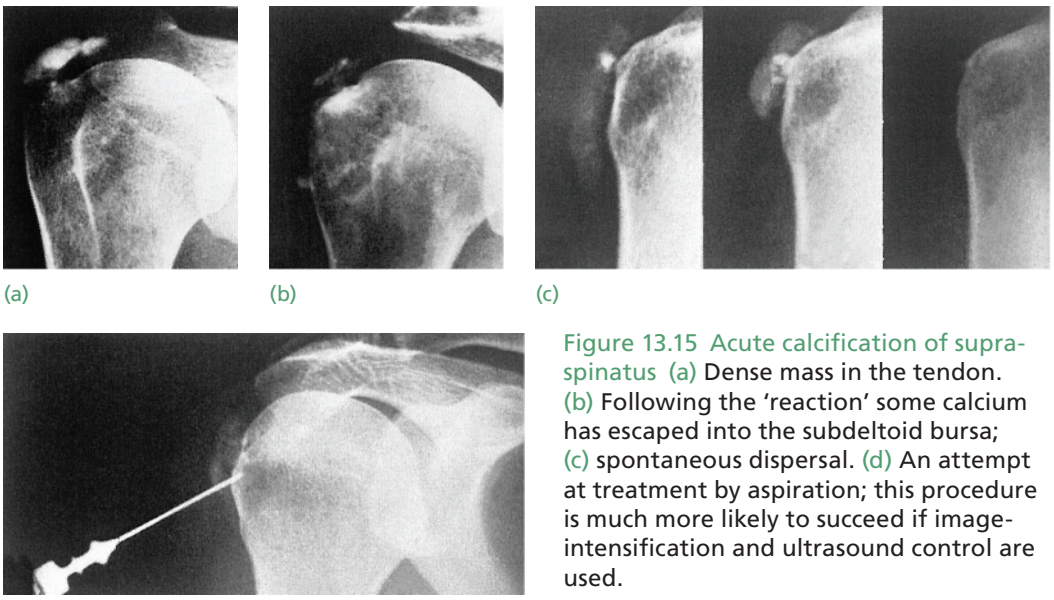


Figure 13.15 Acute calcification of supraspinatus (a) Dense mass in the tendon. (b) Following the 'reaction' some calcium has escaped into the subdeltoid bursa; (c) spontaneous dispersal. (d) An attempt at treatment by aspiration; this procedure is much more likely to succeed if image-intensification and ultrasound control are used.

LESIONS OF THE BICEPS TENDON

Long head of biceps (LHB) pathology is a common cause of shoulder pain. Clinical evaluation is often difficult. It is often associated with degenerative rotator cuff conditions and failure to treat the LHB can be a cause of continuing symptoms.

Tendinitis

The LHB is subject to tenosynovitis because of its anatomy; the tendon, which has a variable origin from the supraglenoid tubercle, has a synovial sheath and follows a constrained path in the bicipital groove.

Bicipital tendinitis usually occurs together with rotator cuff impingement; rarely, it presents as an isolated problem in young people after unaccustomed shoulder strain. Tenderness is sharply localized to the bicipital groove. Two manoeuvres that often cause pain are:

- resisted flexion with the elbow straight and the forearm supinated (*Speed's test*)
- resisted supination of the forearm with the elbow bent (*Yergason's test*).

Rest, local heat and deep transverse friction usually bring relief. If recovery is delayed, a corticosteroid injection will help. For refractory cases, a number of surgical solutions have been described including arthroscopic decompression, biceps tenotomy and biceps tenodesis.

Rupture

Rupture of the tendon of the LHB usually accompanies rotator cuff disruption, but sometimes the biceps lesion is paramount. The patient is usually aged over 50 years. While lifting, he or she feels something snap in the shoulder and the upper arm becomes painful and bruised. Ask the patient to flex the elbow: the detached belly of the biceps forms a prominent lump in the lower part of the arm (Figure 13.16).

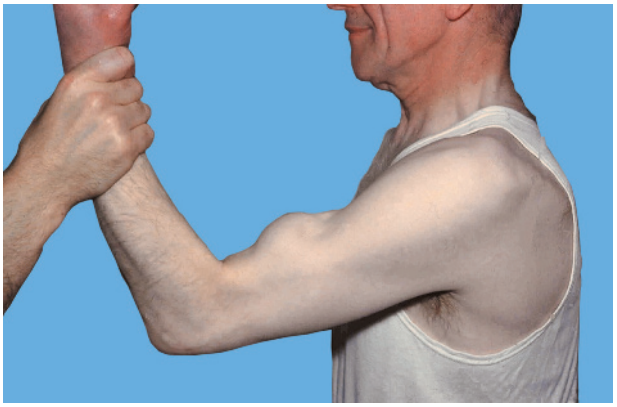


Figure 13.16 Ruptured long head of biceps The lump in the front of the arm becomes even more prominent when the patient contracts the biceps against resistance.

Isolated tears in elderly patients need no treatment, and preceding shoulder symptoms may frequently resolve after rupture. However, if the rupture is part of a rotator cuff lesion – and especially if the patient is young and active – this may be an indication for anterior acromioplasty; at the same time the distal tendon stump can be sutured to the bicipital groove (biceps tenodesis) and the rotator cuff repaired if necessary. Postoperatively the arm is lightly splinted with the elbow flexed for 4 weeks. (Avulsion of the distal attachment of the biceps is discussed in Chapter 14.)

Hypertrophy and intra-articular entrapment (the hourglass biceps)

The biceps tendon sometimes hypertrophies (for example, in association with advanced disease of the rotator cuff) and is unable to slide into the bicipital groove. This causes buckling of the tendon on elevation of the shoulder with entrapment of the tendon between the humeral head and glenoid, leading to pain and a block to terminal elevation. This may also lead to instability of the biceps.

Instability

Both subluxations and dislocations of the LHB have been described. Subluxation is defined as a partial and/or transient loss of contact between the tendon and its groove. Dislocation is the complete and permanent loss of contact between the tendon and the groove; it is usually classified into intra-articular, intra-tendinous and extra-articular subtypes. Dislocation is nearly always associated with a tear of subscapularis, except in the rare cases of extra-articular dislocation in which the tendon is resting anterior to subscapularis.

SLAP LESIONS

Compressive loading of the shoulder in the flexed abducted position (for example, in a fall on the outstretched hand) can damage the superior labrum anteriorly and posteriorly (SLAP). The injury of the superior labrum begins posteriorly and extends anteriorly, stopping before or at the mid-glenoid notch and including the 'anchor' of the biceps tendon to the labrum. Four main types are described:

1 non-traumatic superior labral degeneration, usually in older people and often asymptomatic
2 avulsion of the superior part of the labrum – the commonest type (Figure 13.17)
3 a 'bucket handle' tear of the superior labrum
4 as for type 3 with an extension into the tendon of LHB.

Further subtypes that include associated lesions have also been described.

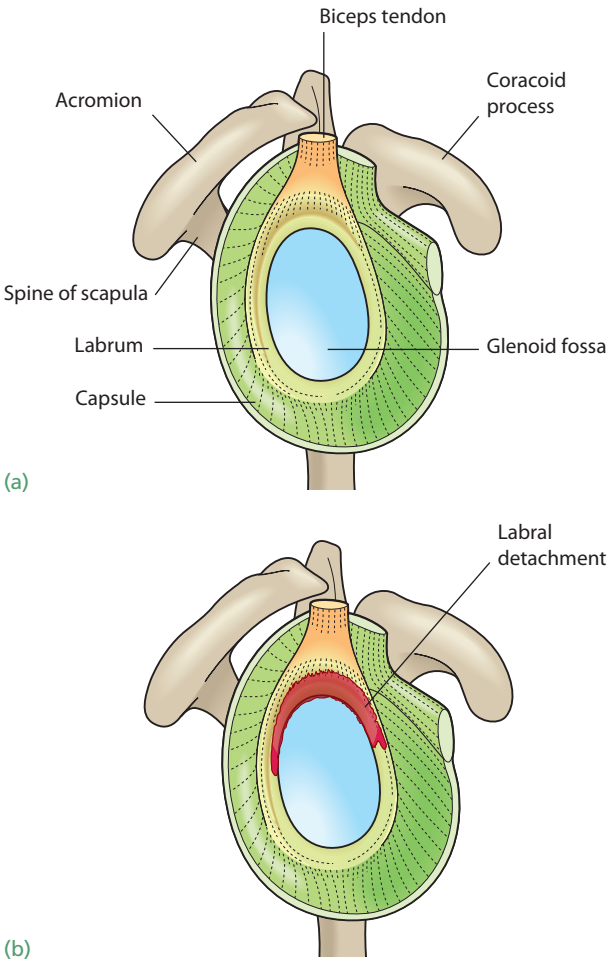


Figure 13.17 SLAP lesions (a) Diagram of the normal anatomy, looking into the glenoid fossa. Note that the biceps tendon takes its origin from the superior part of the labrum. (b) The labrum may tear or become detached from the glenoid. This illustration shows a partial tear. Other types are described in the text.

Clinical features

There is usually a history of a fall on the arm. As the initial acute symptoms settle, the patient continues to experience a painful 'click' on lifting the arm above shoulder height, together with loss of power when using the arm in that position. He or she may also complain of an inability to throw.

O'Brien's test The patient is instructed to flex the arm to 90 degrees with the elbow fully extended and then to adduct the arm 10–15 degrees medial to the sagittal plane. The arm is then maximally internally rotated and the patient resists the examiner's downward force. The procedure is repeated in supination. Pain elicited by the first manoeuvre which is reduced or eliminated by the second signifies a positive test.

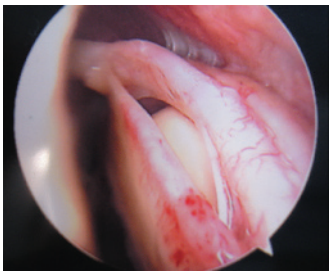


Figure 13.18 SLAP lesions Arthroscopic appearance of a type 3 SLAP lesion.

Imaging

MRI arthrography is the modality of choice though the diagnosis is best confirmed by arthroscopic examination and at the same time the lesion is treated by debridement or repair (Figure 13.18). Different lesions require different surgical procedures.

Treatment

Very few patients with SLAP lesion injuries return to full capability without surgical intervention. Arthroscopic repair of an isolated superior labral lesion is successful in the large majority (91%) of patients. However, the results in patients who participate in overhead sports are not as satisfactory as those in patients who are not involved in overhead sports. Simple lesions are simply debrided. In more significant detachments the labrum is either repaired or excised with a tenotomy or tenodesis of the biceps.

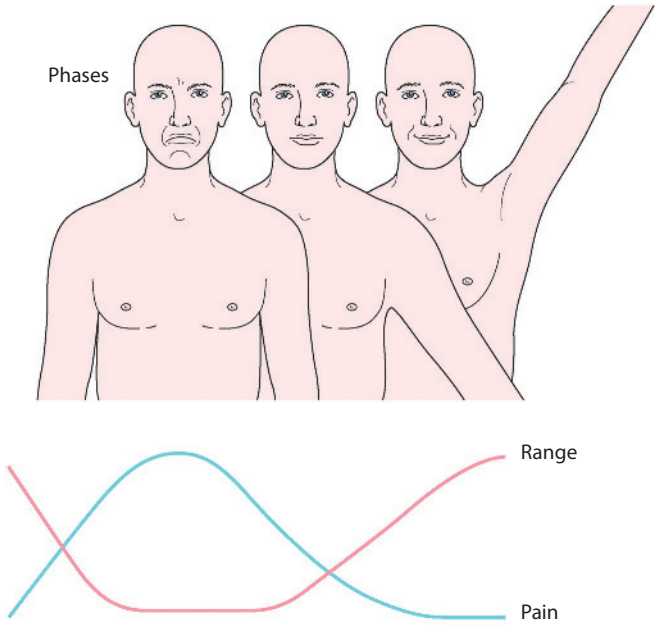
ADHESIVE CAPSULITIS (FROZEN SHOULDER)

The term 'frozen shoulder' (Figure 13.19) should be reserved for a well-defined disorder characterized by progressive pain and stiffness of the shoulder which usually resolves spontaneously after about 18 months. While the condition resolves, and many patients become asymptomatic, up to 40% may develop mild to moderate persistent symptoms. The cause remains unknown.

The histological features are similar to those of Dupuytren's disease, with active fibroblastic and myofibroblastic proliferation in the rotator interval, anterior capsule and coracohumeral ligament. The condition is particularly associated with diabetes, Dupuytren's disease, hyperlipidaemia, hyperthyroidism, cardiac disease and hemiplegia. It occasionally appears after recovery from neurosurgery.

Clinical features

The patient, aged 40–60, may give a history of trauma, often trivial, followed by aching in the arm and shoulder. Pain gradually increases in severity and



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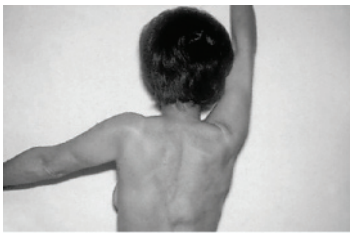

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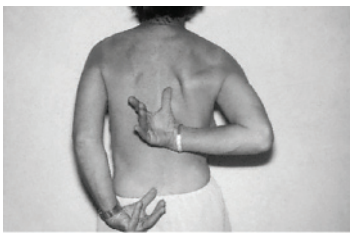


(c)

Figure 13.19 Frozen shoulder (a) Natural history of frozen shoulder. The face tells the story. (b,c) This patient has hardly any abduction but manages to lift her arm by moving the scapula. She cannot reach her back with her left hand.

often prevents sleeping on the affected side. After several months it begins to subside, but as it does so stiffness becomes an increasing problem, continuing for another 6–12 months after pain has disappeared. Gradually movement is regained, but it may not return to normal and some pain may persist.

Apart from slight wasting, the shoulder looks quite normal; tenderness is seldom marked. The cardinal feature is a stubborn lack of active and passive movement in all directions. X-rays are normal unless they show reduced bone density from disuse. Their main value is to exclude other causes of a painful, stiff shoulder.

Diagnosis

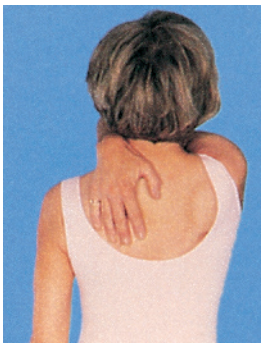
Not every stiff or painful shoulder is a frozen shoulder (Figure 13.20), and indeed the criteria for diagnosing 'frozen shoulder' are controversial. Stiffness occurs in a variety of conditions – arthritic, rheumatic, post-traumatic and postoperative. The diagnosis of frozen shoulder is clinical, resting on two characteristic features:

- painful restriction of movement in the presence of normal X-rays
- a natural progression through three successive phases.

DIFFERENTIAL DIAGNOSIS

When the patient is first seen, the following conditions should be excluded.

Infection In patients with diabetes, it is particularly important to exclude infection. During the first day or two, signs of inflammation may be absent.



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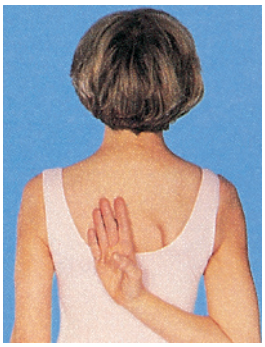

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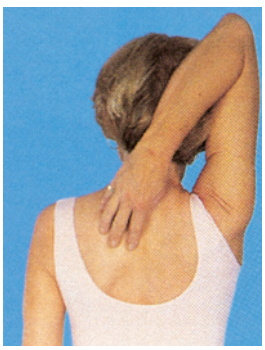


(c)

Figure 13.20 Shoulder pain – the scratch test 'Shoulder' pain may be due to disorders of the shoulder joint itself (for example, glenohumeral arthritis), the acromioclavicular joint (injury or arthritis) or structures around the joint (for example, the rotator cuff syndromes). But it could also be referred from more distant lesions (for example, brachial neuralgia, cervical spondylosis or cardiac ischaemia). If the patient can scratch the opposite scapula in these three ways (a,b,c), the shoulder joint and its tendons are unlikely to be at fault.

Post-traumatic stiffness After any severe shoulder injury, stiffness may persist for some months. It is maximal at the start and gradually lessens, unlike the pattern of a frozen shoulder.

Diffuse stiffness If the arm is nursed over-cautiously (for example, following a forearm fracture), the shoulder may stiffen. Again, the characteristic pattern of a frozen shoulder is absent.

Reflex sympathetic dystrophy Shoulder pain and stiffness may follow myocardial infarction or a stroke. The features are similar to those of a frozen shoulder and it has been suggested that the latter is a form of reflex sympathetic dystrophy. In severe cases the whole upper limb is involved, with trophic and vasomotor changes in the hand (the 'shoulder–hand syndrome').

Treatment

CONSERVATIVE TREATMENT

Conservative treatment aims to relieve pain and prevent further stiffening while recovery is awaited. It is important not only to administer analgesics and anti-inflammatory drugs but also to reassure the patient that recovery is likely.

Exercises are encouraged, the most valuable being 'pendulum' exercises. The role of *physiotherapy* is unproven and the benefits of *steroid injection* are debatable.

Manipulation under general anaesthesia may improve the range of movement. The shoulder is moved gently but firmly into external rotation, then abduction and flexion. At the end, the joint is injected with steroid and local anaesthetic. There are many studies showing rapid improvement and good pain relief. However, this procedure is not without risk. Tearing of the rotator cuff, labral injuries and fractures have been described. An alternative method of treatment is to distend the joint by injecting a large volume (50–200 mL) of sterile saline and steroid under pressure. Arthroscopy has shown that both manipulation and distension achieve their effect by rupturing the capsule.

The results of conservative treatment are subjectively good, with most patients eventually regaining painless and satisfactory function; however, examination is likely to show some residual restriction of movement (especially external rotation) in over 50% of cases.

SURGICAL TREATMENT

The main indication for surgery is prolonged and disabling restriction of movement which fails to respond to conservative treatment.

Arthroscopic capsular release is increasingly employed. New techniques enable the surgeon to release intra-articular, subacromial and subdeltoid adhesions without dividing the subscapularis. Active range of motion can be started immediately. Good results of pain relief and increased range of motion can be expected in the majority over a short time frame.

INSTABILITY OF THE SHOULDER

The shoulder achieves its uniquely wide range of movement at the cost of stability. The humeral head is held in the shallow glenoid socket by the glenoid labrum, the glenohumeral ligaments, the coracohumeral ligament, the overhanging canopy of the coracoacromial arch and the surrounding muscles. Failure of any of these mechanisms may result in instability of the joint.

One must distinguish between *joint laxity* and *joint instability*. Joint laxity implies a degree of translation in the glenohumeral joint which falls within a physiological range and which is asymptomatic. Joint instability is an abnormal symptomatic motion for that shoulder which results in pain, subluxation or dislocation of the joint.

Dislocation is defined as complete separation of the glenohumeral surfaces, whereas *subluxation* implies a symptomatic separation of the surfaces without dislocation.

Pathogenetic classification

The aetiology and classification of shoulder instability is complex, although the Stanmore Instability Classification system developed at the Royal National Orthopaedic Hospital in London is now increasingly used. It recognizes that there are two broad reasons why shoulders become unstable:

- *structural changes* due to major trauma such as acute dislocation or recurrent micro-trauma
- *unbalanced muscle recruitment* (as opposed to muscle weakness) resulting in the humeral head being displaced upon the glenoid.

From a clinical and therapeutic point of view, three polar types of disorder can be identified (Figure 13.21):

- *Type I* – traumatic structural instability
- *Type II* – atraumatic (or minimally traumatic) structural instability
- *Type III* – atraumatic non-structural instability (muscular dyskinesia).

The triangular relationship between these conditions allows for the fact there are intermediate types that lie between the 'poles'; the balance of abnormalities can shift and patients may 'move' from one type to another over time or present with a combination of pathologies (Table 13.1): for example, a purely

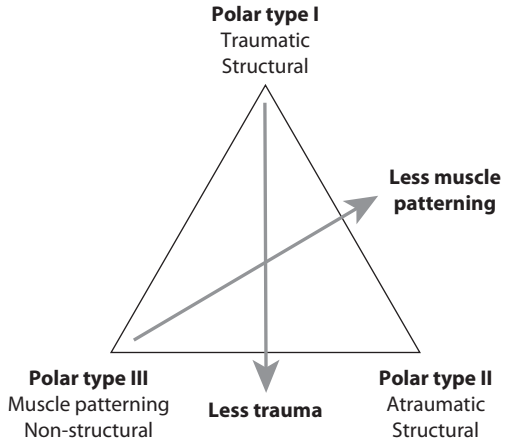


Figure 13.21 Three polar types of disorder

Table 13.1 Pathological changes in each of the polar types

Pathology	Type I	Type II	Type III
Trauma	Yes	No	No
Articular surface damage	Yes	Yes	No
Capsular problem	Bankart lesion	Dysfunctional	Dysfunctional
Laxity	Unilateral	Uni/bilateral	Often bilateral
Muscle patterning	Normal	Normal	Abnormal

structural disorder may, if allowed to persist, become associated with abnormal muscle patterning to the extent that *both* conditions need to be treated and the problems grow in complexity. The system also recognizes that there is a gradation in the opposite direction, from dyskinetic muscle patterning to structural abnormality.

TRAUMATIC ANTERIOR INSTABILITY – POLAR TYPE I

Pathology

This is far and away the commonest type of instability, accounting for over 95% of cases. Traumatic anterior instability usually follows an acute injury in which the arm is forced into abduction, external rotation and extension. In *recurrent dislocation* the labrum and capsule are often detached from the anterior rim of the glenoid (the classic Bankart lesion). In addition there may be an indentation on the posterolateral aspect of the humeral head (the Hill–Sachs lesion), a compression fracture due to the humeral head being forced against the anterior glenoid rim each time it dislocates. In some cases *recurrent subluxation* may alternate with recurrent dislocation. In other cases, the shoulder never dislocates completely and in these the labral tear and bone defect may be absent, although the inferior glenohumeral ligament will be stretched. In patients over the age of 50 years, dislocation is often associated with tears of the rotator cuff.

Clinical features

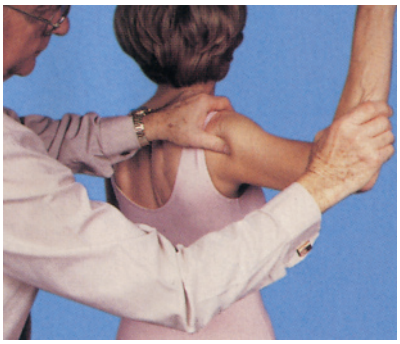
The patient is usually a young adult who gives a history of the shoulder 'coming out', perhaps during a sporting event. The first episode of *acute dislocation* is a landmark and he or she may be able to describe the mechanism precisely: an applied force with the shoulder in abduction, external rotation and extension. The diagnosis may have been verified by X-ray and the injury treated by closed reduction and 'immobilization' in a bandage or sling for several weeks. This may be the first of many similar episodes: *recurrent dislocation* requiring treatment develops in about one-third of patients under the age of 30 and in about 20% of older patients, with an overall redislocation rate of 48%. Some studies have reported instability rates following acute dislocation between 88% and 95% in patients under the age of 20. A greater proportion have instability without actual dislocation.

RECURRENT SUBLUXATION

Symptoms and signs of recurrent subluxation are less obvious. The patient may describe a 'catching' sensation, followed by 'numbness' or 'weakness' – the so-called 'dead arm syndrome' – whenever the shoulder is used with the arm in the overhead position (for example when throwing a ball, serving at tennis or swimming). Pain with the arm in abduction may suggest a rotator cuff syndrome; it is as well to remember that recurrent subluxation may actually cause supraspinatus tendinitis.

On examination, between episodes of dislocation, the shoulder looks normal and movements are full. Clinical diagnosis rests on provoking subluxation. In the *apprehension test*, with the patient seated or lying, the examiner cautiously lifts the arm into abduction, external rotation and then extension; at the crucial moment the patient senses that the humeral head is about to slip out anteriorly and his or her body tautens in apprehension (Figure 13.22). The test should be repeated with the examiner applying pressure to the front of the shoulder; with this manoeuvre, the patient feels more secure and the apprehension sign is negative.

The same effect can be demonstrated by the *fulcrum test*. With the patient lying supine, arm abducted to 90 degrees, the examiner places one hand behind the patient's shoulder to act as a fulcrum over which



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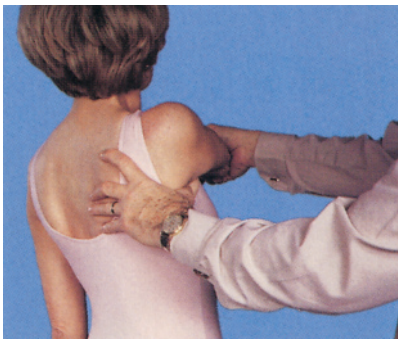
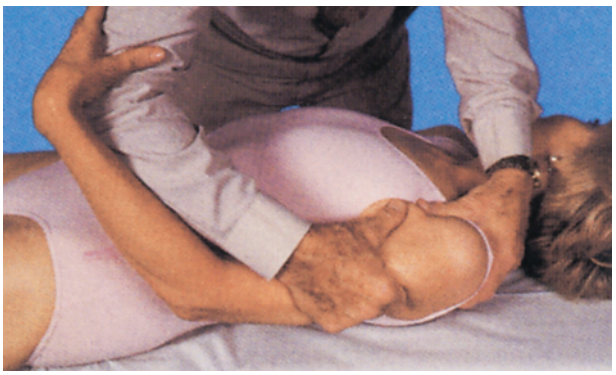


(b)

Figure 13.22 Shoulder instability – the apprehension test (a) This is the apprehension test for anterior subluxation or dislocation. Abduct, externally rotate and extend the patient's shoulder while pushing on the head of the humerus. If the patient feels that the joint is about to dislocate, she will forcibly resist the manoeuvre. (b) Posterior dislocation can be tested for in the same way by drawing the arm forward and across the patient's body (adduction and internal rotation).



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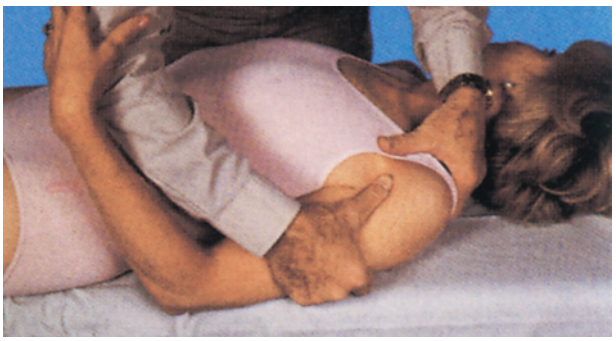


(b)

Figure 13.23 Multidirectional instability (a) The anterior and (b) posterior drawer tests are best performed with the patient lying supine. The amount of movement is compared with that on the unaffected side.

the humeral head is levered forward by extending and laterally rotating the arm; the patient immediately becomes apprehensive.

If instability is marked the *drawer test* may be positive (see Figure 13.23). With the patient supine, the scapula is stabilized with one hand while the upper arm is grasped firmly with the other so as to manipulate the head of the humerus forwards and backwards (like a drawer).

Investigations

Most cases can be diagnosed from the history and examination alone. The Hill–Sachs lesion (when it is present) is best shown by an *anteroposterior X-ray* with the shoulder internally rotated, or in the axillary view. Subluxation is seen in the axillary view. MRI or MR arthrography is useful for demonstrating bone lesions and labral tears.

Arthroscopy is sometimes needed to define the labral tear.

Examination under anaesthesia can help to determine the direction of instability. This forms an essential part of assessing instability. Both shoulders need to be examined. Reports have demonstrated sensitivities and specificities of 100% and 93%, respectively.

Treatment

If dislocation recurs at long intervals, the patient may choose to put up with the inconvenience and simply try to avoid vulnerable positions of the shoulder. There is some evidence that dislocation predisposes to osteoarthritis, although it is probably the initial dislocation rather than recurrence that causes this.

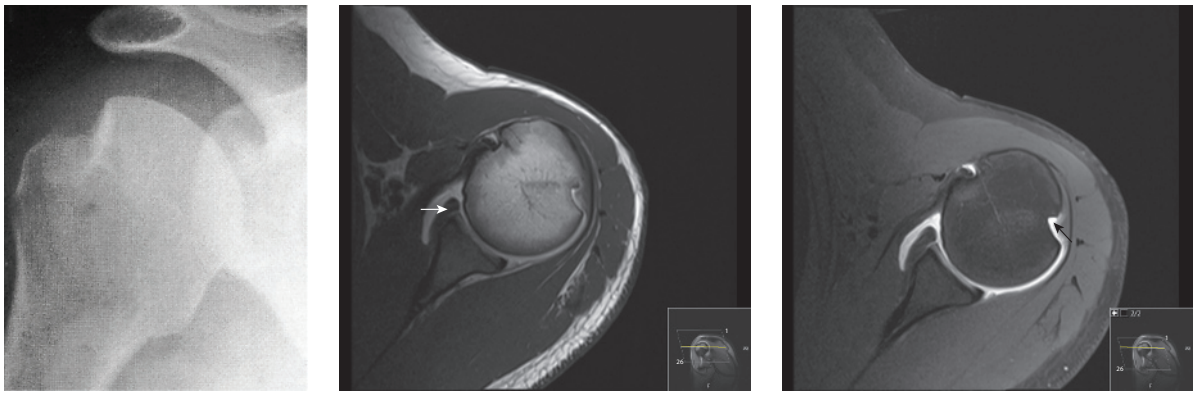
OPERATIVE TREATMENT

The indications for operation include frequent dislocation, especially if this is painful, and recurrent subluxation or a fear of dislocation sufficient to prevent participation in everyday activities, including sport. There is growing evidence to support primary surgery in young adults engaged in highly demanding physical activities following first acute traumatic dislocation. Two types of operation, anatomical and non-anatomical, are employed.

Anatomical repairs These are operations that repair the torn glenoid labrum and capsule, such as the Bankart procedure.

Non-anatomical repairs These procedures are designed to counteract the pathological tendency to joint displacement:

- Operations that shorten the anterior capsule and subscapularis by an overlapping repair (for example, the Putti–Platt operation) – prevent redislocation



(a) (b) (c)

Figure 13.24 Anterior instability – imaging (a) The plain X-ray shows a large depression in the posteriosuperior part of the humeral head (the Hill–Sachs sign). (b,c) MRI shows both a Bankart lesion, with a flake of bone detached from the anterior edge of the glenoid, and the Hill–Sachs lesion (arrows).

but at the cost of significant loss of external rotation. They are now not commonly used.
- Operations that reinforce the anteroinferior capsule by redirecting other muscles across the front of the joint (for example, the Bristow–Laterjet operation in which the coracoid process with its attached muscles is transposed to the front of the neck of the scapula) – produce less restriction of external rotation.
- Operations to correct a reduced retroversion angle of the humeral head by osteotomy.

In general, non-anatomical operations have a more limited role in the management of shoulder instability. While they do not address the underlying pathological changes, they are often used as revision procedures if a soft-tissue repair has not been successful or there are significant bony defects.

If the labrum and anterior capsule are detached, and there is no marked joint laxity, the Bankart operation combined with anterior capsulorrhaphy is the procedure of choice. The labrum is reattached to the glenoid rim with suture anchors or drill holes and, if necessary, the capsule is tightened by an overlapping tuck without shortening the subscapularis. Bankart initially described this as an open operation through the deltopectoral approach; however, arthroscopic techniques have been developed with advanced anchor materials and the development of specialized arthroscopic instruments. With careful patient selection clinical outcomes and recurrence rates of arthroscopic and open stabilization are now comparable; however, after either type of operation there is still a significant recurrence rate (about 20%), usually following another injury. If there is bone loss on either the glenoid aspect or the humeral head, the outcome following arthroscopic surgery is considerably worse, and in selected cases a non-anatomical procedure such as a Latarjet may be more appropriate.

ATRAUMATIC OR MINIMALLY TRAUMATIC INSTABILITY – POLAR TYPES II AND III

The terminology of these groups is somewhat confusing: '*atraumatic instability*' can include entities such as the 'loose shoulder', multidirectional instability, voluntary dislocation and habitual dislocation. In these cases it is often difficult to decide whether the problem is 'structural' or 'non-structural'.

Atraumatic structural instability

This is an acquired multidirectional instability due either to repetitive micro-trauma which has placed undue stress upon the soft tissues or to rapid, forceful movements that contribute to the development of overall laxity of the joint; occasionally a predisposing factor such as glenoid dysplasia is identified.

Atraumatic structural instability is a recognized problem in athletes, particularly swimmers and throwers. They develop symptoms of instability due to overload and fatigue in the stabilizing muscles of the shoulder; dislocation may occur in several different directions. It is doubly important in these cases to rule out the presence of any pathological condition, such as a labral lesion, and to assess whether there is any contributory element of abnormal muscle patterning. This group also includes patients with benign hypermobility syndromes.

Treatment

REHABILITATIVE MEASURES

Dedicated physiotherapy is focused on strengthening the muscles normally involved in stabilizing the shoulder and restoring muscular coordination and control.

Associated problems of muscle patterning are also addressed, and patients may need special instruction in the kinematics of shoulder movements and control of stability, as well as advice about modification of physical activities.

SURGICAL TREATMENT

If rehabilitative measures fail to reduce the problem and the patient is genuinely incapacitated, operative treatment may be required in selected cases – usually some type of capsular plication (which can be performed arthroscopically) or a capsular shift (by open operation).

ATRAUMATIC NON-STRUCTURAL INSTABILITY (ALTERED MUSCLE PATTERNING)

The stability of the shoulder joint throughout its large range of motion comes partly from precise synchronized muscle contractions and relaxations during movement. Each of the muscles moving and stabilizing the shoulder needs to be activated at a specific time in coordination with other protagonistic and antagonistic muscles. If this pattern is altered, instability can occur.

Muscle patterning instability usually occurs in younger patients who can voluntarily slip the shoulder out of joint as a trick movement (habitual subluxation – Figure 13.25), but the shoulder may then go on to dislocate repeatedly (uncontrolled or involuntary dislocation).

Treatment

The aim is to regain normal neuromuscular control and patterning. This can be difficult, time-consuming and require the participation of a full team comprising a specialist shoulder physiotherapist, shoulder surgeon and sometimes an occupational therapist and a psychologist. Treatment follows much the same lines as for atraumatic structural instability but surgery should be avoided if possible.

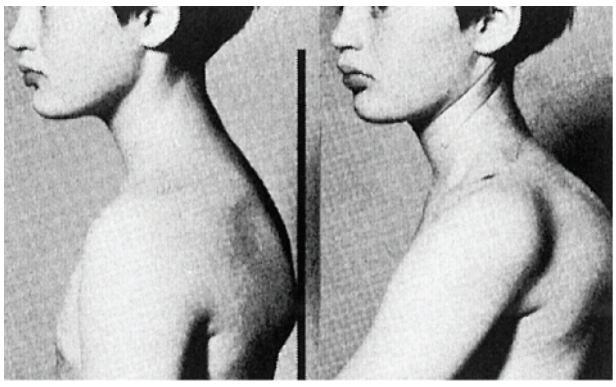
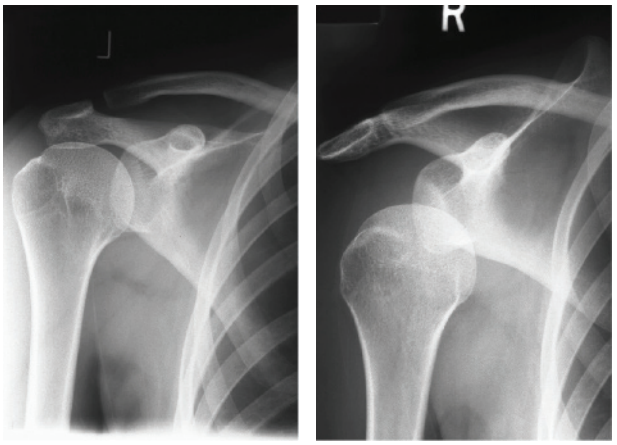


Figure 13.25 Habitual subluxation The clue is in the unconcerned expression.



(a) (b)

Figure 13.26 Inferior subluxation (a) X-ray of a young woman who developed 'clicking' and instability in the right shoulder after recovering from an injury to the neck and right upper limb. Plain X-ray examination showed no abnormality, but when the anteroposterior view was repeated with the patient carrying 10 kg weights in both hands, subluxation due to laxity of the anteroinferior capsule was demonstrated to the right side (b).

INFERIOR SUBLUXATION

Some weeks after an injury to the shoulder girdle a patient sometimes develops a feeling of instability in the shoulder, as if it 'slips out of joint', particularly when carrying something heavy with that arm. X-ray examination of the shoulder may show that the head of the humerus has subluxated inferiorly; if this is not immediately apparent, further views with the patient carrying a 10 kg weight in each hand will show the head of the humerus lying below the glenoid socket on the affected side (Figure 13.26). The condition is due to (temporary) weakness of the shoulder muscles, usually because of prolonged splintage of the arm and lack of exercise.

The condition usually corrects itself after a period of normal muscular activity, but physiotherapy will help to speed up the process. In the occasional case, tissue laxity is more persistent and capsular reefing may be advisable.

POSTERIOR INSTABILITY

Pathology

This condition is usually due to a violent jerk in an unusual position or following an epileptic fit or a severe electric shock. Dislocation may be associated with fractures of the proximal humerus, the posterior capsule is stripped from the bone or stretched, and there may be an indentation on the anterior aspect of the humeral head. Recurrent instability is almost

always a *posterior subluxation* with the humeral head riding back on the posterior lip of the glenoid.

Clinical features

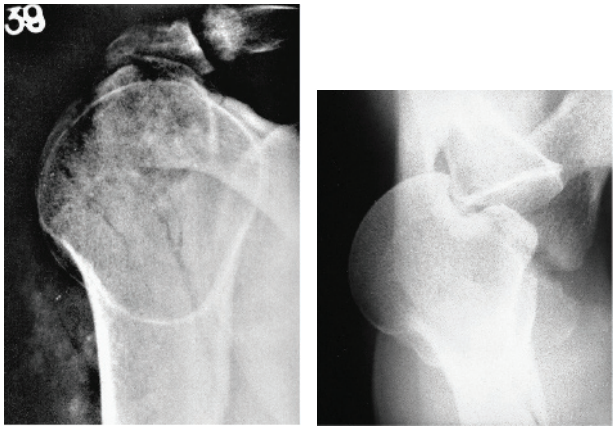
Acute posterior dislocation This is rare, and when it does occur it is often missed. There may be a history of fairly violent injury or an electric shock. On examination the arm is held in internal rotation and attempts at external rotation are resisted. The anteroposterior X-ray may show a typical 'light bulb' appearance of the proximal humerus (the humeral head looks symmetrically bulbous because the shoulder is internally rotated – Figure 13.27). If the arm can be abducted, an axillary view will show the dislocation quite clearly.

Recurrent posterior instability This usually takes the form of subluxation when the arm is used in flexion and internal rotation. On examination, the posterior drawer test (scapular spine and coracoid process in one hand, humeral head pushed backwards with the other) and posterior apprehension test (forward flexion and internal rotation of the shoulder with a posterior force on the elbow) confirm the diagnosis.

Treatment

Recurrent posterior instability due to muscle patterning and proprioceptive problems should be treated with physiotherapy. It is essential that this is undertaken by a therapist trained and experienced in dealing with shoulder instability, as the rehabilitation can be long and arduous.

Surgery should be considered if the primary abnormality is found to be structural (for example, a Bankart lesion, bony lesion or capsular injury). The particular operation depends on the injuries; it is therefore essential to identify the pathology and treat accordingly. No single operation applies to all patients with posterior instability. Soft-tissue reconstructions are the mainstay of treatment. Rarely there is a bone problem, such as excessive glenoid retroversion (shown on CT scan), in which case glenoid osteotomy should be considered. In extreme cases a bony block to posterior translation of the humeral head is employed but failure rates are reported to be high.



(a) (b)

Figure 13.27 Posterior dislocation (a) In the anteroposterior view the humeral head looks globular – the so-called 'light bulb' appearance. (b) In the lateral view one can see the humeral head is lying behind the glenoid fossa, with an impaction fracture on the anterior surface of the head.

DISORDERS OF THE GLENOHUMERAL JOINT

TUBERCULOSIS
(see also Chapter 2)

Tuberculosis of the shoulder is uncommon. Between 1% and 2% of tubercular lesions of the skeleton occur in the shoulder. It usually starts as an osteitis but is rarely diagnosed until arthritis has supervened. This may proceed to abscess and sinus formation (exudative form), but in some cases the tendency is to fibrosis and ankylosis. If there is no exudates, the term 'caries sicca' or dry form is used; however, one suspects that many such cases, formerly diagnosed on the basis of coexisting pulmonary tuberculosis rather than joint biopsy or bacteriological examination, are actually examples of frozen shoulder.

Clinical features

Adults are mainly affected. They complain of a constant ache and stiffness lasting many months or years. The striking feature is wasting of the muscles around the shoulder, especially the deltoid. Depending on the form, there may or may not be swelling present. Systemic features may also be present. In neglected cases a sinus may be present over the shoulder or in the axilla. There is diffuse warmth and tenderness and all movements are limited and painful. Axillary lymph nodes may be enlarged.

X-rays

X-rays show generalized rarefaction, usually with some erosion of the joint surfaces. There may be abscess cavities in the humerus or glenoid, with little or no periosteal reaction (Figure 13.28). The dry form shows narrowing of the joint space with periarticular osteoporosis. MRI can show destructive lesions, changes in the synovium, bone marrow infiltration and the fluid levels of abscesses.

Treatment

In addition to systemic treatment with antituberculous drugs, the shoulder should be rested until

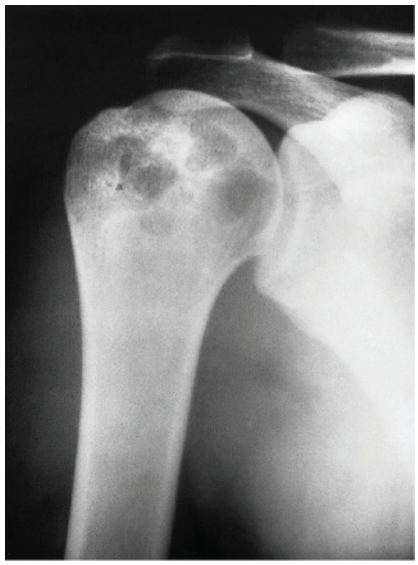


Figure 13.28 Tuberculosis X-ray of the shoulder showing tuberculous abscesses in the head of the humerus.

acute symptoms have settled. Thereafter movement is encouraged and, provided the articular cartilage is not destroyed, the prognosis for painless function is good. If there are repeated flares, or if the articular surfaces are extensively destroyed, the joint should be arthrodesed.

RHEUMATOID ARTHRITIS
(see also Chapter 3)

This is the most common arthropathy to affect the shoulder complex; up to 90% of patients with rheumatoid arthritis have involvement of the acromioclavicular joint, the shoulder joint and the various synovial pouches around the shoulder.

The *acromioclavicular joint* develops an erosive arthritis which may go on to capsular disruption and instability. This is sometimes the first site to be diagnosed from routine X-rays of the chest.

The *glenohumeral joint*, with its lax capsule and folds of synovium, shows marked soft-tissue inflammation. Often there is an accumulation of fluid and fibrinoid particles which may rupture the capsule and extrude into the muscle planes. Cartilage destruction and bone erosion are often severe.

The *subacromial bursa* and the *synovial sheath* of the LHB become inflamed and thickened; often this leads to rupture of the rotator cuff and the biceps tendon.

Clinical features

The patient may be known to have generalized rheumatoid arthritis; occasionally, however, acromioclavicular erosion discovered on an X-ray of the chest is the first clue to the diagnosis.

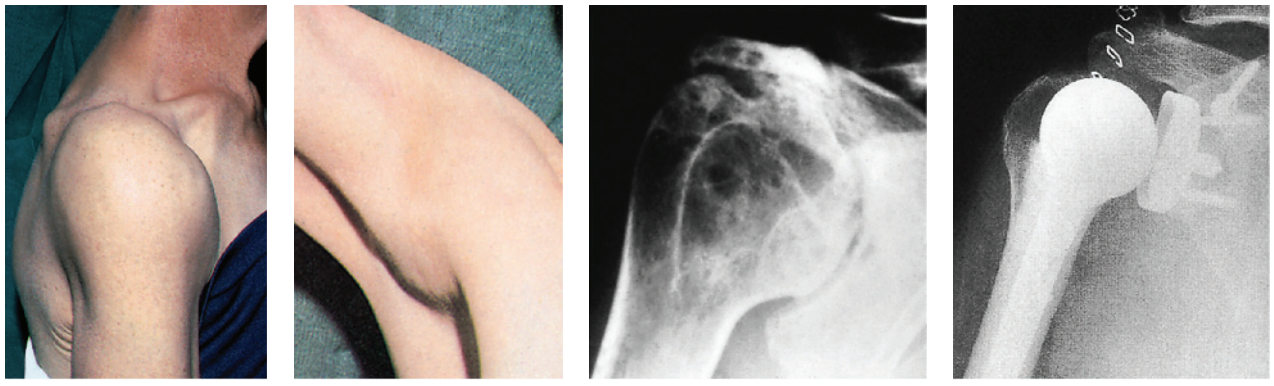
Pain and swelling are the usual presenting symptoms; the patient (usually a woman) has increasing difficulty with simple tasks such as combing her hair or washing her back. Although it may start on one side, the condition usually becomes bilateral.

Synovitis of the joint results in swelling and tenderness anteriorly, superiorly or in the axilla. *Tenosynovitis* produces features similar to those of cuff lesions, including tears of supraspinatus or biceps. Joint and tendon lesions usually occur together and conspire to cause the marked weakness and limitation of movement that are features of the disease. See Figure 13.29.

X-rays

Neer described three radiological patterns:

- *wet* (periarticular erosions, rapid progress, early cuff rupture
- *dry* (subchondral sclerosis, osteophytes, slow progress, cuff intact)
- *resorptive* (marked bone loss, few erosions).



(a) (b) (c) (d)

Figure 13.29 Rheumatoid arthritis (a) Large synovial effusions cause easily visible swelling; small ones are likely to be missed, especially if they present in the axilla (b). (c) X-rays show progressive erosion of the joint. (d) X-ray appearance after total joint replacement.

Treatment

The general treatment of rheumatoid arthritis is discussed in Chapter 3. In the early stages, local treatment in the form of intra-articular injections of steroid may be needed.

If synovitis persists, operative synovectomy is carried out; at the same time, cuff tears may be repaired. Excision of the lateral end of the clavicle may relieve acromioclavicular pain.

In advanced cases pain and stiffness can be very disabling. Provided the rotator cuff is not completely destroyed and there is still adequate bone stock, total joint replacement with an unconstrained prosthesis may be carried out. This operation provides good pain relief, moderate shoulder function and reasonable durability. Surface replacement arthroplasty has comparable outcomes to total joint replacement but is not suitable for severely damaged joints in which the humeral head is insufficient or too soft.

If the rotator cuff is destroyed, or bone erosion very advanced, reverse shoulder replacement may be preferable.

OSTEOARTHRITIS

Osteoarthritis (OA) of the glenohumeral joint is more common than is generally recognized affecting more than 30% of patients over 60 years of age. Primary OA is when there are no predisposing factors but secondary OA may be secondary to local trauma, recurrent subluxation or long-standing rotator cuff lesions. Often chondrocalcinosis is present as well but it is not known whether this predisposes to osteoarthritis or appears as a sequel to joint degradation. It is characterized by a gradual progressive breakdown of the articular cartilage and other joint surfaces.

Clinical features

The patient is usually aged 50–60 years and may give a history of injury, shoulder dislocation or a previous painful arc syndrome. There is usually little to see but shoulder movements are restricted in all directions. Pain is common, particularly at night.

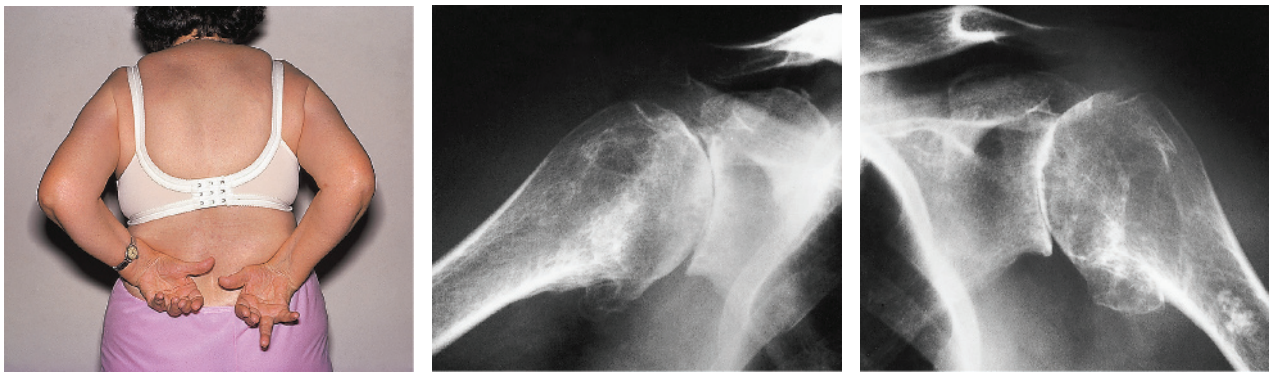
X-rays

X-rays show distortion of the joint, bone sclerosis and osteophyte formation; the articular 'space' may be narrowed or may show calcification (Figure 13.30).

Treatment

Analgesics and anti-inflammatory drugs relieve pain, and exercises may improve mobility. Most patients manage to live with the restrictions imposed by stiffness, provided pain is not severe. However, if both shoulders are involved, the disability can be severe. There is little evidence to support the routine use of steroid injection. These may give short-term relief and can be useful as a diagnostic aid. Arthroscopic washout and debridement has gained popularity and some studies report decreased pain and increased movement. There is insufficient evidence to support its routine use, however.

In advanced cases, if pain becomes intolerable, shoulder arthroplasty is justified. Arthroplasty is discussed in more detail later in this chapter. It may not always improve mobility much, but it does relieve pain. The alternative is arthrodesis, although this is much less commonly performed now than it was.



(a) (b) (c)

Figure 13.30 Osteoarthritis of the shoulder (a) This woman has advanced osteoarthritis of both shoulders. Movements are so restricted that she has difficulty dressing herself and combing her hair. (b,c) X-rays show the severe degree of articular destruction.

RAPIDLY DESTRUCTIVE ARTHROPATHY (MILWAUKEE SHOULDER)

Occasionally, in the presence of long-standing or massive cuff tears, patients develop a rapidly progressive and destructive form of osteoarthritis in which there is severe erosion of the glenohumeral joint, the acromion process and the acromioclavicular joint – what Neer and his colleagues called a *cuff tear arthropathy*. The changes are now attributed to hydroxyapatite crystal shedding from the torn rotator cuff and a synovial reaction involving the release of lysosomal enzymes (including collagenases) which lead to cartilage breakdown. A similar condition is seen in other joints such as the hip and knee. The shoulder disorder, however, has come to be known as *Milwaukee shoulder*, after the city from whence McCarty hailed.

Clinical features

The patient is usually aged over 60 years and may have suffered with shoulder pain for many years. Over a period of a few months the shoulder becomes swollen and increasingly unstable. On examination, there is marked crepitus in the joint and loss of active movements.

X-rays

X-rays show severe erosion of the articular surfaces, subluxation of the joint and calcification in the soft tissues (Figure 13.31).

Treatment

Resurfacing arthroplasty relieves pain and allows good rotations at waist level but will not improve abduction, because the rotator cuff is disrupted and the joint is unstable. It is quick and minimally invasive, retaining bone stock and keeping options open for future revision or arthrodesis.

Reverse shoulder arthroplasty in cuff tear arthropathy allows better elevation in the presence of a well-functioning deltoid as it depends less on the status of the cuff. Problems may occur in the long-term follow-up regarding progressive glenoid loosening due to the so-called 'inferior notching', which is supposed to be a result of an impingement at the inferior glenoid rim followed by increased polyethylene wear and progressive osteolysis. It is thus advisable to avoid reverse shoulder arthroplasty in the younger patient.

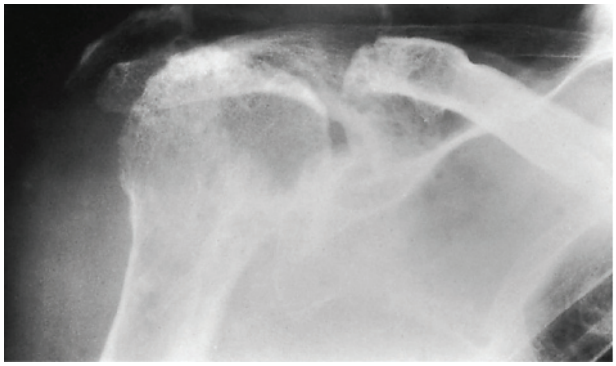


Figure 13.31 Milwaukee shoulder X-ray showing a destructive arthropathy with marked swelling and calcification in the soft tissues around the shoulder.

OSTEONECROSIS

The shoulder is the second most common site of steroid-induced osteonecrosis (Figure 13.32). The condition may also be seen in association with marrow storage disorders, sickle-cell disease and caisson disease, or following irradiation of the axilla.

The clinical features and diagnosis are discussed in Chapter 6. Articular collapse occurs more slowly than in weight-bearing joints and operative treatment can usually be delayed for several years. If this should become necessary, joint replacement is the method of choice.

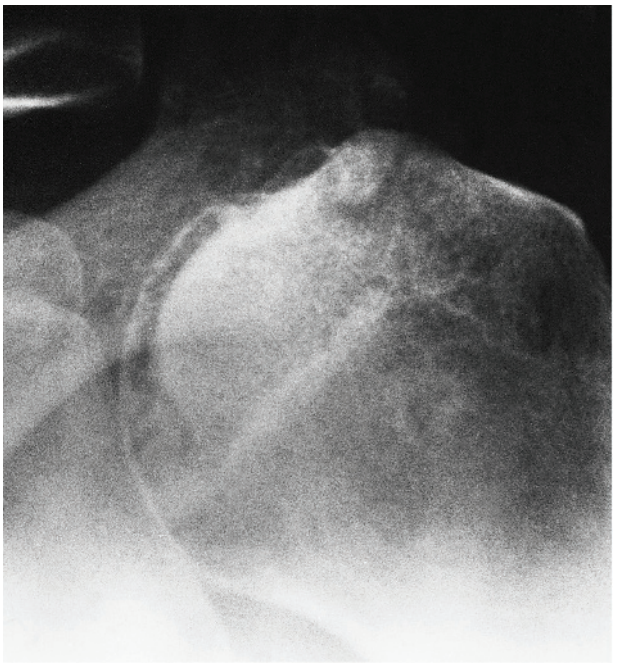


Figure 13.32 Osteonecrosis A young woman with systemic lupus erythematosus was treated with large doses of rednisolone. She developed pain in one hip and one shoulder. X-ray of the shoulder shows the classic features of osteonecrosis, including a long subarticular fracture of the humeral head.

DISORDERS OF THE SCAPULA AND CLAVICLE

CONGENITAL ELEVATION OF THE SCAPULA

The scapulae normally complete their descent from the neck by the third month of foetal life; occasionally one or both scapulae remain incompletely descended. Associated abnormalities of the cervical spine are common and sometimes there is a family history of scapular deformity.

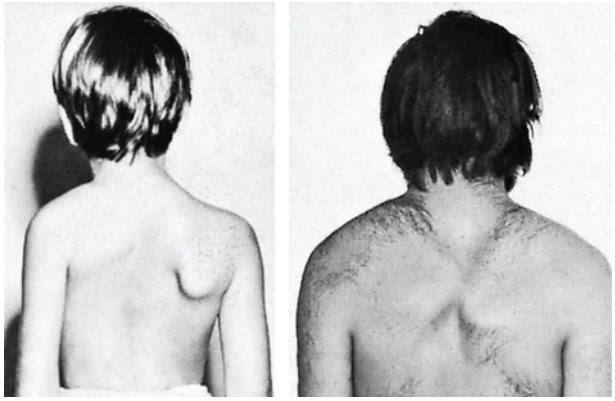
Clinical features

Two similar, and possibly related, conditions are encountered: *Sprengel's deformity* and *Klippel–Feil syndrome* (Figure 13.33).

Sprengel's deformity Deformity is the cardinal symptom and it may be noticed at birth. The shoulder on the affected side is elevated; the scapula looks and feels abnormally high, smaller than usual and somewhat prominent; occasionally both scapulae are affected. The neck appears shorter than usual and there may be kyphosis or scoliosis of the upper thoracic spine. Shoulder movements are painless but abduction and elevation may be limited by the fixation of the scapula.

X-rays will show the elevated scapula and any associated vertebral anomalies; sometimes there is also a bony bridge between the scapula and the cervical spine (the omovertebral bar).

Klippel–Feil syndrome This is usually a more widespread disorder. There is bilateral failure of scapular descent associated with marked anomalies of the cervical spine and failure of fusion of the occipital bones. Patients look as if they have no neck; there is a low hairline, bilateral neck webbing and gross limitation of neck movement. This condition should not be confused with bilateral shortness of the sternomastoid muscle in which the head is poked forward and the chin thrust up; the absence of associated congenital lesions is a further distinguishing feature.



(a) (b)

Figure 13.33 Scapular disorders (a) Sprengel shoulder; (b) Klippel–Feil syndrome.

Treatment

Mild cases are best left untreated. Surgical treatment aims to decrease deformity and improve shoulder function. In children under 6 years of age, the scapula can be repositioned by releasing the muscles along the vertebral and superior borders of the scapula, excising the supraspinous portion of the scapula and the omovertebral bar, pulling the scapula down, then reattaching the muscles to hold it firmly in its new position.

In older children, this carries a risk of brachial nerve compression or traction between the clavicle and first rib; here it is safer merely to excise the supraspinous portion of the scapula in order to improve the appearance but without improving movement.

Before undertaking any operation the cervical spine should be carefully imaged in order to identify any abnormalities of the odontoid process or base of skull.

CLEIDOCRANIAL DYSOSTOSIS

This is an autosomal dominant disorder characterized by hypoplasia or aplasia of the clavicles and flat bones (pelvis, scapulae and skull). Those affected have a typical appearance, with drooping shoulders, an usually narrow chest and the ability to bring the shoulders together across the front of the chest.

X-rays These show hypoplasia or complete absence of the clavicles, and sometimes also of the scapulae. Other skeletal defects, which occur in varying degree, are delayed closure of the fontanelles, brachycephaly, underdevelopment of the pelvis, coxa vara and scoliosis.

Treatment Despite the widespread defects, treatment is usually unnecessary and patients enjoy good function.

CONGENITAL PSEUDARTHROSIS OF THE CLAVICLE

The typical clinical picture is that of a child with a painless lump in the mid-shaft of the clavicle. This always occurs on the right side, except in the presence of dextrocardia.

X-rays These show the break in the clavicle, which usually heals only after excision of the 'nonunion' and bone grafting.

Treatment If it is required, treatment is by excision of the pseudarthrosis and bone grafting across the gap.

SCAPULAR INSTABILITY

Winging of the scapula (Figure 13.34) can be due to weakness of the serratus anterior muscle. It results in asymmetry of the shoulders but the deformity may not be obvious until the patient tries to contract the serratus anterior against resistance. This may limit active elevation, but more commonly presents with fatigue pain and deformity.

There are several causes of weakness or paralysis of the serratus anterior muscle:

- neuralgic amyotrophy
- injury to the brachial plexus (a blow to the top of the shoulder, severe traction on the arm or carrying heavy loads on the shoulder)
- direct damage to the long thoracic nerve (for example, during radical mastectomy or first rib resections)
- fascioscapulohumeral muscular dystrophy.

Disability is usually slight and is best accepted. However, if function is noticeably impaired, it is possible to stabilize the scapula by transferring the sternal portion of pectoralis major and attaching it via a fascia lata graft to the lower pole of the scapula; or the scapula can be fixed to the rib cage to provide the deltoid and the rotator cuff muscles with a stable base from which to control the shoulder.

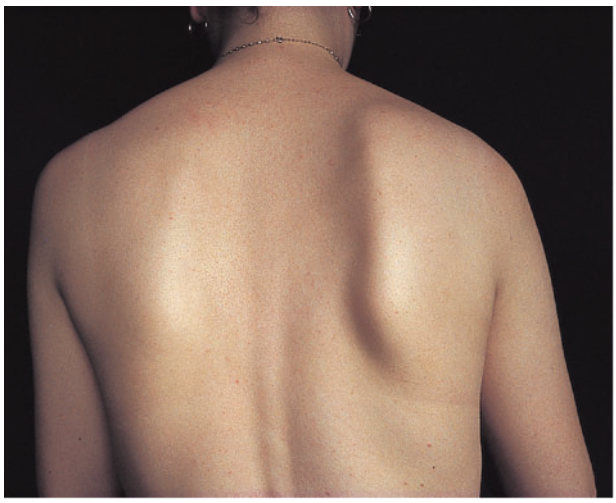


Figure 13.34 Winged scapula This young woman's right scapula was somewhat prominent even at rest, but here the 'winging' is enhanced by having her thrust her arms forcibly against the wall.

A less obvious, but sometimes more disabling, form of scapular instability may follow *injury* to the *spinal accessory nerve* (for example, following operations in the posterior triangle of the neck). The trapezius muscle is an important stabilizer of the shoulder and loss of this function results in weakness and pain on active abduction against resistance. Early recognition may permit nerve repair or grafting.

GRATING (SNAPPING) SCAPULA

Asymptomatic scapulothoracic crepitus is found in about a third of healthy persons. People with symptoms complain of grating or clicking on moving the arm; the condition is often painless but annoying, although it does sometimes become painful. Usually no cause is found, though bony, muscular and bursal abnormalities have been blamed.

X-rays Tangential X-ray views of the scapula should be obtained to exclude an osteochondroma on the undersurface of the scapula. A CT scan with three-dimensional reconstruction can be helpful; if an osteochondroma is present, the lesion can be excised. If a bony lesion is not identified, conservative treatment is usually adopted.

SEPTIC ARTHRITIS OF THE STERNOCLAVICULAR JOINT

This condition is rare except in drug abusers following intravenous injections, and as a secondary complication of sternoclavicular haemarthrosis following trauma. Local signs may be misleadingly mild, but persistent pain, swelling and tenderness associated with systemic signs of infection should arouse suspicion.

Imaging *X-rays* are usually normal until fairly late when they may show erosion of the sternoclavicular joint and the adjacent bone. If infection is suspected, further imaging of the joint will be required, such as *MRI* or *CT*, which will allow the extent of any spread of infection or bony destruction to be identified. *Radioscintigraphy* is able to identify multifocal septic arthritis.

Investigations If infection is suspected, blood cultures and aspiration of the joint will be required. A wide range of organisms have been found to cause infection at this site.

Treatment If frank pus is present in the joint, an arthrotomy with formal washout will be required. If there is delay in diagnosis or institution of the correct treatment, rupture of the joint capsule may occur with tracking of pus into the chest wall, retrosternum or superior mediastinum.

STERNOCLAVICULAR HYPEROSTOSIS

Several individually uncommon disorders are associated with pain and swelling over the clavicle or the sternoclavicular joint. They are often confused, although certain characteristic features permit appropriate differentiation in the majority of cases.

Condensing osteitis of the clavicle

This is usually seen in women of 20–40 years who present with pain at the medial end of the clavicle, which is aggravated by abducting the arm. The clavicle may be thickened and tender. *X-rays* reveal sclerosis, and radionuclide scanning shows increased activity in the affected bone.

The condition may be no more than a reaction to the mechanical stress of excessive lifting activities, and treatment consists simply of avoiding such activities. Of greater importance is the need to distinguish it from the other hyperostotic disorders.

Condensing osteitis shares both morphological and radiological features with osteitis of the ilium and osteitis of the pubis. It has been noted that all of these bones have a fibrocartilaginous covering which may explain the predilection of the condition for those sites. It can run a relapsing and remitting course but usually resolves.

Sternocostoclavicular hyperostosis

This condition in some ways resembles condensing osteitis, but it is seen in slightly older people (both men and women) and is usually bilateral. Patients develop pain, swelling and tenderness over the sternoclavicular region and *X-rays* show hyperostosis of the medial ends of the clavicles, the adjacent sternum, the anterior ends of the upper ribs and the soft tissues in between. Vertebrae also may be affected and the ESR may be increased; little wonder that it has been suggested that this is a type of seronegative spondarthropathy.

Biopsy is of little help; the histological changes are non-specific and microorganisms have not been identified. A peculiarity which links this condition with the next is an association with pustular lesions on the palms and soles (palmoplantar pustulosis) and pustular psoriasis.

Subacute or chronic multifocal osteomyelitis

Multifocal osteomyelitis usually occurs in children and adolescents; the clavicle and lower limb metaphyses are sites of predilection. It may present as a painful, fusiform swelling of the clavicle and X-rays show thickening and sclerosis of the medial third of the bone.

Like sternocostoclavicular hyperostosis, it is sometimes associated with palmoplantar pustulosis. The diagnosis is strongly suggested if pustulosis is present, otherwise it usually emerges gradually as other sites become affected over the course of the next year or two, and X-rays show the typical lytic areas in the metaphyses and/or epiphyses close to the physis. There is no effective treatment; the lesions almost invariably heal spontaneously over a period of months or years, the only trace of the condition being the thickened bone ends.

OSTEOARTHRITIS OF THE ACROMIOCLAVICULAR JOINT

Osteoarthritis of the acromioclavicular joint is common in middle-aged and older people. It generally affects two groups of people. The first are people in their thirties who are involved in sports or carry out heavy manual activities. Early presentation in this group may be damage to the cartilaginous disc. The second group are older people where the condition is due to degenerative changes.

Predisposing factors are trauma (subluxation of the joint) and occupational stress (habitually carrying weights on the shoulder or working with pneumatic hammers and drills), but the condition also occurs in the absence of any suggestive history. The patient may complain of 'shoulder pain', but if you ask him or her to point, he or she will direct your attention to the prominent bump at the outer end of the clavicle; tenderness is sharply localized to this area (Figure 13.35).

Figure 13.35 Osteoarthritis of the acromioclavicular joint Osteophytic thickening of the acromioclavicular joint produced a small (but very tender) bump on top of the left shoulder. Occasionally the joint capsule herniates, producing a large 'cyst' over the acromioclavicular joint.

Shoulder movements are usually not restricted (unless the shoulder joint itself is involved) but there may be pain at the extremes of abduction and flexion – giving cross arm pain and higher arc pain.

X-rays These show the characteristic features of osteoarthritis; the changes are often bilateral, even though only one side may be hurting. In some cases, the condition is discovered while examining the patient for an impingement syndrome; indeed, acromioclavicular OA may *cause* impingement.

Treatment The initial treatment is non-surgical with activity modification analgesics or steroid injections. If this is ineffectual, pain may be relieved by excision of the lateral end of the clavicle. This procedure can now be performed arthroscopically. Trimming of the bony roughness, or excision of the outer end of the clavicle, may also be needed during subacromial decompression for rotator cuff impingement.

OPERATIONS

Rotator cuff surgery and shoulder stabilization are described in the relevant sections.

Arthroscopy

Arthroscopy is a useful technique for the *diagnosis* of periarticular and intra-articular disorders, such as rotator cuff disruption and instability. At the same time a *biopsy* can be taken which may assist in the diagnosis of synovial disorders such as rheumatoid arthritis or pigmented villonodular synovitis.

Arthroscopic surgery is now well established. There has been a transition over the last 20 years from its use in diagnosis to that of repair and reconstructive procedures. It is the first-line surgical option for subacromial decompression, acromioclavicular joint excisions, debridement of rotator cuff tears and release of frozen shoulder. Arthroscopic repair of Bankart lesions produces results comparable to those obtained by open surgery.

Arthroplasty of the shoulder

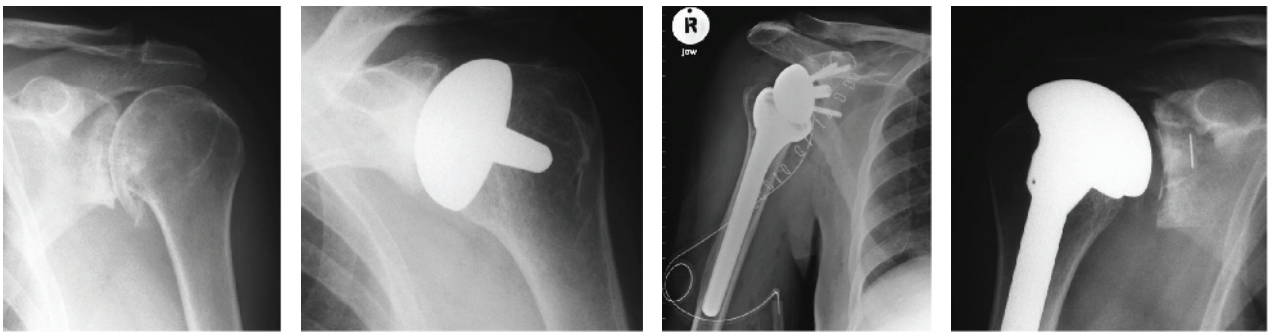
Shoulder replacement (Figure 13.36) was initially introduced by Neer in the 1950s for the treatment of proximal humeral fractures. Subsequent modifications and the introduction of glenoid resurfacing broadened the indications to include other disease processes, including end-stage glenohumeral osteoarthritis and rheumatoid arthritis. If non-operative treatment fails, the two surgical options commonly considered are humeral head replacement (HHR) and total shoulder replacement (TSR). The optimal treatment choice, however, remains controversial.

Indications

The indications for arthroplasty are:

- osteoarthritis causing pain and loss of movement
- rheumatoid arthritis
- complex fractures of the proximal humerus
- avascular necrosis of the humeral head
- tumours of the proximal humerus
- severe arthritis with cuff arthropathy.

The choice of procedure lies between total shoulder replacement, humeral head replacement (hemiarthroplasty) – which can be stemmed or resurfacing – and more constrained shoulder replacements such as the reverse polarity shoulder replacements. The relative merits of total shoulder arthroplasty and hemiarthroplasty are not clear. Glenoid resurfacing is contraindicated if inadequate bone stock or irreparable rotator cuff tears (or both) are present. Hemiarthroplasty affords the benefits of decreased operation time, blood loss and technical



(a) (b) (c) (d)

Figure 13.36 Shoulder replacements (a,b) Osteoarthritis and a resurfacing arthroplasty. (c) Early postoperative X-ray of a reverse polarity shoulder replacement. (d) Total shoulder replacement with replacement of the glenoid.

difficulty which would otherwise attend glenoid exposure and resurfacing. On the other hand, individual studies have reported less consistent pain relief with isolated humeral head replacement. With isolated humeral head replacement, the glenoid can undergo progressive erosion over time, often leading to deteriorating results.

Relative indications for TSR in patients with glenohumeral arthritis include loss of articular cartilage or incongruent osseous surfaces, with normal or reparable rotator cuff tendons. TSR requires more operative time and is technically more challenging than hemiarthroplasty, and the procedure introduces the concern of glenoid loosening, the most common complication. However, proponents of TSR suggest it may yield more consistent pain relief and a better range of motion.

In a systematic review of the literature there is a suggestion that overall TSR may yield superior results; however, it remains unclear if one procedure is significantly better than the other.

Complications

The commonest, in order of frequency, are loosening of the components, glenohumeral instability, rotator cuff failure, periprosthetic fracture, infection and implant failure. Glenoid fixation remains a challenge; lucent lines around the glenoid component are very common but are not always symptomatic.

Outcome

This depends largely on the indications for surgery. Arthroplasty for fractures, avascular necrosis or proximal humeral tumours gives good pain relief and shoulder movement, although power is always diminished.

Where there is more extensive joint destruction and disruption of the soft tissues (for example, in rheumatoid arthritis), pain relief is still excellent but the range of movement is only moderately improved. The greater the integrity of the surrounding soft tissues (and especially the rotator cuff), the more stable the new joint will be, and thus the better the outcome of the operation. In severe cuff failure, reverse geometry arthroplasty has been used with reasonable success in the short term in elderly patients, although further research is needed to assess longevity and continued functional improvement.

Arthrodesis

Arthrodesis of the glenohumeral joint is now seldom performed, but it is still a useful operation for severe shoulder dysfunction.

Indications

The indications for shoulder arthrodesis are:

- paralysis of the scapulohumeral muscles
- infective disorders of the glenohumeral joint (including tuberculous arthritis)
- advanced erosive arthritis with massive disruption of the rotator cuff
- failed total shoulder arthroplasty
- uncontrolled instability.

The operation

A prerequisite is stable and powerful scapulothoracic movement because, with a fused shoulder, 'movement' is achieved entirely by rotation of the scapula on the thorax. A number of techniques have been reported with extra-articular arthrodesis, intra-articular arthrodesis and a combination of both. Internal fixation has been used more frequently in recent years. Extra-articular arthrodesis is primarily a historic procedure that was used before the antibiotic era to treat tuberculosis.

A variety of methods of internal fixation for intra-articular arthrodesis have been described. It is generally agreed that internal fixation is desirable because it maintains the position of the arthrodesis and can decrease the length of time spent in plaster immobilization.

The optimal position is 30 degrees of flexion, 30 degrees of abduction and 30 degrees of internal rotation. A thermoplastic orthosis needs to be worn for 6 weeks.

Outcome

Despite the restriction of glenohumeral movement, postoperative function is surprisingly good; and of course the joint is free of pain!

Complications include non-union, infection, malposition often with too much internal rotation, prominence of the internal fixation and fracture of the humerus.

NOTES ON APPLIED ANATOMY

Joints

The anatomy of the shoulder is uniquely adapted to allow freedom of movement and maximum reach for the hand.

Five 'articulations' are involved:

- the glenohumeral joint
- the pseudojoint between the humerus and the coracoacromial arch
- the sternoclavicular joint
- the acromioclavicular joint
- the scapulothoracic articulation.

Stability

The shallow glenohumeral articulation has little inherent stability because the glenoid surface area is only one-quarter that of the humeral articular surface. The extent to which the socket is deepened by the labrum may seem trivial, but it must be significant because labral tears are associated with dislocation. Stability depends mainly on the integrity of the ligaments and capsule. The muscles provide kinetic stability: during abduction the rotator cuff muscles draw the head of the humerus firmly into its socket while the deltoid elevates the arm.

Rotator cuff

The rotator cuff is a sheet of conjoint tendons closely applied over the top of the shoulder capsule and inserting into the greater tuberosity of the humerus. It is made up of subscapularis in front, supraspinatus above and infraspinatus and teres minor behind. The 'rotator' muscles have an important function in stabilizing the head of the humerus by pulling it firmly into the glenoid whenever the deltoid lifts the arm forwards or sideways. The rotator interval lies between the supraspinatus and infraspinatus tendons.

Arching over the cuff is a fibro-osseous canopy – the coracoacromial arch – formed by the acromion process posterosuperiorly, the coracoid process anteriorly and the coracoacromial ligament joining them. Separating the tendons from the arch, and allowing them to glide, is the subacromial bursa. Of the four cuff tendons, the supraspinatus is the most exposed; it runs over the top of the shoulder under the anterior edge of the acromion and the adjacent acromioclavicular joint, with the intra-articular portion of the biceps tendon closely applied to its deep surface.

Movement

Abduction and flexion of the shoulder look simple; in fact, they are very complex movements involving all the joints of the shoulder girdle. Imagine what would happen if the deltoid muscle acted alone in abducting the shoulder. Because of the relatively unstable fulcrum, the deltoid would simply shrug the arm upwards at the side of the body. In reality, the rotator cuff muscles, particularly the supraspinatus, draw the head of the humerus firmly into the socket and slightly downwards, thus allowing the deltoid to act as a true abductor.

The first 30 degrees of abduction occurs almost entirely at the glenohumeral joint with slight movement of the clavicle at the sternoclavicular joint. From 30 to 90 degrees of abduction the scapula gradually comes into play, with about one-third of the movement coming from the scapula rotating on the thorax. From 90 to 180 degrees, the movement is mainly scapulothoracic and for this reason it is termed 'elevation' rather than 'abduction'. As the arm rises above shoulder height, it rolls into external rotation so that the greater tuberosity clears the projecting acromion. The sternoclavicular joint participates in movements close to the trunk (for example, shrugging or bracing the shoulders); the acromioclavicular joint moves in the last 60 degrees of abduction.

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The elbow

14

Adam C. Watts & David Warwick

CLINICAL ASSESSMENT

Symptoms

Pain from the elbow may be localized or diffuse, extending into the forearm. Localized pain over the lateral or medial epicondyle of the humerus is usually due to tendonitis ('tennis elbow' or 'golfer's elbow' respectively). Certain activities such as lifting may trigger or exacerbate the pain. Pain from the dorsal aspect of the elbow is likely to be due to olecranon bursitis or rarely triceps tendinopathy; anterior elbow pain is likely to be due to distal biceps tendinopathy. Pain in childhood or adolescence requires further investigation to rule out osteochondritides. In older patients osteoarthritis is a common cause of symptoms. *Remember that pain in the elbow is sometimes referred pain from the cervical spine.*

Stiffness, which means loss of range of movement, may develop slowly and will not be noticed until it is relatively marked. It can be very disabling, making it hard for the patient to reach their mouth (loss of flexion) or perform simple toileting (loss of supination).

Swelling of the elbow if diffuse may be due to injury, inflammation or infection. A sudden symmetrical loss of range suggests an increase of fluid within the joint which may be synovial fluid (effusion), blood or pus. A localized soft-tissue lump on the point of the olecranon is likely to be an olecranon bursitis. A rubbery lump more distal may be due to gouty tophus or rheumatoid nodule.

Deformity is uncommon except in rheumatoid arthritis and after trauma. Always ask about previous injuries.

Instability may present as pain or locking – the feeling of the elbow getting stuck in a position temporarily. It is usually due to trauma or destructive joint disease.

Ulnar nerve symptoms (tingling, numbness, clumsiness or weakness of the hand) may occur in elbow disorders because of the nerve's proximity to the joint.

Loss of function may be significant with elbow pathology because the role of the elbow is to position the hand in space to manipulate the environment and bring objects towards the individual.

Signs

Both upper limbs should be completely exposed including the shoulder girdle, and it is essential to look at the back of the elbow as well as the front. The neck, shoulder and hands often need to be examined too.

Look

With both upper limbs exposed the arms are held by the patient's side with the elbow extended and palms facing forward (the anatomical position). In this position the forearms are normally angled slightly outward relative to the line of the arm – a valgus or *carrying angle* of 5–15 degrees. 'Varus' or 'valgus' *deformity* is determined by angular deviation towards the body or away beyond those limits or, in unilateral abnormalities, by comparison with the normal side. Note that the carrying angle cannot be assessed reliably if there is a flexion contracture of the elbow.

Varus and valgus deformities (*cubitus varus* and *cubitus valgus*) are usually the result of trauma around the elbow. A varus deformity can be demonstrated by asking the patient to abduct the shoulders to 90 degrees with the palms facing forwards: the arm takes on the appearance of a rifle butt (*gunstock deformity*).

Look for scars around the elbow, in particular posterior to the medial epicondyle that may be related to previous ulnar nerve surgery.

Feel

Start by identifying the most obvious bony landmarks: the olecranon process posteriorly, the medial and lateral epicondyles and the head of the radius just distal

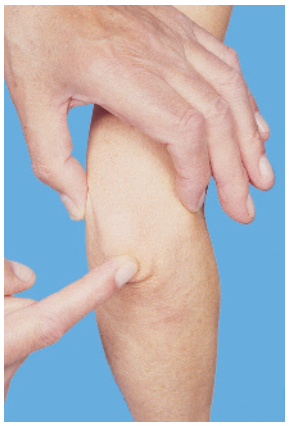


Figure 14.1 Examination Feeling begins with the skin. Is there undue warmth? Next, feel the bony landmarks. With the elbow flexed, the tips of the medial and lateral epicondyles and the olecranon process form an isosceles triangle. With the elbow extended, they lie transversely in line with each other. These relationships may be disturbed by trauma.

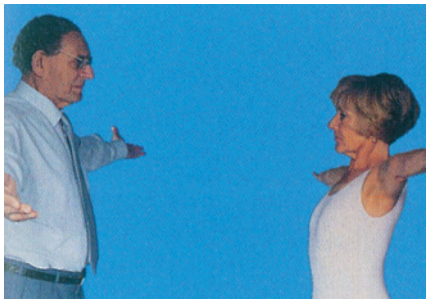
to the lateral epicondyle; pronating and supinating the forearm makes it easier to find the mobile radial head and lateral joint line (Figure 14.1). The ulna can be palpated throughout its length, the radius only at its proximal end and in the distal third of the forearm.

The back of the elbow is palpated for warmth and swelling (signs of an olecranon bursitis) and subcutaneous nodules (a feature of rheumatoid arthritis or gout). Feel more widely for synovial thickening and fluid (fluctuation on each side of the olecranon). Swelling of the joint is most easily appreciated by inspecting and palpating the area on the lateral aspect of the elbow demarcated by the olecranon, lateral epicondyle and radial head – the lateral soft spot. The ulnar nerve is very superficial behind the medial epicondyle and here it can be rolled under the fingers to feel if it is thickened or hypersensitive.

Last of all, feel for tenderness and try to determine which structure is affected.

Move

Active and passive flexion and extension are compared on the two sides (Figure 14.2). A normal range of movement is from zero degrees (an absolutely straight arm)



(a)

(b)

(c)

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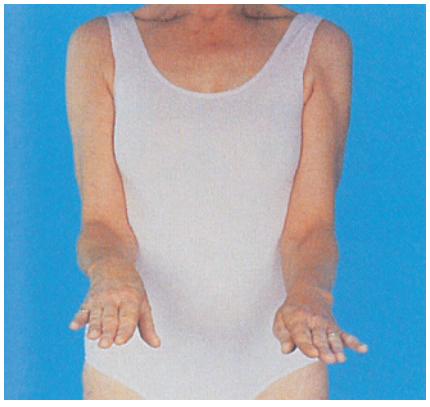

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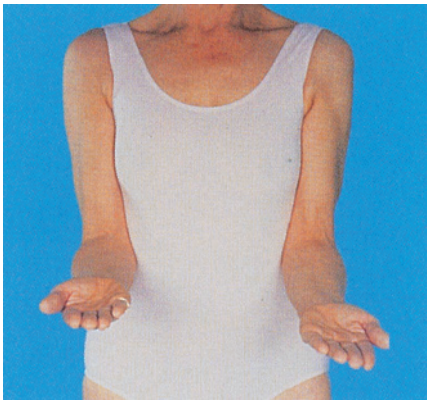


(f)

Figure 14.2 (a,b) The best way to examine active movement is to stand in front of the patient and show them what to do. (c,d) The normal range of flexion is from 0° (full extension) to 140° (full flexion). (e,f) To test pronation and supination, ask the patient to tuck their elbows tightly in to the body with the elbows flexed to 90° and then turn their hands from palm up to palm down. The normal range is 90° in both directions, although this is a composite of forearm and carpal rotation.

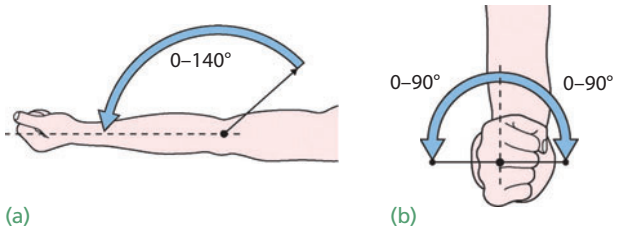


Figure 14.3 Normal range of movement (a) The extended position is recorded as 0° and any hyperextension as a minus quantity; flexion is full when the arm and forearm make contact. (b) From the neutral position the palm rotates 90° into pronation and 90° into supination. This is a composite of radioulnar motion (80°) and carpal rotation (10°).

to approximately 140 degrees depending on the size of the arm muscles. Some people with lax joints can hyperextend their elbow beyond zero, which by convention is recorded as a minus value. Pronation and supination of the forearm is tested with the elbow flexed to 90 degrees and the elbow held in by the patient's side to avoid compensatory shoulder movement (Figure 14.3); 80 degrees in both directions from neutral is normal. Note that this movement occurs not just at the elbow joint but within the whole forearm. *Stability* is assessed with the elbow after trauma by flexing the elbow to 30 degrees to unlock the olecranon from the olecranon fossa, maximally externally rotating the humerus to stabilize the shoulder joint and applying a valgus force to assess the medial ligament and then fully internally rotating the humerus and applying a varus force to assess the lateral ligament.

General examination

Clinical examination should include the neck and shoulder (that are sources of referred pain to the elbow) and the hand (for signs of nerve dysfunction).

Imaging

Plain X-rays

Standard X-rays would include an AP and lateral view. The position and integrity of each bone is noted, then the joint line and space. Attention should be paid to the soft-tissue shadow to look for swelling. There may be calcification in the soft tissues in chronic cases of tennis or golfer's elbow. Fluid in the elbow joint will cause elevation of the anterior and posterior fat pads in the coronoid and olecranon fossa that can be seen on the lateral radiograph as a 'sail sign'.

In children the epiphyses are largely cartilaginous and the articular relations often have to be deducted from the shape and position of the emerging secondary ossification centres. The age at which they appear is remembered by the mnemonic CRITOL in which the O stands for olecranon which appears at about 10 years and there is a 2-year age gap between each letter, thus: Capitellum – 2 years; Radial head – 4 years; Internal (medial) epicondyle – 6 years; Trochlea – 8 years; Olecranon – 10 years; Lateral epicondyle – 12 years.

Computed tomography

CT is mainly used for planning trauma reconstruction but can be used, with or without arthrography, to look for loose bodies or detail changes in osteoarthritis.

Magnetic resonance imaging

MRI is used mainly for the investigation of soft-tissue lesions around the elbow such as ligament tears and tendinopathies. The addition of arthrography improves sensitivity for detecting loose bodies and chronic ligament lesions. In adolescents with lateral elbow pain MRI can be used to identify osteochondritis dissecans. The distal biceps tendon is best viewed using flexion abduction supination (FABS) MRI scans that allow the whole tendon to be seen in a single view.

Ultrasound

Ultrasound is used for dynamic investigation around the elbow and to guide injections.

CONGENITAL DISORDERS

Congenital dislocation of the radial head

The radial head can dislocate anterior or posterior to the capitellum. Controversy exists around whether this is truly congenital or an acquired developmental disorder. It has been proposed that dorsal dislocations, which will often present as a lump on the lateral side of the elbow, are more likely to be truly congenital. The radial head becomes dome-shaped with chronic dislocation due to unrestrained growth. Function is usually good but pain can develop later in life due to abnormal loading on the capitellum. Excision can be performed as an open or arthroscopic procedure.

Congenital synostosis

Congenital deficiencies of the forearm bones are occasionally associated with fusion of the humerus to the radius or ulna. This disabling condition is, fortunately, very rare. Osteotomy can be used to alter the position of the hand if it is functionally unfavourable.

Proximal radioulnar synostosis causes loss of rotation, but the elbow flexion is retained and the inconvenience is often only moderate. Surgery to regain rotation rarely succeeds. A rotational osteotomy can give a more suitable angle of pronation–supination tailored to the individual's needs.

ACQUIRED DEFORMITIES

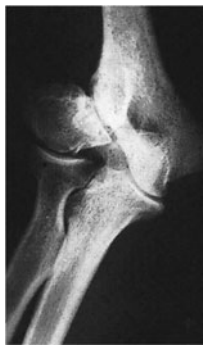
Cubitus valgus

The normal carrying angle of the elbow is 5–15 degrees of valgus; more than this or an asymmetry between the two sides is regarded as a valgus deformity (Figure 14.4). The most common cause is long-standing non-union of a fractured lateral condyle; the deformity may be associated with marked prominence of the medial condylar outline. In an elbow with cubitus valgus there is an increased risk of a delayed or tardy ulnar nerve palsy; years after the causal injury the patient notices a weakness of the hand with numbness or tingling of the ulnar fingers. The deformity itself requires no treatment but the affected nerve should be transposed to lie in front of the medial epicondyle to reduce the distance it has to travel.

Cubitus varus ('gunstock' deformity)

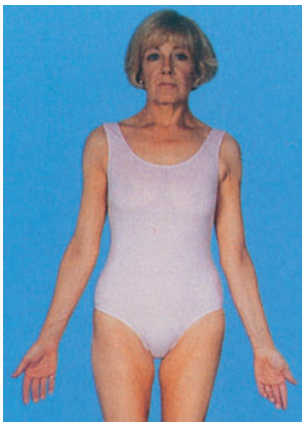
The deformity is most obvious when the elbow is extended and the arms are elevated (Figure 14.5). The most common cause is malunion of a supracondylar fracture in childhood. The deformity can be corrected by a closing wedge osteotomy of the humerus, but this is best left until skeletal maturity.

(a)



(b)

Figure 14.4 Cubitus valgus (a) This man has excessive valgus of the right elbow but his main complaint was of weakness and deformity in the hand, which was caused by a tardy ulnar nerve palsy secondary to the elbow deformity. (b) Valgus deformity from an un-united fracture of the lateral condyle.



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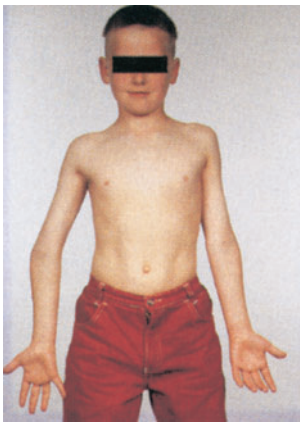

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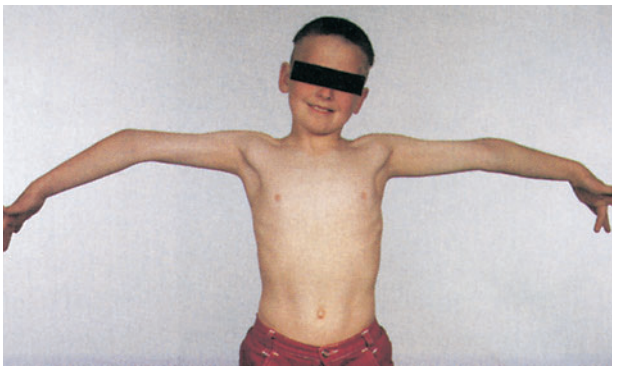
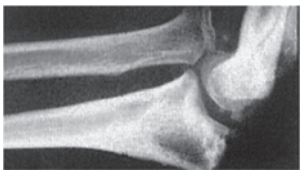


(c)

Figure 14.5 Cubitus varus (a) Note that the elbows are normally held in approximately 10° of valgus (the carrying angle). (b) This young boy ended up with varus angulation after a supracondylar fracture of the distal humerus. The deformity is much more obvious (c) when he raises his arms (gunstock deformity) and increases his risk of developing posterolateral rotatory instability and a snapping triceps.

Subluxation of the radial head

As discussed previously this can be an acquired deformity due to trauma or skeletal dysplasias in which the ulna is disproportionately short (e.g. multiple osteochondromas) (Figure 14.6). It usually causes little disability and attempts to relocate the radial head with skeletal distraction usually fail. Radial head excision can be performed after skeletal maturity.



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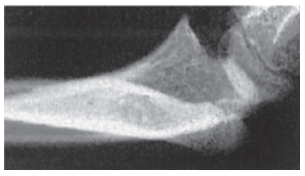


(b)

Figure 14.6 Dislocated radial head (a) Anterior dislocation from an old Monteggia fracture; (b) posterior dislocation, most likely congenital.

If the subluxation is due to an unreduced Monteggia fracture dislocation, and it is identified in reasonable time, open reduction and realignment of the ulna together with soft-tissue reconstruction may improve function.

Pulled elbow

Dislocation of the radial head from the annular ligament is a fairly common injury in children under the age of 6 years. There may be a history of the child being jerked by the arm and subsequently complaining of pain and inability to use the arm. The arm is held more or less immobile with the elbow fully extended and the forearm pronated; any attempt to supinate the forearm is resisted. The diagnosis is essentially clinical, although radiographs are usually obtained to exclude a fracture.

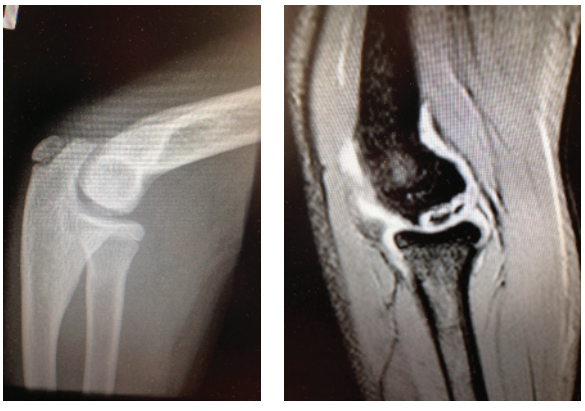
If the history and clinical picture are suggestive, an attempt should be made to reduce the subluxation or dislocation. Simple analgesia is provided and, while the child's attention is diverted, the elbow is extended and pronated (the alternative technique of supination and flexion is more painful and less successful); the radial head is relocated with a snap. The child should be re-examined after 15 minutes and should be using the arm comfortably. The parents should be advised of a 5% risk of redislocation.

OSTEOCHONDRITIS DISSECANS

The capitellum is one of the common sites of osteochondritis dissecans. This may be due to repeated stress but can occur spontaneously. The diagnosis should be excluded in any active adolescent complaining of lateral elbow pain. They may complain of loss of range of movement, usually equal loss of flexion and extension, due to an effusion of the elbow. A history of locking may be elicited due to a loose body in the elbow from fragmentation of the articular surface.

X-rays may show fragmentation or, at a later stage, flattening of the capitellum (Figure 14.7). *CT and MRI* are more useful for defining the lesion.

Treatment is symptomatic. The lesion can heal and symptoms resolve with a repeat MRI at 3 months to assess resolution. If the patient has a loose body, this should be removed arthroscopically. A stable osteochondral lesion measuring less than 4 mm in depth on CT can be fixed back.



(a) (b)

Figure 14.7 Osteochondritis dissecans (a) The capitellum is fragmented and slightly flattened. (b) Sometimes the fragment separates and lies in the joint.

LOOSE BODIES

Loose bodies in the elbow may be due to: (1) acute trauma; (2) osteochondritis dissecans; (3) synovial chondromatosis; or (4) osteoarthritis. The patient may complain of sudden locking of the joint in a flexed position. This may be followed by stiffness due to an effusion. A loose body is rarely palpable.

Loose bodies may be visible on plain *X-rays*. In osteochondritis dissecans the fragmentation of the capitellum may be visible and the radial head may be enlarged. *MR arthrogram* is the investigation of choice in most cases to determine the number of loose bodies present (Figure 14.8). In osteoarthritis *CT* may be used to identify where osteophytes may be limiting movement.

Loose bodies are most easily removed arthroscopically but open surgical techniques can be used.

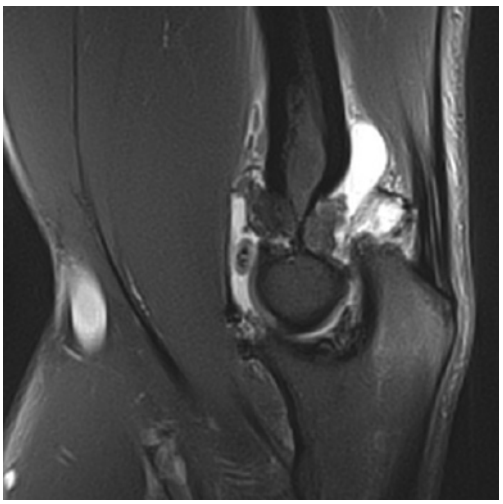


Figure 14.8 Loose body MR arthrogram showing a loose body in the elbow joint.

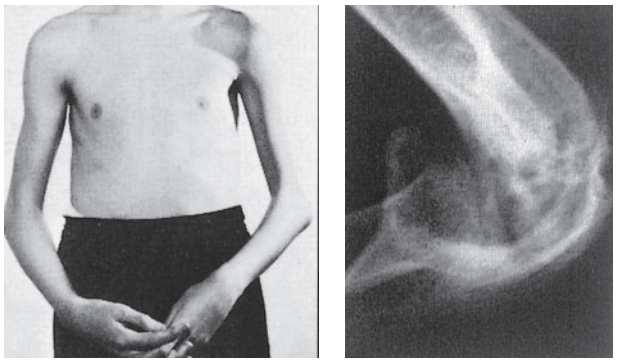
TUBERCULOSIS

Clinical features

The elbow is affected in about 10% of patients with skeletal tuberculosis (Figure 14.9). Although the disease begins as synovitis or osteomyelitis, patients are rarely seen until arthritis supervenes. The onset is insidious with a long history of aching and stiffness. The most striking physical sign is the marked wasting. While the disease is active the joint is held flexed, looks swollen, and feels warm and diffusely tender; movement is considerably limited and accompanied by pain and spasm. Always palpate for supratrochlea and axilliary lymph nodes; they may be enlarged.

X-rays

These show typical features of periarticular osteoporosis and joint erosion. There may also be subchondral cystic lesions.



(a) (b)

Figure 14.9 Tuberculosis of the elbow Muscle wasting is marked (a) and bone destruction extensive (b).

Other investigations

Aspiration, synovial biopsy and microbiological investigation will usually confirm the diagnosis.

Treatment

General antituberculous treatment is essential. The elbow is rested in a splint positioned at 90 degrees of flexion until the acute symptoms subside. Later, a collar and cuff may suffice and movement is encouraged.

Chronic pain, stiffness or deformity may be troublesome enough to justify elbow replacement. Arthrodesis is an option but there is no suitable position to fuse an elbow for good function.

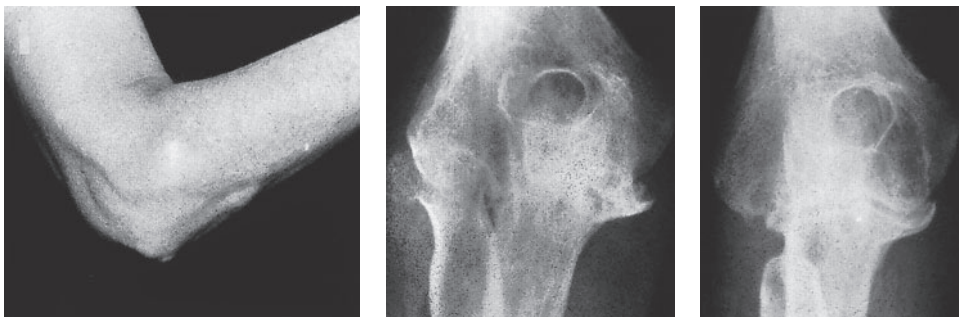
RHEUMATOID ARTHRITIS

The elbow is involved in more than 50% of patients with polyarticular rheumatoid arthritis, and in the majority of cases the condition is bilateral. With improvements in medical management the need for surgical treatment is on the decline.

Clinical features

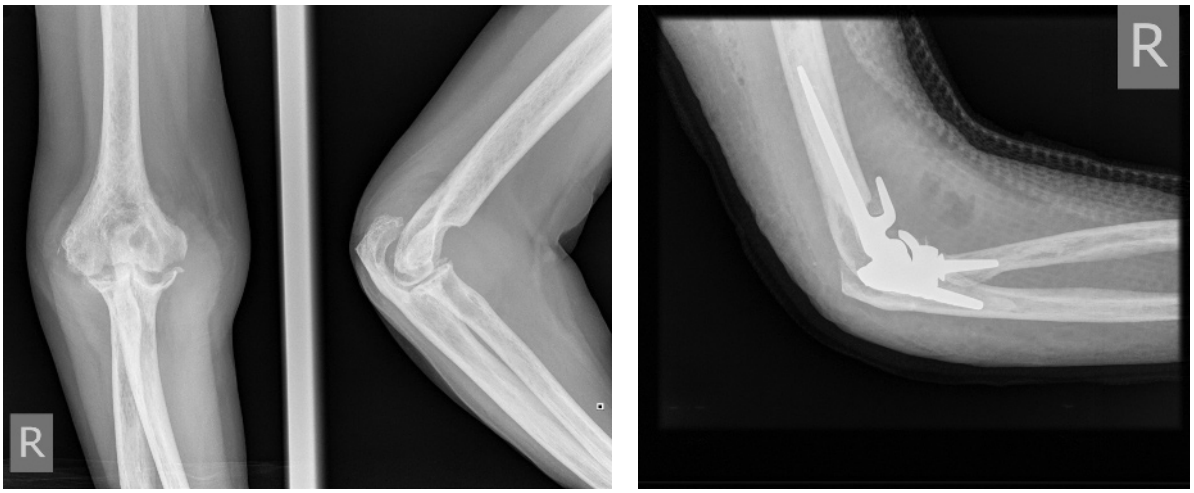
Olecranon bursitis and rheumatoid nodules are often found on the back of the elbow even if the joint itself is not affected. With joint involvement synovitis gives rise to pain, tenderness and swelling, especially over the lateral aspect of the radiohumeral joint (Figure 14.10). Later the entire elbow may be swollen. Movement becomes restricted and, if bone destruction is marked, the joint becomes unstable.

Synovial swelling occasionally causes ulnar nerve or posterior interosseous nerve compression, with symptoms and signs in the wrist and hand. It is important to distinguish these features from those of local weakness and tendon ruptures due to generalized disease.



(a) (b) (c)

Figure 14.10 Rheumatoid arthritis (a) This rheumatoid patient has nodules over the olecranon and a bulge over the radiohumeral joint; (b) his X-rays show deformity of the radial head and marked erosion of the rest of the elbow. (c) Excision of the radial head combined with synovectomy relieved the pain and improved elbow movement.



(a) (b)

Figure 14.11 Total elbow replacement (a) Severe rheumatoid arthritis of the elbow. (b) X-ray after joint replacement.

X-rays

X-rays reveal periarticular osteopenia, bone erosions, with gradual destruction of the radial head and widening of the greater sigmoid notch of the ulna. In some, large synovial extensions penetrate the articular surface and appear as 'cysts' in the proximal radius and ulna. Increasingly, due to improved medical care, rheumatoid patients present with a radiographic picture that more closely resembles osteoarthritis.

Treatment

NON-OPERATIVE TREATMENT

In addition to general treatment with a ladder of anti-rheumatoid medication, the elbow should be splinted during periods of active synovitis. Local injections of corticosteroid preparations may reduce pain and swelling – at least temporarily.

OPERATIVE TREATMENT

If, despite adequate conservative treatment, painful synovitis persists, synovectomy is worthwhile. This can be performed as an open or arthroscopic procedure. The open procedure is performed through a lateral approach and is often combined with radial head excision when the head is eroded by synovitis. This permits greater access to the elbow joint. Arthroscopy allows a more thorough synovectomy, but boundaries between synovium, capsule and muscle are often poorly defined so the risk of nerve injury is high. These operations relieve pain but rarely slow progression of the disease. After 5–6 years, progressive erosion results in instability of the ulnohumeral joint and a recurrence of pain.

Progressive joint destruction and instability may call for reconstructive surgery. Excision arthroplasty and fusion are rarely employed because they produce poor functional outcomes. Joint replacement (Figure 14.11) is usually successful in relieving pain and maintaining a functional range of movement. Approximately 85% of implants will last to a minimum of 10 years, but the operation is difficult to perform and prone to complications such as infection, instability, dislocation, ulnar neuropathy and aseptic loosening of the implants.

GOUT AND PSEUDOGOUT

The elbow is a favourite site for gout. Tophi may appear as nodules on the extensor aspect of the forearm near the elbow, and the olecranon bursa can become rapidly painful, swollen and inflamed in an acute attack. The erythema and swelling may spread and are easily mistaken for cellulitis or joint infection. The serum uric acid level may be raised but a normal level does not exclude the diagnosis. Aspirate from the bursa will contain urate crytals but meticulous technique must be followed to prevent sinus formation or superinfection.

Treatment is with high dose anti-inflammatory preparations.

Similar attacks occur in pseudogout, due to the deposition of calcium pyrophosphate crystals, which can be identified on aspirate.

Chronic calcium pyrophosphate arthropathy should always be suspected when 'osteoarthritic' changes appear spontaneously in an unusual site such as the elbow; X-rays may show additional features such as chondrocalcinosis and periarticular calcification (Figure 14.12). The diagnosis can be confirmed by demonstrating the typical positively birefringent crystals in fluid aspirated from the joint. Treatment is as for osteoarthritis.

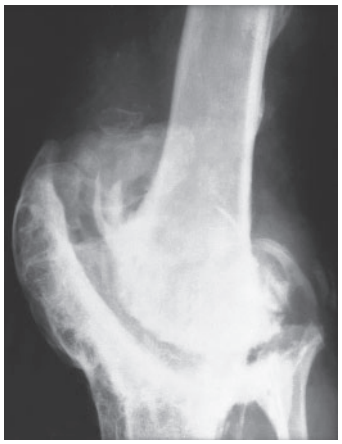


Figure 14.12 Pyrophosphate arthropathy
Destructive arthritis and typical flared osteophytes in a patient with generalized pyrophosphate arthropathy.

OSTEOARTHRITIS

Symptomatic osteoarthritis of the elbow is uncommon and is frequently associated with previous pathology – fracture, dislocation, loose body, osteochondral defect in adolescence, inflammatory arthritis or gout. Primary osteoarthritis of the elbow is more common in male manual workers and frequently presents in the sixth decade of life. When part of a polyarticular disorder, calcium pyrophosphate deposition disease should be considered.

Clinical features

The most common complaint from patients with elbow osteoarthritis is loss of range of movement.

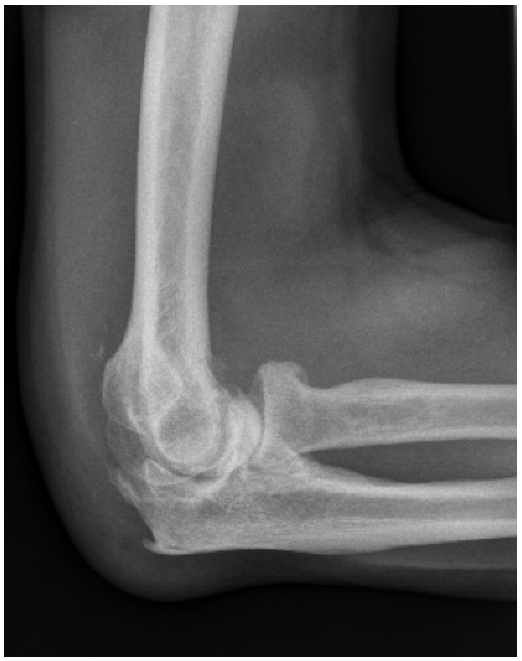


Figure 14.13 Osteoarthritis X-ray with osteophytes, joint narrowing, sclerosis and loose bodies typically seen after trauma or in manual workers.

Loss of extension is cosmetically unappealing when walking and loss of flexion may inhibit functions such as shaving, brushing teeth and eating. Pain is less common but may be present – most commonly this is lateral elbow pain arising from an arthritic radiocapitellar joint. Examination reveals local tenderness, swelling, crepitus and restriction of movement. Signs of ulnar nerve entrapment may be present due to osteophytes encroaching on the cubital tunnel.

X-rays

These show joint space narrowing (particularly of the radiocapitellar joint), sclerosis, osteophytes and frequently loose bodies (Figure 14.13). In advanced arthritis the radial head subluxates anteriorly due to loss of congruence with the capitellum. Chondrocalcinosis and periarticular calcification are typical of pyrophosphate arthropathy.

Treatment

This is aimed at symptom control. Pain is addressed with activity modification, and non-steroidal anti-inflammatory medication and intra-articular steroid injections may provide relief for painful flares. Stiffness does not usually require treatment but, if there are functional limitations, arthroscopic or open joint debridement can be performed with removal of osteophytes and capsular release. Loose bodies are most easily removed arthroscopically. Ulnar nerve entrapment may require *in situ* decompression and cheilectomy (removal of osteophytes) or subcutaneous ulnar nerve transposition.

In advanced cases where pain cannot be controlled in any other way, joint replacement or prosthetic arthroplasty can be considered, but patients are advised to limit use of the arm to light activity only to reduce the risk of implant loosening, the most common cause for joint revision.

Application of a temporary external fixator with joint distraction with or without interposition arthroplasty (implanting soft-tissue grafts between the bone ends) may delay the need for a joint replacement but increase the risks for any subsequent arthroplasty procedure, and the patient should be counselled accordingly.

NEUROPATHIC ARTHROPATHY

Neuropathic arthritis is seen in syringomyelia and diabetes mellitus. Sometimes neurological features predominate and the diagnosis may be known; occasionally the patient presents with progressive instability of the elbow as the architecture is eroded. The joint may be markedly swollen and hypermobile, with

coarse crepitation on passive movement, or it may be completely flail.

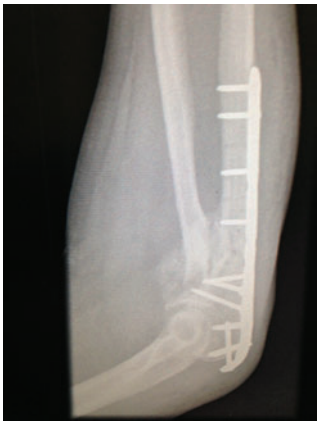
The condition must be distinguished from other causes of flail elbow, such as advanced rheumatoid arthritis and unreduced fracture dislocations.

Treatment

Treatment consists of splintage to maintain stability. Arthrodesis usually fails and is functionally disabling. Joint arthroplasty is technically difficult and prone to early failure in this setting.

STIFFNESS OF THE ELBOW

Stiffness of the elbow may be due to *congenital abnormalities* (synostosis or arthrogryposis), *infection*, *inflammatory arthritis*, *osteoarthritis* or the late effects of *trauma* (Figure 14.14). Most of these conditions are dealt with in other chapters. Here consideration is given to post-traumatic stiffness, which is an important cause of stiffness.



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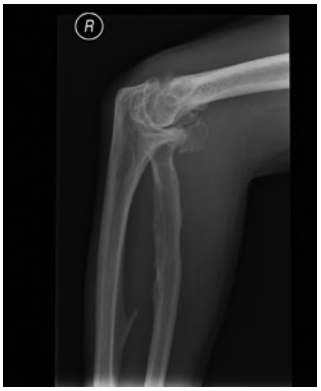

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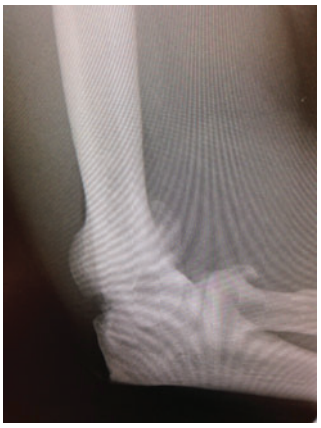

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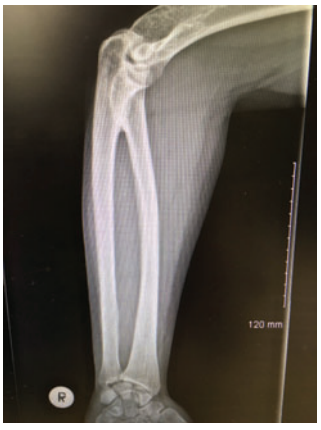


(d)

Figure 14.14 Elbow stiffness (a) Heterotopic ossification; (b) post-trauma; (c) osteoarthritis; (d) congenital synostosis.

Post-traumatic stiffness

The elbow is particularly prone to post-traumatic stiffness. Identifiable causes can be either extrinsic (e.g. soft-tissue contracture or heterotopic bone formation), intrinsic (e.g. intra-articular adhesions or articular incongruity) or a combination of these. Clinical assessment should include examination of all the joints of the upper limb as well as an evaluation of the functional needs of the patient. Elbow movement should be assessed with the use of a goniometer. Most of the activities of daily living can be managed with a restricted range of elbow motion: flexion from 30 to 130 degrees and pronation and supination of 50 degrees each. Any greater loss is likely to impair function.

Treatment

NON-OPERATIVE TREATMENT

The most effective treatment is prevention, by early movement through a functional range. Prolonged immobilization, even beyond a week, can promote stiffness. If motion does not improve after injury, serial splinting may help. Physiotherapy aids recovery with active open and closed chain exercises. Passive manipulation of the elbow should be avoided.

OPERATIVE TREATMENT

The indication for operative treatment is failure to regain a functional range of movement at 6 months after injury, as range is unlikely to improve significantly

over that time. There are a few caveats: the limb as a whole should be useful; there should be no overriding neurological impairment; and the patient should be cooperative and motivated.

Surgical treatment is dictated by the identified cause of stiffness. Capsular release or capsulectomy (open or arthroscopic) may restore a satisfactory range of movement. If X-rays show that bone incongruity is blocking motion, this must be addressed. Consideration should be given to instability and non-union as causes of stiffness, and to neurological causes such as ulnar nerve entrapment. Heterotopic ossification can be excised before maturation without adverse results but prophylaxis with radiotherapy or indomethacin is required.

Surgery is only a small part of the treatment of stiffness with important consideration given to perioperative pain control, the use of continuous passive motion devices and the importance of appropriate early physiotherapy.

Post-traumatic radioulnar synostosis sometimes follows internal fixation of fractures of the radius and ulna. It is treated by resection when the synostosis has matured, followed by diligent physiotherapy.

RECURRENT ELBOW INSTABILITY

Recurrent elbow instability should be categorized as posterolateral rotatory instability (PLRI), valgus extension overload or recurrent frank dislocation, which is rare. PLRI is the most common recurrent elbow instability and occurs as a result of injury to the lateral collateral ligament complex of the elbow, which permits the forearm to rotate posterolaterally off the humerus with subluxation of the radial head (Figure 14.15). Lateral elbow pain is common and may be associated with locking or 'clunking' of the elbow. Various tests have been described including the varus stress test, push-up test and table-top test.

Treatment

The lateral collateral ligament can be repaired, if identified acutely, or reconstructed with tendon autograft, allograft or synthetic graft with satisfactory results.

TENDINOPATHIES AROUND THE ELBOW

The elbow is prone to painful maladaptive degeneration of the tendon attachments. These are typically of insidious onset but precipitating factors may be identified from a careful history.

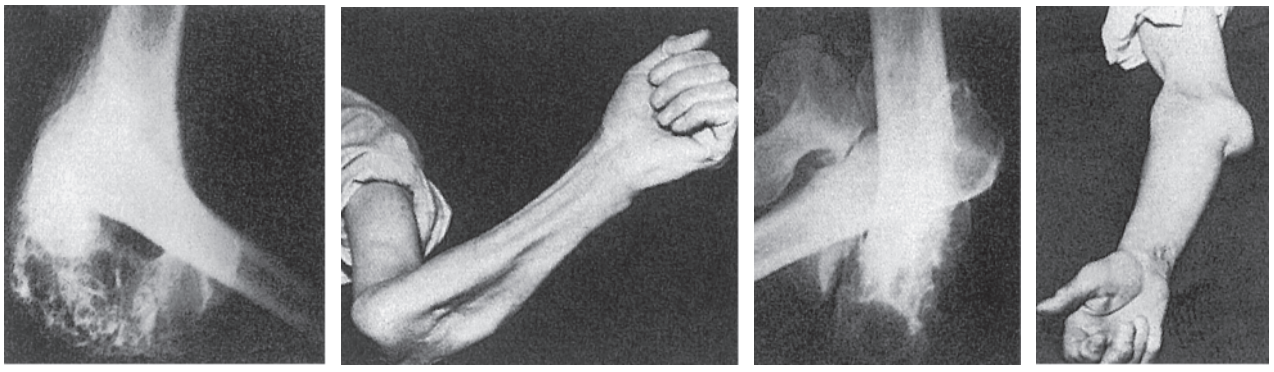
LATERAL EPICONDYLOSIS (TENNIS ELBOW)

This is the most common tendinopathy, with pain arising from the origin of extensor carpi radialis brevis (ECRB) at the lateral epicondyle (Figure 14.16). While tennis players may be susceptible, most cases occur in those without a history of racquet use. Sudden traumatic onset is suggestive of an acute tendon rupture that often requires surgical repair.

Clinical features

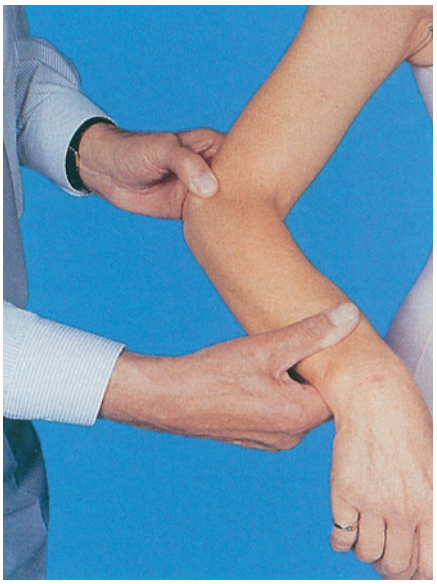
The patient is usually an active person aged 40 to 55. Lateral elbow pain comes on gradually, often after a period of unaccustomed activity. Radiation down the forearm and elbow stiffness first thing in the morning are common. The pain is often aggravated by lifting, such as lifting a laptop out of a bag, lifting a kettle or shaking hands. The elbow usually looks normal but slight swelling may be visible. The patient will typically have a full range of movement.

Tenderness is elicited by palpation over the front of the lateral epicondyle and by provocative manoeuvres such as resisted middle finger extension (Maudsley's test) or elbow extension in pronation with a flexed wrist (Mill's sign).



(a) (b) (c) (d)

Figure 14.15 Flail elbow (a,b) Following gunshot wound; (c,d) neuropathic arthritis.



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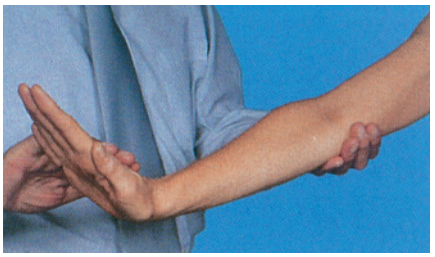

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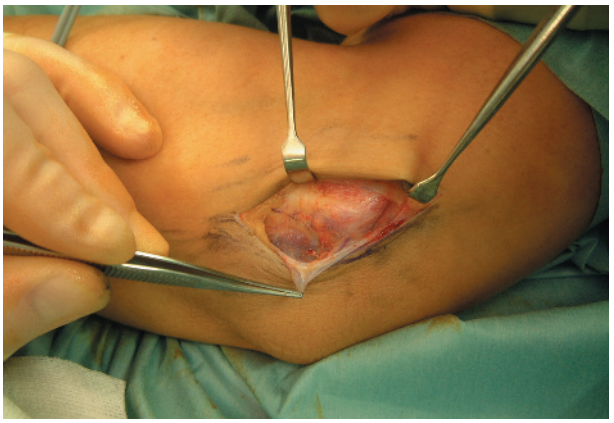


(c)

Figure 14.16 Tennis elbow (a) Tenderness over the anterior aspect of the lateral epicondyle; (b) pain provoked by resisted wrist extension; (c) tennis elbow surgery – the abnormal extensor carpi radialis brevis origin is debrided.

Imaging is not required except at the extremes of age, with a history of trauma, or a history of mechanical symptoms such as locking.

Diagnosis

In patients with long-standing symptoms that do not respond to treatment, the possibility of a painful radial tunnel syndrome, radiocapitellar plica or PLRI should be considered. Ultrasound or MRI can be used to confirm the diagnosis.

Treatment

The natural history for tennis elbow is for spontaneous resolution within 12 months in 90% of cases.

NON-OPERATIVE TREATMENT

Activity modification and physiotherapy will speed up recovery. This means identification and avoidance of precipitating factors, and an eccentric loading regime for the common wrist extensors. A static wrist splint may help with pain control. Steroid injection will provide short-term pain relief but recurrence rates are high and the elbow is more likely to be painful in the long term. Autologous blood product injection may have a role.

OPERATIVE TREATMENT

If the pain persists despite adequate non-surgical measures, tendon debridement can provide pain relief in approximately 85% of cases. Surgery can be performed as an open procedure with an average of 6 weeks recovery, or using arthroscopic technique with a faster recovery of 2 weeks but a higher risk of nerve injury.

Medial epicondylosis (golfer's elbow)

This is similar to tennis elbow but only one-fifth as common and more resistant to treatment. In this case it is the pronator teres origin that is affected. Tenderness over the medial epicondyle and pain on resisted forearm pronation in extension are diagnostic. Ulnar nerve entrapment and medial ligament injury should be excluded.

Treatment is as for tennis elbow, but the outcome of surgery is less predictable and more prone to complications.

Distal biceps tendinopathy

This is an increasing problem due to the growing number of individuals going to the gym to lift weights into their forties and fifties. It is one of the few causes of anterior elbow pain, with pain reproduced on resisted forearm supination.

Treatment is as for tennis elbow, except that, if surgery is required, the tendon is reattached after debridement (a substantial undertaking).

Baseball pitcher's elbow

Repetitive, vigorous throwing activities can cause damage to the bones of soft-tissue attachments around the elbow. In adolescents traction apophysitis of the medial epicondyle, or little leaguer's elbow, became so prevalent that limits are now placed on the number of pitches that juniors are permitted to make. *Treatment* is rest and avoidance of pitching activities.

In adults, persistent valgus strain causes attenuation of the medial collateral ligament and posteromedial impingement – valgus extension overload. This often requires arthroscopic debridement and medial ligament reconstruction.

AVULSION OF THE DISTAL BICEPS TENDON

The typical patient is a man in his forties who feels sudden pain and weakness at the front of the elbow after strenuous effort. The patient will often report a 'tearing sensation' and that the biceps muscle belly has moved proximally in the arm.

Diagnosis is usually straightforward, with absence of the biceps tendon on palpation of the antecubital fossa with the patient holding the arm flexed with the shoulder abducted to 90 degrees and the forearm supinated (O'Driscoll's hook test). If there is doubt, prompt flexion abduction supination (FABS) MRI or ultrasound is used to investigate.

Treatment

Operative repair is not always necessary, but the patient should be counselled that they will have persistently reduced supination strength and possible persistent cramping pain in the biceps if left untreated. Operative repair is best performed early (within 2 weeks of injury) and numerous techniques have been reported, but button fixation is the strongest repair technique, permitting early rehabilitation. Outcomes are generally good with restoration of strength to near-normal levels.

BURSITIS

The olecranon bursa sometimes becomes enlarged as a result of repeated pressure or friction; this used to be called 'student's elbow'. If the enlargement is a nuisance, the fluid can be aspirated. The bursitis may be a result of infection in one-fifth of cases and differentiation can be difficult.

The most common non-traumatic cause is gout (Figure 14.17); there may be a sizeable lump with calcification on X-ray. In rheumatoid arthritis the bursa may become enlarged and sometimes 'rubbery' nodules can be felt over the proximal ulna. In both conditions other joints are likely to be affected too.

Recurrent olecranon bursitis can be treated by surgical excision but wound healing can be a problem.

OPERATIONS

Arthroscopy

Arthroscopy of the elbow is technically demanding because of the proximity of major nerves.

Indications

The role of elbow arthroscopy is expanding. Current use is for joint washout for infection, removal of loose bodies, capsular release, removal of osteophytes, excision of plica, synovectomy, radial head excision, tennis elbow release and fracture fixation.

Technique

The risk of this operation is devastating injury to the ulnar nerve, median nerve and posterior interosseous nerve, each of which lies less than a centimetre from the joint and very close to the portals used for access. Special training, a thorough knowledge of the anatomy and specialist techniques are required. Synovectomy and capsulectomy carry the greatest risk.

Arthroplasty

A complex anatomy and biomechanics make it more challenging to repeat the success stories of hip and knee replacement, but in circumstances of severe intractable pain or instability it may be the best option available.

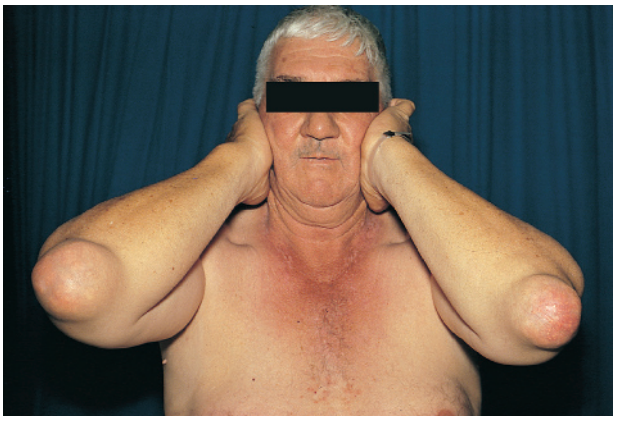


Figure 14.17 Olecranon bursitis The enormous red lumps over the points of the elbows are enlarged olecranon bursae; the ruddy complexion completes the typical picture of gout.

Indications

The primary indication is rheumatoid arthritis but improvements in medical management have resulted in an overall decline in the number of elbow replacements being performed. In trauma, total joint arthroplasty is being superseded by hemiarthroplasty for unreconstructable distal humerus fractures. It is occasionally suitable for the treatment of osteoarthritis. Elbows that are ankylosed (e.g. due to previous infection) can be successfully salvaged with elbow replacement.

One should think carefully before advocating this operation to patients who intend to return to heavy work or leisure activity or to those with single joint involvement.

Design

Earlier constrained (single-axis hinge) implants had a high failure rate due to loosening. Unlinked designs are more technically challenging and demand careful attention to soft-tissue balance due to risks of instability, however 90% good results can be achieved in carefully selected patients (those with good bone stock and competent ligaments). Semi-constrained implants are the most widely used and may allow some of the forces to be absorbed by the soft tissues while maintaining some intrinsic stability.

Outcome

The majority of patients with an elbow replacement can expect relief of pain and a functional range of movement. Ten-year survival rates as high as 80% have been achieved in patients with rheumatoid arthritis (perhaps the joint is protected because of poor function in the rest of the limb) whereas the survival rate for those with osteoarthritis or trauma sequelae is not so good. A good outcome can also be achieved in selected trauma patients (older individuals with low demand) with hemiarthroplasty or total elbow replacement.

Complications

The operation has a relatively high complication rate, particularly ulnar nerve palsy, aseptic loosening and infection.

Arthrodesis

Arthrodesis of the elbow is rarely indicated. It is technically difficult and very disabling. Even with normal wrist and shoulder function, it is not possible to fuse the elbow in a position that would facilitate both feeding (i.e. 100 degrees flexion) and perineal hygiene (about 45 degrees flexion). Compression plating is the most straightforward and stable technique.

NOTES ON APPLIED ANATOMY

The elbow needs to be able to convey the hand upwards to the head and mouth, downwards to the perineum and legs, and also to a wide variety of positions for manipulation of the environment around us. Elbow function cannot be considered in isolation from forearm function as both are closely integrated. A varied combination of flexion and extension with pronation and supination is clearly needed. Although the normal elbow is capable of full extension, flexion to about 130 degrees and 90 degrees of both pronation and supination, the *functional range of movement* is 30–130 degrees of flexion and 50 degrees of both pronation and supination.

The forearm is normally in slight valgus relative to the upper arm, the average carrying angle being about 11 degrees. The complex geometry of the joint allows for the fact that, when the elbow is flexed, the forearm comes to lie directly upon the upper arm. The carrying angle may be altered by malunion of a fracture or damage to a physis, resulting in cubitus valgus or cubitus varus.

The joint acts as a 'sloppy hinge', permitting a few degrees of valgus/varus movement and some rotational laxity. Stability is provided by: (1) the relative conformity of the humeral trochlea with the olecranon; (2) the lateral collateral ligament complex; and (3) the medial collateral ligament (particularly the anterior part). The radial head is a secondary constraint to valgus instability; it can be excised when necessary as long as the medial collateral ligament, humeroulnar articulation and interosseous membrane are intact. The elbow is not a 'non-weight-bearing' joint – forces up to three times body weight pass across it with normal use.

Pronation and supination take place at the proximal and distal radioulnar joints, with a small amount of movement in the wrist too. The movement is often supplemented by rotation at the shoulder. The radiocapitellar joint is held in position by the strong annular (orbicular) and collateral ligament that embraces the head and neck of the radius but is not attached to it. The capsule of the elbow is attached to the annular ligament but is not attached to the radius. The circular and slightly concave upper surface of the radius ensures that in all positions of rotation it retains adequate contact with the capitellum.

Nerves

The ulnar nerve passes behind the medial condyle of the humerus; it may be compromised if there is marked cubitus valgus. Distal to the condyle the nerve is closely applied to the elbow capsule, and there also it may be compromised if the joint is osteoarthitic.

On the lateral side of the elbow the radial nerve passes between brachialis and brachioradialis. It then splits to become the sensory superficial radial nerve and motor posterior interosseous nerve. The latter passes beneath extensor carpi radialis brevis and then between two parts of the supinator muscle; it is vulnerable to injury during surgical approaches to the proximal part of the radius.

In front of the elbow lie the brachialis muscle and also the median nerve in company with the great vessels; these relationships make an anterior approach to the elbow challenging.

The wrist

15

David Warwick & Roderick Dunn

The wrist and hand function together, for all practical purposes, as a single articulated unit. The hand would be unable to perform its range of complicated movements without the reciprocal movement, positioning and stabilizing action of the wrist. Loss of movement at the wrist limits the manipulative skill of the fingers and thumb; and pain in the wrist makes it impossible to grip or pinch with full strength. Disorders of the wrist and hand are often interrelated and therefore, in the clinical setting, these two units should be examined and analysed together. However, for the sake of emphasis, they are treated here in two separate chapters.

CLINICAL ASSESSMENT

Symptoms

Pain may be localized to the radial side (especially in de Quervain's disease and thumb base arthritis), to the ulnar side (e.g. in distal radioulnar joint arthritis and pisotriquetral arthritis) or to the dorsum (in radiocarpal arthritis, Kienböck's disease and occult dorsal wrist ganglion).

Stiffness is often not noticed until it is severe in the flexion–extension plane; loss of rotation is noticed earlier and can be very disrupting.

Swelling may signify involvement of either the joint or the tendon sheaths or a ganglion.

Deformity is a late symptom except after trauma or radial nerve palsy. Ask if it is localized to a particular site (e.g. an overly prominent head of ulna, suggesting subluxation of the distal radioulnar joint) or involving the posture of the wrist as a whole (progressive radial deviation in advanced rheumatoid arthritis, RA).

Loss of function refers mainly to the hand, though the patient may be aware that the problem lies in the wrist.

Clicks are common and usually of no relevance; *clunks* with pain or weakness may signify instability.

Signs

Examination of the wrist is not complete without also examining the elbow, forearm and hand. Both upper limbs should be completely exposed.

Look

The skin is inspected for scars. Swelling, sweating, colour changes and waxiness suggest complex regional pain syndrome. Both wrists and forearms are compared to see if there is any deformity. If there is swelling, note whether it is diffuse or localized to one of the tendon sheaths. Look also at the hands and fingers to see if there are any related abnormalities.

The posture of the wrist at rest and during movement varies with different positions of the hand and fingers. This is discussed in the opening sections of Chapter 16.

Feel

Palpation of the wrist will yield valuable information only if the surface anatomy is thoroughly understood (see Figure 15.1). Tender areas must be accurately localized

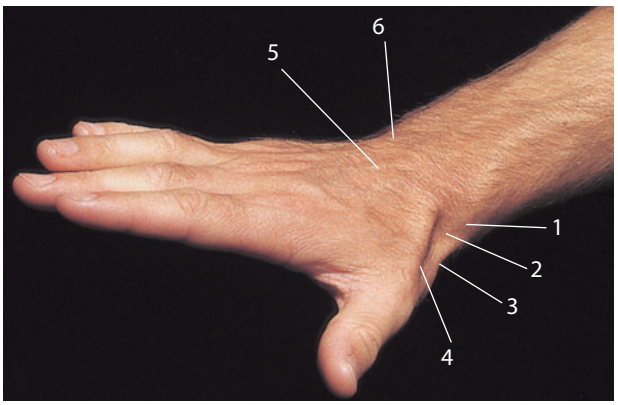
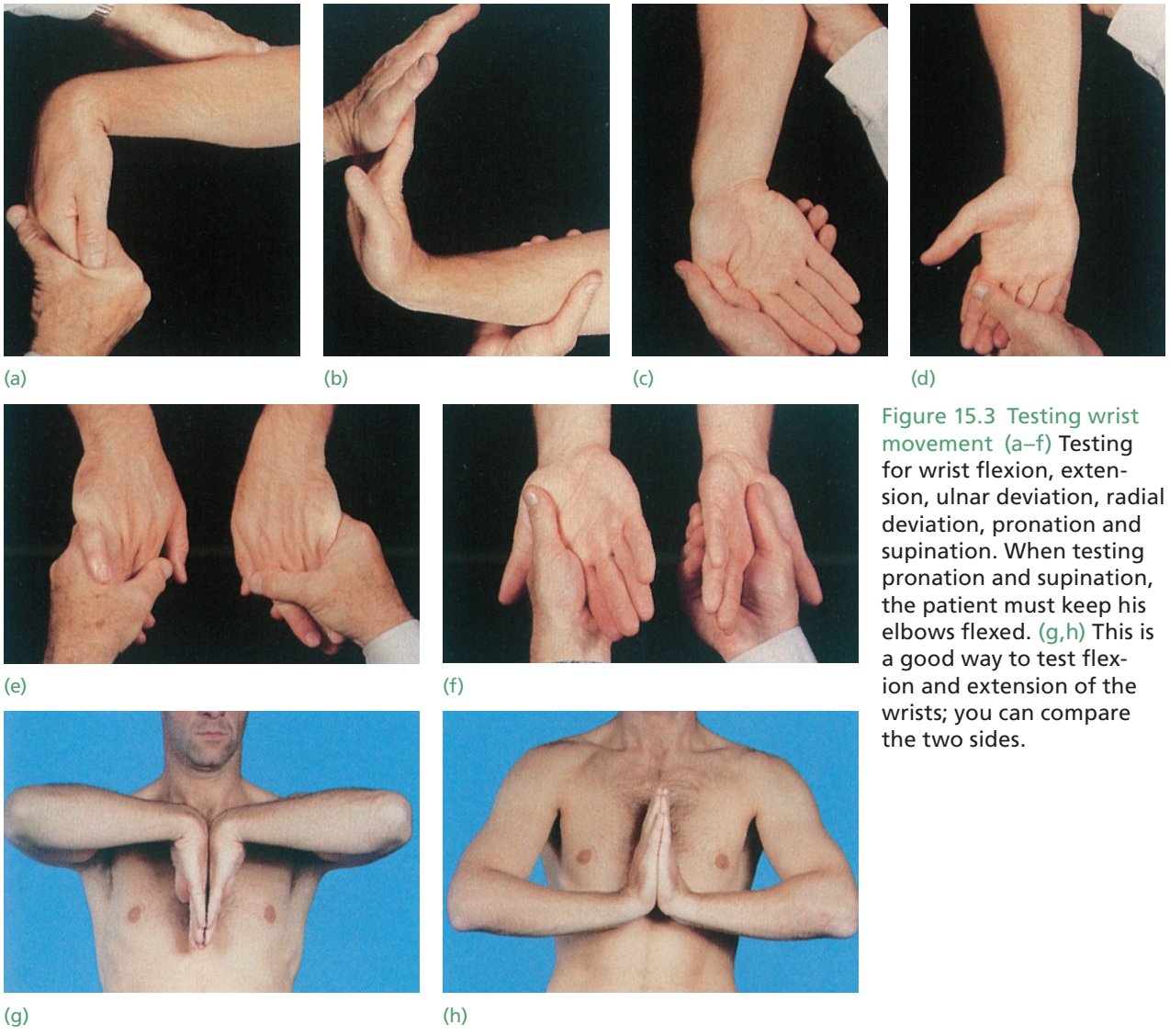


Figure 15.1 Tender points at the wrist (1) Tip of the radial styloid process; (2) anatomical snuffbox, bounded on the radial side by the extensor pollicis brevis (3) and on the ulnar side by the extensor pollicis longus (4); (5) the extensor tendons of the fingers; (6) the head of the ulna.

(a) (b) (c)

Figure 15.2 Radial corner pain (a) Tenderness at the tip of the radial styloid suggests de Quervain's disease (tenovaginitis of the combined sheath for extensor pollicis brevis and abductor pollicis longus). This diagnosis can be confirmed by Finkelstein's test. Hold the patient's hand with his thumb tucked firmly into the palm; then turn the wrist into full ulnar deviation; in a positive test, this will elicit sharp pain in the affected sheath. (b) Tenderness in the anatomical snuffbox is typical of a scaphoid injury. (c) Tenderness just distal to the head of the ulna is found in extensor carpi ulnaris tendinitis.



(a) (b) (c) (d)

(e) (f)

(g) (h)

Figure 15.3 Testing wrist movement (a–f) Testing for wrist flexion, extension, ulnar deviation, radial deviation, pronation and supination. When testing pronation and supination, the patient must keep his elbows flexed. (g,h) This is a good way to test flexion and extension of the wrists; you can compare the two sides.

and the various landmarks compared with those of the normal wrist. The site of tenderness may be diagnostic (Figure 15.2): for example in de Quervain's disease (tip of radial styloid), scaphoid fracture (anatomical snuffbox), carpometacarpal osteoarthritis (base of first metacarpal), Kienböck's disease (over the lunate), triangular fibrocartilage complex (over the head of the ulna and in the dent on the ulnar side of the wrist just palmar to the ulnar styloid – the foveal sign) and localized tenosynovitis of any of the wrist tendons. At the same time note if the skin feels unduly warm or sweaty.

If the head of the ulna seems abnormally prominent on the dorsum of the wrist, stress the distal radioulnar joint by pressing down firmly on the ulnar prominence; if it moves up and down, the joint is unstable (this is aptly named the 'piano-key sign').

Move

See Figures 15.3 and 15.4.

Passive movements To compare passive dorsiflexion of the wrists the patient places his palms together in the position of prayer, then elevates his elbows. Palmar flexion is examined in a similar way. Radial and ulnar deviation are measured in either the palms-up or the palms-down position. With the elbows at right angles and tucked in to the sides, pronation and supination are assessed.

While testing passive movements, the presence of abnormal 'clunks' should be noted; they may signify some form of carpal instability.

Active movements Ask the patient to pull the hand backwards to its limit (*extension*), then forwards as far as possible (*flexion*), and then sideways to right and left (*radial and ulnar deviation*). Active *pronation and supination* should be performed with the patient's elbows tucked tightly into the waist. These movements are then repeated but carried out against resistance, to test for muscle power. Finally, grip strength is measured, preferably using a mechanical dynamometer. Loss of power may be due to pain, tendon rupture or muscle weakness.

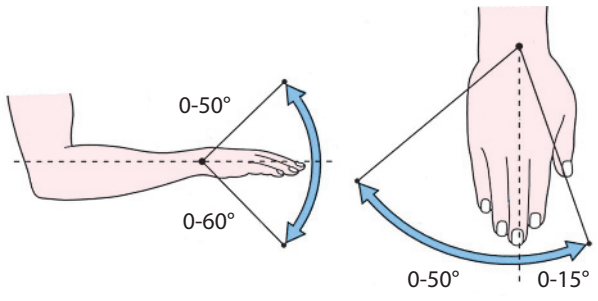


Figure 15.4 Normal range of movement From the neutral position dorsiflexion is slightly less than palmarflexion. Most hand functions are performed with the wrist in ulnar deviation; normal radial deviation is only about 15°.

Provocative tests

Special tests are needed to assess stability of the carpal articulations. The *lunotriquetral joint* is tested by pinching the lunate with one hand, the triquetral–pisiform with the other, and then applying a sheer stress: pain or clicking suggests an incompetent lunotriquetral ligament. The *pisotriquetral joint* is tested by pushing the pisiform radialwards against the triquetrum. Stability of the *scapholunate joint* is tested by pressing hard on the palmar aspect of the scaphoid tubercle while moving the wrist alternately in abduction and adduction: pain or clicking on abduction (radial deviation) is abnormal (Watson's sign). The central portion of the *triangular fibrocartilage* is tested by pushing the wrist medially then flexing and extending it under load to elicit pain (the grind test). The *distal radioulnar joint* is tested for stability by holding the radius and then balloting the ulnar head up and down. These tests are mentioned again in the section on carpal instability.

Imaging

X-rays

Anteroposterior and lateral views are obtained routinely. Note the position and shape of the individual carpal bones and whether there are any abnormal spaces between them (Figure 15.5). Examine the congruency of the proximal and midcarpal joint (Gilula's lines).

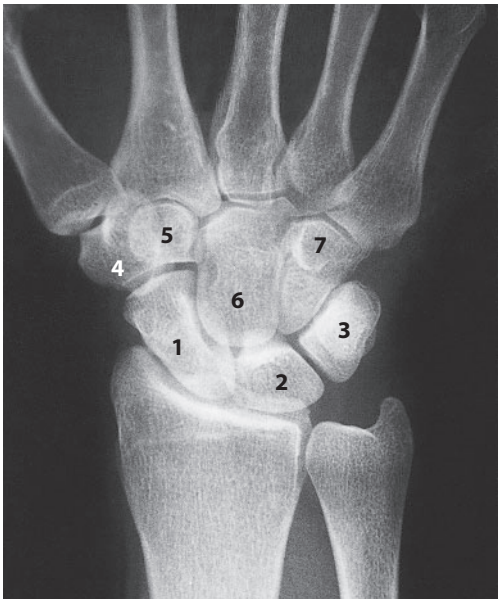


Figure 15.5 X-ray Note the shape and position of the bones which make up the normal carpus: (1) scaphoid, (2) lunate, (3) triquetrum overlain by pisiform, (4) trapezium, (5) trapezoid, (6) capitate, (7) hamate.

Then look for evidence of joint space narrowing, especially at the radiocarpal joint and the carpometacarpal joint of the thumb. The wrist X-ray should be taken in a standard position of mid-pronation with the elbow at 90 degrees; often both wrists must be X-rayed for comparison. Special views may be necessary to show a scaphoid fracture or carpal instability. Moving the wrist under image intensification is useful to investigate some cases of carpal instability.

Computed tomography

CT is the ideal method for assessing congruity of the distal radioulnar joint, fractures of the hook of hamate, and alignment of scaphoid fractures prior to surgery for non-union or malunion.

Magnetic resonance imaging

MRI is particularly useful for detecting changes associated with scaphoid fractures, avascular necrosis of the lunate (Kienböck's disease), occult dorsal ganglia and intraosseous ganglia. The thickness of the cuts may be too large to detect injury to thin structures such as the lunotriquetral ligament, scapholunate ligament or triangular fibrocartilage but modern scanners have ever-increasing sensitivity and specificity.

Arthrography

The wrist contains three separate compartments – the radiocarpal joint, the distal radioulnar joint and the midcarpal joint. Defects in the triangular fibrocartilage, scapholunate ligaments or lunotriquetral ligaments can be identified by arthrography. High-resolution MRI or CT arthrography is effective in showing cartilage loss, TFCC perforations and interosseous ligament tears.

Radionuclide scan

A localized area of increased activity may reveal an osteoid osteoma, an occult scaphoid fracture or early osteoarthritis. This test has been surpassed by MRI.

Fluoroscopy

Fluoroscopic examination may be needed to demonstrate some patterns of carpal instability.

Arthroscopy

The wrist is suspended by finger traps, inflated with saline and inspected through specific portals into the radiocarpal joint, the ulnocarpal joint and the midcarpal joint. Ligament tears, articular cartilage damage, osteoarthritis, occult ganglia, synovitis and triangular fibrocartilage lesions can be recognized and in some cases treated.

CONGENITAL ANOMALIES OF THE WRIST AND HAND

Abnormalities occurring in the upper limb anlage during the first 3 months of embryonic life may affect more than one segment (or indeed the whole) of the developing limb, and congenital anomalies can occur together in the forearm, wrist and hand. For this reason, we have dealt with the subject under a single heading.

The embryonic arm buds appear about 4 weeks after fertilization and from then on the limbs develop progressively in three axes, from proximal to distal, radial to ulnar and dorsal to palmar. By 6 weeks the digital rays begin to appear and then develop in concert with the general mesenchymal differentiation that gives rise to the primitive skeleton and muscles. Growth and apoptosis (genetically programmed cell death) result in modelling of the limbs and the formation of joints and separate digits. The process is more or less complete by the end of the eighth week after fertilization, at which time primary ossification centres begin to appear in the long bones. Ossification centres in the epiphyses and carpal bones do not emerge until after birth, so X-rays in the neonatal period must be interpreted with this in mind.

Malformations may occur during embryonic development because of defective formation or incomplete separation of mesenchymal components, the former accounting for partial or complete absence of a part and the latter for coalitions between adjacent elements.

Congenital limb anomalies can be sporadic or syndromic (associated with other abnormalities). The overall incidence of congenital upper-limb anomalies is estimated to be about 1 in 600 live births, and many are not severe enough to require operative treatment.

Malformations may be caused by heritable genetic mutations, or intrauterine damage (e.g. drugs, infection or ionizing radiation); in the majority of cases the cause is unknown.

Classification

In 2014, the International Federation of Societies for Surgery of the Hand (IFSSH) reclassified congenital hand and upper-limb anomalies, using the Oberg, Manske and Tonkin (OMT) classification. The OMT system replaces the previous Swanson classification, and will be updated regularly on the IFSSH website. OMT uses four main categories: (I) malformations; (II) deformations; (III) dysplasias; (IV) syndromes. The malformations category divides conditions into two main groups: (A) the entire upper limb, and

(B) the hand plate. These groups are then divided into four subgroups of abnormal axis formation/differentiation: (1) proximal to distal axis, (2) radial to ulnar axis, (3) dorsal to ventral axis, and (4) unspecified axis, and there are a number of further subcategories in each.

General considerations

Initial consultation

The parents and child are likely to be anxious, and may previously have been given conflicting information by non-specialist clinicians. There may be issues of maternal guilt, parental anger and resentment, as well as unrealistic expectations about the outcome and possibilities of surgery. It is important to gain the confidence of the family at the initial consultation; remember that these children are likely to be long-term patients.

They must be given a *diagnosis*, an indication of *prognosis*, *reassurance about the future* and a *long-term plan of treatment*, including a *schedule of surgery*, which may require staged operations over a number of years. Many manage well throughout life with untreated congenital anomalies. It follows that in some conditions the indication for surgery is clear, for example release of ring constrictions causing limb ischaemia in a neonate. In others, such as treatment of adolescent camptodactyly, decision making may be more difficult.

Clinical examination

Children are often shy in clinic. For those who are old enough to talk to you, some key questions can help to initiate conversations: for example, who is your best friend at school, what is your favourite subject at school, which sports team do you support?

Psychological factors should always be considered; teasing usually starts around the age of 6, and the early involvement of clinical psychologists may be helpful. The clinic waiting room provides a useful opportunity for families to share experiences, as do patient support groups.

The clinic should be held in a child-friendly setting. White coats, crowded clinic rooms, and other intimidating hospital paraphernalia should be avoided. Toys encourage children to play in an unconstrained manner, which allows close observation of their hand function (it is impossible to make young children demonstrate hand movements). It may be easier to examine a child while he or she is sitting on the parent's lap.

Absence of skin creases at joints indicates lack of movement, either due to lack of motors, or stiffness (tight soft tissues, abnormal or absent joint).

Investigations

It may be useful to obtain *radiographs* of the contralateral normal limb for comparison.

Remember that many congenital wrist and hand anomalies are part of a larger syndrome. Surgeons should not assume that associated conditions have been investigated fully and should always check if this is the case – and, if necessary, refer to other specialists. For example, skeletal, cardiac, haematological, gastrointestinal, renal or craniofacial anomalies are commonly associated with radial dysplasia, and these should be sought. Children with radial dysplasia (and/or thumb hypoplasia) or thumb duplication should also be tested for Fanconi anaemia.

Genetic counselling should be made available for inherited or unusual conditions, and it may be helpful in reaching a diagnosis.

Indications for operative treatment

Whenever the need for operative treatment is considered, four general principles should be borne in mind:

- *Function* – Improved function is the primary goal of surgery. The hand must be considered in the context of the whole upper limb as well as other systemic conditions.
- *Progression of deformity with growth* – Decide whether deformities will progress with growth. For example, syndactyly between digits of unequal length will lead to deviation of the longer toward the shorter digit, and for this reason they should be treated early before deformity becomes established.
- *Appearance* – The hand is second only to the face in self-consciousness of appearance. If it looks normal, a child is more likely to use it normally. If it looks abnormal, the child may hide it away. This principle is known as 'dynamic cosmesis'.
- *Pain* – Most congenital upper-limb malformations are not painful. One exception is ring constriction syndrome, in which the fingertips may be tight and painful with poor soft-tissue cover. Although as a general principle we try to preserve digital length, these are improved by minimal shortening to provide pain-free, durable soft-tissue cover and a more pleasing appearance.

Congenital upper-limb conditions

Transverse deficiency

This can exist anywhere between the shoulder and the phalanges. The most common levels of absence are at the proximal forearm and mid-carpus, then at

the metacarpals and humerus. Associated anomalies are unusual.

Proximal forearm Children adapt surprisingly well to congenital limb absence, particularly as they still have sensibility and proprioception at the distal end of the abbreviated limb, the lack of which limits prosthetic use. Some may learn to use prosthetics, both for function and cosmesis, and uptake is likely to increase with improved technology.

Transverse arrest of digits Children with vestigial digits (*symbrachydactyly* – see Figure 15.8a) have good function, but they can also be treated by microvascular transfer of one or more toes if there are proximal enabling structures available (skin, tendons and nerves) with good outcomes. The results of non-vascularized transfer of a toe phalanx into the existing skin envelope are disappointing, and this technique is falling out of favour.

Intersegmental deficiency

Very rarely an intercalary segment in the upper limb fails to develop and the forearm or hand may be attached directly to the trunk, or the hand is attached to the humerus. This condition, also known as *phocomelia*, may affect more than one limb and is sometimes associated with craniofacial deformities. For the upper limb, there is no satisfactory treatment other than prosthetics.

Radial longitudinal deficiency

Radial dysplasia is rare (incidence approximately 1 in 50 000 live births) and may involve structures from the elbow to the thumb (Figure 15.6). It can be syndromic (Holt–Oram syndrome, Fanconi anaemia, thrombocytopenia absent radius) or sporadic. It does occur as an isolated abnormality but can also be associated with other conditions including vertebral anomalies, anal atresia, cardiovascular anomalies, tracheo-oesophageal fistula, renal anomalies and other limb defects (embodied in the acronym VACTERL).

The infant is born with the wrist in marked radial deviation – hence the use of the term '*radial club hand*' and half are affected bilaterally.

There are four types: short distal radius (Type I), hypoplastic radius (Type II), partial absence of radius (Type III), and complete absence of the radius (Type IV); often the thumb, scaphoid and trapezium also fail to develop normally.

Treatment Mild radial dysplasia is treated from birth by gentle stretching and splintage, best done by the parents. More serious cases can be treated by distraction, prior to radialization with a tension-free soft-tissue correction. This has less effect on growth of the carpus and distal ulna than the older technique of 'centralizing' the ulna on the carpus. Prolonged pin fixation and splintage is still required to avoid recurrence of the deformity.

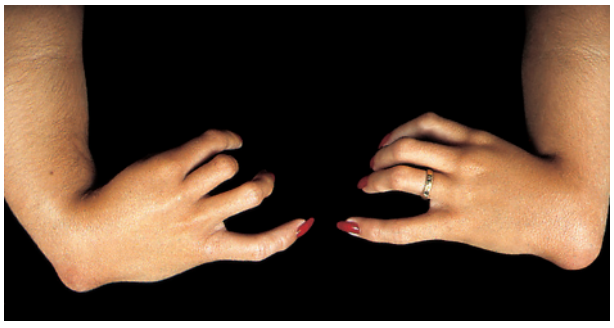
Attention must be paid to the elbow; if the joint is stiff, the radially deviated wrist can actually be advantageous, as the child can then get the hand to his or her mouth (for eating) and the perineum (for toilet care). Surgical correction of the wrist in these cases can result in a functional disaster.

THUMB HYPOPLASIA

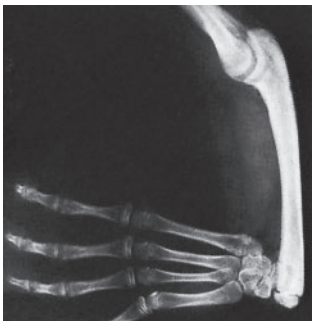
Thumb hypoplasia ranges from a slightly small thumb to complete absence (see Figure 15.8f). Thumbs are reconstructable if there is a basal joint, requiring, in the worst cases, MCP joint stabilization (by ulnar collateral ligament reconstruction or chondrodesis), widening of the first web space using local flaps, and an opposition transfer (usually the Huber transfer using abductor digiti minimi). When there is no basal joint or the thumb is absent, pollicization of the index finger can be performed (as long as the index finger is not also hypoplastic, when it will not make a good thumb).

Ulnar longitudinal deficiency

This is approximately half as common as radial dysplasia (1 in 100 000), and about half of these patients have associated musculoskeletal problems. Although



(a)



(b)

Figure 15.6 Radial longitudinal deficiency (a) Bilateral. (b) X-ray showing that the entire radius is absent.

most cases are sporadic, it is also associated with a few syndromes.

Children may present with ulnar deviation of one or both wrists, due to partial or complete absence of the ulna; in addition, some of the carpal bones may be absent and the ulnar rays of the hand may be missing. With growth the radius elongates disproportionately and becomes bowed; ultimately the radial head may dislocate.

The diagnosis may be difficult when there are absent ulnar digital rays in the hand, and the ulna is present. In such cases, there may also be syndactyly between the radial digits, all of which can be in the plane of the palm, providing only side-to-side pinch (and not opposition) between the thumb and index fingers.

Treatment During the first few months stretching and splinting may be helpful. If wrist deformity and radial bowing are progressive and severe, surgery may be advisable and consists of excision of any tethering ulnar anlage and osteotomy of the radius. If the radial head has dislocated and elbow movement is restricted, the radial head can be excised; if the forearm is unstable, the distal radius can be fused to the proximal ulna to make a one-bone forearm.

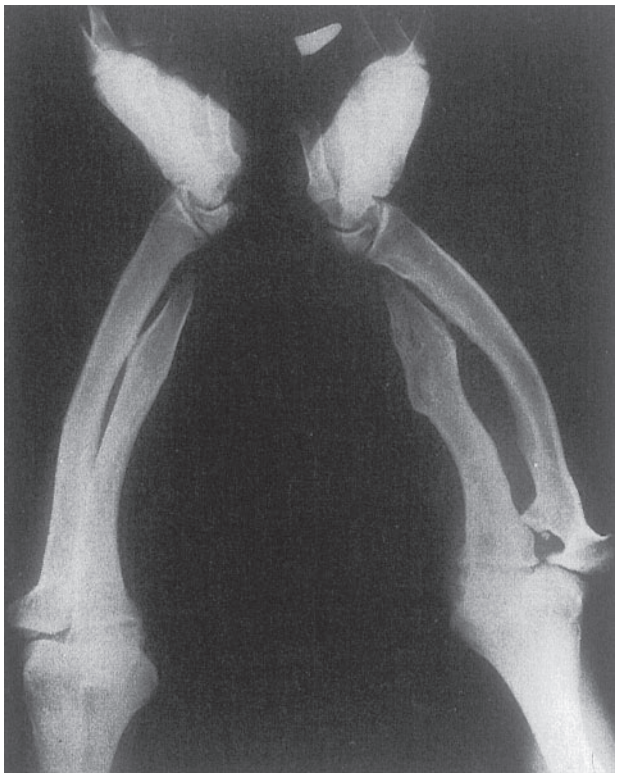


Figure 15.7 Distal ulnar deformity The X-ray characteristically shows a tapering, carrot-shaped distal end of the ulna. This bilateral case was due to hereditary multiple exostoses; there is bilateral bowing of the radius and on the right side the radial head has subluxated.

SECONDARY ULNAR DYSPLASIA

A similar but milder deformity sometimes occurs in children over the age of about 10 years who were born with hereditary multiple exostoses or dyschondroplasia. If the distal ulna is affected in these conditions, growth at the distal physis may be retarded; the distal ulna tapers and is short. If the radius remains unaffected and goes on growing normally, it becomes bowed and the radial head tends to subluxate or dislocate (see Figure 15.7). In most cases the elbow and forearm are completely stable and no treatment is needed (except, possibly, for cosmetic reasons).

Cleft hand (central dysplasia)

True cleft hand presents with a V-shaped cleft in the centre of the hand which may be associated with the absence of one or more digits, a central transverse metacarpal (which widens the cleft with growth), syndactyly of digits bordering the cleft, and a narrow first web space (Figure 15.8d). It is often familial (autosomal dominant inheritance), may be unilateral or bilateral, and can be associated with 'cleft feet'. Other anomalies, such as cleft lip, cleft palate and congenital heart disease, can be present. The condition differs from *symbrachydactyly*, previously known as 'atypical cleft hand', which is not heritable and not associated with other anomalies.

Surgery is complex, having to deal with closure of the cleft (sometimes requiring transposition of metacarpals), reconstruction of the first web space using redundant skin from the cleft, and correction of other anomalies in the adjacent digits.

Polydactyly

Polydactyly ('extra digits') may occur on the radial (pre-axial), the ulnar (post-axial) or the central part of the hand.

Ulnar (post-axial) polydactyly or duplication of the little finger is the most common congenital anomaly of the hand. It is often inherited and is much more common in Afro-Caribbean and Asian populations than in Caucasians. The extra digit is usually only attached by skin pedicle containing the neurovascular bundle, and may be removed under local anaesthesia; this is easiest when the child is less than 4 months old. If a phalanx or entire digit is duplicated, removal and soft-tissue reconstruction should be performed when the child is older, under formal operating theatre conditions.

Radial (pre-axial) polydactyly or thumb duplication (Figure 15.8e,g) usually follows the pattern described by Wassell, with a bifid distal phalanx in type I up to a triphalangeal duplicate thumb and metacarpal

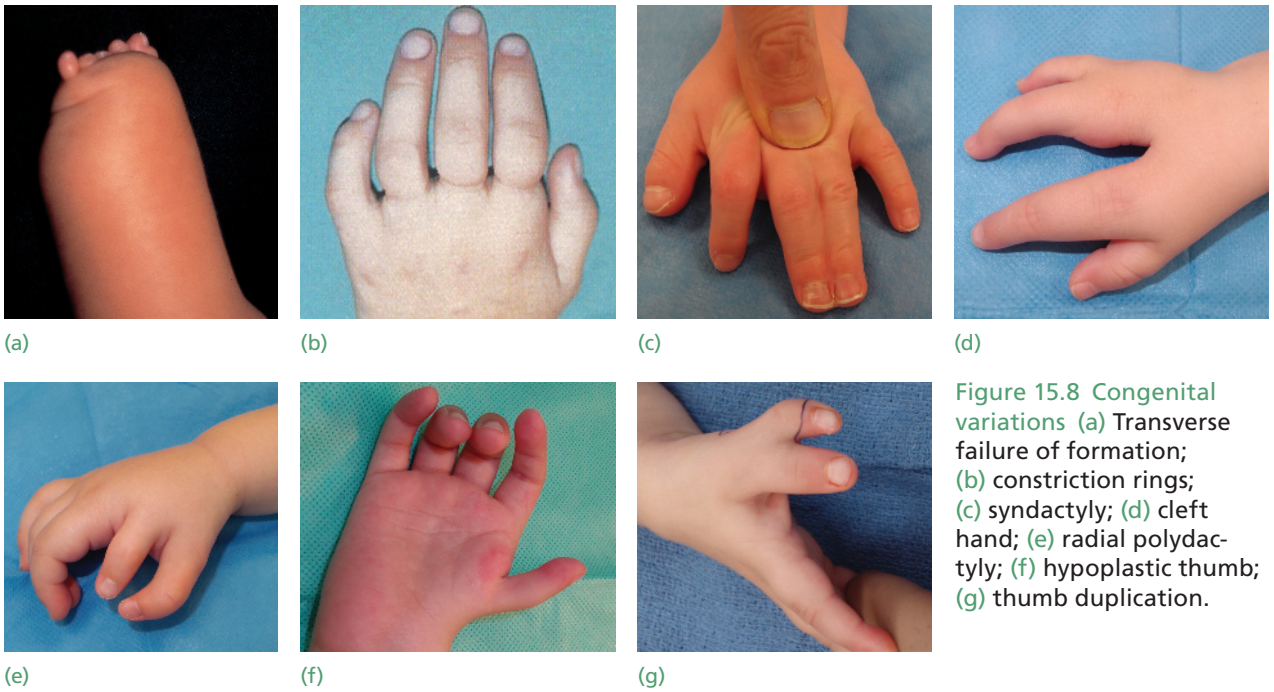


Figure 15.8 Congenital variations (a) Transverse failure of formation; (b) constriction rings; (c) syndactyly; (d) cleft hand; (e) radial polydactyly; (f) hypoplastic thumb; (g) thumb duplication.

in type VII. Duplicate thumb correction is complex and requires corrective osteotomies, preservation of collateral ligaments and tendons, and readjustment of the skin envelope. It is not simply a matter of excising the smaller digit.

Central duplication is rare and familial and may be associated with syndactyly (*synpolydactyly*).

Macrodactyly

Macrodactyly must be distinguished from other forms of enlarged digits (neurofibromatosis, multiple enchondromatosis, vascular malformations). There are two forms: *static* (present at birth and growing proportionately with other digits) and *progressive* (enlargement of a digit in early childhood, growing faster than other digits with deformity).

The condition is rare, and the majority of cases are unilateral, affecting the index, middle, thumb, ring or little finger, in order of frequency. The median or ulnar nerve is often enlarged and may become compressed. Surgical correction is extremely difficult and generally unrewarding. It includes debulking, epiphyseal arrest (when the digit has reached adult size) and nerve excision and grafting. Unfortunately, amputation may often be the best option but this must be handled tactfully, and is of course more difficult when multiple digits are involved.

Brachydactyly

Brachydactyly (undergrowth) is common and may be part of a wider syndrome (e.g. Turner's syndrome). It can affect a single bone, a digit or an entire limb.

Syndactyly

Syndactyly means conjoined digits (Figure 15.8c), and occurs in about 1:2000 live births. It may be *simple* (soft tissue only) or *complex* (skin and bone), *complete* (affecting the entire web) or *incomplete* (only part of the web).

Mild, incomplete syndactyly of central digits may need no treatment. *Treatment* of complete syndactyly involves separation of the conjoined structures, using a dorsal flap to reconstruct the web space, zigzag incisions to separate the fingers and avoid scar contractures, and full-thickness skin grafts for the skin defects. Nail folds can be reconstructed using local flaps as described by Buck-Gramcko.

When multiple digits are involved, only one side of each digit should be separated at a time to avoid potential injury of both digital arteries (which may also be hypoplastic). Syndactyly of border digits (thumb and index, ring and little fingers) can cause progressive deformity with growth and requires early surgical reconstruction. Division of syndactyly between the ring and little fingers is relatively straightforward, but in the thumb and index finger, consideration must be given to creating a first web space.

Symphalangism

This term describes congenital stiffness of the proximal interphalangeal joints of the fingers. These joints are abnormal and the fingers are underdeveloped. Surgical intervention is usually unrewarding.

Poland's syndrome

This is a sporadic condition in which there can be hypoplasia affecting the whole upper limb girdle

including the shoulder and chest wall (rib, muscle, breast, nipple). In girls, the breast abnormality may only become apparent in at puberty.

The hand in Poland's syndrome is characteristically small with brachydactyly, syndactyly and symphalangism.

Camptodactyly

'Bent finger' is a flexion deformity of the proximal interphalangeal joint, usually of the little finger. It may be an isolated condition or part of a syndrome. It may be inherited or sporadic, and two-thirds of cases are bilateral.

The condition presents as two groups: those occurring in infancy and affecting males and females equally, and those presenting in adolescence, mainly affecting females. There is often an abnormal muscle insertion (usually one of the lumbricals), and there may be a characteristic abnormal radiographic appearance of the head of the proximal phalanx.

The mainstay of *treatment* is splinting. Surgery may be indicated if the deformity is marked or is a severe nuisance. Soft-tissue releases and/or muscle transfers are advocated by some surgeons but the results can be disappointing.

Clinodactyly

In this condition a digit is bent sideways (radially or ulnarwards), usually due to an abnormally shaped middle phalanx – the so-called 'delta phalanx' – in which the epiphysis is C-shaped or 'longitudinally bracketed'. It usually affects the little finger and is often inherited and bilateral. The child should be examined for other defects as it occurs as part of many syndromes. Severe cases can be treated by corrective osteotomy and bone grafting.

The condition must be distinguished from the *Kirner deformity*, in which the distal phalanx of the little finger has a palmoradial curvature. This usually presents in adolescence and most cases do not require treatment.

Constriction ring syndrome

Also known as amniotic band syndrome, the aetiology of this condition is thought to be early, *in utero* rupture of the amniotic membrane and the formation of constricting amniotic membrane strands. The condition can affect both the upper and lower limbs, and there may also be associated foot deformities.

In the hand, there are constriction rings around digits or limbs, and short fingers, often with bulbous or pinched tips (Figure 15.8b). There may be *acrosyndactyly*, where the distal phalanges are joined at tips, but the fingers are separate more proximally.

In severe cases, the part of the limb or digit distal to the constriction ring may necrose, or be threatened by ischaemia requiring urgent release, and long-term growth is compromised.

Treatment consists of excision of the constricting band and multiple Z-plasties, as well as other soft-tissue reconstruction.

Congenital clasped thumb

Infants with this condition hold their thumbs persistently in the palm, under the flexed fingers. It appears to be due to weakness or absence of the extensor tendons, and there may also be a contraction of the first web space, which makes it a little more difficult to treat. It may present as an isolated problem or as part of a syndrome.

The diagnosis should not be made before the fourth month as it is normal for infants to hold their thumbs in the palm before then.

Treatment is initially by splintage and stretching. If this fails, surgical correction by first web-space release can be performed, combined with an immediate or staged extensor indicis tendon transfer (depending on how tight the first web space is).

Congenital trigger thumb

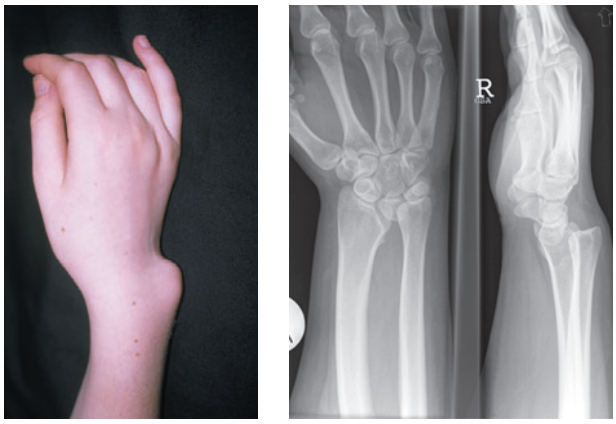
Care should be taken to distinguish this condition from congenital clasped thumb syndrome. It is unlikely that it is a truly congenital disorder but it may occur within a few months after birth. It appears to be a form of stenosing tenovaginitis of flexor pollicis longus, with a palpable thickening of the tendon (known as the *Notta's nodule*) at the level of the A1 pulley over the MCP joint. Triggering often resolves spontaneously but, if the condition persists, it can be treated surgically by division of the A1 pulley of the flexor tendon sheath, taking care to avoid damage to the oblique pulley.

Madelung's deformity

In this deformity, which may be either congenital or post-traumatic, the lower radius curves forwards (ventrally), carrying with it the carpus and hand but leaving the lower ulna sticking out as a lump on the back of the wrist (Figure 15.9).

The congenital form of the disorder may appear as an isolated entity or as part of a generalized dysplasia; although the abnormality is present at birth, the deformity is rarely seen before the age of 10 years, after which it increases until growth is complete. Function is usually excellent.

If deformity is severe, the lower end of the ulna may be shortened; this is sometimes combined with osteotomy of the radius. Excision of the physeal tether and replacement with a free fat graft is an alternative in certain cases.



(a) (b) (c)

Figure 15.9 Madelung's deformity (a) Note the prominent ulnar head and radial tilt; (b) characteristic X-ray showing the increased slope of the radius and (c) subluxation of the ulna.

Synostosis

Failure of embryological separation of skeletal components can result in conjoined normal-looking bones or fused (unseparated) joints. This may occur at any level from the fingers to the humerus and can be longitudinal (e.g. humeroulnar synostosis) or transverse (e.g. proximal radioulnar synostosis or carpal coalitions). The condition may appear in isolation or as part of a wider syndrome. If there is no significant loss of function, operative treatment is unnecessary. If important movements are affected (e.g. fusion at the elbow joint), osteotomy and repositioning of the limb in a more favourable position may be considered. Carpal fusions usually need no treatment.

Arthrogryposis multiplex congenita (AMC)

Arthrogryposis is described in Chapter 10. Part or the whole of the upper limb may be affected, giving rise to muscle weakness and joint contractures. The shoulders are usually adducted, the elbows stiff, the wrists and fingers flexed, and the thumbs clasped in adduction and flexion. The overlying skin is smooth and devoid of the normal creases.

Treatment is by early stretching and splinting; later joint releases and tendon transfers may be used, starting from proximal to distal. The choice of muscles for tendon transfers must be considered in the context of the patient's ability to walk, and the need to transfer to and from a wheelchair.

Other generalized syndromes

Many generalized disorders involve the upper limbs. Examples include Down's syndrome (short little fingers), Marfan's syndrome (long fingers, camptodactyly), neurofibromatosis (macrodactyly) and cerebral palsy. The hand problems will require specialized treatment in their own right, in addition to management of the general disorder.

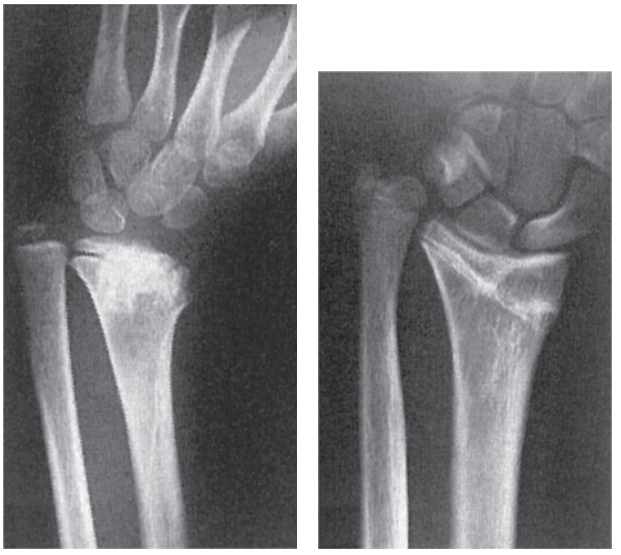
ACQUIRED DEFORMITIES OF THE WRIST

Physeal injury

Fracture-separation of the distal radial epiphysis may result in partial fusion of the physis, with pain and asymmetrical growth deformity of the wrist (Figure 15.10). The bony bridge crossing the physis, if it is small, may be excised and replaced by a fat graft. Once growth slows down the deformity can be corrected by a suitable osteotomy, if necessary combined with soft-tissue release; a circular frame apparatus can be used for this.

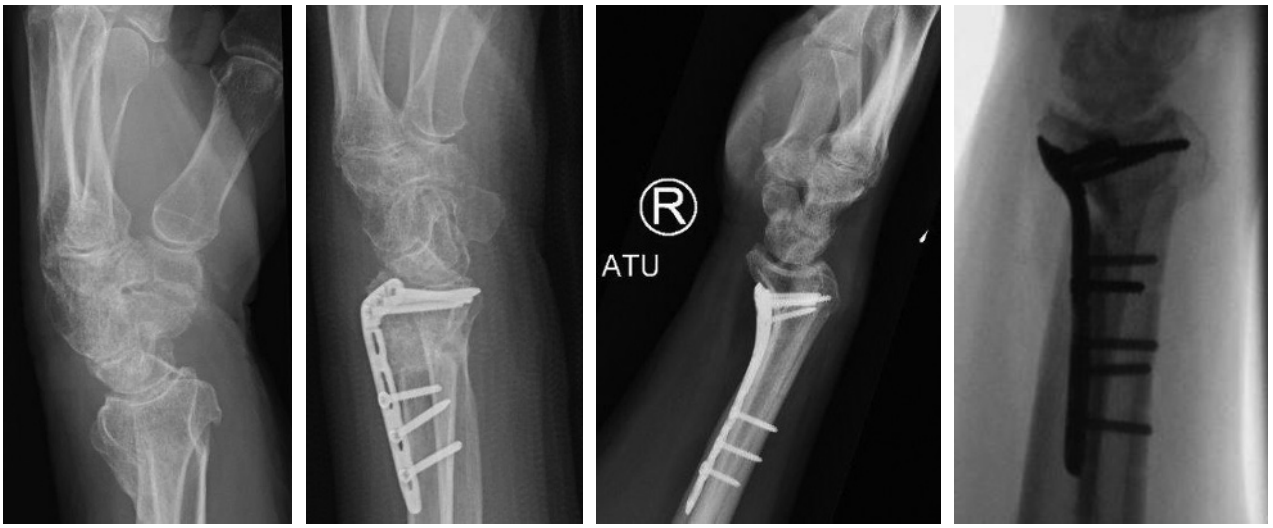
Forearm fractures

After a Colles' fracture, radial deviation, posterior angulation and prominence of the radial head are common. These deformities may be unsightly but cause little disability. Subluxation of the distal radioulnar joint may result in prominence of the ulnar head, painful rotation and loss of pronation or supination. This should be treated by reconstructing the distal radius; *the ulnar head should never be excised*. Abnormal angulation of the radius may lead to midcarpal malalignment with pain and loss of grip strength. A radial osteotomy is then necessary; the bone fragments are fixed with a locking plate and bone graft is added (Figure 15.11).



(a) (b)

Figure 15.10 Growth plate arrest (a) Impacted physeal fracture; (b) later arrest of radius, relative overgrowth of ulna.



(a) (b) (c) (d)

Figure 15.11 Distal radius osteotomy (a) Preoperative dorsal tilt; (b) postoperative correction with dorsal plate and bone graft; (c) preoperative view, fracture healed with volar tilt and rotation reduced; (d) osteotomy has been revised, proper congruity of sigmoid notch achieved.

Rheumatoid deformities

The typical rheumatoid deformity is radial deviation of the wrist, swelling of the extensor tendons, dorsal prominence of the ulnar head and sometimes tendon rupture. The carpus falls into flexion and supination as the ulnar side sags forwards away from the prominent ulnar head.

'Drop wrist'

Radial nerve palsy causes the wrist to drop into flexion and active extension is lost. With a posterior interosseous nerve palsy, the wrist will extend radialwards because extensor carpi radialis longus function is preserved.

If the nerve does not recover, tendon transfers will greatly improve function (see Chapter 11).

CHRONIC INSTABILITY OF THE WRIST

Movements of the wrist and hand are interdependent, the wrist providing appropriate mobility and stability to position and steady the hand for the remarkable range of actions and tactile sensibility employed in our daily activities. Abnormalities of wrist mechanics are a common source of functional disability; this is seen most often in rheumatoid arthritis, in association with congenital laxity and after local trauma.

Articulations of the wrist

The wrist comprises three movable joints: the *distal radioulnar joint*, the *radiocarpal joint* (between the radius and the proximal row of carpal bones) and the *midcarpal joint* (between the proximal and distal rows of carpal bones) (Figure 15.12). There is further movement between the lunate and the adjacent triquetrum and scaphoid which flex and extend so the wrist tilts ulnarwards or radialwards.

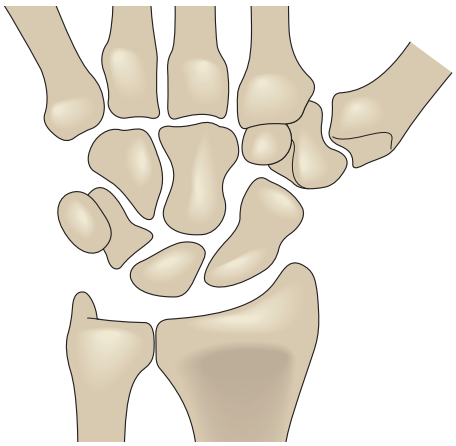


Figure 15.12 The carpal joints This diagram shows the radiocarpal joint between the radius and the proximal row of carpal bones and the midcarpal joint between the proximal and distal rows of carpal bones. The proximal row is an intercalcated segment.

THE DISTAL RADIOULNAR JOINT (DRUJ)

The distal radius and ulna are linked to each other by the interosseous membrane, the capsule of the DRUJ and the *triangular fibrocartilage complex (TFCC)* (Figure 15.13). The head of the ulna articulates congruently with the sigmoid notch of the distal radius; movement at the joint occurs by the radius both

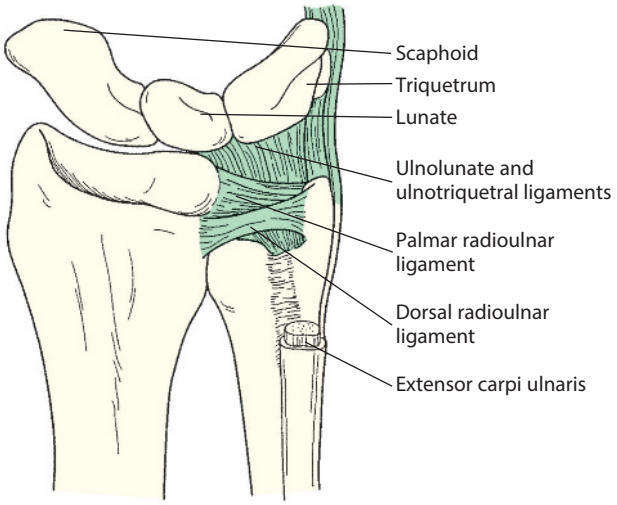


Figure 15.13 The distal radioulnar joint (DURJ)
The joint incorporates the triangular fibrocartilage complex. The fibrocartilaginous plate is connected at its apex to the base of the ulnar styloid process and laterally to the inferomedial ridge of the radius. Its outer fibres blend with those of the ligaments around the ulnar aspect of the wrist.

rotating and sliding in an arc around the head of the ulna during pronation and supination of the forearm. Interposed between the head of the ulna and the carpus is a fibrocartilaginous disc, a fan-shaped structure spreading from an apical attachment at the base of the ulnar styloid process to the rim of the radial sigmoid notch. Its dorsal and volar edges are coextensive with the dorsal and palmar radioulnar ligaments; further attachments to the joint capsule, the ulnotriquetral and ulnolunate ligaments, the ulnar collateral ligament and the sheath of the extensor carpi ulnaris tendon complete the fibrocartilage complex. The peripheral attachments of the TFCC have a good vascular supply and can heal after injury; the central area of the triangular plate is avascular and tears do not heal.

THE RADIOCARPAL AND MIDCARPAL JOINTS

Movements in the sagittal plane (flexion and extension) occur at both the radiocarpal and midcarpal joints. Movements in the frontal plane (adduction or ulnar deviation and abduction or radial deviation) occur mainly at the radiocarpal joint, but they inevitably involve also the scaphoid, which has to flex forwards as the trapezium moves towards the radial styloid during abduction.

The bones of the distal carpal row (hamate, capitate, trapezium and trapezoid) are joined by ligaments to each other and to the bases of the metacarpals. Although there is some movement of the fourth and fifth carpometacarpal joint, there is very little movement in the remaining carpometacarpal articulations.

The distal row articulates through the midcarpal joint with the bones of the proximal row (triquetrum, lunate and scaphoid), which are likewise held together by stout interosseous ligaments. Because these bones have no muscles attaching to them, their position is determined by the way they all fit together and by the constraints of the interosseous ligaments. The proximal row is, in a sense, 'interposed' between the forearm bones and the hand bones and is called an *intercalated segment*.

The articular surface of the radius slopes obliquely forwards at 11 degrees and ulnarwards at 22 degrees; the radial styloid is about 11 mm distal to the ulnar styloid (the 'rule of elevens'). With the wrist in the neutral position, tightening of the long muscles will tend to drag the carpus down the slope, and when the wrist is pulled into abduction this tendency is increased. By contrast, when the wrist is adducted about 30 degrees, muscle pull draws the carpus most securely into the radial 'socket'. This is, in fact, the '*position of function*' (or maximum stability) and there is a natural inclination to adopt this position during power grip. This action is mediated by flexor carpi ulnaris (which is why it is unwise to choose that muscle for a tendon transfer).

The scaphoid is potentially the most unstable of all the carpal bones. As the wrist flexes and extends, so does the scaphoid bone; the lunate and triquetrum follow passively, guided by the interosseous ligaments. With abduction, the space between the trapezium and radial styloid closes down so the scaphoid moves out of the way by flexing palmarwards and sliding ulnarwards. During adduction, the scaphoid tilts dorsally and slides radially. As the wrist abducts and adducts, the helical surface of the hamate also causes the triquetrum to move. Different wrists have different ratios of flexing/extending or sliding of the scaphoid (so-called 'column' and 'row' wrists).

Instability of the distal radioulnar joint

Chronic instability of the distal radioulnar joint (Figure 15.14) may result from malunion of the distal radius or radial shaft, from avulsion of the TFCC from its insertion into the base of the ulnar styloid, rheumatoid arthritis or excision of the distal end of the ulna. The previous history is therefore important. Fracture of the radial shaft is associated with dislocation of the distal radioulnar joint (Galeazzi fracture-dislocation); after reduction of the radius, one must be certain that the radioulnar joint also is reduced.

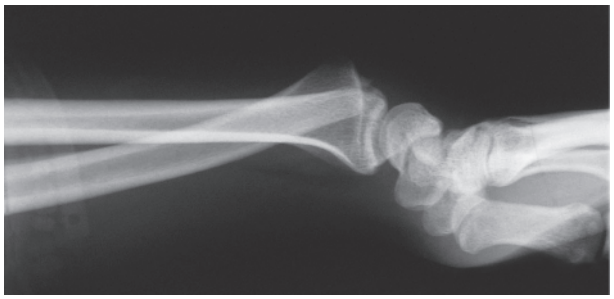


Figure 15.14 Ulnar head instability The ulna remains in position (it is fixed at the elbow) and the radius has subluxated forwards.

The patient complains of painful restriction of pronation and supination, clunking and undue prominence of the ulnar head. There may be tenderness directly over the radioulnar joint and the dent just in front of the ulnar styloid (the *foveal sign*) due to peripheral avulsion of the TFCC. The unstable ulna can be 'balloted' by holding the patient's forearm in neutral and pushing sharply upon the prominent head of the ulna (the *piano-key sign*). Remember that the ulna head is stable (as it is fixed to the hinge joint of the elbow); it is in fact the radius which is unstable in relation to the ulna. Always compare with the other side as quite marked laxity can be normal in some people.

Imaging

X-ray examination may show evidence of previous fractures (e.g. distal radius malunion, ulnar styloid avulsion), previous surgery or rheumatoid arthritis. CT in pronation and supination demonstrates bone incongruity; MRI and arthroscopy may demonstrate avulsion of the insertion of the TFCC into the base of the ulnar styloid.

Treatment

Treatment depends on the cause. If the radius has malunited, then an osteotomy is needed. If the ulnar stylois is avulsed, then it can be fixed and bone grafted; if the TFCC is avulsed, it is reattached by arthroscopic or open methods. Ulnar shortening osteotomy can tighten the ulnar corner and improve stability in those with ligamentous laxity; a tendonweave to reproduce the volar and dorsal radioulnar ligaments is the most reliable but difficult reconstruction. *The ulnar head must never be excised to treat instability; that will only worsen the problem.*

If the ulnar head has previously (and usually unwisely) been removed, soft-tissue reconstructions are unreliable and ulnar head replacement will usually be needed to restore stability. A special constrained device has been developed with promising results.

Longitudinal instability of the radius and ulna

Fracture of the radial head is sometimes accompanied by disruption of the interosseous membrane and dislocation of the distal radioulnar joint (the Essex-Lopresti lesion). Excision of the radial head can lead to proximal migration of the radius and ulnocarpal impaction (see below); whenever possible the radial head should be preserved or replaced by a metal implant. Chronic longitudinal instability causes ulnar-sided wrist pain and loss of grip strength.

Treatment of the distal radioulnar joint symptoms is generally unsatisfactory. A combination of radial head replacement and an ulnar shortening osteotomy sometimes improves symptoms. There is increasing interest in reconstructing the interosseous ligament. Radioulnar fusion is sometimes employed as the only salvage procedure but this is a disabling procedure as there is no perfect position (if fixed in supination, one cannot type; if fixed in pronation, one cannot carry a tray).

Disorders of the triangular fibrocartilage complex

Clinically significant disorders of the TFCC can be divided into traumatic and degenerative conditions. There are two distinctly different portions: the peripheral part which attaches to the base of the ulnar styloid, disruption of which causes instability and pain, and the central portion, which acts as a load diffuser between the forearm and the proximal carpal row, perforation of which can occur by natural degeneration or a fall.

Traumatic disruption

There may be a history of a fall on the outstretched hand or a twisting injury of the forearm. The patient complains of pain, and sometimes clicking or even instability in the distal radioulnar joint, particularly on twisting the wrist. In a peripheral tear of the TFCC, the foveal sign is usually positive and there is pain and clunking on rotation of the forearm. The ballotment test (piano-key sign) is positive. Symptoms of central traumatic perforation can be reproduced by holding the wrist in adduction and compressing the ulnar head against the carpus then moving backwards and forwards (the *grind test*) and by eliciting tenderness over the ulnar head. The diagnosis is confirmed by MRI with arthrography and, most sensitively, by arthroscopy.

Treatment Peripheral tears can be reattached by either open or arthroscopic techniques with a reasonable expectation that they will heal. If this fails,

a tendon reconstruction is needed. Central traumatic tears, in the absence of ulnocarpal impaction (see below), are best managed by arthroscopic debridement.

Ulnocarpal impaction and TFCC degeneration

The TFCC tends to degenerate with age; usually this is asymptomatic. However, progressive degenerative change may be associated with a relatively long ulna, impaction of the ulnar head against the ulnar side of the lunate and ulnocarpal arthritis (the *ulnocarpal impaction syndrome*) (Figure 15.15). X-ray examination (standard views with the shoulder abducted 90 degrees, the elbow flexed 90 degrees and the forearm in midpronation–supination) may show a relatively long ulna ('*positive ulnar variance*') and in late cases there may be arthritic changes in the ulna–lunate–triquetral articulation.

Treatment Initial treatment is with simple analgesics, splintage and steroid injections. Many settle with time and so surgery should not be precipitous. If this is not successful, the long ulna is shortened using a special jig and compression plate. A better alternative for just 2–3 mm of positive variance is an arthroscopic excision of the distal dome of the ulnar head. *The ulnar head itself should never be excised for this condition.*

Chronic instability of the radiocarpal and intercarpal joints

Abnormal movement between the carpus and the forearm bones, or between individual carpal bones, results from loss of the bony relationships and/or ligamentous constraints which normally stabilize the wrist. The initiating cause is usually some type of injury – a wrist sprain with ligament damage, subluxation or dislocation at one of the radiocarpal or intercarpal joints or a fracture of one of the wrist bones – but chronic instability may also arise insidiously in erosive joint disorders such as rheumatoid arthritis.

Patterns of carpal instability

Acute carpal injuries are dealt with in Chapter 26. Here we shall consider the problems associated with chronic carpal instability (Figure 15.16). The disorder affects mainly the intercalated segment (proximal carpal row) of the wrist. The common patterns are listed here.

Dorsal intercalated segment instability (DISI) Following a fracture of the scaphoid or rupture of the scapholunate ligament (scapholunate dissociation), the lunate no longer passively follows the scaphoid. The scaphoid tends to flex and the lunate assumes its default position of extension (dorsal tilt).

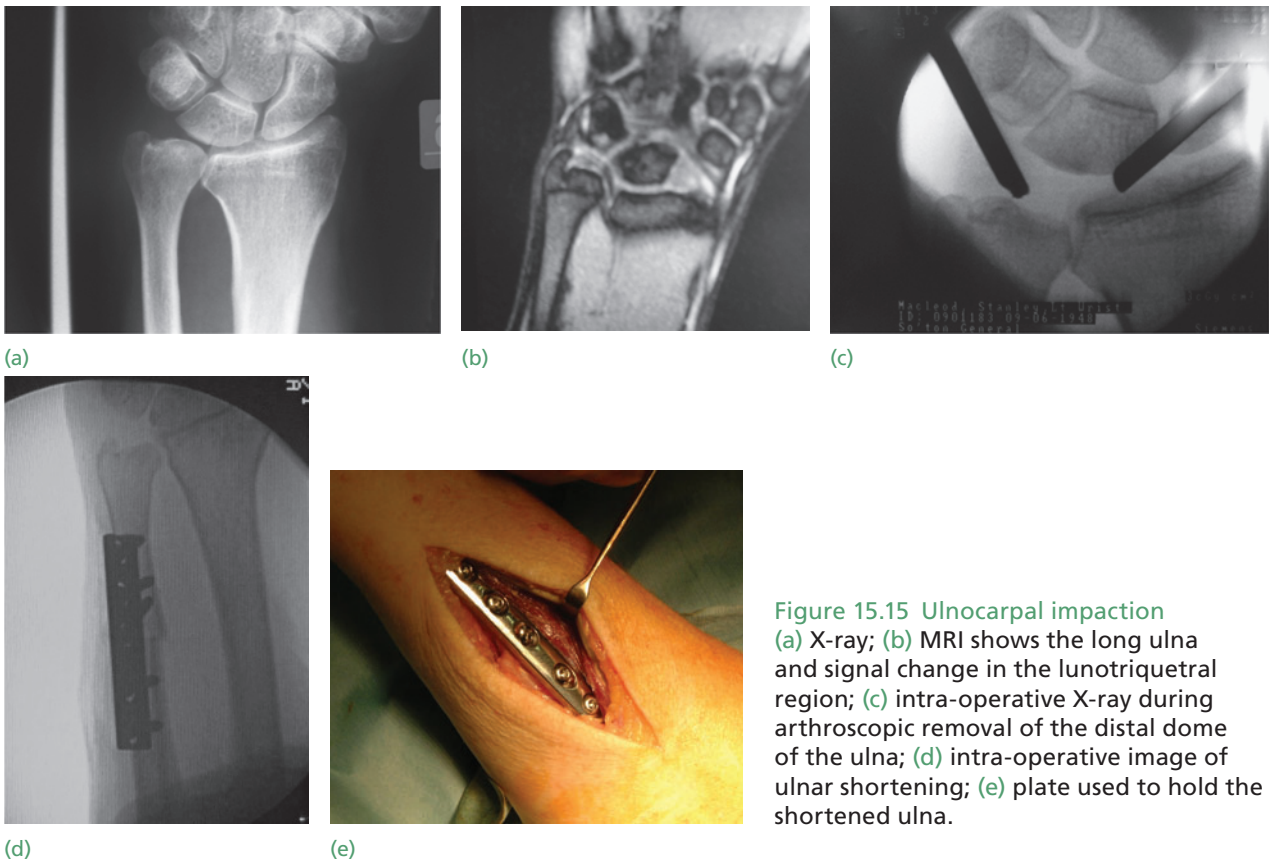


Figure 15.15 Ulnocarpal impaction
(a) X-ray; (b) MRI shows the long ulna and signal change in the lunotriquetral region; (c) intra-operative X-ray during arthroscopic removal of the distal dome of the ulna; (d) intra-operative image of ulnar shortening; (e) plate used to hold the shortened ulna.

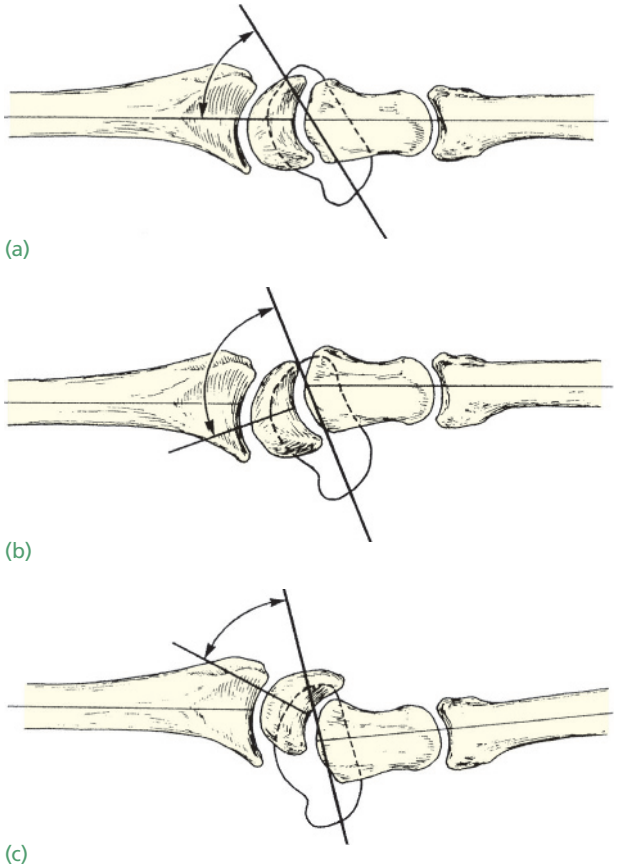


Figure 15.16 Carpal instability The relationships of the carpal bones in (a) the normal wrist; (b) DISI and (c) VISI.

The deformity may occur spontaneously with natural degeneration of the ligament; it is also associated with scaphoid–trapezium–trapezoid (STT) arthritis.

Volar intercalated segment instability (VISI) Less commonly, the lunotriquetral ligament is ruptured. The lunate, unrestrained by the triquetrum, but still controlled by the scaphoid, tends to flex while the capitate tends to extend.

Midcarpal instability This usually emerges as a chronic problem, associated with generalized ligamentous laxity. The proximal and distal rows become unstable through the midcarpal joint.

Adaptive midcarpal instability If a distal radius fracture heals with the radial articular surface tilted dorsally, then the proximal carpal row tends also to tilt dorsally and the midcarpal joint flexes to maintain the palm in line with the forearm. This is painful and grip is reduced.

Radiocarpal translocation Chronic synovitis and articular erosion (as in RA) gradually leads to attenuation of the wrist ligaments and subluxation of the entire radiocarpal joint. In advanced RA the carpus usually shifts ulnarwards and simultaneously deviates into abduction and supination.

Clinical features of carpal instability

The patient with scapholunate or lunotriquetral incompetence presents with pain and weakness of the wrist, and sometimes also clunking during movement or gripping actions. It is important to enquire about any previous injury, however trivial it may have seemed at the time.

On examination, there may be generalized tenderness over the carpus from synovitis or more localized tenderness, for example at the scapholunate junction or over the scaphoid itself. Grip strength is reduced. Provocative tests are useful.

Watson's test for scapholunate incompetence Thumb pressure is applied to the volar aspect of the wrist over the distal pole of the scaphoid – this restores the alignment of the volar-tilted scaphoid. While maintaining this position, the wrist is moved alternately into adduction and abduction. A painful 'clunk' occurs as the proximal pole of the scaphoid subluxes dorsally.

Lunotriquetral ballottement With one hand the examiner grasps and stabilizes the lunate between index finger and thumb. With the other thumb he presses on the pisiform/triquetrum to produce a shearing motion between lunate and triquetrum. If there is pain and excessive movement, this suggests incompetence of the lunotriquetral ligament.

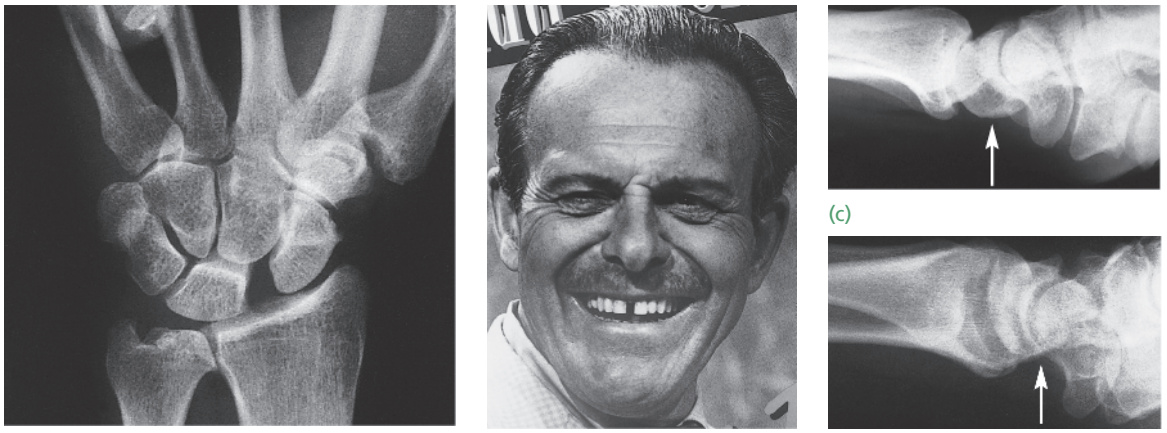
Pivot shift test The examiner grasps the patient's forearm with one hand and the patient's hand with the other; he then compresses the wrist axially while moving it from abduction to adduction. A painful 'clunk' suggests midcarpal instability.

X-rays

An anteroposterior X-ray may show an old or new scaphoid fracture. There may be widening of the scapholunate interval (the *Terry-Thomas sign*); if the scaphoid is flexed, it will look foreshortened and the tubercle may appear as a dense 'ring' in the bone (Figure 15.17).

A true lateral view is examined to assess the relative alignment of the distal radius, the lunate, capitate and scaphoid. In a normal wrist, the articular surfaces of the radius, lunate and capitate are parallel. In the DISI deformity, the capitate axis is shifted dorsally but it flexes relative to the lunate, the lunate tilts backwards and the scaphoid flexes; the scapholunate angle is greater than 70 degrees. In a VISI deformity, the lunate is flexed forward and the scapholunate angle is less than 30 degrees; the capitate tilts dorsally.

There is often a step in the lunotriquetral surface (disruption of Gilula's arc). In an anteroposterior 'clenched fist view' the scaphoid is seen to flex and a scapholunate gap becomes more apparent.



(a) (b) (c) (d)

Figure 15.17 Carpal instability (a) A year after 'straining' his wrist this patient was still complaining of pain; the X-ray shows a gap between the scaphoid and the lunate (the Terry-Thomas sign) and rotation of the scaphoid. (b) The actor Terry Thomas with the trademark gap between his front teeth (reproduced by permission; © United Artists Inc.) (c) In the lateral view the lunate is tilted dorsally and the scaphoid ventrally (DISI); compare this with (d), an example of VISI, showing volar tilt of the lunate.

Anteroposterior views with the wrist adducted and abducted emphasize scapholunate gaps and abnormal scaphoid flexion (the ring sign), particularly when compared with X-rays of the other side.

Further investigations

Image intensification This helps to define the site of instability in difficult cases.

Arthrography Leakage of contrast through incompetent scapholunate or lunotriquetral spaces will show.

MRI MRI will reveal any associated injuries, such as a scaphoid fracture. The scapholunate and lunotriquetral interosseous ligaments are so slim that the resolution of MRI scanning may be inadequate to detect significant injuries.

Arthroscopy of the radiocarpal and midcarpal joints This is the best method for demonstrating carpal instability. Ligament tears, certain patterns of instability, synovitis and damaged articular cartilage can be detected.

Treatment

Scapholunate and lunotriquetral dissociation The best results are obtained if the ligaments heal in an anatomical position. The diagnosis should, therefore, be made as soon as possible after injury; this requires a high index of suspicion. The surgeon should be alerted by a history of wrist pain following a fall on the outstretched hand and a finding of midcarpal tenderness. MRI or arthroscopy may be needed to secure the diagnosis. The ligaments are repaired, the bones stabilized with K-wires and the wrist held in a cast for at least 2 months.

Patients seen more than 3 months after injury will require a more extensive type of carpal reduction and ligament reconstruction. For scapholunate incompetence various reconstructions have been described. Perhaps the three ligament tenodesis ('3T') is the most reliable. Half of flexor carpi radialis tendon is passed through a drill hole in the scaphoid and then secured across the back of the carpus. This pulls up the flexed scaphoid from its flexed position and tightens the carpus transversely.

If the displacement cannot be reduced, or if soft-tissue repair fails or if osteoarthritis supervenes, then a salvage operation is needed. The options include a proximal row carpectomy (if the lunate–capitate junction is preserved) or a scaphoid excision with fusion of the hamate, capitate, triquetrum and lunate (if the lunate–capitate joint is arthritic or if the patient needs the strongest wrist especially in torsion). Another option is the radioscapholunate fusion, which is more complex but may be stronger and more durable.

The best treatment for *lunotriquetral instability* has not been established. Reconstruction with tendon grafts is not totally reliable. A lunotriquetral fusion is stronger but securing union is difficult.

Symptomatic midcarpal instability Treatment includes proprioceptive training (a gyroscopic device can help). Intractable symptoms may respond to arthroscopic shrinkage of the capsule with a diathermy probe. The alternative of a ligament reconstruction is unreliable, and midcarpal fusion causes very significant loss of movement (about 50%).

Dorsal malunion of the distal radius A dorsal tilt deformity that is symptomatic may be treated by a corrective osteotomy of the distal radius (Figure 15.11); normal carpal alignment should be restored unless there are fixed changes from leaving the treatment for too long.

KIENBÖCK'S DISEASE

Robert Kienböck, in 1910, described what he called 'traumatic softening' of the lunate bone. This is a form of ischaemic necrosis, probably due to chronic stress or injury, though one cannot be certain about this. It has been suggested that relative shortening of the ulna ('negative ulnar variance') predisposes to stress overload of the lunate between the distal edge of the radius and the carpus, but this has not been proven convincingly.

Pathology

As in other forms of ischaemic necrosis, the pathological changes proceed in four stages (Table 15.1 shows the radiological appearance and treatment options for each pathological stage):

- *Stage 1* – ischaemia without naked-eye or radiographic abnormality
- *Stage 2* – trabecular necrosis with reactive new bone formation and increased radiographic density, but little or no distortion of shape
- *Stage 3* – collapse of the bone
- *Stage 4* – disruption of radiocarpal congruence and secondary osteoarthritis.

Clinical features

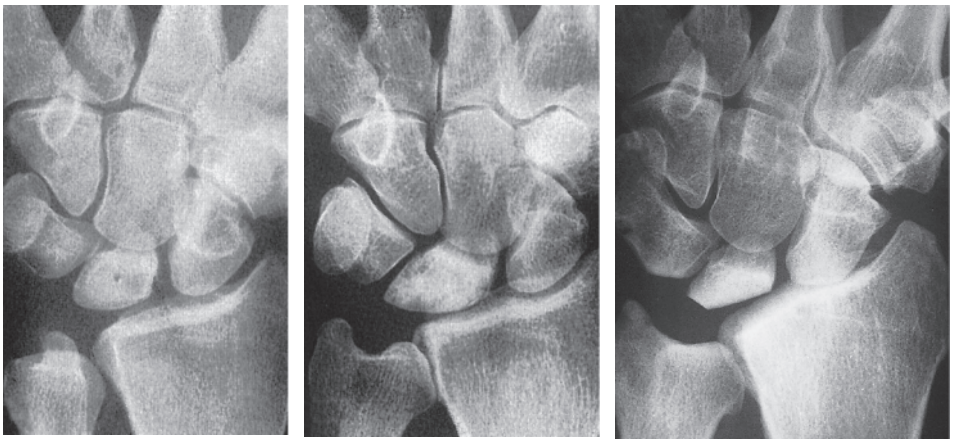
The patient, usually a young adult, complains of ache and stiffness; only occasionally is there a history of acute trauma. Tenderness is localized over the lunate and grip strength is diminished. In the later stages wrist movements are limited and painful.

Imaging

X-rays at first show no abnormality, but radioscintigraphy may reveal increased activity. Later X-rays may show either mottled or diffuse density of the bone, and later still the bone looks intensely sclerotic and irregular in shape or squashed (Figure 15.18). The capitate migrates proximally into the space left by the collapsing lunate, and the scaphoid flexes forward. Eventually, there are osteoarthritic changes in the wrist. Ulnar variance should be assessed by standardized X-ray examination with the

Table 15.1 Keinbock's disease: staging and treatment

Stage	X-ray/MRI	Treatment options
1	Normal X-ray, changes on MRI	Splint Vascularized bone graft
2	Lunate sclerosis in plain X-ray, fracture lines sometimes present	Vascularized bone graft If negative ulnar variance: radial shortening If positive ulnar variance: radial tilting or capitate shortening
3a	Fragmented lunate, height preserved	If negative ulnar variance: radial shortening If positive ulnar variance: radial tilting or capitate shortening Proximal row carpectomy Scaphocapitate fusion STT fusion
3b	Collapse of lunate, proximal migration of capitate, fixed scaphoid flexion	Proximal row carpectomy Scaphocapitate fusion STT fusion
4	Arthritis	Proximal row carpectomy Total wrist fusion Wrist replacement



(a) (b) (c)

Figure 15.18 Kienböck's disease (a) In stage 2, the bone shows mottled increase of density but is still normal in shape. (b) In stage 3, density is more marked and the lunate looks slightly squashed. (c) In stage 4, the bone has collapsed and there is radiocarpal osteoarthritis. In all three, the ulna looks disproportionately short.

shoulder abducted to 90 degrees, the forearm in neutral rotation and the wrist in neutral flexion–extension. As the lunate collapses, the relative length of the capitate from third metacarpal bone to distal radius increases.

MRI is the most reliable way of detecting the early changes (Figure 15.19). A gadolinium-enhanced MRI scan will demonstrate the condition even if plain X-rays are normal.

Treatment

NON-OPERATIVE TREATMENT

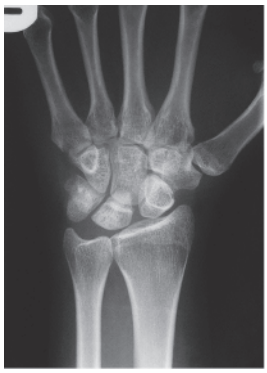
The natural history is often benign with tolerable symptoms despite severe radiological changes. Destructive surgery should therefore be undertaken with caution. In early cases, splintage of the wrist for 6–12 weeks relieves pain and possibly reduces mechanical stress. If bone healing catches up with ischaemia, the lunate may remain virtually undistorted; this is more likely in very young patients. However, if pain persists, and more importantly if the bone begins to flatten, operative treatment may be indicated (Table 15.1).

OPERATIVE TREATMENT

In its earliest stages, before collapse, the bone can be revascularized with a pedicled bone graft or vascular bundle implantation.

While the wrist architecture is only minimally disturbed (i.e. up to early stage 3), it seems rational to aim for a reduction of carpal stress. Options include shortening the radius (the most common procedure and apparently fairly effective when the radius is a little too long – Figure 15.20a)); other more sophisticated options include shortening the capitate and, for those with a neutral ulna variance, tilting the radius radialwards.

Once the bone has collapsed, the options are limited. A wrist neurectomy is worth trying; this will preserve



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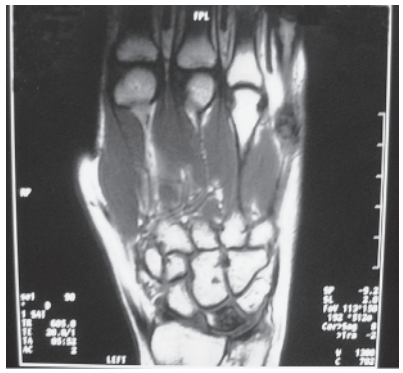

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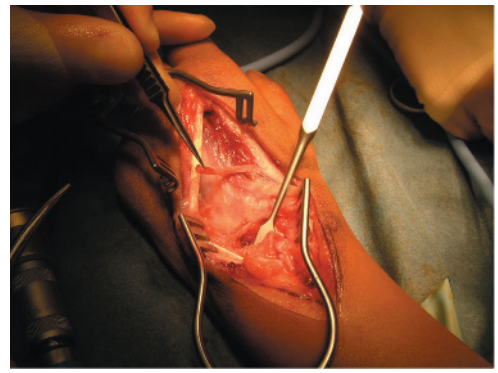
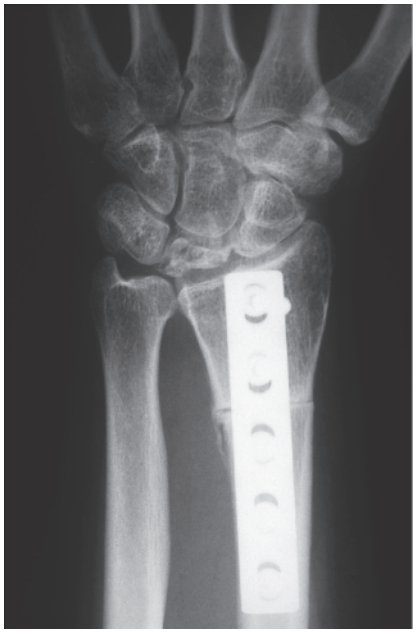


(c)

Figure 15.19 Kienböck's disease stage 1 (a) Not seen on X-ray; (b) seen on MRI scan; (c) treated by vascular bundle implantation.



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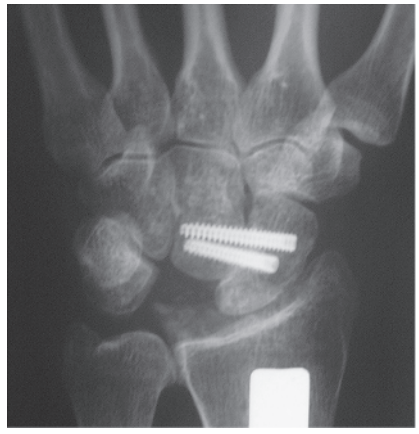

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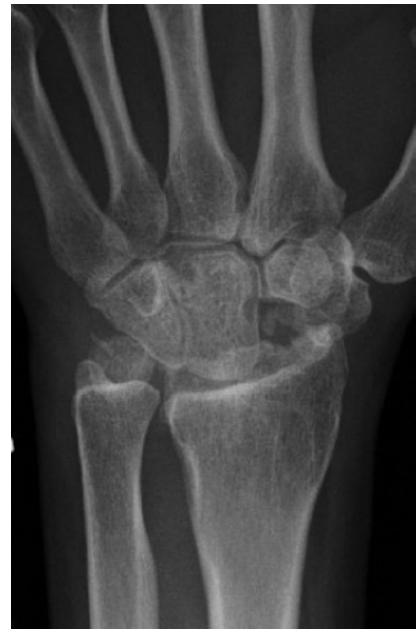


(c)

Figure 15.20 Kienböck's disease-treatment (a) Radial shortening; (b) scaphocapitate fusion; (c) proximal row carpectomy.

movement yet reduce pain. Lunate replacement by a silicone prosthesis, once popular, gives poor long-term results and particle shedding is liable to cause synovitis. A scaphocapitate fusion (Figure 15.20b) or excision of the proximal row of the carpus (Figure 15.20c) may improve function, but in the long term these may not prevent the occurrence of osteoarthritis.

If pain and restriction of movement become intolerable, *radiocarpal arthrodesis* is the one reliable way of providing a stable, pain-free wrist. Wrist replacement is an alternative in individuals with lesser demands.

PREISER'S DISEASE

This is a very rare condition in which the scaphoid undergoes spontaneous avascular necrosis. Early disease, diagnosed on MRI, can respond to vascularized bone grafting. Later symptomatic disease with advanced destruction of the scaphoid would need joint-preserving surgery (proximal row carpectomy (PRC) or scaphoidectomy–four-corner fusion (4CF)).

TUBERCULOSIS
(see also Chapter 2)

At the wrist, tuberculosis is rarely seen until it has progressed to a true arthritis. Pain and stiffness come on gradually and the hand feels weak. The forearm looks wasted; the wrist is swollen and feels warm (Figure 15.21). Involvement of the flexor tendon compartment may give rise to a large fluctuant swelling that crosses the wrist into the palm (compound palmar ganglion). In a neglected case there may be a sinus. Movements are restricted and painful.

X-rays show localized osteoporosis and irregularity of the radiocarpal and intercarpal joints; there may also be bone erosion.

Diagnosis

The condition must be differentiated from rheumatoid arthritis. Bilateral arthritis of the wrist is nearly always rheumatoid in origin, but when only one wrist is affected the signs resemble those of tuberculosis. X-rays and serological tests may establish the diagnosis, but often a biopsy is necessary.

Treatment

Antituberculous drugs are given and the wrist is splinted. If an abscess forms, it must be drained. If the wrist is destroyed, systemic treatment should be continued until the disease is quiescent and the wrist is then arthrodesed.

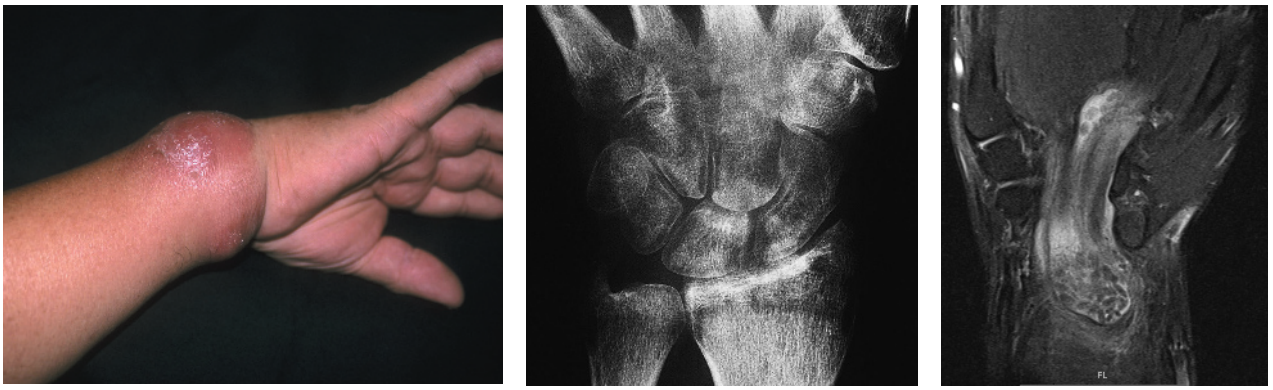
RHEUMATOID ARTHRITIS
(see also Chapter 3)

After the metacarpophalangeal (MCP) joints, the wrists and distal radioulnar joints are the most common sites of rheumatoid arthritis. Wrist and hand should always be considered together when dealing with this condition.

Pathology

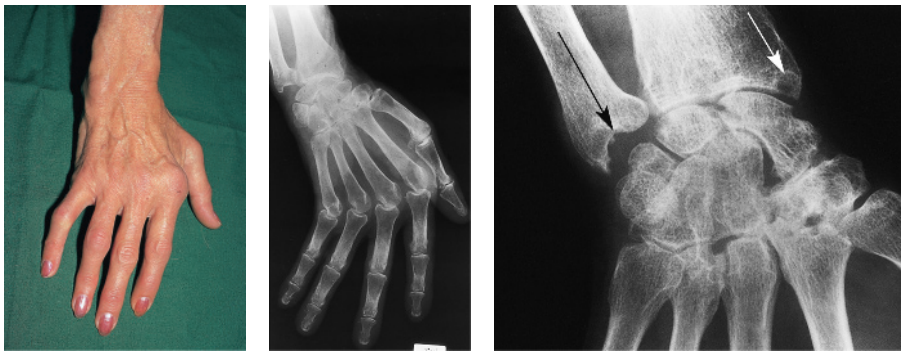
In the early stages, the characteristic features are synovitis of the joints and tendon sheaths. If the disease persists, the distal radioulnar joint (DRUJ), radiocarpal joint and intercarpal joints become eroded; this, together with attenuation of the ligaments and tendons, leads to instability and progressive deformity of the wrist and hand (Figure 15.22).

The ulnar side of the carpus gradually shifts towards flexion and volar subluxation, causing the head of the ulna to jut out prominently on the dorsum of the wrist. The proximal carpal row slides



(a) (b) (c)

Figure 15.21 Tuberculosis of the wrist (a) Pointing abscess; (b) X-ray showing diffuse osteoporosis; (c) MRI showing flexor synovium affected by TB.



(a) (b) (c)

Figure 15.22 Rheumatoid arthritis (a) Typical zigzag deformity in established rheumatoid arthritis. The wrist is deviated radialwards and the fingers ulnarwards. (b) X-ray of the same patient. (c) Enlarged X-ray view – note the characteristic erosions at the distal ends of the radius and ulna (arrows).

ulnarwards and the metacarpal bones deviate radialwards, which mechanically predisposes to a reciprocal ulnar deviation of the fingers – a cardinal feature of the 'rheumatoid hand'. At the same time, the scaphoid falls into marked flexion because of erosion of the interosseous ligaments and loss of carpal height. The combination of instability and erosive tenosynovitis eventually leads to tendon rupture – typically one or more of the long extensor tendons.

An unstable wrist means a weak hand; deformities of the MCP joints are almost invariably associated with complementary deformities of the wrist.

Clinical features

Early symptoms are pain, swelling and stiffness of the wrists. At first the swelling is usually localized to the common extensor tendon sheath or the extensor carpi ulnaris, but as time progresses the joints become thickened and tender. Swelling of the synovium in the carpal tunnel may cause median nerve compression.

Gradually the wrist becomes unstable as the articular surfaces erode and ligaments become attenuated. Early infiltration of tendons may lead to weakness of wrist extension and flexion. Instability of the DRUJ aggravates the apparent dorsal protrusion of the ulnar head, which can often be jogged up and down by pressing upon it with your thumb (the *piano-key sign*).

Tendon lesions are common in the late stage. The first to rupture is usually the extensor digiti minimi (Figure 15.23), followed by the extensor communis tendons of the little and ring fingers. Extensor pollicis longus tendon is also vulnerable. The flexor tendons also sometimes rupture, either within the digital sheaths or in the cramped confines of the carpal tunnel.

The proximal joints in both upper limbs should be examined as well. It is important to know whether the arm is able to place the wrist and hand in functional positions.

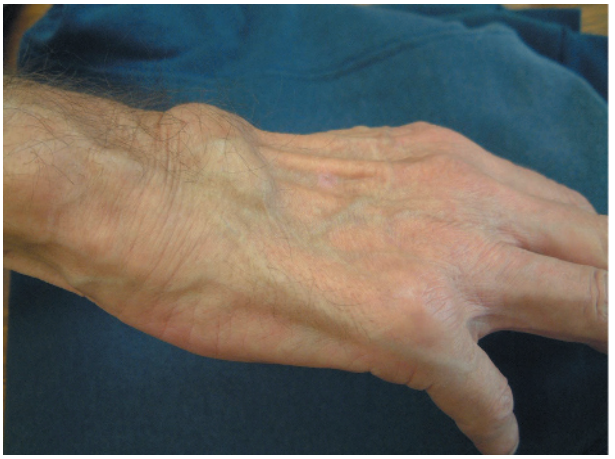


Figure 15.23 Rheumatoid arthritis Synovitis around the ulnar head with rupture of extensor digiti minimi.

X-rays

Typical signs are peri-articular osteoporosis and erosion of the ulnar styloid and the radiocarpal and intercarpal joints (Figure 15.24). In most cases the hands also will be affected, but there is a well-recognized group of patients (mostly elderly men) in whom the wrists carry the brunt of the disease.

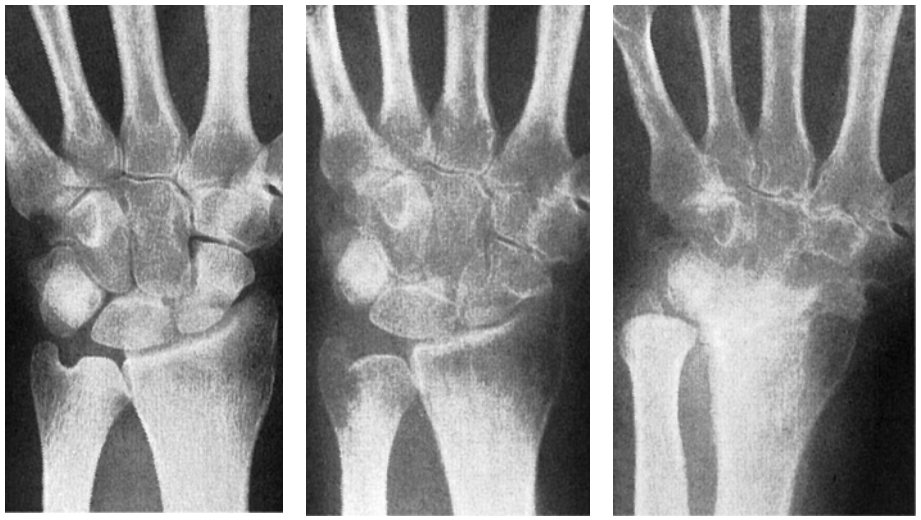
Treatment

EARLY STAGE DISEASE

During the early stages of rheumatoid arthritis the objectives of treatment are to relieve pain and counteract synovitis. In addition to systemic treatment, synovitis of the wrist and/or tendons will be helped by intermittent splintage and intrasynovial injections of corticosteroid preparations.

ESTABLISHED DISEASE

As joint erosion makes its appearance, the focus turns increasingly to the safeguarding of joint stability and the prevention of deformity.



(a) (b) (c)

Figure 15.24 Rheumatoid arthritis (a) At first, the X-rays show only soft-tissue swelling. (b) Two years later, this patient shows early bone changes – peri-articular osteoporosis and diminution of the joint space. (c) Five years later still, bone erosions and joint destruction are marked.

Extensor tenosynovectomy and soft-tissue stabilization of the wrist may forestall further deterioration. Through a dorsal longitudinal incision the extensor retinaculum is exposed and carefully dissected but left attached at the radial side. The thickened synovium around the extensor tendons, as well as any bony protrusions on the back of the wrist, are removed. The preserved extensor retinaculum is then placed beneath the tendons to further reduce the risk of later tendon rupture.

If the *radioulnar joint* is involved, synovectomy can be combined with excision of the ulnar head and transposition of the extensor carpi radialis longus to the ulnar side of the wrist (to counteract the tendency to radial deviation). Fusion of the lunate to the radius (*Chamay procedure*) prevents ulnar slide of the carpus.

Flexor tenosynovitis is not as obvious as extensor tendon involvement. It may present as *carpal tunnel syndrome* – median nerve compression by swollen tendons in the carpal tunnel – which requires open release of the flexor retinaculum and tenosynovectomy. Obvious bony protrusions in the floor of the carpal tunnel (due to carpal collapse) should be removed and the raw area covered with a soft-tissue flap. Bear in mind that median nerve symptoms in patients with rheumatoid arthritis may be caused by pathology in the proximal part of the limb or the cervical spine, so these patients should always undergo nerve conduction studies and electromyography before the carpal tunnel decompression.

LATE DISEASE

In the late stage tendon ruptures at the wrist, joint destruction, instability and deformity may require reconstructive surgery.

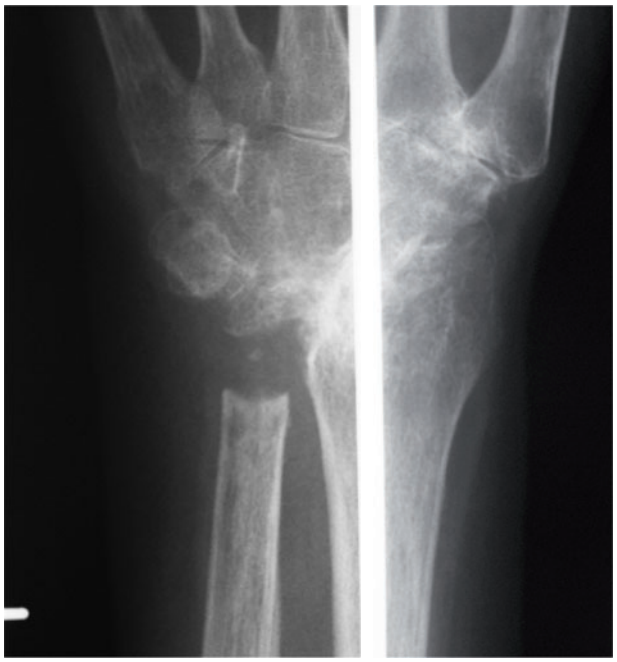
Ruptured extensor tendons can seldom be repaired; side-to-side suture of a distal tendon stump to an adjacent tendon, tendon grafting or tendon transfer gives a satisfactory if not perfect result.

Rupture of the flexor pollicis longus tendon in the carpal tunnel may be caused by scuffing of the tendon against the distal pole of the scaphoid or the edge of the trapezium – the so-called 'Mannerfelt lesion'. Repair or grafting is complex and may disappoint. In low demand individuals, the simplest way of dealing with this problem is to fuse the thumb interphalangeal joint and rely on the other motors to manipulate this important digit.

Painful joint destruction, instability and deformity can be dealt with by either joint replacement or arthrodesis. Arthroplasty using a silicone 'spacer' has been abandoned due to a very high failure rate with silicone synovitis erosion of bone stock. Total wrist replacement with a metal–polyethylene device is becoming more reliable, but is only suitable for those with well-preserved bone stock and low functional demands.

Arthrodesis is widely considered to be the best option for dealing with painful instability in the radiocarpal joint. If the wrist is already 'fusing' itself spontaneously, simple stabilization with a Steinman pin passed between the second and third metacarpals, across the carpus and into the distal radius is all that is needed (Figure 15.25). Bone grafts are not necessarily added but can be taken from the ulnar head if it is excised. For patients with better bone stock, pin fixation is inadequate; formal arthrodesis with a wrist fusion plate is preferable (see Figure 15.29). In this group, ulnar head replacement rather than ulnar head excision should be considered if the patient has high demands from their upper limb.

As a general rule, wrist deformities should be corrected before hand deformities. Furthermore the dominant wrist should, if possible, be fused in slight extension to provide reliable power grip, while the non-dominant wrist is fused in some flexion (or replaced) so as to provide the posture needed for perineal care.



(a) (b)

Figure 15.25 Rheumatoid arthritis wrist fusion Surgical fusion using a long intramedullary pin. The ulnar head has been excised.

OSTEOARTHRITIS OF THE WRIST

Osteoarthritis of the wrist appears at three main sites: the radiocarpal joint, the distal radioulnar joint and the first carpometacarpal joint. Since these usually present as distinct syndromes, they are considered separately.

Radiocarpal osteoarthritis

Osteoarthritis of the radiocarpal joint is uncommon and, when it does occur, it is sometimes a late sequel to infection or an injury such as an intra-articular fracture of the distal radius, an ununited or malunited fracture of the scaphoid, scapholunate ligament rupture or Kienböck's disease; yet it should be borne in mind that, while trauma of all kinds is common, only a fraction of all such injuries lead to arthritis in later life. Most cases of radiocarpal arthritis are probably spontaneous.

Clinical features

The patient may have forgotten the original injury. Years later he or she complains of pain and stiffness. At first these symptoms occur intermittently after use; later they become more constant, with frequent exacerbations or recurrent 'wrist sprains'. The appearance may be normal but there is often swelling over the back of the wrist, and movements are limited and painful.

X-rays

Typical features are narrowing of the radiocarpal joint, subchondral sclerosis and osteophyte formation at the margins of the joint. A predisposing cause, such as an old fracture or Kienböck's disease, may be apparent.

Treatment

CONSERVATIVE MEASURES

Many patients have very advanced radiological changes yet few symptoms. Analgesic medication and rest in a splint are often sufficient treatment. However, if pain is intolerable or if function is seriously disturbed (e.g. if the patient is unable to grip firmly or lift moderately heavy objects), surgical options have to be considered.

SURGICAL TREATMENT

Surgical treatment options are summarized in Figure 15.26.

Partial excision of the radial styloid Osteoarthritis following a scaphoid fracture may be limited to that part of the joint. In that case excision of the tip of the radial styloid process is helpful, but no more than 7 mm must be removed to avoid destabilizing the carpus (Figure 15.27). This can be done by open or arthroscopic means and at the same time a partial wrist denervation may be performed.

OPERATIONS ON CARPAL BONES

Proximal row carpectomy (PCR) or four-corner fusion (4CF) For advanced changes, surgery can help but wrist movement should be preserved if possible. The procedure depends upon the pattern of arthritis. Very often the radius–lunate joint is preserved, which allows either the entire proximal row of carpal bones to be removed (proximal row carpectomy – the head of the capitate then articulates on the lunate fossa of the radius) or scaphoid removal with four-corner fusion (the lunate–capitate–hamate–triquetrum are fused with wires, a circular plate or buried screws) (Figure 15.28).

The outcome of these procedures is similar (about 60% grip strength, 60% movement). Proximal row carpectomy is easier to perform and risks fewer complications; four-corner fusion may give a more stable grip in torsion.

Total arthrodesis of the wrist This is occasionally necessary. The radiocarpal and intercarpal joints are decorticated, bone graft is impacted and a plate is fixed to the third metacarpal and the distal radius. Contouring the plate to 15 degrees of dorsiflexion improves grip strength.

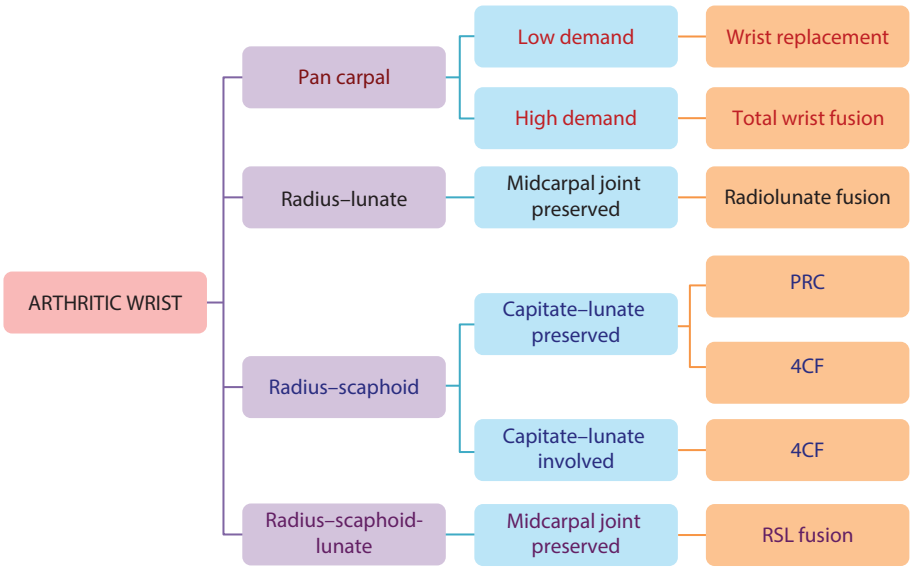


Figure 15.26 Surgical treatment for radiocarpal osteoarthritis PRC: proximal row carpectomy; 4CF: four-corner fusion.

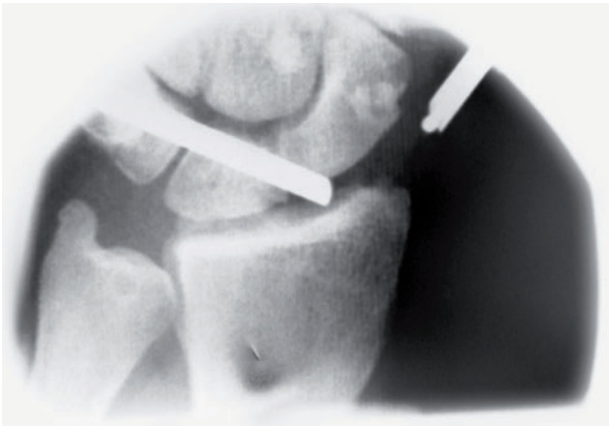
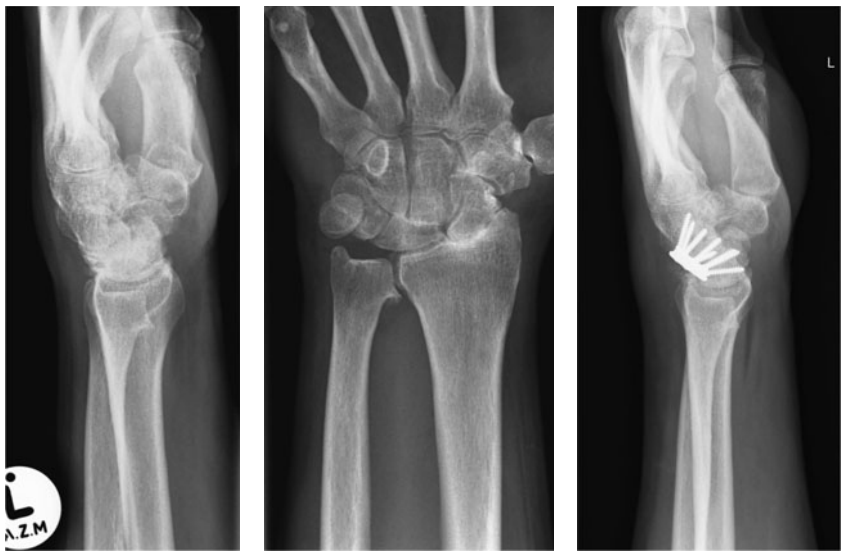


Figure 15.27 Radiocarpal arthritis Early stage treated by arthroscopic radial styloidectomy.

Arthroplasty Wrist replacement with metal or polythene implants is becoming more reliable, although at present this operation is reserved for those with low functional demands (Figure 15.29). Long-term survivorship studies have yet to show whether replacement arthroplasty will hold up in patients with higher demands.

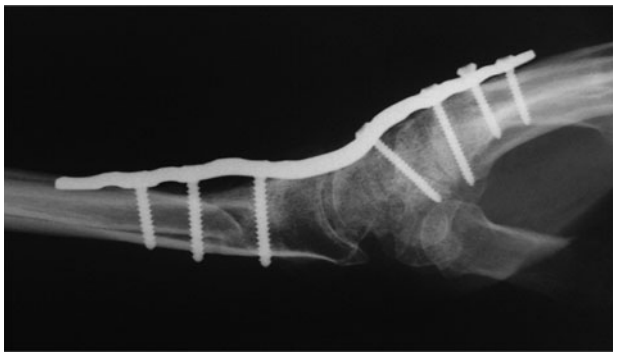
Distal radioulnar arthritis

Progressive destruction of the distal radioulnar joint is a characteristic feature of severe rheumatoid arthritis. Degenerative changes are seen in primary or secondary osteoarthritis (possibly following marked and long-standing instability of the joint).



(a) (b) (c)

Figure 15.28 Radiocarpal arthritis (a,b) The so-called 'SLAC wrist' – scapholunate advanced collapse; (c) treated by scaphoid excision and four-corner fusion.



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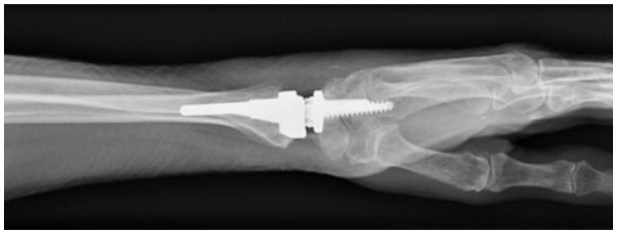

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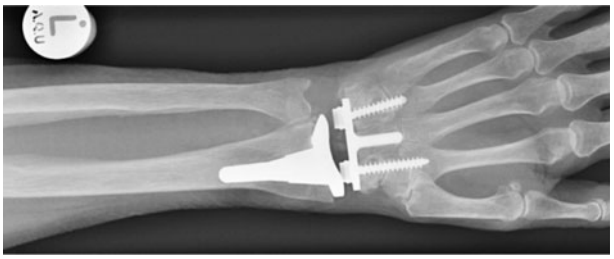


(c)

Figure 15.29 Wrist arthritis (a) Total wrist fusion; (b,c) wrist replacement.

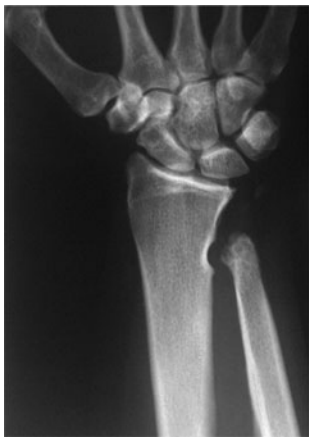
If pain and loss of function cannot be controlled by conservative measures, the patient may benefit from ulnar head replacement (although, like all implants in the hand and wrist, there is a risk of significant early complications and the durability is uncertain). Operations that involve excision of the ulnar head (Darrach's, Sauve Kapandji) should be undertaken with extreme caution and avoided if possible because of the high risk of causing severe and intractable instability (Figure 15.30).

Carpometacarpal osteoarthritis

Osteoarthritis of the trapeziometacarpal joint is common in postmenopausal women. It is often accompanied by Heberden's nodes of the finger joints, in which case it is usually bilateral and part of a generalized osteoarthritis.

Clinical features

The patient, usually a middle-aged or older woman, complains of diffuse pain around the base of her thumb. Pinch and grip are weakened. On examination, the joint is swollen and in advanced cases is held in an adducted position, with prominence of the subluxed metacarpal base. With more established fixed adduction of the thumb base, the metacarpophalangeal joint hyperextends to provide a competent thumb–index span. The carpometacarpal joint is tender and the 'grind test' (compressing and rotating the metacarpal longitudinally against the trapezium) is often painful. Stressing the joint by the examiner pushing the metacarpal head into adduction against the side of the index metacarpal, and by the examiner extending the thumb metacarpal backwards parallel to the index metacarpal, are the most sensitive tests.



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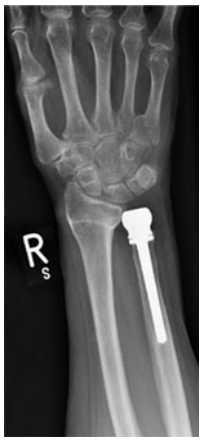

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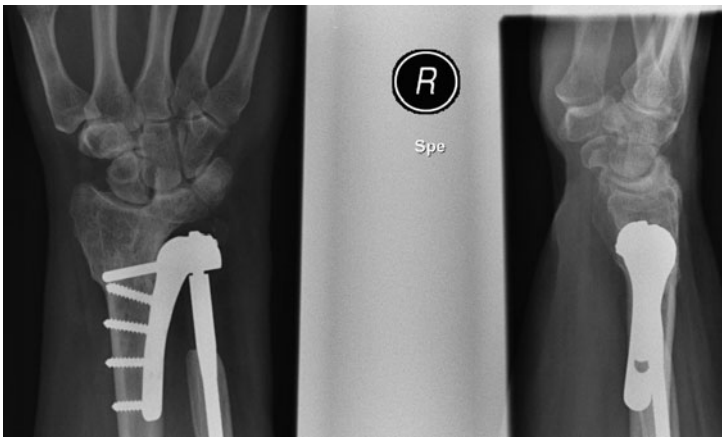


(c)

Figure 15.30 Distal radioulnar joint arthritis – operative treatment (a) Excising too much of the distal ulna may cause painful radioulnar impingement. (b) One alternative is an ulnar head replacement. (c) Total DRUJ replacement for arthritis and instability.

Imaging

X-rays show narrowing and then lateral subluxation of the trapeziometacarpal joint. *MRI and arthroscopy* are useful in early cases when the diagnosis is in doubt.

Treatment

Asymptomatic radiological disease is common, affecting 60% of women over 60. Most symptomatic patients can be treated by reassurance that it will settle, anti-inflammatory preparations, local corticosteroid injections and temporary splintage. There may be a role for intra-articular hyaluronidase. If these measures fail to control pain, or if instability becomes marked, operative treatment is considered (Figure 15.31).

SURGICAL TREATMENT

Joint-preserving operations In early cases, joint-preserving operations are helpful for about 70% of patients. The options are either *extension osteotomy* or *ligament reconstruction*. These procedures alter the joint forces and thus improve pain and function.

Excisional arthroplasty Excision of the trapezium gives pain relief and return of function, though thumb pinch is always weak. The bone can be removed through either the palmar approach or the anatomical snuffbox, taking care not to damage the superficial radial nerve, the radial artery or the flexor carpi radialis tendon. Attempts have been made to prevent postoperative collapse of the joint and proximal migration of the metacarpal by rerouting a slip of flexor carpi radialis or abductor pollicis longus tendon. The benefit of this extra intervention has not been established.

Replacement arthroplasty Replacement arthroplasty using a silicone spacer has a high complication rate and the results are unpredictable. Metal-on-polyethene implants and pyrocarbon implants are also available but they are experimental and long-term durability is uncertain. Many devices have had unacceptable failure rates and have thus been abandoned. There is no evidence that implants are better than trapeziectomy, except that the earlier recovery is quicker with an implant.

Arthrodesis Arthrodesis of the trapeziometacarpal joint relieves pain, but the restriction of movement and high failure rate are distinct drawbacks. The scaphotrapezial joint should be normal.

If the metacarpophalangeal joint has been secondarily damaged by hyperextension, then either a sesamoid arthrodesis (which restores flexion but preserves movement) or fusion (at 25 degrees for stable pinch) is indicated.

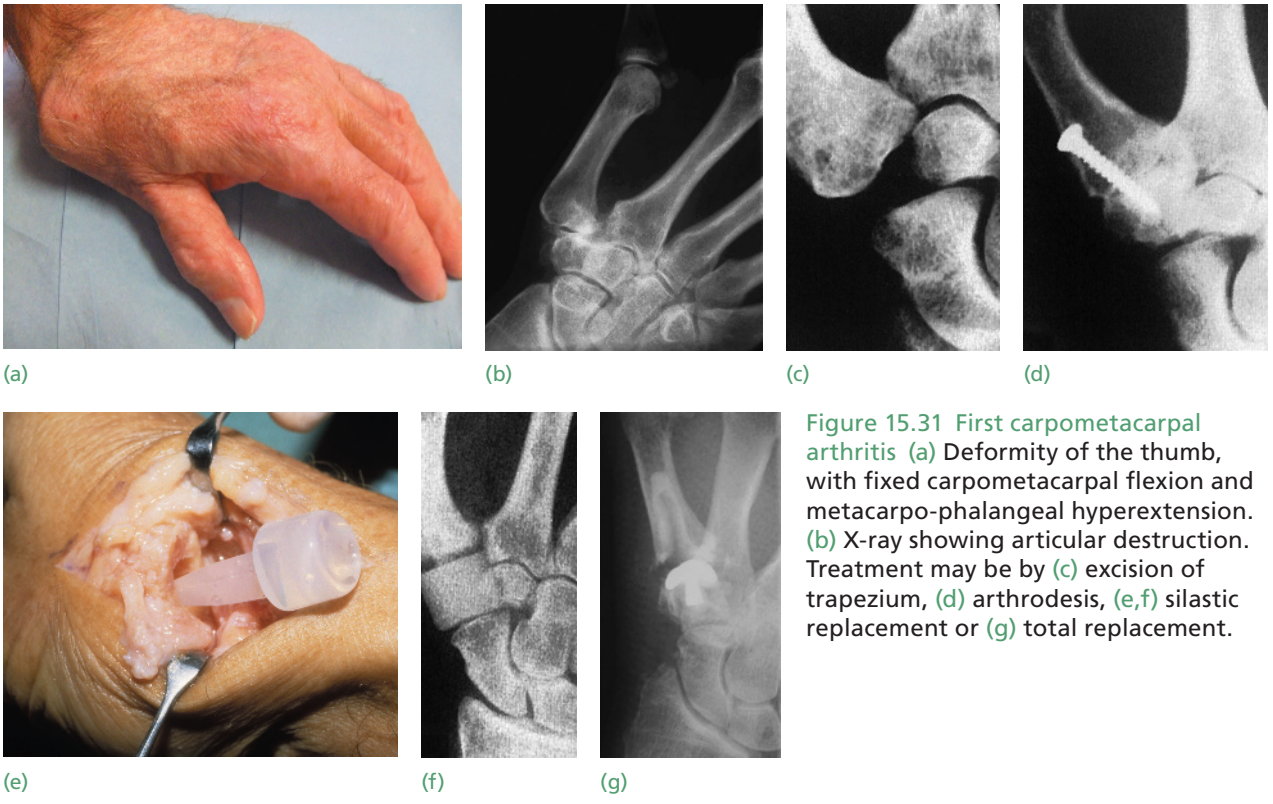


Figure 15.31 First carpometacarpal arthritis (a) Deformity of the thumb, with fixed carpometacarpal flexion and metacarpo-phalangeal hyperextension. (b) X-ray showing articular destruction. Treatment may be by (c) excision of trapezium, (d) arthrodesis, (e,f) silastic replacement or (g) total replacement.

Scaphoid–trapezium–trapezoid (STT) arthritis

The joint between the distal end of the scaphoid and the underside of the trapezium and trapezoid ('the triscaphe joint') can develop arthritis either in isolation or in association with arthritis of the carpometacarpal joint.

Late middle-aged females are most commonly affected. The patient points to the front of the scaphoid tubercle as the source of the pain (whereas in carpometacarpal arthritis the patient points to the back of the thumb base).

Treatment

Treatment is initially along standard lines – adaptive measures, anti-inflammatory medication, cortisone injection and splintage. Injections are best performed under ultrasound or fluoroscopic control.

Patients with severe symptoms may benefit from surgery (Figure 15.32). However, there is as yet no completely satisfactory operation. *STT fusion* removes the painful joint but is technically difficult. *Excision of the distal pole of the scaphoid* is easier but can cause midcarpal collapse unless only the smallest amount is removed. *Trapeziectomy with undercutting of the trapezoid* is straightforward, especially if there is concomitant trapeziometacarpal arthritis. *Pyrocarbon interposition arthroplasty* is an experimental option, but whether this has any immediate or long-term advantage over simple excision is unknown.

Pisotriquetral arthritis

This unusual condition usually occurs spontaneously. Occasionally it follows trauma. Pain is felt over the front of the ulnar side of the hand, worse when the FCU tendon (for which the pisiform is a sesamoid) is loaded – for example, holding a heavy iron.

Pushing backwards on the pisiform is painful, as would be lunotriquetral pathology; to distinguish it, the key physical sign is pain and crepitus when stressing the pisiform radialwards with the examiner's thumb.

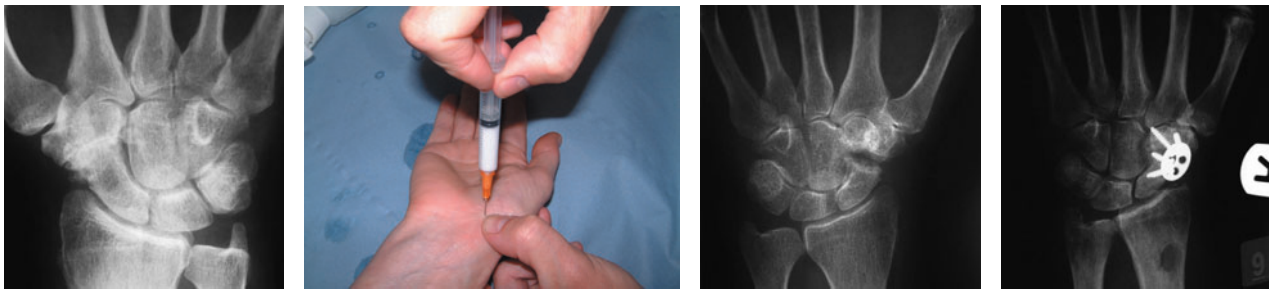
Normal *X-rays* do not show the disease; a lateral in 25 degrees supination is needed (e.g. Figure 15.33a). If any doubt, then a *CT or MRI* will secure the diagnosis.

Treatment

The pain may settle with time and a splint. Treatment with a steroid injection may be helpful. Occasionally, surgical excision is undertaken with excellent results unless the immediately adjacent ulnar nerve is damaged.

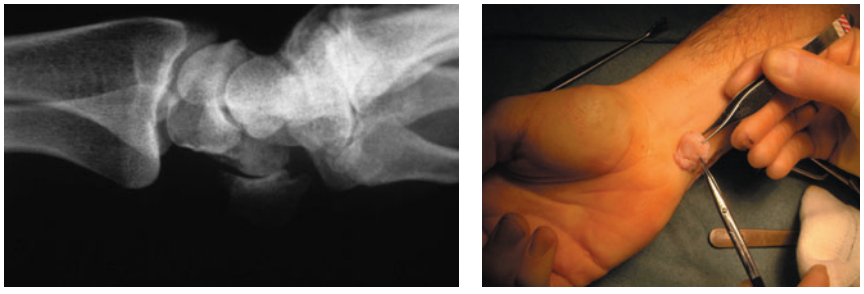
TENDON PATHOLOGY

The extensor retinaculum has six compartments which transmit tendons lined with synovium. Tenosynovitis can be caused by unaccustomed overuse but sometimes it occurs spontaneously. The resulting synovial



(a) (b) (c) (d)

Figure 15.32 Scaphoid–trapezium–trapezoid arthritis (a) Changes on X-ray; (b) steroid injection; (c) distal pole of scaphoid excision (do not remove too much!); (d) STT fusion.



(a) (b)

Figure 15.33 Pisotriquetral arthritis (a) Shown on 25° supinated view. (b) Pisiform excision gives good results – but beware the ulnar nerve!

inflammation causes secondary thickening of the sheath and stenosis of the compartment, which further compromises the tendon. Early treatment, including rest, anti-inflammatory medication and injection of corticosteroids, may break this vicious circle.

The first dorsal compartment (abductor pollicis longus and extensor pollicis brevis) and the second dorsal compartment (extensor carpi radialis brevis) are most commonly affected. The flexor tendons are affected far less frequently.

De Quervain's disease

Pathology

This condition, first described in 1895, is caused by reactive thickening of the sheath around the extensor pollicis brevis and abductor pollicis longus tendons within the first extensor compartment. It may be initiated by overuse but it also occurs spontaneously, particularly in middle-aged women, and sometimes during pregnancy.

Clinical features

The patient is usually a woman aged 40–50, who complains of pain on the radial side of the wrist. There may be a history of unaccustomed activity such as pruning roses or wringing out clothes. Sometimes there is a visible swelling over the radial styloid and the tendon sheath feels thick and hard. Tenderness is most acute at the very tip of the radial styloid.

The pathognomonic sign is elicited by *Finkelstein's test* (Figure 15.34): the examiner places the patient's thumb across the palm in full flexion and then, holding the patient's hand firmly, turns the wrist sharply into adduction. In a positive test this is acutely painful; repeating the movement with the thumb left free is relatively painless. Resisted thumb extension (hitch-hiker's sign) is also painful.

The *differential diagnosis* includes arthritis at the base of the thumb, scaphoid non-union and the intersection syndrome (see below).

Treatment

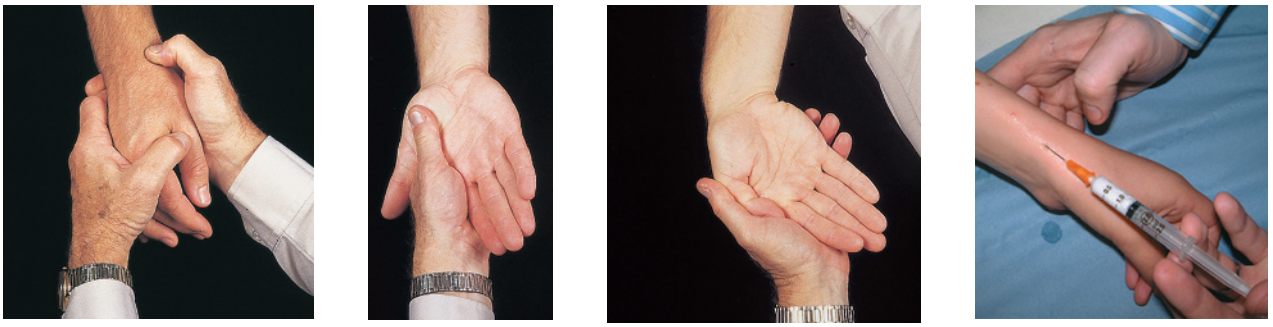
The early case can be relieved by a corticosteroid injection into the tendon sheath, sometimes combined with hand therapy (ultrasound, frictions, splintage). Resistant cases need an operation, which consists of slitting the thickened tendon sheath. Sometimes there is duplication of tendons and even of the sheath, in which case both sheaths need to be divided. Care should be taken to prevent injury to the dorsal sensory branches of the radial nerve, which may cause intractable dysaesthesia.

Intersection syndrome

This condition, otherwise known as *crossover syndrome* or *peritendinitis crepitans*, is characterized by pain, swelling and crepitus over the tendons of extensor pollicis brevis and abductor pollicis longus 4–6 cm proximal to the extensor retinaculum. It is found in weightlifters, canoeists and rowers. It should be distinguished clinically from de Quervain's disease. The condition is generally attributed to friction between these tendons (the so-called 'outcropping tendons') and the underlying longitudinally aligned extensor tendons, leading to an adventitious bursa or a tenosynovitis. There is usually an associated tenosynovitis within the second extensor compartment containing extensors carpi radialis longus and brevis.

Treatment

Treatment involves rest, splintage, steroid injection and, in resistant cases, surgical widening of the second compartment and exploration of the intersection.



(a) (b) (c) (d)

Figure 15.34 De Quervain's disease (a) There is point tenderness at the tip of the radial styloid process. (b,c) Finkelstein's test: Ulnar deviation with the thumb left free is relatively painless (b) but, if the movement is repeated with the thumb held close to the palm, (c) the pull on the thumb causes intense pain. (d) Injecting the tendon sheath.

Other sites of extensor tenosynovitis

Overuse tenosynovitis of *extensor carpi radialis brevis* (the most powerful extensor of the wrist) or *extensor carpi ulnaris* may cause pain and point tenderness just medial to the anatomical snuffbox or immediately distal to the head of the ulna, respectively (see Figure 15.2). Splintage and corticosteroid injections are usually effective.

The common extensor compartment is occasionally irritated by direct trauma. Patients present with pain and crepitus on the dorsum of the wrist; flexing and extending the fingers produces a fine, palpable crepitus over the common extensor compartment. *Treatment* is by rest and splintage of the wrist.

Extensor tenosynovitis is also a common feature of rheumatoid disease.

Flexor tendinitis

Except in specific inflammatory disorders such as rheumatoid arthritis, the flexor tendons are rarely affected.

Flexor carpi radialis tendinitis (Figure 15.35) causes pain on the front of the wrist alongside the scaphoid tubercle; symptoms are reproduced by resisted wrist flexion. Tenderness is sharply localized and should be distinguished from that of de Quervain's disease or osteoarthritis of the basal joint of the thumb.

Flexor carpi ulnaris can become inflamed near its insertion into the pisiform. Occasionally X-rays show calcific deposits around the sheath.

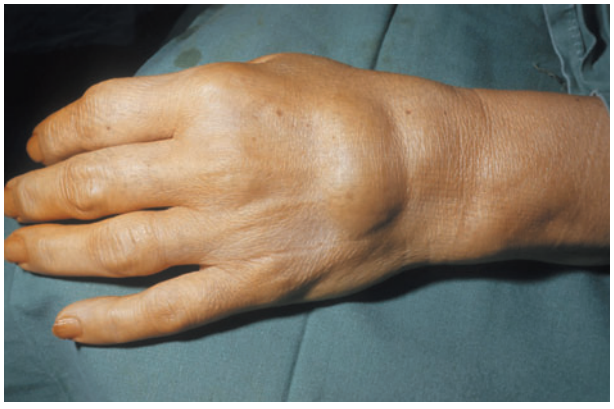
Treatment of these conditions is the same as for the other types of tenosynovitis.

Extensor carpi ulnaris instability

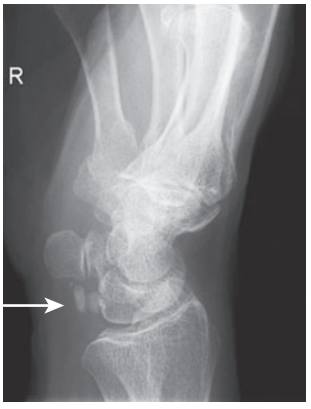
The extensor carpi ulnaris (ECU) tendon is a wrist extensor in pronation and a wrist adductor in supination. It is held against the back of the ulna head by its own sheath. This sheath can give way after a sudden supination stress (usually in tennis or rugby). The tendon then clunks as the wrist passes from pronation to supination. Surgical repair is usually required and involves either a direct reattachment or a reconstruction using a patch of retinaculum.

Occupational pain disorders

Inappropriate terms such as *repetitive stress injury* and *cumulative trauma disorder* have been used in the past for a controversial syndrome comprising ill-defined and unusually disabling pain around the wrist and forearm (and sometimes the entire limb) which is usually ascribed to a particular work practice. There is no good evidence to suggest that using the hands causes harm – after all, training makes muscles and tendons less vulnerable to damage. Exceptions include excessive vibration which can lead to carpal tunnel syndrome and hand–arm vibration syndrome (neurological damage to the fine nerves supplying the fingertips and Raynaud's phenomenon). Other defined and treatable conditions such as carpal tunnel syndrome, thumb base arthritis, tenosynovitis from sudden unaccustomed use and de Quervain's should be excluded and treated accordingly. Epidemiological studies suggest that these conditions are no more common among keyboard operators than in the general population. What has fuelled the controversy surrounding the 'occupational' disorders is their apparent severity and intractability compared with other types of overuse syndrome and the potential rewards for successful litigation. There are often social and psychological aspects which confound the picture. The term 'work-relevant upper limb disorder' is preferred as it acknowledges that the symptoms are noticed at work but it does not imply causation.



(a)



(b)

Figure 15.35 Other types of tendinitis
(a) Rheumatoid; (b) calcific tendinitis of flexor carpi ulnaris.

SWELLINGS AROUND THE WRIST

Ganglion cysts

Pathology

The ganglion cyst is the most common swelling in the wrist. It arises from leakage of synovial fluid from a joint or tendon sheath and contains a glairy, viscous fluid. Although it can appear anywhere around the carpus, it usually develops on the dorsal surface of the scapholunate ligament. Palmar wrist ganglia usually arise from the scapholunate or scaphoid–trapezium–trapezoid joint.

Clinical features

The patient, often a young adult, presents with a painless lump, though occasionally there is slight ache and weakness. The lump is well defined, cystic and not tender; it can sometimes be transilluminated. It does not move with the tendons. The back of the wrist is the commonest site; less frequently a ganglion emerges alongside the radial artery on the volar aspect (Figure 15.36). Occasionally a small, hidden ganglion is found to be the cause of compression of the deep (muscular) branch of the ulnar nerve.

Treatment

Treatment is usually unnecessary. The lump can safely be left alone; it often disappears spontaneously. However, it can be aspirated to reassure the patient. A complex volar ganglion might be better aspirated under ultrasound control due to the proximity of the radial artery and vulnerable nerves. If it becomes troublesome – and certainly if there is any pressure on a nerve – operative removal is justified. Even then it may recur with embarrassing persistence; it is not easy to ensure that the root is removed.

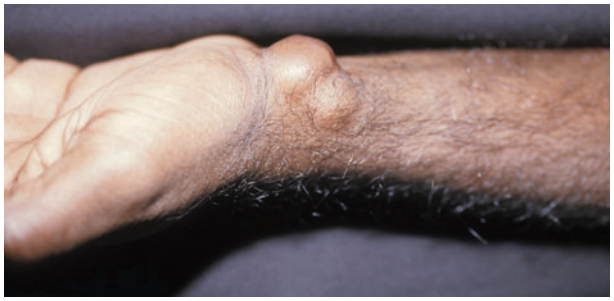


Figure 15.36 Volar wrist ganglion

Occult ganglion or dorsal synovial impingement

Sometimes patients complain of pain in the back of the wrist, provoked by extending the wrist. On examination there is a discrete tender point over the back of the midcarpus and the pain is reproduced by full passive wrist extension. *Ultrasound*, or preferably *MRI*, will show either a small ganglion (Figure 15.37) or thickening of the synovium at the radiocarpal or midcarpal joint.

Treatment should be initially with a steroid injection; if that fails, then arthroscopic excision may succeed.

Extensor tenosynovitis

Localized swelling of a tendon sheath on the dorsum of the wrist sometimes occurs in rheumatoid disease and can be mistaken for a 'cyst'.

Carpometacarpal boss

A firm round swelling over the back of the second and third carpometacarpal joint is sometimes seen in a young adult (Figure 15.38). It is not always tender. It is thought that it may be caused by some instability at the joint.

Treatment involves reassurance; the lump can be excised but if it recurs, the underlying joint should be fused.

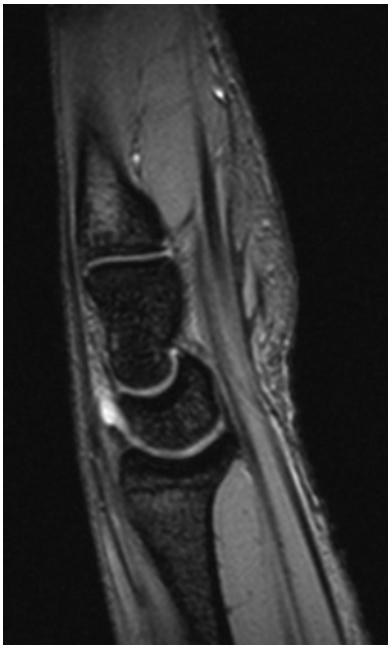
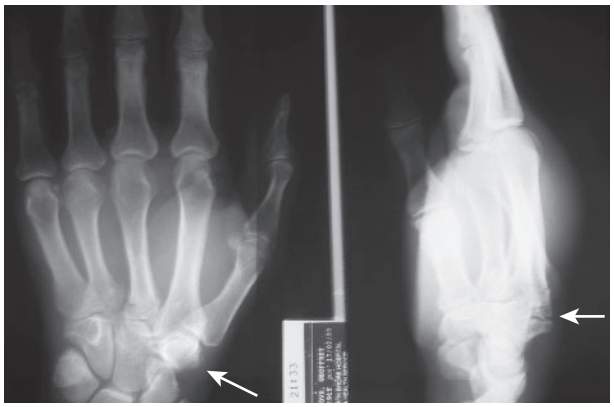


Figure 15.37 Occult ganglion
MRI shows an occult ganglion, best treated arthroscopically.



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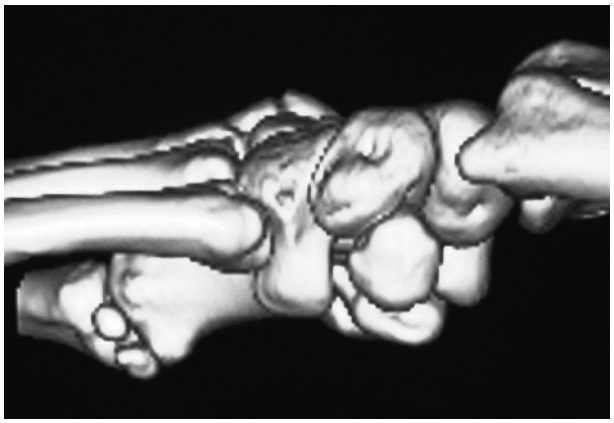


(b)

Figure 15.38 Carpometacarpal boss (a) X-ray; (b) three-dimensional CT.

'Compound palmar ganglion'

This lesion is neither a ganglion nor compound. Chronic inflammation distends the common sheath of the flexor tendons both above and below the flexor retinaculum. Rheumatoid arthritis and tuberculosis are the commonest causes. The synovial membrane becomes thick and villous. The amount of fluid is increased and it may contain fibrin particles moulded by repeated movement to the shape of melon seeds. The tendons may eventually fray and rupture.

Clinical features

Pain is unusual but paraesthesia due to median nerve compression may occur. The swelling is hourglass in shape, bulging above and below the flexor retinaculum; it is not warm or tender; fluid can be pushed from one part to the other (cross-fluctuation).

Treatment

If the condition is tuberculous, general treatment is begun. The contents of the sac are evacuated, streptomycin is instilled and the wrist rested in a splint. If these measures fail, the entire flexor sheath is dissected out. Complete excision is also the best treatment when the cause is rheumatoid disease.

CARPAL TUNNEL SYNDROME

The carpal tunnel syndrome, due to median nerve compression under the flexor retinaculum of the wrist, is described together with other nerve compression disorders in Chapter 11.

NOTES ON APPLIED ANATOMY

In most positions of the forearm the styloid process of the radius is more distal than that of the ulna, but with the forearm supinated the two processes are at approximately the same level. This relationship, known as *ulnar variance*, may be altered as a result of growth abnormalities or injury. The ulna appears longer with the wrist in pronation as the radius passes obliquely across the forearm.

Relative shortness of the ulna appears as an anatomical variant in association with Kienböck's disease. *Relative overlength* is associated with ulna–carpal impaction syndrome (central TFCC perforations and late ulnocarpal arthritis).

Gilula's arcs These lines are helpful radiographic indicators. They are illustrated in Figure 15.39. The congruent lines between the distal radius/ulna and proximal carpal row, and between the proximal and distal carpal row, are disturbed in midcarpal instability, Kienböck's disease and dislocation of the wrist.

Normal radiographic anatomy

Normal angles (Figure 15.40):

- Radial tilt: 22 degrees
- Palmar tilt: 11 degrees
- Scapholunate angle: 30–65 degrees

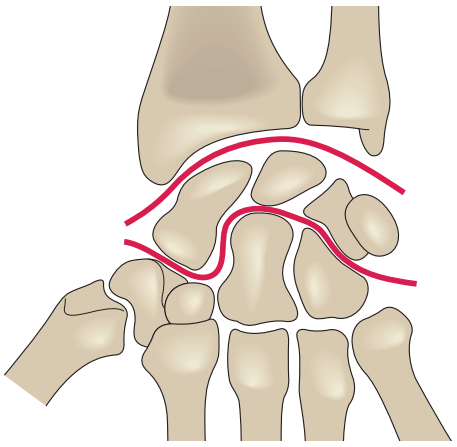


Figure 15.39 Gilula's arc

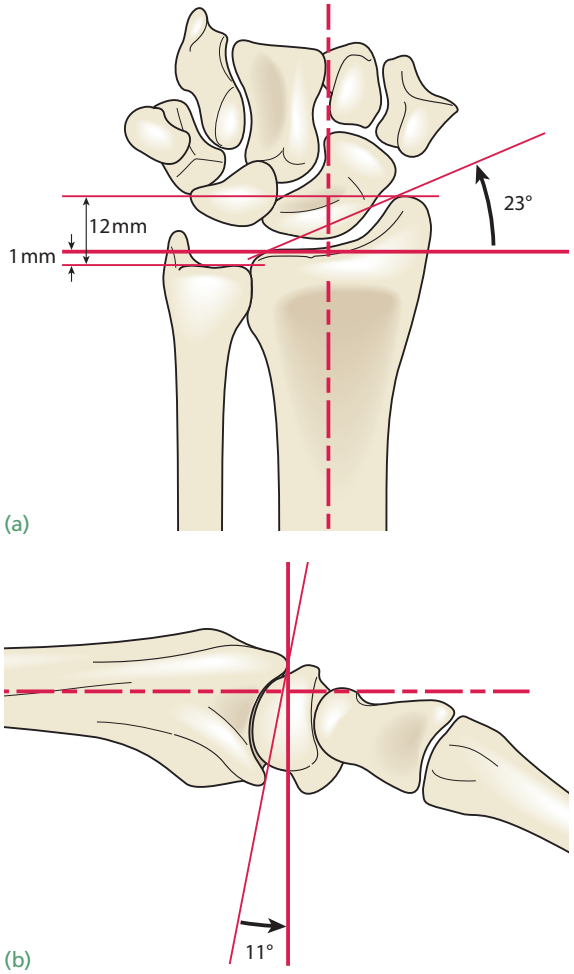


Figure 15.40 Angle of the distal radius

Carpal height The ratio between the distal edge of the capitate and proximal edge of the lunate/third metacarpal is usually 0.54 ± 0.03. The ratio is reduced in carpal collapse (e.g. with Kienböck's disease and scapholunate ligament failure).

Just distal to the radial styloid is the scaphoid, immediately beneath the anatomical snuffbox, which is one of the key areas for localizing tenderness. Tenderness at the distal end of the snuffbox may incriminate the carpometacarpal joint of the thumb. More proximal tenderness, at the tip of the radial styloid, is characteristic of de Quervain's disease. Dorsal to the snuffbox the oblique course of extensor pollicis longus exposes it to damage by a careless incision.

The carpal bones are arranged in two rows, with the pisiform as the odd man out. The scaphoid crosses both rows. The scaphoid, trapezium and thumb combine to function almost as a separate entity, a 'jointed strut', with independent movement; degenerative arthritis of the wrist occurs most commonly in the joints of this strut.

Kinematics

Wrist flexion The proximal row and distal row flex and ulnar deviate.

Wrist extension The proximal row and distal row extend and radially deviate.

Radial deviation There are two synchronized movements. First, the proximal row flexes (to prevent scaphoid blocking flexion between radial styloid and trapezium); distal row slightly extends. Second, the scaphoid slides ulnarwards, pushing lunate and triquetrum. There is variable flexion and sliding between individuals known as 'column wrist' and 'row wrist'.

Ulnar deviation The proximal row extends and distal row slightly flexes.

Radial–ulnar deviation is provided 60% by the midcarpal and 40% by the radiocarpal/ulnocarpal joints.

Flexion-extension This is about 50% midcarpal and 50% radiocarpal.

Range of movement

Normally the arc of flexion–extension is 110–150 degrees; radial tilt is 30 degrees and ulnar tilt 45 degrees. The *functional range* is about 10 degrees flexion to 30 degrees extension. The 'dart-throwers' motion' is now recognized as an important aspect of function – i.e. the ability to move the wrist alternately into radial tilt–extension and ulnar tilt–flexion. Many routine activities rely on this particular synchronized motion; tendon transfers and partial wrist fusions should be designed to maintain these movements whenever the pathology allows.

Surgical anatomy of the nerves

On their volar aspect the carpal bones form a concavity roofed over by the carpal ligament; in the tunnel lie the flexor tendons and the median nerve. The thenar branch of the nerve (supplying the all-important thenar muscles) is in danger if, during a decompression operation, the carpal ligament is divided too far radially. On the dorsoradial side of the wrist, branches of the superficial radial nerve are vulnerable (beware during operations for ganglia, de Quervain's, thumb carpometacarpal joint). On the ulnar side, the close relationship of the ulnar nerve to the pisiform and hamate hook must be borne in mind. Operations at the distal end of the ulna threaten the dorsal branch of the ulnar nerve which runs anteriorly about 3 cm proximal to the ulnar styloid.

Ossification of the wrist bones

The ossific centre for the distal radius epiphysis appears at age 2 and fuses at age 16–18. The other

bones develop ossification centres in clockwise order (looking at the right hand from behind, fully pronated, i.e. face down): capitate (1 month), hamate (1 year); triquetrum (2–3 years); lunate (4 years); scaphoid (4–6 years); trapezium (4–6 years); trapezoid (4–6 years); pisiform (8–10 years).

NOTE: In an adolescent, the incompletely ossified scaphoid can be mistaken for a scapholunate dissociation.

Ligaments of the wrist

The extrinsic ligaments are discrete consolidations of the capsule. The palmar ligaments are stronger than the dorsal (Figure 15.41).

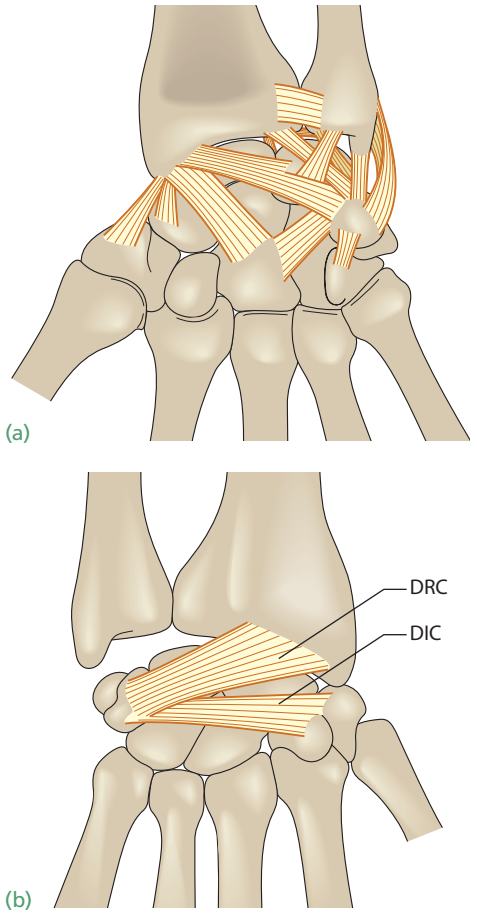


Figure 15.41 Ligaments of the wrist (a) Palmar ligaments of the wrist; (b) dorsal ligaments of the wrist. DRC = dorsal radiocarpal ligament; DIC = dorsal intercarpal ligament.

EXTRINSIC CARPAL LIGAMENTS (DORSAL)

- *Dorsal radiocarpal ligaments* (radioscaphoid, radiotriquetral). Rupture contributes to VISI.
- *Dorsal intercarpal ligament* (triquetrum to scaphoid and trapezoid). Available as a donor for tenodesis against palmar rotation of scaphoid.

EXTRINSIC CARPAL LIGAMENTS (PALMAR)

- *Radioscaphocapitate ligament.* Attaches to palmar edge of radial styloid. Fulcrum for scaphoid flexion. Divided then carefully repaired during palmar approach to scaphoid. Readily seen in arthroscopy. Beware removing attachment by enthusiastic radial styloidectomy.
- *Long radiolunate ligament.* Restrains lunate from palmar dislocation.
- *Ligament of Testut (radioscapholunate).* Synovial fold, no stabilizing function. Landmark for scapholunate ligament in wrist arthroscopy.
- *Short radiolunate ligament.* From ulnar edge of distal radius to lunate, blends ulnarwards with the ulnolunate ligament.
- *Ulnocarpal ligament.* Ulnocapitate, ulnolunate, ulnotriquetral. Blend into volar radiolunate ligament (i.e. anterior limb of TFCC). Ulnotriquetral ligament blends into sub-sheath of extensor carpi ulnaris (also part of TFCC).

SPACE OF POIRIER

This is the gap between the lunate and midcarpal joint through which the lunate can dislocate anteriorly.

INTRINSIC (INTEROSSEOUS LIGAMENTS)

- *Scapholunate interosseous ligament:* C-shaped, thickest dorsally.
- *Lunotriquetral:* C-shaped, thickest palmarwards.
- *Capitate–hamate; trapezium–capitate; trapezium–trapezoid.*

Blood supply of the wrist

There are dorsal and palmar arches, supplied by the radial artery, ulnar artery and anterior interosseous artery. These can be used as flaps to vascularize the scaphoid and lunate.

The hand

16

David Warwick

The hand is (in more senses than one) the medium of introduction to the outside world. Its unique repertoire of prehensile movements, grasp, pinch, hook-action and tactile acuity sets us apart from all other species. We can think of the hand as a sophisticated tool, but it is also an organ of communication, used for gesturing and expressing a range of emotions from anxiety and fear to submission and helplessness, scorn and hatred, determination and control, or tenderness and love. We are more aware of our hands than of any other part of the body; when they go wrong, we know about it from a very early stage.

CLINICAL ASSESSMENT

Symptoms

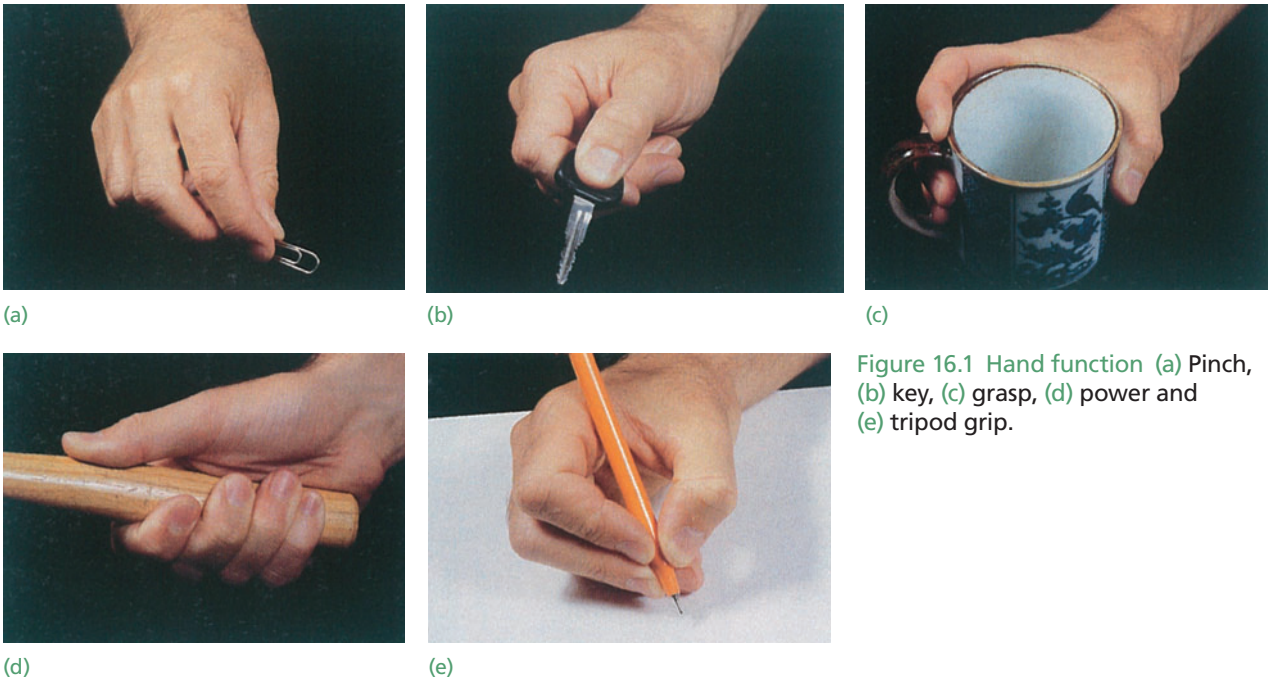
Pain may be felt in the palm, the thumb or the finger joints. Remember, though, that a poorly defined pain may be referred from the neck, shoulder or mediastinum.

Deformity may appear suddenly (e.g. due to tendon rupture) or slowly (suggesting bone or joint pathology, a soft-tissue contracture or a postural defect due to a nerve lesion).

Swelling may be localized (and, if associated with throbbing pain, is almost certainly due to infection) or it may be evident in many joints simultaneously. Ask whether the swelling is constant or intermittent, and how long it has been present.

Sensory symptoms and motor weakness provide well-defined clues to neurological disorders. A precise description of the affected area tells us a great deal about the level of the lesion.

Loss of function takes various forms. The patient may have difficulty handling eating utensils, holding a cup or glass, grasping a doorknob (or a crutch), dressing or (most trying of all) attending to personal hygiene. Equally important is loss of function due to sensory change in the fingers.



(a) (b) (c) (d) (e)

Figure 16.1 Hand function (a) Pinch, (b) key, (c) grasp, (d) power and (e) tripod grip.

Signs

Both upper limbs should be bared for comparison. Before focusing on the hands, take a quick look at the shoulders and elbows and their range of movement. Also ask which is the dominant hand. A rapid assessment can be carried out in a few minutes. A full examination needs patience and meticulous attention to detail.

Look

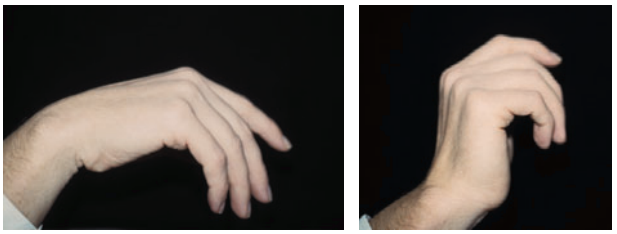
Note how the patient holds the hand and uses it during the interview; the resting posture may be suggestive of nerve or tendon damage. Ask the patient to place both hands on the table in front of you, with the palms first upwards and then downwards. The skin may be scarred, altered in colour, dry or moist, and hairy or smooth. Puckering and ridging of the skin in the palm, sometimes extending into one of the fingers, are cardinal signs of Dupuytren's contracture. Deformity of the fingers and the presence of any lumps should be noted. Swelling may arise in the subcutaneous tissues, in a tendon sheath or in a joint. Do not forget to look at the nails as they may show signs of atrophy or disease: for example, psoriasis, which is sometimes associated with a typical arthropathy, or a 'grooved' nail which is a telltale feature of a ganglion cyst at the nail bed.

If multiple joints are involved, take careful note of their distribution. Characteristically, rheumatoid arthritis causes swelling of the proximal joints – metacarpophalangeal (MCP) and proximal interphalangeal (PIP) – while osteoarthritis affects mainly the distal interphalangeal (DIP) joints.

Compare the thenar eminences of the two hands and look for wasting on one or other side (a sign of median nerve dysfunction).

Posture in different resting positions While looking at the patient's hands, observe their resting posture in different positions. Normally, with the palm upwards, the fingers fall into a gentle cascade with the MCP joints slightly flexed – about 30 degrees in the index, ranging to 70 degrees in the little. The interphalangeal (IP) joints similarly lie in increasing flexion from index to little. When the hand is turned palm downwards, the fingers straighten out, again in a gentle cascade with greater extension on the index finger than the little finger. If the regular cascade is interrupted, then a tendon is probably either divided or stuck. If the cascade is normal but active movements are not possible, then a nerve injury should be suspected.

Note also that there is a reciprocal relationship between the position of the wrist and the resting position of the fingers (Figure 16.2). Normally, as the wrist drops into flexion the fingers automatically tend to straighten, and when the wrist is pulled into extension the fingers flex slightly; contractures of the long flexors will cause the fingers to curl tightly in flexion when the wrist is extended.



(a) (b)

Figure 16.2 Passive tenodesis Note the resting position of the fingers with the wrist (a) flexed, (b) extended.

Feel

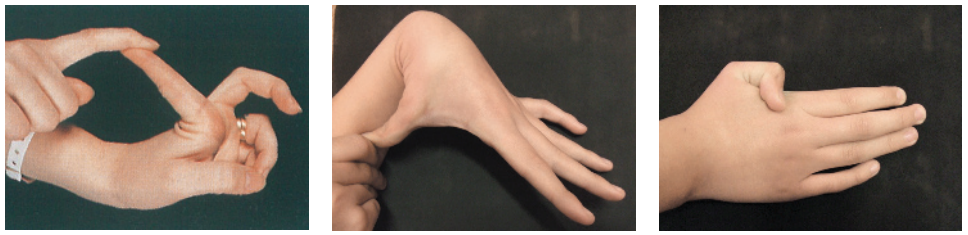
The temperature and texture of the skin are noted and the pulse is felt. Swelling or thickening may be in the subcutaneous tissue, a tendon sheath, a joint or one of the bones. If a nodule is felt, the underlying tendon should be moved (by flexing and extending the relevant finger) to discover if the nodule is attached to the tendon or its sheath. This will also reveal whether the tendon glides smoothly or whether it gets stuck momentarily with finger in flexion and then snaps free as the finger is extended (the 'trigger finger' effect). Any point of tenderness should, if possible, be accurately localized to a particular structure.

Move

Passive movements There is a good argument for starting with passive movements, so that you can see whether all the little finger joints are *capable* of moving before testing the *patient's ability* to move them. The thumb and each finger are examined in turn and the range of movement recorded. Note whether the movement causes pain.

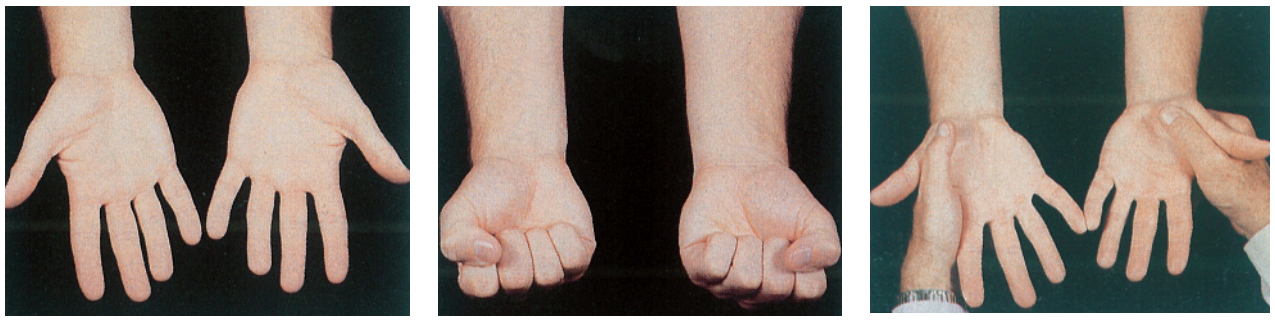
Some degree of passive hyperextension at the MCP joints (tested by gently pushing each finger dorsalwards to its limit) is normal, but anything more than 90 degrees of (hyper)extension is suggestive of generalized joint laxity (Figure 16.3); the diagnosis can be confirmed by testing the range of extension in other joints such as the thumbs, elbows and knees.

Active movements Ask the patient to place both hands with the palms facing upwards, to *extend* the fingers and thumbs fully and then to curl them into full *flexion* as if making a gentle fist (Figure 16.4). A 'lagging finger' is immediately obvious, though



(a) (b) (c)

Figure 16.3 Joint laxity (a) in the fingers, (b,c) in the thumb.



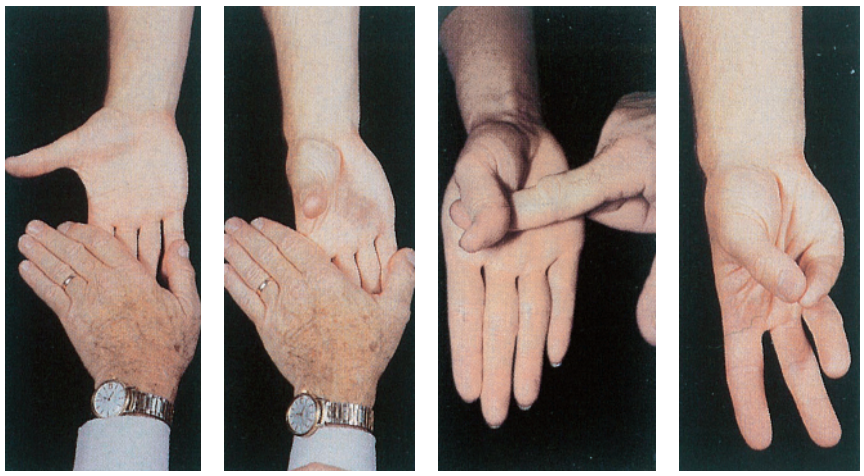
(a) (b) (c)

Figure 16.4 Gross active movement (a) Full extension. (b) Full flexion. (c) A good test for abductor power is to have the patient spread his or her fingers as strongly as possible; slowly push the hands together until the tips of the little fingers are forcefully opposing one another; the weaker one will collapse.

it still remains to establish whether this is due to a stiff joint, a defective tendon or loss of motor power. Active movements at each of the MCP, PIP and DIP joints will have to be examined.

Abduction and adduction When the MCP joints are held in extension, they are able to move sideways in the plane of the flattened hand; this is because, in the extended position, the collateral ligaments of the MCP joints are somewhat lax. Spreading the fingers apart is denoted as *abduction* and bringing them back to the neutral position (all the fingers side by side) is *adduction*. Active power can be roughly gauged by having the patient abduct the fingers forcibly and the examiner then pressing against the spread-out index and little fingers, trying to force them back to the neutral position. A better way is to ask the patient to spread the fingers of both hands to the maximum; the examiner then grasps the patient's hands, pushes them towards each other and forces the two little fingers against each other. The weaker (non-dominant) side will normally give way first, but if the difference in one or other hand is very marked it signifies true abductor weakness, a sign of ulnar nerve or T1 root dysfunction.

Thumb movements Movements of the thumb and their nomenclature are unusual, comprising (as they do) the combined mobility of both the first carpometacarpal (CMC) and the first MCP joint (Figure 16.5).



(a) (b) (c) (d)

Figure 16.5 Thumb movements You should have no difficulty defining the planes of movement if you follow this routine: (a) hold the patient's hand flat on the table and instruct him or her to 'stretch to the side' (extension), (b) 'point to the ceiling' (abduction), (c) 'pinch my finger' (adduction) and (d) 'touch your little finger' (opposition).

With the hand lying flat, palm upwards, six types of movement are observed:

- *extension* (sideways movement in the plane of the palm)
- *abduction* (upward movement at right angles to the palm)
- *adduction* (pressing against the palm)
- *flexion* (sideways movement across the palm)
- *opposition* (touching the tips of the fingers)
- *retroposition* (lifting the thumbs backwards behind the plane of the hand).

Weakness of abduction (tested simply by pressing against the abducted thumb of each hand) is a cardinal feature of median nerve dysfunction. In advanced cases there will also be obvious wasting of the thenar eminence.

Pain, deformity and loss of motion at the base of the thumb (the first CMC joint) are common symptoms of osteoarthritis.

Testing the muscles and tendons

Flexion of the fingers is motivated mainly by *flexor digitorum profundus (FDP)* and *flexor digitorum superficialis (FDS)*; these muscles also assist in flexion of the MCP joints but the main MCP flexors are the *intrinsic muscles*. Active mass flexion can be tested by asking the patient to curl his or her fingers into flexion so as to engage them in the examiner's fingers in a tug of strength. However, the patient's flexors can also be tested independently, as follows (and see Figure 16.6).

To test for *flexor digitorum profundus* in an individual finger, the PIP joint is held and immobilized in extension and the patient is then asked to bend the tip of the finger.

To test *flexor digitorum superficialis*, the flexor profundus must first be inactivated, otherwise one cannot tell which tendon is flexing the PIP joint. This is done by grasping all the fingers, except the one being examined, and holding them firmly in full extension; because the profundus tendons share a common muscle belly, this manoeuvre automatically prevents *all* the profundus tendons from participating in finger flexion. The patient is then asked to flex the isolated finger which is being examined; this movement must be activated by flexor digitorum superficialis. There are two exceptions to this rule: first, the little finger sometimes has no independent flexor digitorum superficialis. Second, the index finger often has an entirely separate flexor profundus, which cannot be inactivated by the usual mass action manoeuvre; instead, flexor superficialis is tested by asking the patient to pinch hard with the DIP joint in full extension and the PIP joint in full flexion; this position can be maintained only if the superficialis tendon is active and intact.

Since the thumb has only a single IP joint, the *flexor pollicis longus* is tested by immobilizing the thumb MCP joint and then asking the patient to flex the IP joint.

The *long extensors* are tested by asking the patient to extend the MCP joints. Inability to do this usually signifies either paralysis or tendon rupture; occasionally, a long extensor tendon may simply have slipped off the knuckle into the interdigital gutter (a common occurrence in rheumatoid arthritis due to sagittal band rupture).

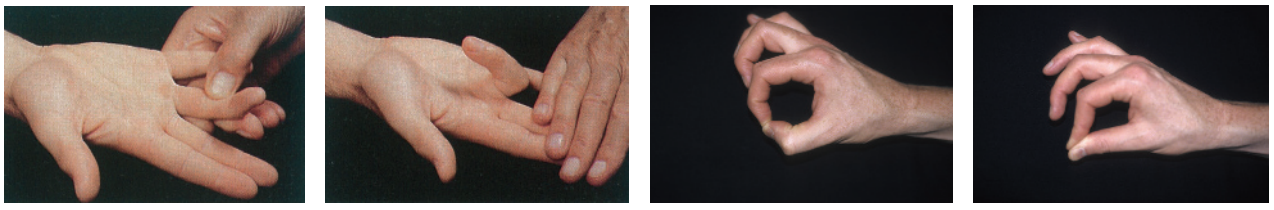
The *intrinsic muscles (lumbricals and interossei)* can act uniquely to flex the MCP joints with the IP joints held simultaneously in extension (i.e. preventing the long flexors from acting). Ask the patient to extend the fingers with the MCP joints flexed (the 'duckbill' position). The interossei also motivate finger abduction and adduction.

Grip strength

Grip strength is an important indicator of hand and wrist function. A painful wrist will result in a weak hand. Loss of finger function due to pain, stiffness, instability or weakness will also reduce grip. Grip strength should be measured with a mechanical dynamometer; if this is not available, an indication can be derived from having the patient squeeze a partially inflated sphygmomanometer cuff (normally a pressure of 150 mmHg can be achieved easily). Pinch grip also should be measured using a specific pinch gauge.

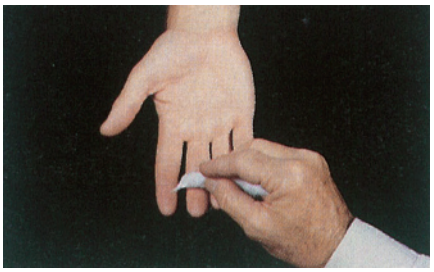
Neurological assessment

If symptoms such as numbness, tingling or weakness exist – and in all cases of trauma – a full neurological examination of the upper limbs should be carried out,

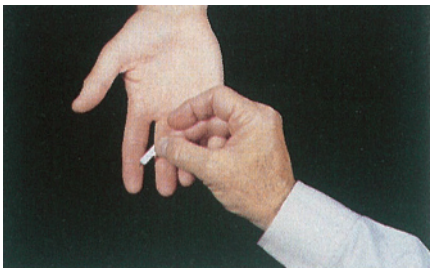


(a) (b) (c) (d)

Figure 16.6 Testing for (a) FDP lesser fingers, (b) FDS lesser fingers, (c) FDP index, (d) FDS index.



(a)



(b)

Figure 16.7 Neurological assessment (a) Light touch, (b) pinprick.

testing power, reflexes and sensation. Further refinement is achieved by testing monofilament detection, two-point discrimination, vibration sensibility, proprioception and stereognosis (tactile discrimination) (Figure 16.7).

Functional tests

Ultimately it is function that counts; patients learn to overcome their defects by ingenious modifications and trick movements. Function can be measured subjectively using patient-completed scales, but objective tests are more reliable. There are several types of grip, which can be tested by giving the patient a variety of tasks to perform: picking up a pin (precision grip), holding a sheet of paper (pinch), holding a key (sideways pinch), holding a pen (chuck grip), holding a bag handle (hook grip), holding a glass (span) and gripping a hammer handle (power grip). Stereognosis is evaluated using Moberg's pick-up test. The patient is instructed to pick up a number of small objects and place them in a box; the procedure is timed and efficiency of the affected hand is compared with that of the 'good' hand.

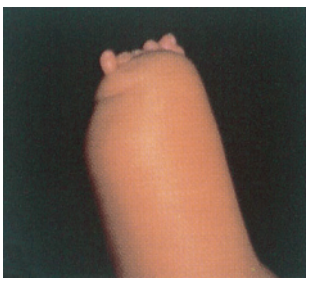
Each finger has its special task: the thumb and index finger are used for pinch. The index finger is also an important sensory organ; slight loss of movement matters little but, if sensation is abnormal, the patient probably will not use the finger at all. The middle finger controls the position of objects in the palm. The ring and little fingers are used for power grip; any loss of movement here will affect function markedly.

Stiffness is poorly tolerated in the little finger whereas instability is less worrisome; the opposite is true for the thumb and index finger.

Dexterity is lost in severe carpal tunnel syndrome (median nerve compression) because of the combination of thenar weakness, reduced sensation, diminished stereognosis and proprioception.

CONGENITAL HAND ANOMALIES

The incidence of congenital upper limb abnormalities is estimated to be about 1 in 600 live births (Figure 16.8). Some are confined to the hand but in most cases the wrist and forearm are involved as well. We have therefore covered congenital anomalies of the wrist and hand as a single subject in Chapter 15.



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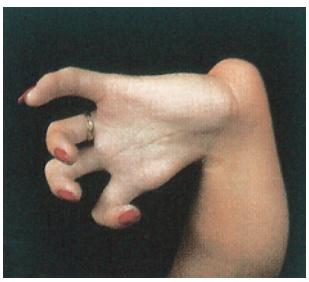

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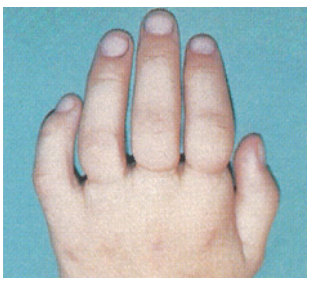

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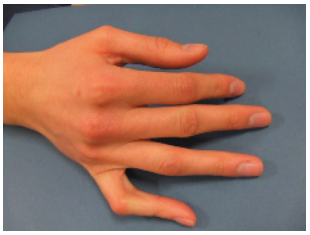

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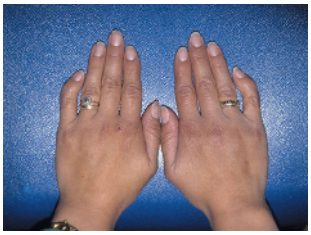


(e)

Figure 16.8 Congenital variations (a) Transverse failure, (b) radial club hand and absent thumb, (c) constriction rings, (d) camptodactyly, (e) clinodactyly.

ACQUIRED DEFORMITIES

Deformity of the hand may result from acquired disorders of the skin, subcutaneous tissues, muscles, tendons, joints, bones or neuromuscular function. Often there is a history of trauma or infection or concomitant disease; at other times the patient is unaware of any cause.

Problems arise for three main reasons: (1) the defect may be unacceptable simply because of its unsightly appearance; (2) function is impaired; and (3) the deformed part becomes a nuisance during daily activities.

Assessment and management of hand deformities demands a detailed knowledge of functional anatomy and, in particular, of the normal mechanisms of balanced movement in the wrist and fingers.

Skin contracture

Cuts and burns of the palmar skin are liable to heal with contracture. *Surgical incisions should never cross skin creases perpendicularly;* they should lie more or less parallel or oblique to them, or in the mid-axial line of the fingers. A useful alternative is a zigzag incision with the middle part of the Z in the skin crease. Longitudinal wounds can also be closed as Z-plasties.

Established contractures may require excision of the scar, Z-plasty of the remaining skin, skin grafts, a pedicled flap and occasionally a free flap.

Superficial palmar fascia (Dupuytren's) contracture

The superficial palmar fascia (palmar aponeurosis) fans out from the wrist towards the fingers, sending extensions across the MCP joints to the fingers. Hypertrophy and contracture of the palmar fascia may lead to puckering of the palmar skin and fixed flexion of the fingers. The condition is dealt with later in this chapter.

Muscle contracture

VOLKMANN'S ISCHAEMIC CONTRACTURE

Contracture of the forearm muscles may follow circulatory insufficiency due to injuries at or below the elbow. Shortening of the long flexors causes the fingers to be held in flexion; they can be straightened only when the wrist is flexed so as to relax the long flexors. Sometimes the picture is complicated by associated damage to the ulnar or median nerve (or both). If disability is marked, some improvement may be obtained by lengthening the shortened tendons, or by excising the fibrosed muscles and restoring finger movement with tendon transfers.

SHORTENING OF THE INTRINSIC MUSCLES

Shortening of the intrinsic muscles in the hand produces a characteristic deformity: flexion at the MCP joints with extension of the IP joints and adduction of the thumb (the so-called *'intrinsic-plus' hand*). Slight degrees of deformity may not be obvious, but can be diagnosed by *Bunnell's 'intrinsic-plus' test*: with the MCP joints pushed passively into hyperextension (thus putting the intrinsics on stretch), it is difficult or impossible to flex the IP joints passively; if the MCP joints are then placed in flexion, the IP joints can be passively flexed.

The causes of intrinsic shortening or contracture are: (1) spasticity (e.g. in cerebral palsy); (2) volar subluxation of the MCP joints (e.g. in rheumatoid arthritis); (3) scarring after trauma or infection; and (4) shrinkage due to ischaemia. Moderate contracture can be treated by resecting a triangular segment of the intrinsic 'aponeurosis' at the base of the proximal phalanx (*Littler's operation*).

Tendon lesions

MALLET FINGER

This results from injury to the extensor tendon of the terminal phalanx. It may be due to direct trauma but more often painlessly follows an innocent event when the fingertip is forcibly bent during active extension, perhaps while tucking the blankets under a mattress or trying to catch a ball. The terminal joint is held flexed and the patient cannot straighten it, but passive movement is normal. With the extensor mechanism unbalanced, the PIP joint may become hyperextended ('swan-neck').

X-rays are taken to show or exclude a fracture. If there is a fracture but minimal subluxation of the joint, it is treated by splintage with the DIP joint in extension for 6 weeks. Operative treatment is considered only if there is a large fragment (>50%) and subluxation of the DIP joint. Otherwise surgery is ill-advised, as the complication rate is high and it is unlikely to improve the outcome.

A mallet finger without bone injury is treated with a plastic splint with the DIP joint in extension for 8 weeks, followed by 4 weeks of night splintage. This treatment may still work if presentation is delayed for a few weeks. The great majority do very well. Old lesions need treatment only if the deformity is marked and hand function seriously impaired. The options include fusion for painful arthritic joints or tendon reconstruction. If a flexible swan-neck deformity accompanies the mallet finger, a simple central slip tenotomy is usually effective.

RUPTURED EXTENSOR POLLICIS LONGUS

The long thumb extensor may rupture after fraying or ischaemia where it crosses the wrist (e.g. after a Colles' fracture, or in rheumatoid arthritis). The distal phalanx drops into flexion; it can be passively extended, and there may still be weak active extension because of thenar muscle insertion into the extensor expansion; however, the thumb cannot be actively elevated backwards above the plane of the hand (retroposition). Direct repair is unsatisfactory and a tendon transfer, using the extensor indicis, is needed. The results are satisfactory in over 90% of cases.

DROPPED FINGER

Sudden loss of finger extension at the MCP joint is usually due to tendon rupture at the wrist (e.g. in rheumatoid arthritis). Because direct repair is not usually possible, the distal portion can be attached to an adjacent finger extensor or a tendon transfer performed.

Occasionally the deformity is due to catching of the collateral ligament on a metacarpal osteophyte or rupture of the sagittal band which centralizes the tendon over the back of the knuckle.

BOUTONNIÈRE DEFORMITY

This lesion presents as a flexion deformity of the PIP joint and extension of the DIP joint. It is due to interruption or stretching of the central slip of the extensor tendon where it inserts into the base of the middle phalanx. The lateral slips separate and the head of the proximal phalanx thrusts through the gap like a button through a buttonhole. Ironically, while English speakers call it a '*boutonniere*' deformity, the French refer to it as '*le buttonhole*'.

The usual causes are direct trauma or rheumatoid disease. Initially the deformity is slight and passively correctable; later the soft tissues contract, resulting in fixed flexion of the proximal and hyperextension of the DIP joint (Figure 16.9). Early diagnosis is therefore important; an impending deformity should be suspected in anyone with tenderness or a cut over the dorsum of the PIP joint, especially if they cannot actively extend the IP joint with the MCP joints and wrist flexed.

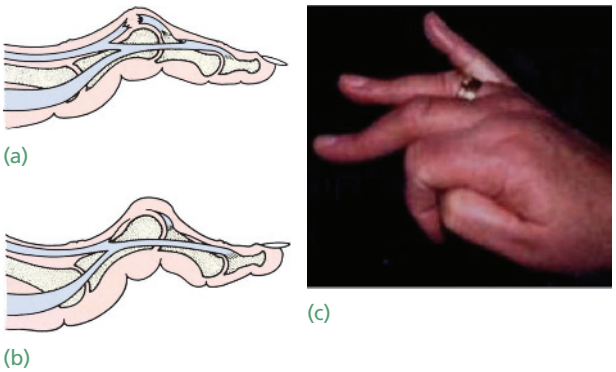


Figure 16.9 Boutonnière deformity (a) When the middle slip of the extensor tendon first ruptures there is no more than an inability to extend the PIP joint. (b) Gradually the lateral slips slide volarwards, the knuckle pops through the 'buttonhole' and the DIP joint is pulled into hyperextension. (c) Clinical appearance.

In the early post-traumatic case, splinting the PIP joint in full extension for 6 weeks usually leads to healing; the DIP joint must be moved passively to prevent the lateral bands from sticking. Open injuries of the central slip should be repaired, with the joint protected by a K-wire for 3 weeks.

For later cases where the joint is still passively correctible, several operations have been invented (suggesting that none is too reliable). The easiest and probably most successful procedure is to divide the extensor tendon just proximal to its insertion into the distal phalanx. This allows the extensor mechanism to move proximally, thus enhancing PIP extension and diminishing DIP extension.

Long-standing fixed deformities are extremely difficult to correct and may be better left alone.

SWAN-NECK DEFORMITY

This is the reverse of the boutonnière deformity; the PIP joint is hyperextended and the DIP joint flexed. The deformity can be reproduced voluntarily by lax-jointed individuals. The clinical disorder has many causes, with two things in common: imbalance of extensor versus flexor action at the PIP joint and laxity of the palmar plate. Thus it may occur: (1) if the PIP extensors overact (e.g. due to intrinsic muscle spasm or contracture, after mallet finger, or following volar subluxation of the MCP joint); (2) if the PIP flexors are inadequate (inhibition or division of the flexor superficialis); or (3) if the palmar plate of the PIP joint fails (in rheumatoid arthritis, lax-jointed individuals or trauma). If the deformity is allowed to persist, secondary contracture of the intrinsic muscles, and eventually of the PIP joint itself, makes correction increasingly difficult and ultimately impossible.

Treatment depends on the cause and whether or not the deformity has become fixed. If the deformity corrects passively, then a simple figure-of-eight ring splint to maintain the PIP joint in a few degrees of flexion may be all that is required; if this works but cannot be tolerated, then tenodesis of the PIP joint works well. The options are either to attach one slip of flexor digitorum superficialis to the proximal phalanx, which prevents hyperextension, or to reroute a lateral band anteriorly so it becomes a flexor rather than an extensor of the PIP joint. If the intrinsics are tight, they are released.

If the deformity is fixed, it may respond to gentle manipulation supplemented by temporary K-wire fixation in a few degrees of flexion; if not, lateral band release from the central slip may be needed. The dorsal

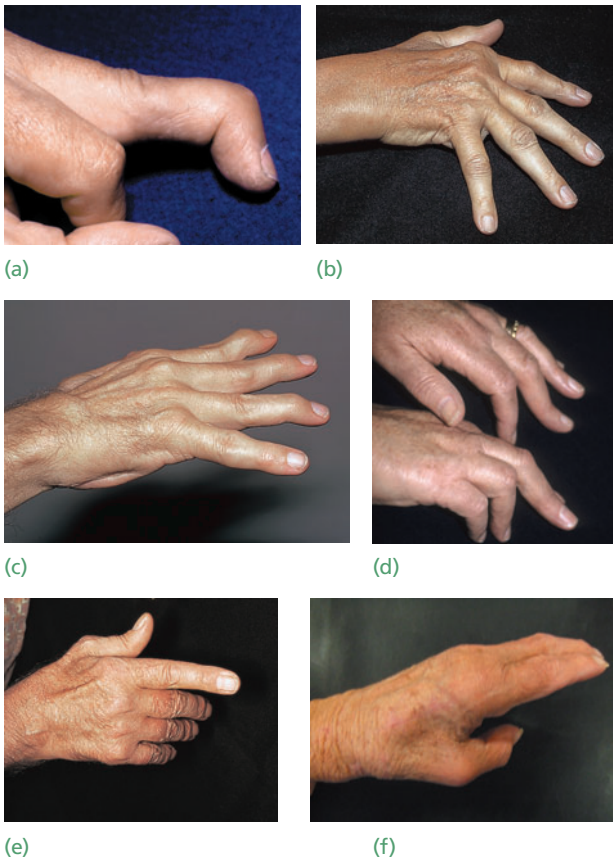


Figure 16.10 Deformities due to tendon lesions (a) Mallet finger. (b) Dropped fingers due to extensor tendon ruptures at the wrist. (c) Swan-neck deformities. (d) Boutonnière deformities. (e) Rupture of extensor pollicis brevis. (f) Rupture of extensor pollicis longus.

skin may not close directly after correction. If the swan-neck deformity is secondary to a mallet finger, the latter should be addressed as described above.

If function is severely impaired and does not respond to one of the above measures, the joint is arthrodesed in a more acceptable position.

Joint disorders

RHEUMATOID ARTHRITIS

Rheumatoid arthritis causes multiple, symmetrical deformities of both hands, typically ulnar deviation of the MCP joints and boutonnière or swan-neck deformities of the proximal finger joints.

JUVENILE IDIOPATHIC ARTHRITIS

The pattern of involvement is different from that of adult disease. The wrists tend to develop ulnar (rather than radial) deviation, the MCP joints develop flexion contractures (rather than ulnar drift), and the IP joints also become fixed in flexion (swan-neck deformities are rare). The hands are small because of premature fusion of the physis.

The mainstay of treatment is medical. Long-term splintage of the hand is helpful and synovectomy is sometimes needed. Later, wrist fusion, MCP joint replacement and IP joint fusion also have a role, usually after skeletal maturity.

PSORIATIC ARTHRITIS

Erosive arthropathy of the IP joints leads to profound weakness and instability; the PIP joints may develop fixed flexion deformities. If the disease progresses, psoriatic arthritis can devastate the small joints of the hand ('arthritis mutilans') resulting in severe, and sometimes bizarre, deformities of the IP and MCP joints. The nails are often pitted (onychodystrophy) and skin lesions (a guttate or pustular rash) may be evident. Occasionally joint fusion is needed to relieve pain and to provide stability in a functional position.

SYSTEMIC LUPUS ERYTHEMATOSUS

This autoimmune disease, affecting women five times more frequently than men, causes soft-tissue slackening with extensor tendon dislocation, ulnar deviation at the MCP joints and swan-neck deformities of the fingers. Soft-tissue corrections tend to fail with time and eventually fusions may be needed to maintain function.

SCLERODERMA

Typically the fingers are smooth-skinned and stiff (sclerodactyly), with flexion deformities of the IP joints. Raynaud's phenomenon and painful ulcers may develop. Early on, physiotherapy and splinting help; in the later stages, joint fusion in a functional position and digital sympathectomy to relieve ulcers may be needed. Painful calcific deposits can be excised but wound breakdown is a risk.

OSTEOARTHRITIS

Osteoarthritis, by contrast, affects mainly the DIP joints. It is common in postmenopausal women and may cause deformity. The thumb CMC joint is another common site, and this may result in adduction of the first metacarpal and flexion of the first CMC joint. Treatment is discussed in Chapter 15.

GOUT

Gouty swellings (tophi) and finger deformities are sometimes mistaken for rheumatoid disease. However, the lesions tend to be asymmetrical and the X-ray appearances are distinctive. The diagnosis can be confirmed by identifying urate crystals in the tophaceous material. Curiously, gout and rheumatoid arthritis hardly ever occur in the same patient. In addition to systemic treatment, evacuation of a tophus (or tophi) is sometimes advisable.

TRAUMA

Fractures may go on to malunion and joints may become stiff and swollen. This subject is dealt with in Chapter 27.

Bone lesions

A variety of bone lesions (acute infection, tuberculosis, malunited fractures, infantile rickets, tumours) may cause metacarpal or phalangeal deformity. X-rays usually show the abnormality. In addition to treating the pathological lesion, deformity may need correction by osteotomy with internal fixation.

Neuromuscular disorders

SPASTIC PARESIS

Cerebral palsy, head injury and stroke may result in typical deformities of the hand (Figure 16.11). The 'intrinsic-plus' posture is easily recognized. Another common disability is 'thumb-in-palm'; the tendency to adduct and flex the thumb into the palm is increased by activity, especially finger flexion. Releasing the adductor pollicis from the third metacarpal may improve the appearance, but normal thumb pinch is rarely restored.

OTHER NEUROLOGICAL DISORDERS

Poliomyelitis, leprosy, syringomyelia and Charcot–Marie–Tooth disease may cause hand deformities. If there is only partial involvement, tendon transfer may be feasible.

PERIPHERAL NERVE LESIONS

The postural deformities are so characteristic that the diagnosis should seldom be in doubt (see Chapter 11). The most common are drop wrist and drop fingers (radial nerve palsy), a simian thumb and pointing index finger (median nerve palsy), and partial claw hand (ulnar nerve palsy). The distribution of sensory loss helps to establish the site of the lesion.

THE 'INTRINSIC MINUS' HAND

Among the late neurological defects, *intrinsic paralysis* is particularly disabling. The 'intrinsic minus' hand shows wasting of the small muscles and moderate clawing, with extension of the MCP and partial flexion of the IP joints. If all the intrinsics are affected (e.g. after poliomyelitis or a combined low median and ulnar nerve injury), the thumb lies flat at the side of the hand and cannot be opposed. In ulnar nerve palsy only the ring and little fingers are clawed, because the index and middle lumbricals are supplied by the median nerve; these muscles continue to flex the MCP joints and extend the IP joints. Thumb opposition is retained but thumb pinch is unstable because index-finger abduction (first dorsal interosseous) and thumb adduction (adductor pollicis) are weak; loss of thumb adduction is compensated for by exaggerated IP flexion during strong pinch (*Froment's sign*).

The objectives of *treatment* are:

1. *stabilization of the MCP joints in flexion* – this can be achieved dynamically by a tendon transfer (e.g. flexor superficialis into the intrinsic tendon) or statically by looping a slip of flexor digitorum superficialis around the flexor pulley (*Zancolli's operation*)
2. *restoration of index abduction* – to provide stable pinch (e.g. by abductor pollicis longus transfer to first dorsal interosseous and ECRB to adductor pollicis, both with a tendon graft
3. *restoration of thumb opposition* (if it is lost) – by a tendon transfer looped around a fascial or tendon pulley and attached to the radial edge of the proximal phalanx of the thumb.

Before any of these operations, stiff finger joints must be made mobile.

DUPUYTREN'S CONTRACTURE

This is a nodular hypertrophy and contracture of the superficial palmar fascia (palmar aponeurosis). The condition has a complex inheritance with several gene loci involved. It is most common in caucasians of Northern European descent. It is more common in males than females; the prevalence increases with age, but onset at an early stage usually means

(a) (b) (c) (d)

Figure 16.11 Spastic contracture – hand deformities (a,b,c) cerebral palsy, and (d) head injury with brain damage.

aggressive disease. There is a high incidence in epileptics receiving phenytoin therapy; associations with diabetes, epilepsy, smoking, alcoholic cirrhosis, AIDS and pulmonary tuberculosis have also been described. There is a contentious and weak association with injury and vibration to the wrist and hand.

Pathology

The essential problem in Dupuytren's disease is a genetically determined proliferation of myofibroblasts. After an initial proliferative phase, fibrous tissue within the palmar fascia and fascial bands within the fingers contracts, causing flexion deformities of the MCP and PIP joints (Figure 16.12). Fibrous attachments to the skin lead to puckering. The digital nerve is displaced or enveloped, but not invaded, by fibrous tissue. Occasionally the fascia of the foot or penis (Ledderhose's disease (Figure 16.13b) and Peyronie's disease respectively) is affected.

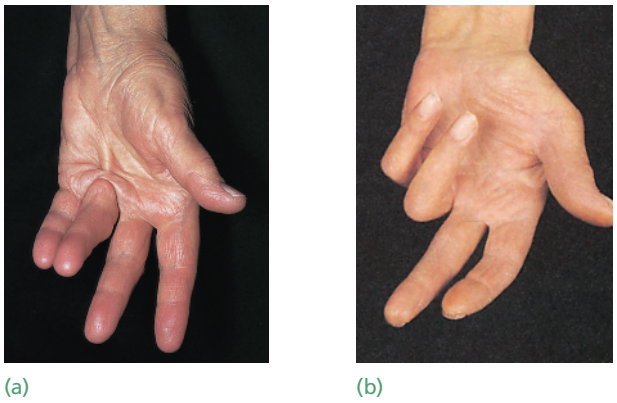


Figure 16.12 Dupuytren's disease Contractures may occur at (a) palmar crease, (b) proximal interphalangeal joint, (c) thumb web, (d) little finger.

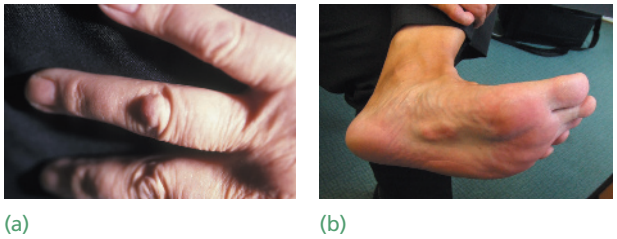


Figure 16.13 Dupuytren's disease – other manifestations (a) Garrod's pads, (b) Ledderhose's nodules.

Clinical features

The patient complains of a nodular thickening in the palm or finger. The palm of the ring finger and the PIP of the little finger are the most commonly involved. Pain may occur early on but is seldom a marked feature. Often both hands are involved, one more than the other. Gradually the nodules may thicken and contract, forming cords. If the subcutaneous cords extend into the fingers, they may produce flexion deformities at the MCP and PIP joints. Sometimes the dorsal knuckle pads are thickened (Garrod's pads – Figure 16.13a). About 60% of patients give a family history.

Diagnosis

Dupuytren's contracture must be distinguished from skin contracture (where the previous laceration is usually obvious), tendon contracture (in which the finger deformity changes with wrist position) and PIP joint contracture (in which there may be a history of clinodactyly or joint injury).

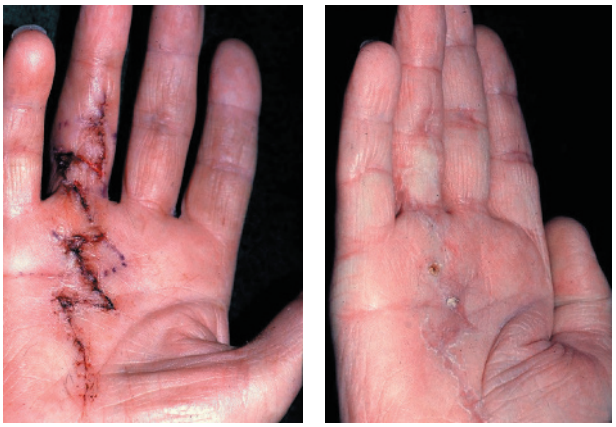
Treatment

Treatment is indicated if the deformity is a nuisance or rapidly progressing. Once a 30 degree angle is reached at the PIP or MCP, the patient usually cannot get the hand flat on the table (the '*table-top test*') and starts to be aware of functional problems. Most treatments are effective at this earlier stage but, in particular, PIP joint contractures can become irreversible if left too long. The aim is reasonable, but not always complete, correction.

Radiotherapy This is an option for a very small group with painful widespread proliferative disease. Dry skin is a common side effect.

Percutaneous needle fasciotomy An isolated discrete cord across the front of the MCP joint can be divided by scratching several times through the skin with a bevelled needle. This can be very gratifying for selected cords with a very quick recovery. However, no fibrous matrix is removed and the altered cells remain; recurrence is very high.

Collagenase clostridium histiolyticum (Xiapex, Xiaflex) This is a 'surgical drug' which is injected into the cord and dissolves it (Figure 16.14c). Since it does not damage the nerves or blood vessels, it is safer than surgery; however, the palm can be painful for a few days, and the skin may blister or split (although recovers promptly). If injected too deep, the flexor tendon can rupture, so training and skill are required. The drug allows very prompt recovery – a few days rather than a few weeks with surgery. Recurrence is an issue, again because the cells are not removed.



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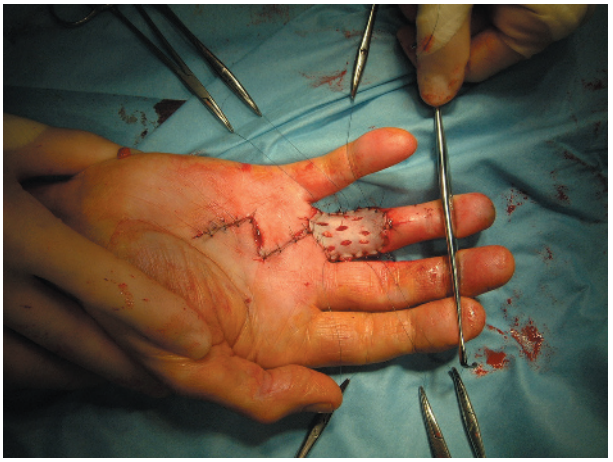

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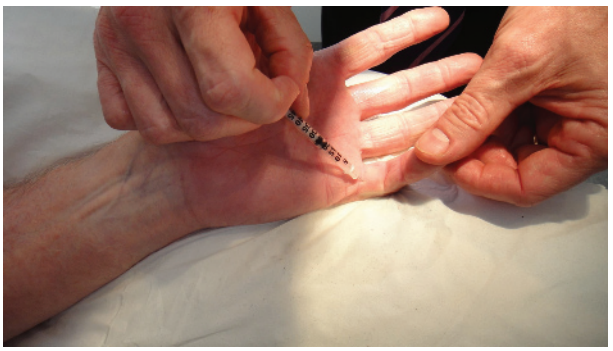


(c)

Figure 16.14 Dupuytren's disease – treatment
(a) Z-plasty in the hand shortly after operation and 2 weeks later when healing is almost complete; (b) skin graft in theatre; (c) collagenase injection.

Surgical fasciectomy If the disease is more extensive, the affected area is approached through a longitudinal or a Z-shaped incision and, after carefully freeing the nerves and blood vessels, the cords are excised. Skin closure may be facilitated by multiple Z-plasties (Figure 16.14a). This provides more skin to cover the increased length of the corrected finger. The wound in the palm can be left open and will soon heal with dressings. This makes skin closure easier and allows any haematoma (which may predispose to stiffness) to escape. Postoperative splints are not routinely required.

Skin grafting If there is severe skin involvement (particularly in surgery for recurrent disease), if there is a strong family history, or if the patient is particularly young, then skin grafting should be considered (Figure 16.14b). The operation is fairly straightforward; the advantage is that recurrence is very rare (because all the culpable cells as well as their fibrous matrix are excised).

Salvage Joint fusion or amputation is occasionally advisable for severe, recurrent disease in the little finger.

TRIGGER FINGER (DIGITAL TENOVAGINOSIS)

A flexor tendon may become trapped by thickening at the entrance to its sheath; on forced extension it passes the constriction with a snap ('triggering'). A secondary nodule can develop on the tendon. The underlying cause is unknown but the condition is certainly more common in patients with diabetes. People with rheumatoid disease may develop synovial thickening or intratendinous nodules which can also cause triggering. Occupational factors, though sometimes blamed, are unlikely to be causative.

Clinical features

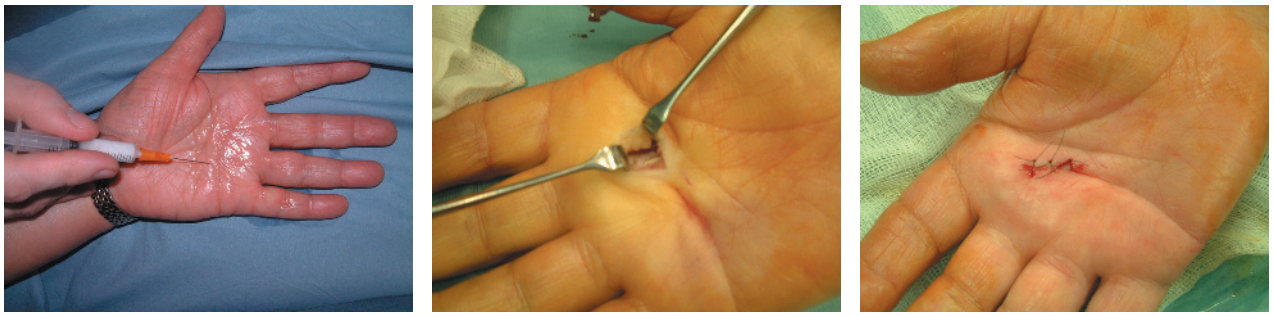
Any digit may be affected, but the thumb, ring and middle fingers most commonly; sometimes several fingers are affected. The patient notices a click as the finger is flexed; when the hand is unclenched, the affected finger initially remains bent at the PIP joint but with further effort it suddenly straightens with a snap. A tender nodule can be felt in front of the MCP joint and the click may be reproduced at this site by alternately flexing and extending the finger.

INFANTILE TRIGGER THUMB

Parents sometimes notice that their baby or infant cannot extend the thumb tip. The diagnosis is often missed, or the condition is wrongly taken for a 'dislocation'. Very occasionally the child grows up with the thumb permanently bent. This condition must be distinguished from the rare *congenitally clasped thumb* in which both the IP joint and the MCP joint are flexed because of congenital insufficiency of the extensor mechanism (see Chapter 15).

Treatment

In adults, early cases may be cured by an injection of corticosteroid carefully placed at the mouth of the tendon sheath. Recurrent triggering up to 6 months later occurs



(a) (b) (c)

Figure 16.15 Trigger finger (a) Injection of steroid; (b,c) operative treatment.

in over 30% of patients – particularly younger patients and those with diabetes, who may then need a second injection. Refractory cases need operation, through an incision over the distal palmar crease, or in the MCP crease of the thumb – the A1 section of the fibrous sheath is incised until the tendon moves freely (Figure 16.15).

In babies it is worth waiting until the child is about 3 years old, as spontaneous recovery often occurs. If not, then the pulley is released.

Care should be taken to avoid injury to the digital neurovascular bundles during surgery. The risk is greatest in the thumb (where the nerves are close to the midline) and the index finger (where the radial digital nerve crosses the tendon).

In patients with rheumatoid arthritis, the fibrous pulley must be carefully preserved; damage to this structure will predispose to ulnar deviation of the fingers. Flexor synovectomy with excision of one slip of flexor digitorum superficialis is preferred.

RHEUMATOID ARTHRITIS
(see also Chapter 3)

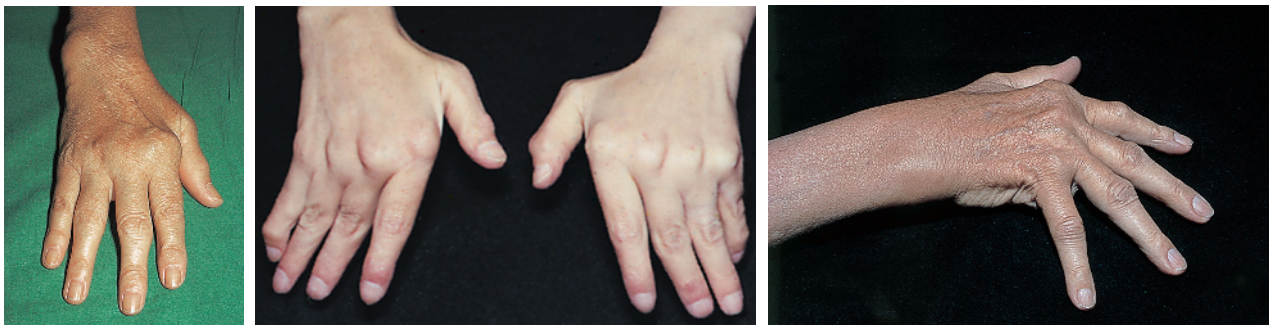
The hand, more than any other region, is where rheumatoid arthritis carves its story. The early stage is characterized by synovitis of the joints and tendon sheaths. If the disease progresses, joint and tendon erosions prepare the ground for mechanical derangement. In the late stage, joint destruction, attenuation of the ligaments and tendon ruptures lead to instability and progressive deformity.

With the advent of biological treatment such as anti-TNF agents, the need for surgical treatment has diminished considerably.

Clinical features

Stiffness and swelling of the fingers are early symptoms; the patient may mention that the wrist also is swollen (Figure 16.16). Sometimes the first symptoms are typical of carpal tunnel compression, caused by flexor tenosynovitis at the wrist.

Examination may reveal swelling of the MCP and PIP joints, giving the fingers a spindle shape; both hands are affected, more or less symmetrically. Swelling of tendon sheaths is usually seen on the dorsum of the wrist and along the ulnar border (extensor carpi ulnaris); thickened flexor tendons may also be felt on the volar aspect of the proximal phalanges. The joints are tender and crepitus may be felt on moving the tendons. Joint mobility and grip strength are diminished.



(a) (b) (c)

Figure 16.16 Rheumatoid arthritis – clinical features (a) Early case with typical features: radial deviation of the wrist; subluxation of the radioulnar joint; swollen MCP joints and ulnar deviation of the fingers. (b) More advanced changes, including subluxation of the MCP joints. (c) Dropped fingers due to rupture of extensor tendons at the wrist.

As the disease progresses, early deformities make their appearance: slight radial deviation of the wrist and ulnar deviation of the fingers, correctable swan-neck deformities of some fingers, an isolated boutonnière or the sudden appearance of a drop finger or mallet thumb (from extensor tendon rupture).

In the late stage, long after inflammation may have subsided, established deformities are the rule: the carpus settles into radial tilt and volar subluxation; there is marked ulnar drift of the fingers and volar dislocation of the MCP joints, often associated with multiple swan-neck and boutonnière deformities. These 'rheumatoid deformities' are so characteristic that they allow the diagnosis to be made at first glance. When the abnormalities become fixed, functional loss may be so severe that patients can no longer dress or feed themselves.

General features

The hand should not be considered in isolation. Its functional interaction with the wrist and elbow is crucial and, in a generalized disorder such as rheumatoid disease, the condition of all the upper limb joints and the cervical spine should be carefully assessed.

Weakness Rheumatoid hands are weak because of a combination of generalized muscular weakness, pain inhibition, tendon malalignment or rupture, joint stiffness and nerve compression.

Rheumatoid nodules These are associated with aggressive disease in seropositive patients. They tend to occur at pressure areas (e.g. the pulps of the fingers and the radial side of the index finger).

Z-collapse If one of two adjacent joints changes direction, then the overlying long tendons will pull the other joint into the opposite direction. In rheumatoid arthritis, this is typified by radial tilt of the wrist with ulnar drift of the MCP joints, the boutonnière deformity and the swan-neck deformity.

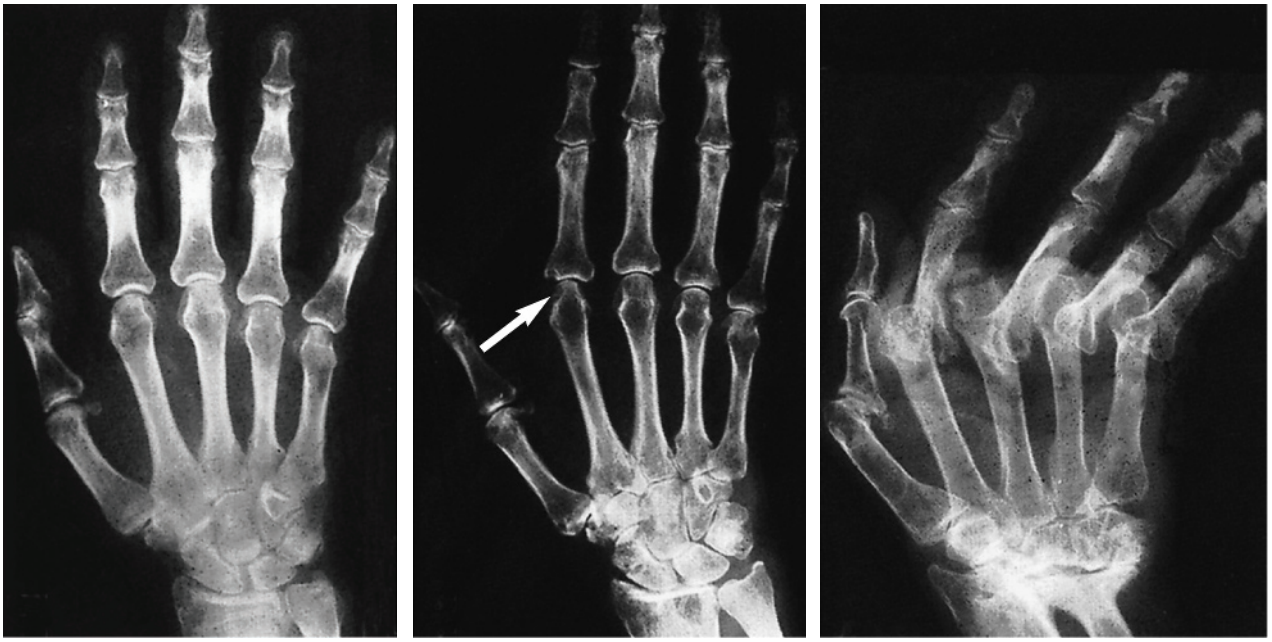
X-rays

During the early stage X-rays show only soft-tissue swelling and osteoporosis around the joints. Later, one can usually discern joint 'space' narrowing and small peri-articular erosions; these are commonest at the MCP joints and in the styloid process of the ulna. In advanced cases, articular destruction may be marked, affecting the MCP, PIP and wrist joints almost equally. Joint deformity and dislocation are common (Figure 16.17).

Treatment

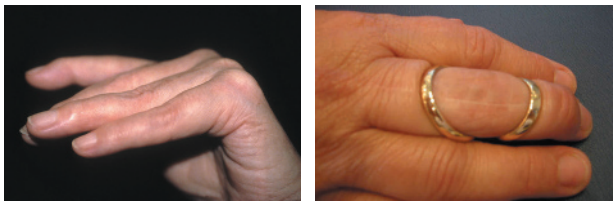
EARLY STAGE DISEASE

Treatment is directed essentially at controlling the systemic disease and the local synovitis. In addition to general measures, static splints may reduce pain and swelling (Figure 16.18). These splints are not corrective but are designed to rest inflamed joints and tendons; in



(a) (b) (c)

Figure 16.17 Rheumatoid arthritis – X-ray changes (a) Early on, the X-rays may show no more than soft-tissue swelling and juxta-articular osteoporosis. (b) A later stage showing characteristic punched-out juxta-articular erosions at the second and third metacarpophalangeal joints. The wrist is now also involved. (c) In the most advanced stage, the metacarpophalangeal joints are dislocated and the hand is severely deformed.



(a) (b)

Figure 16.18 Rheumatoid arthritis – treatment
(a) Swan-neck deformity; (b) swan-neck 'figure of eight' splint.

mild cases they are worn only at night, in more active cases during the day as well. Persistent synovitis of a few joints or tendon sheaths may benefit from local injections of corticosteroid with local anaesthetic. Only small quantities are injected (e.g. 0.5 mL for an MCP joint or flexor tendon sheath and 1 mL for the wrist). This should not be repeated more than two or three times. A boggy flexor tenosynovitis may not respond to this limited therapeutic assault; operative synovectomy may be needed. If carpal tunnel symptoms are present, the transverse carpal ligament is divided and, if necessary, a flexor synovectomy performed.

ESTABLISHED DISEASE

As the disease progresses it becomes increasingly important to prevent deformity. Uncontrolled synovitis of joints or tendons requires operative synovectomy followed by physiotherapy. Excision of the distal end of the ulna, synovectomy of the common extensor sheath and the wrist, and reconstruction of the soft tissues on the ulnar side of the wrist may arrest joint destruction and progressive deformity.

Early instability and ulnar drift at the MCP joints can be corrected by excising the inflamed synovium, tightening the capsular structures and releasing the ulnar pull of the intrinsic tendons. Mobile boutonnière and swan-neck deformities can be treated with splints; if they progress or are fixed, surgery may be needed. Isolated tendon ruptures are repaired or bypassed by appropriate tendon transfers. These procedures are followed by splintage and hand therapy.

Destruction of the MCP joints without ulnar drift can be treated with surface replacement (cobalt-chrome on polyethylene or pyrocarbon).

LATE DISEASE

In late cases deformity is combined with articular destruction; soft-tissue correction alone will not suffice. For the MCP and IP joints of the thumb, arthrodesis gives predictable pain relief, stability and functional improvement. The MCP joints of the fingers can be excised and replaced with Silastic 'spacers', which improve stability and correct deformity. Replacement of IP joints gives less predictable results; if deformity is very disabling (e.g. a fixed swan-neck), it may be better to settle for arthrodesis in a more functional position. At the wrist, painless stability can be regained by fusion of the radiocarpal, mid-carpal and CMC joints. Wrist replacement with metal–plastic implants, while providing some movement, may well fail; the loss of bone stock that accompanies failure means that salvage can be very difficult.

The thumb in rheumatoid arthritis

The combination of soft-tissue failure and joint erosion leads to characteristic deformities of the thumb: rupture of flexor pollicis longus tendon, a boutonnière lesion at the MCP joint, CMC instability, swan-neck deformity and ulnar collateral ligament instability.

Depending on the deformity, the patient's demands and the condition of the rest of the hand, treatment may involve various combinations of splintage, tendon repair, joint fusion, excision arthroplasty and joint replacement.

Treatment options are summarized in Box 16.1.

Metacarpophalangeal deformities

Chronic synovitis of the MCP joints results in failure of the palmar plate and the collateral ligaments. The powerful flexor tendons drag the proximal phalanx palmarwards, causing subluxation of the joint. The deformity may be aggravated by primary or secondary intrinsic muscle tightness.

The most obvious deformity of the rheumatoid hand is ulnar deviation of the MCP joints. There are several reasons for this: palmar grip and thumb pressure naturally tend to push the index finger ulnarwards; weakening of the collateral ligaments and the first dorsal interosseous muscle reduces the normal resistance to this force; the wrist is usually involved and, as it collapses into radial deviation, the MCP joints automatically veer in the opposite direction (the so-called 'zigzag mechanism'); once ulnar drift begins, it becomes self-perpetuating due to tightening of the ulnar intrinsic muscles and stretching of the radial intrinsics and the adjacent capsular structures. As the sagittal bands fail, the extensor tendon slips ulnarwards and palmarwards, accentuating the deformity even further.

At an early stage, before joint destruction and soft-tissue instability, synovectomy may relieve pain (Figure 16.19) but the joint usually stiffens somewhat. When ulnar drift has started, splintage may maintain function and retard progression. With marked deformity but little joint damage, a soft-tissue reconstruction (reefing of the radial sagittal bands, tightening of the radial collateral ligament with intrinsic muscle release and transfer) can give a satisfactory and fairly durable correction.

Once there is marked damage to the joint surface, replacement with a Silastic spacer, along with the soft-tissue reconstruction, is recommended (Figure 16.20). There is no point in correcting the MCP joints unless

BOX 16.1 MANAGEMENT OF THUMB DEFORMITIES IN RHEUMATOID ARTHRITIS

Ruptured FPL

- If painless: leave alone
- If painful: tendon graft, flexor digitorum sublimus transfer or IP fusion

Simple boutonnière deformity

- If passively correctible: cortisone injection to MCP joint and splintage
- MCP joint synovectomy and extensor realignment unreliable
- If MCP joint fixed but IP joint passively correctible and CMC joint mobile: fuse MCP joint
- If MCP joint and IP joint fixed: fuse IP joint and either fuse or replace MCP joint

Boutonnière with CMC joint failure

- Trapeziectomy and CMC joint stabilization, with MCP joint and IP joint treated as above

Arthritis mutilans

- Arthrodesis with interposition bone graft

Swan-neck deformity

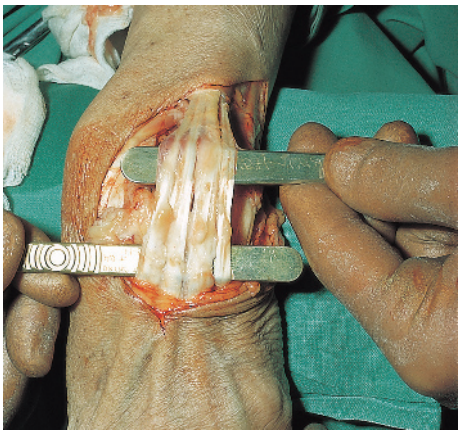
- CMC joint failure causes adduction contracture of thumb base and MCP joint hyperextension
- If deformity severe: trapeziectomy with soft-tissue reconstruction or fusion of MCP joint

Failure of ulnar collateral ligament (like 'gamekeeper's thumb')

- Synovitis attenuates ulnar collateral ligament. Pinch grip causes increasing deformity
- Ligament reconstruction (if bone and soft-tissue quality allow) or MCP joint fusion

Swan-neck with MCP joint and CMC joint preserved

- Synovitis of MCP joint causes hyperextension with secondary passive flexion of IP joint
- Treat by palmar plate advancement, or if soft tissues tenuous, MCP fusion



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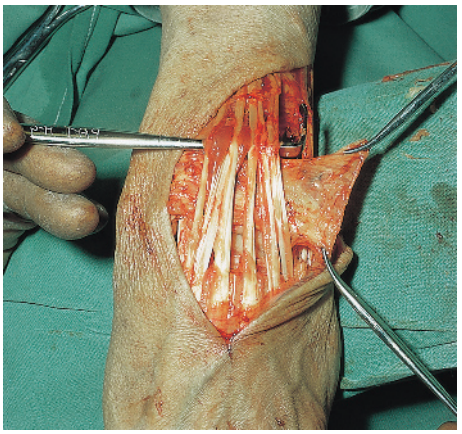
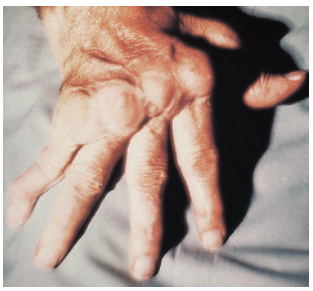


(b)

Figure 16.19 Rheumatoid arthritis – synovectomy Synovitis of the common extensor sheath will eventually damage the tendons. (a) Here, after synovectomy, one can see nodules on several tendons. (b) The sheath itself is preserved intact and laid beneath the tendons to cover the back of the joint and provide a bed upon which the tendons can move.



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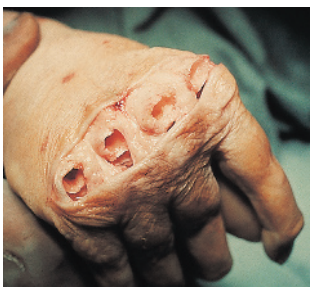

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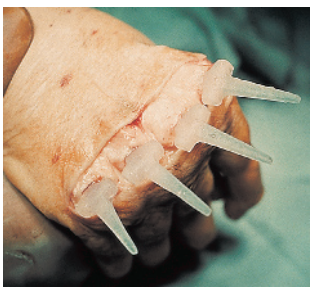

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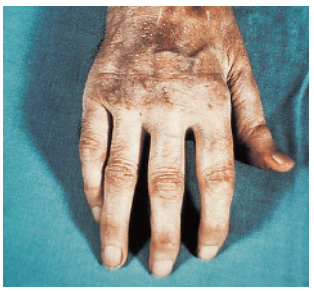


(d)

Figure 16.20 Rheumatoid arthritis – joint replacement (a) Before operation there is subluxation and deformity of all the finger MCP joints. (b,c) The eroded metacarpal heads are excised and flexible spacers inserted. (d) Postoperative result.

any wrist deformity is also corrected; the tendency to zigzag deformity will otherwise lead to recurrence of the ulnar drift.

Finger deformities

Boutonnière Synovitis in the proximal IP joint causes elongation or rupture of the central slip which passes over the back of the joint before inserting into the base of the middle phalanx. The lateral bands slip away from the central slip and pass in front of the axis of rotation of the proximal joint but remain behind the axis in the distal joint, to form the characteristic deformity.

Early, correctable deformity responds to splinting and synovectomy; later, central slip reconstruction (an unpredictable procedure) may be required; simple division of the distal insertion is a simpler, and often effective, alternative. In fixed deformities, or those with joint damage, fusion or replacement is considered.

Swan-neck Chronic synovitis may lead to swan-neck deformity by one or more of the following mechanisms: failure of the palmar plate of the PIP joint; rupture of the flexor digitorum superficialis; dislocation or subluxation of the MCP joint and consequent tightening of the intrinsic muscles.

Treatment depends on a careful analysis of the cause and will include figure-of-eight splintage, tendon transfer, intrinsic release and occasionally fusion (see Table 16.1).

Tenosynovitis and tendon rupture

Extensor tendons Extensor tendon rupture is a common complication of chronic synovitis. Extensor digiti minimi is usually the first to go and predicts rupture of the other tendons. Treatment consists of suturing the distal tendon stump to an adjacent tendon, inserting a bridge graft (e.g. palmaris longus) or performing a tendon transfer (e.g. extensor indicis proprius). Synovectomy and excision of the distal ulna may also be necessary.

Table 16.1 Types of swan-neck deformity in rheumatoid arthritis

Type	Description	Treatment
Type I	PIP joint flexible, independent of MCP position (i.e. Bunnell's test negative) Due to palmar plate failure at PIP joint ± failure of flexor digitorum superficialis	Palmar plate tenodesis or lateral band transfer
Type II	PIP joint flexibility dependent on MCP position Intrinsic muscle tightness Bunnell's test: with MCP joint passively	Release intrinsic muscles
Type III	PIP joint stiff regardless of MCP position Due to contracture of joint	Manipulation; release lateral band from central slip
Type IV	Destruction of PIP joint	Fusion

Flexor tendons Flexor tenosynovitis is one of the earliest and most troublesome features of rheumatoid disease. The restriction of finger movement is easily mistaken for arthritis; however, careful palpation of the palm and the nearby joints will quickly show where the swelling and tenderness are located. Secondary problems include carpal tunnel syndrome, triggering of one or more fingers and tendon rupture. Synovitis of the flexor digitorum superficialis also contributes to the swan-neck deformity.

If carpal tunnel release is needed, the operation should include a flexor tenosynovectomy. If the flexor tendons are bulky (best felt over the proximal phalanges) and joint movement is limited, then flexor tenosynovectomy should improve movement and, just as important, should prevent tendon rupture. Triggering, likewise, should be treated by tenosynovectomy rather than simple splitting of the sheath. Rupture of flexor digitorum profundus is best treated by distal IP joint fusion. Rupture of flexor pollicis longus (due to attrition against the underside of the distal radius or flexor synovitis) can be treated either by tendon grafting or by fusion of the thumb IP joint.

OSTEOARTHRITIS

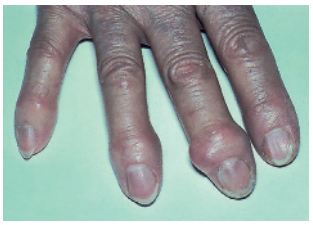
Eighty per cent of people over the age of 65 have radiological signs of osteoarthritis in one or more joints of the hand (Figure 16.21); fortunately, most of them are asymptomatic.

DISTAL INTERPHALANGEAL JOINTS

Osteoarthritis of the DIP joints is very common in postmenopausal women. It often starts with pain in one or two fingers; the distal joints become swollen and tender, the condition usually spreading to all the fingers of both hands. On examination there is bony thickening around the joints (Heberden's nodes) and some restriction of movement.

Treatment is usually symptomatic. If pain and instability are severe, a cortisone injection will give temporary relief. Joint fusion is a good solution. The angle of fusion is debatable. Intramedullary double-pitched screws are effective and avoid the problems of percutaneous wires. However, the final position is one of extension which slightly reduces grip in the little and ring fingers.

Mucous cysts sometimes protrude between the extensor tendon and collateral ligament of an osteoarthritic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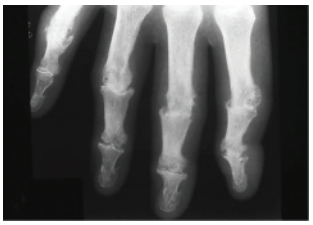
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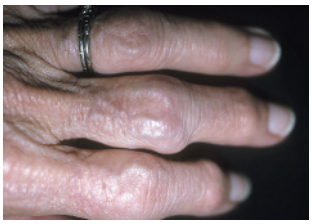
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Figure 16.21 Osteoarthritis (a,b) The common picture is one of 'knobbly finger-tips' due to involvement of the DIP joints (*Heberden's nodes*). (c) In some cases the PIP joints are affected as well (*Bouchard's nodes*).

DIP joint. They press on the germinal matrix of the nail, causing an unsightly groove. They occasionally ulcerate and septic arthritis can develop. If the cyst is too bothersome, excision of the cyst with the underlying osteophyte is effective. With luck, the nail will recover as well.

PROXIMAL INTERPHALANGEAL JOINTS

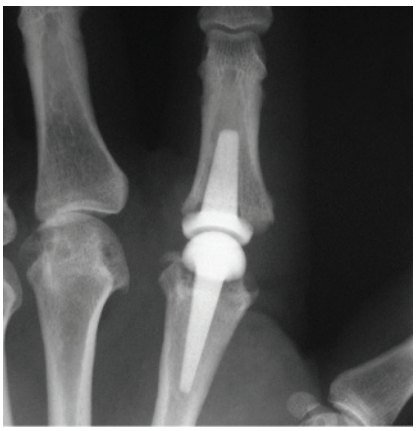
Not infrequently some of the PIP joints are involved (Bouchard's nodes). These are strongly associated with osteoarthritis elsewhere in the body (polyarticular OA). The joints are swollen and tend to deviate ulnarwards due to mechanical pressure in daily activities.

Treatment is usually non-operative. Pain usually settles over time. A steroid injection is very helpful for a flare-up. If the joint is very painful or unstable, surgery is considered (Figure 16.22). Fusion restores reliable, pain-free pinch in the *index and middle finger PIP joints*; if movement is required, an anatomically contoured joint (pyrocarbon or cobalt-chrome on polyethylene) can be used although the results are unpredictable: some patients do very well; others have problems with deformity, instability or stiffness. Long-term durability is unknown. Fusion of the *ring and little fingers* compromises grip and so joint replacement is usually preferable. Silastic hinges are easiest and cheapest and are perhaps most reliable, but anatomic implants made from pyrocarbon or cobalt-chrome on polyethylene are available.

METACARPOPHALANGEAL JOINTS

This is an uncommon site for osteoarthritis. When it does occur, a specific cause can usually be identified: previous trauma, infection, gout or haemochromatosis.

Treatment is initially non-operative with the use of analgesics, splints or local injections. Fusion of the thumb MCP gives excellent results; however, in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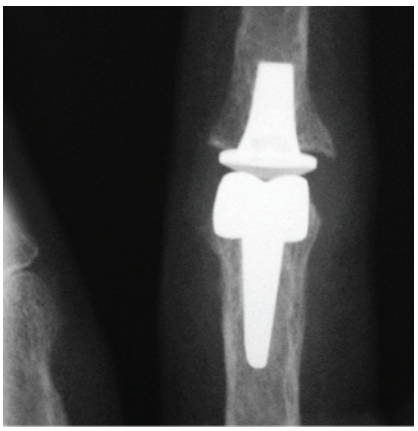
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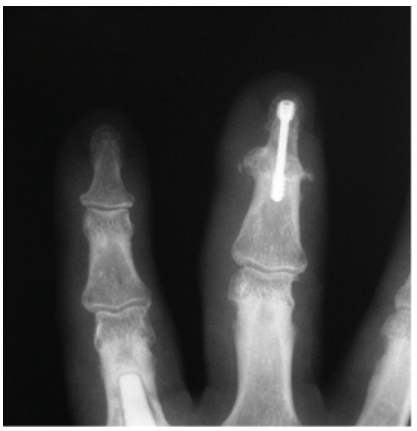
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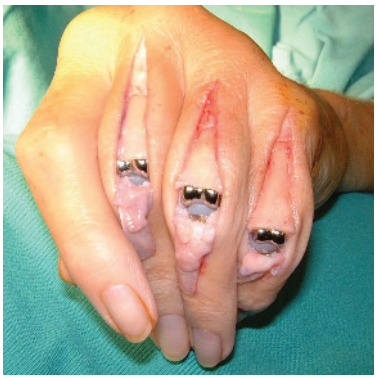
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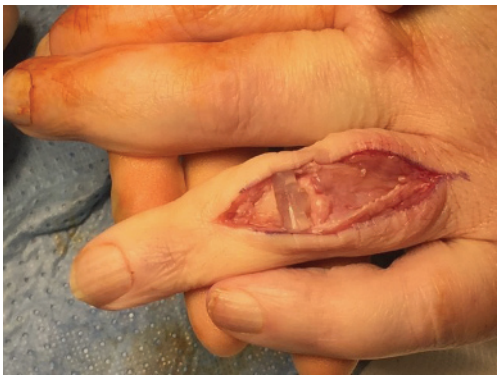
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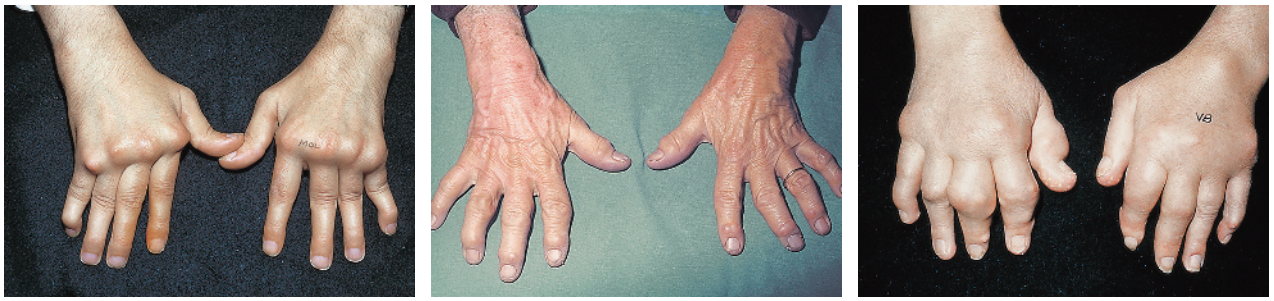
Figure 16.22 Osteoarthritis – operative treatment
(a) Pyrocarbon MCP joint replacement; (b) PIP joint replacement; (c) arthrodesis of the DIP joint; (d) cobalt-chrome on polythene PIP replacement; (e) silastic PIP replacement.



(a) (b) (c)

Figure 16.23 Swollen fingers Always be on the alert for 'lookalikes'. The clues (in most cases) are: (a) proximal joints = rheumatoid arthritis; (b) distal joints = osteoarthritis; (c) asymmetrical joints = gout.

the fingers this operation has serious functional consequences and is to be avoided. The MCP joints can be replaced with pyrocarbon or cobalt-chrome on polyethylene implants, with encouraging early and mid-term results.

CARPOMETACARPAL JOINT OF THE THUMB

This is discussed in Chapter 15.

CARPOMETACARPAL JOINT OF THE RING AND LITTLE FINGERS

These joints can become arthritic after a fracture dislocation. Because the fourth and fifth CMC joints normally flex forwards during power grip, pain can be disabling, particularly in patients engaged in heavy manual work. If a steroid injection fails to give improvement, surgery (usually fusion) is indicated.

ACUTE INFECTIONS OF THE HAND

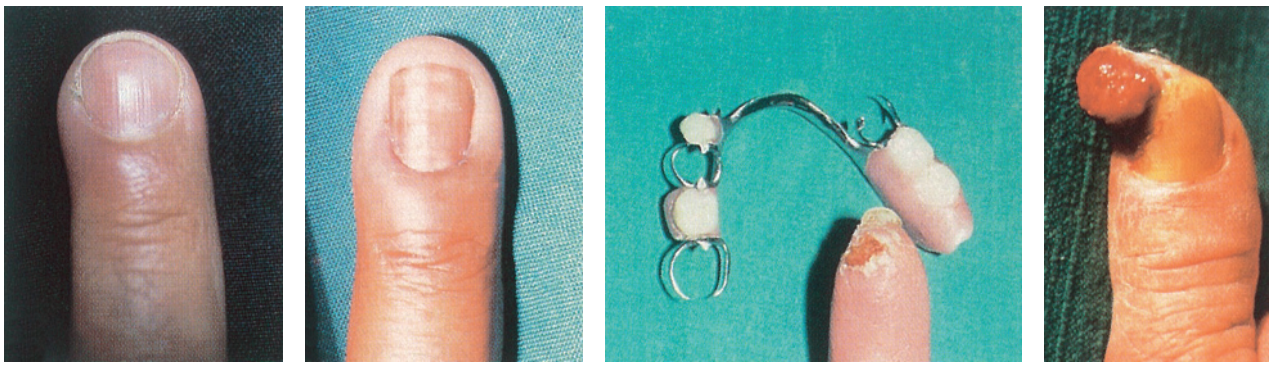
Infection of the hand is frequently limited to one of several well-defined compartments: under the nail-fold (paronychia); the pulp space (felon) and in the subcutaneous tissues elsewhere; the deep fascial spaces; tendon sheaths; and joints (Figure 16.24). Usually the cause is a staphylococcus which has been implanted during fairly trivial injury. However, cuts contaminated with unusual organisms account for about 10% of cases.

Pathology

Here, as elsewhere, the response to infection is an acute inflammatory reaction with oedema, suppuration and increased tissue tension. In closed tissue compartments (e.g. the pulp space or tendon sheath) pressures may rise to levels where the local blood supply is threatened, with the risk of tissue necrosis. In neglected cases infection can spread from one compartment to another and the end result may be a permanently stiff and useless hand. There is also a danger of lymphatic and haematogenous spread; even apparently trivial infections may give rise to lymphangitis and septicaemia.

Clinical features

Usually there is a history of trauma (a superficial abrasion, laceration or penetrating wound), but this may have been so trivial as to pass unnoticed. A few hours or days later the finger or hand becomes painful and



(a) (b) (c) (d)

Figure 16.24 Acute infections (1) (a) Acute nail-fold infection (paronychia); and (b) chronic paronychia. (c) Pulp-space infection (felon or whitlow) of the thumb due to a prick injury on the patient's own denture. (d) Septic granuloma. (Courtesy of Professor S. Biddulph.)

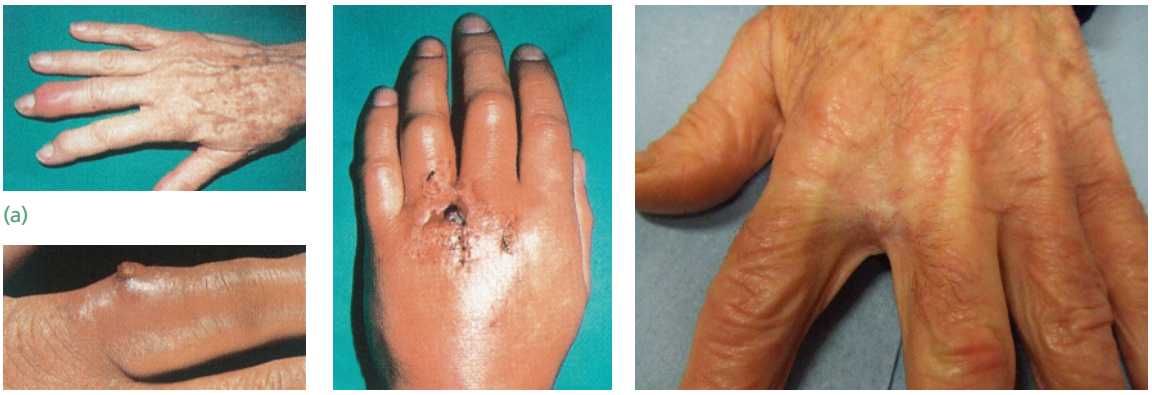


Figure 16.25 Acute infections (2) (a) Septic arthritis of the terminal interphalangeal joint following a cortisone injection. (b) Infected insect 'bite'. (c) Septic human bite resulting in acute infection of the fourth metacarpophalangeal joint. (Courtesy of Professor S. Biddulph.) (d) Web-space infection.

swollen. There may be throbbing and sometimes the patient feels ill and feverish. Ask if he or she can recall any causative incident: a small cut or superficial abrasion, a prick injury (including plant thorns) or a local injection. Also, do not forget to enquire about predisposing conditions such as diabetes mellitus, intravenous drug abuse and immunosuppression.

On examination the finger or hand is red and swollen, and it is usually exquisitely tender over the site of tension (Figure 16.25). However, in immune-compromised patients, in the very elderly and in babies, local signs may be mild. With superficial infection the patient can usually be persuaded to flex an affected finger; with deep infections active flexion is not possible. The arm should be examined for lymphangitis and swollen glands, and the patient examined more generally for signs of septicaemia.

X-ray examination may disclose a foreign body but is otherwise unhelpful in the early stages of infection. However, a few weeks later there may be features of osteomyelitis or septic arthritis, and later still of bone necrosis.

If pus becomes available, this should be sent for bacteriological examination.

Diagnosis

In making the diagnosis, several conditions must be excluded: an *insect bite or sting* (which can closely mimic a subcutaneous infection), a *thorn prick* (which, itself, can become secondarily infected), acute *tendon rupture* (which may resemble a septic tenosynovitis) and *acute gout* (which is easily mistaken for septic arthritis).

Plant-thorn injuries are extremely common and the distinction between secondary infection and a non-septic reaction to a retained fragment can be difficult. Rose thorn and blackthorn are the usual suspects in the UK, but any plant spine (including cactus needles) can be implicated. The local inflammatory response sometimes leads to recurrent arthritis or tenosynovitis, which is arrested only by removing the retained fragment. If the condition is suspected, the fragment may be revealed by ultrasound scanning or MRI. Secondary infection with unusual soil or plant organisms may occur.

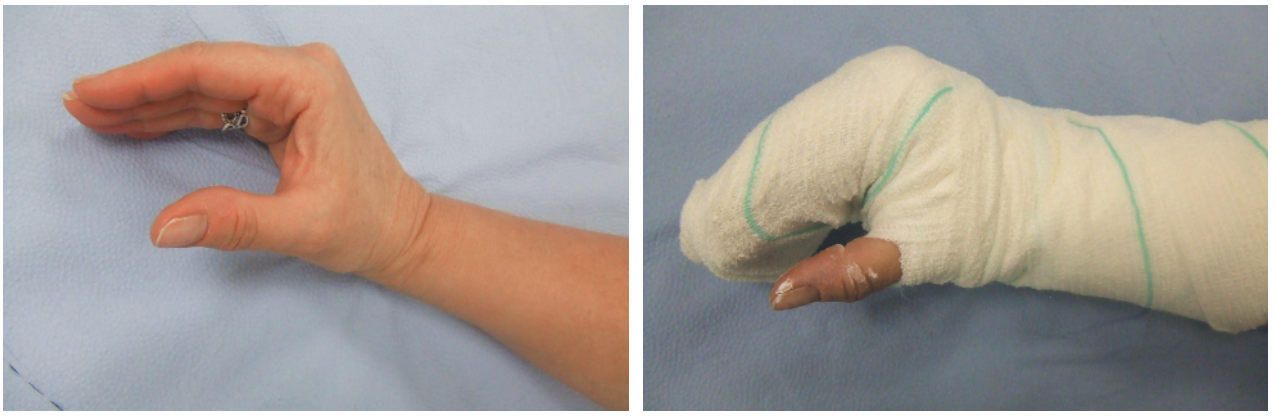
Principles of treatment

Superficial hand infections are common; if their treatment is delayed or inadequate, infection may rapidly extend, with serious consequences. The essentials of treatment are:

- antibiotics
- rest, splintage and elevation
- drainage
- rehabilitation.

Antibiotics As soon as the clinical diagnosis is made, and preferably after a specimen has been taken for Gram stain and culture, antibiotic treatment is started – usually with flucloxacillin or a cephalosporin. If bone infection is suspected, fusidic acid may be added. For bites (which should always be assumed to be infected) a broad-spectrum penicillin is advisable. Agricultural injuries risk infection by anaerobic organisms and it is therefore prudent to add metronidazole. The interim antibiotic may later be changed when the bacterial sensitivity is known.

Rest, splintage and elevation In a mild case the hand is rested in a sling. In a severe case the patient is admitted to hospital; the arm is held elevated in an overhead sling while the patient is kept under observation. Analgesics are given for pain. *The hand must be splinted in the position of safe immobilization* with the wrist slightly extended, the MCP joints in full flexion, the IP joints extended and the thumb in abduction (Figure 16.26).



(a) (b)

Figure 16.26 The position of safe immobilization The knuckle joints are 90° flexed, the finger joints extended and the thumb abducted. This is the position in which the ligaments are at their longest and splintage is least likely to result in stiffness.

Drainage If treated within the first 24–48 hours, many hand infections will respond to antibiotics, rest, elevation and splintage.

If there are signs of an abscess – throbbing pain, marked tenderness and toxaemia – the pus should be drained (Figure 16.27). A tourniquet and either general or regional block anaesthesia are essential. The hand should be exsanguinated by elevation only; an exsanguinating bandage may spread the sepsis. The incision should be planned to give access to the abscess without causing injury to other structures *but never at right angles across a skin crease*. When pus is encountered, it must be carefully wiped away and a search made for deeper pockets of infection. Necrotic tissue should be excised. The area is thoroughly washed out and, in some cases, a catheter may be left in place for further, postoperative, irrigation (e.g. in cases of flexor tenosynovitis). The wound is either left open or lightly sutured, and it is then covered with a nonadhesive dressing and gauze. The pus obtained is sent for culture.

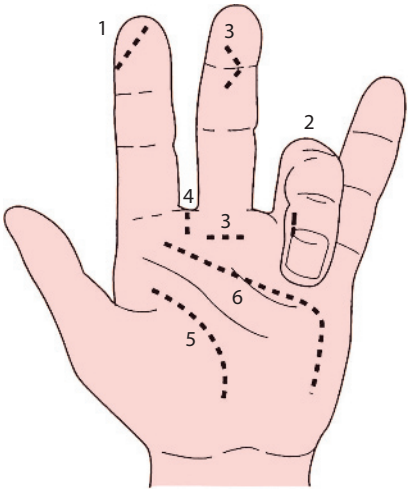


Figure 16.27 Infections The incisions for surgical drainage are shown here: (1) pulp space (directly over the abscess); (2) nail-fold (it may also be necessary to excise the edge of the nail); (3) tendon sheath; (4) web space; (5) thenar space; (6) mid-palmar space.

At the end of the operation the hand is splinted in the position of safe immobilization. A removable splint will permit repeated wound dressings and exercises. A sling is used to keep the arm elevated.

The hand should be re-examined within the next 24 hours to ensure that drainage is effective; if it is not, further operative drainage may be needed. Inadequate drainage of acute infection may lead to chronic infection.

Postoperative rehabilitation As soon as the signs of acute inflammation have settled, movements must be started under the guidance of a hand therapist, otherwise the joints are liable to become stiff. For the first few days the resting splint is reapplied between exercise sessions.

Nail-fold infection (paronychia)

Infection under the nail-fold is the commonest hand infection; it is seen most often in children, or in older people after rough nail-trimming. The edge of the nail-fold becomes red and swollen and increasingly tender. A tiny abscess may form in the nail-fold; if this is left untreated, pus can spread under the nail.

At the first sign of infection, treatment with antibiotics alone may be effective. However, if pus is present, it must be released by an incision at the corner of the nail-fold in line with the edge of the nail; a pledget of paraffin gauze is used to keep the nail-fold open. If pus has spread under the nail, part or allof the nail may need to be removed.

Chronic paronychia Chronic nail-fold infection may be due to (1) inadequate drainage of an acute

infection, or (2) a fungal infection, which requires specific treatment. Topical or oral antifungal agents are used to eradicate fungal infection but, failing this, or for chronic bacterial infection, the nail bed may have to be laid open ('marsupialized'); care should be taken to avoid damaging the germinal nail matrix.

Pulp infection (felon)

The distal finger pad is essentially a closed fascial compartment filled with compact fat and subdivided by radiating fibrous septa. A rise in pressure within the pulp space causes intense pain and, if unrelieved, may threaten the terminal branches of the digital artery which supply most of the terminal phalanx.

Pulp-space infection is usually caused by a prick injury; blackthorn injuries are particularly likely to become infected. The most common organism is *Staphylococcus aureus*. The patient complains of throbbing pain in the fingertip, which becomes tensely swollen, red and acutely tender.

If the condition is recognized very early, antibiotic treatment and elevation of the hand may suffice. Once an abscess has formed, the pus must be released through a small incision over the site of maximum tenderness. If treatment is delayed, infection may spread to the bone, the joint or the flexor tendon sheath.

Postoperatively the finger is dressed with a loose packing of gauze; antibiotic treatment is modified if the results of culture and sensitivity so dictate, and is continued until all signs of infection have cleared. The wound will gradually heal by secondary intention.

Herpetic whitlow The herpes simplex virus may enter the fingertip, possibly by auto-inoculation from the patient's own mouth or genitalia, or by cross-infection during dental surgery. Small vesicles form on the fingertip, then coalesce and ulcerate. The condition is self-limiting and usually subsides after about 10 days, but it may recur from time to time.

Herpetic whitlow should not be confused with a staphylococcal felon. Surgery is unhelpful and may be harmful, exposing the finger to secondary infection. Aciclovir may be effective in the early stages.

Other subcutaneous infections

Anywhere in the hand a blister, a superficial cut or an insect 'bite' may become infected, causing redness, swelling and tenderness. A local collection of pus should be drained through a small incision over the site of maximal tenderness (but never crossing a skin crease or the web edge); in the finger, a mid-lateral incision is suitable. It is important to exclude a deeper pocket of pus in a nearby tendon sheath or in one of the deep fascial spaces.

Tendon sheath infection (suppurative tenosynovitis)

The tendon sheath is a closed compartment extending from the distal palmar crease to the DIP joint. In the thumb and little finger, the sheaths are co-extensive with the radial and ulnar bursae, which envelop the flexor tendons in the proximal part of the palm and across the wrist; these bursae also communicate with Parona's space in the lower forearm.

Pyogenic tenosynovitis is uncommon but dangerous. It usually follows a penetrating injury, the commonest organism being *Staphylococcus aureus*; however, *Streptococcus* and Gram-negative organisms are also encountered.

The affected digit is painful and swollen; it is usually held in slight flexion, is very tender, and the patient will not move it or permit it to be moved (see Box 16.2). Early diagnosis is based on clinical findings; X-rays are unhelpful but ultrasound scanning may be useful.

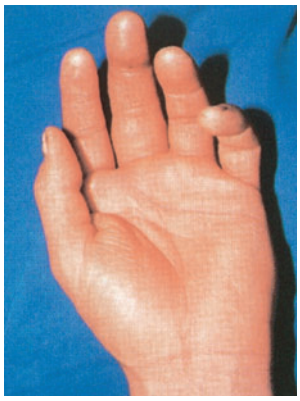
Delayed diagnosis results in a progressive rise in pressure within the sheath and a consequent risk of vascular occlusion and tendon necrosis. In neglected cases infection may spread proximally within the radial or ulnar bursa, or from one to the other (a *'horse-shoe' abscess*); it can also spread proximally to the flexor compartment at the wrist and into Parona's space in the forearm. Occasionally this results in median nerve compression.

Treatment Treatment must be started as soon as the diagnosis is suspected. The hand is elevated and splinted and antibiotics are administered intravenously – ideally a broad-spectrum penicillin or a systemic cephalosporin. If there is no improvement after 24 hours, surgical drainage is essential. Two incisions are needed, one at the proximal end of the sheath and one at the distal end; using a fine catheter, the sheath is then irrigated (always from proximal to distal) with irrigation solution. Additional, proximal, incisions may be needed if the synovial bursae are infected.

Postoperatively the hand is swathed in absorbent dressings and splinted in the position of safe immobilization. The dressings should not be too bulky, as this will make it difficult to ensure correct positioning

BOX 16.2 KANAVEL'S SIGNS OF FLEXOR SHEATH INFECTION

Flexed posture of digit
Tenderness along the course of the tendon
Pain on passive finger extension
Pain on active flexion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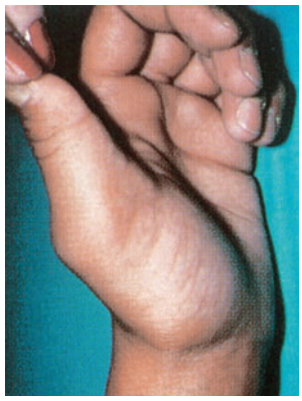
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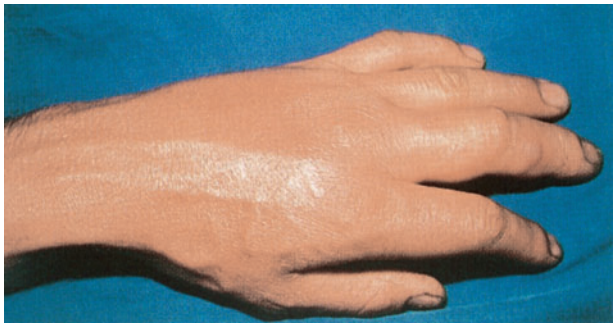
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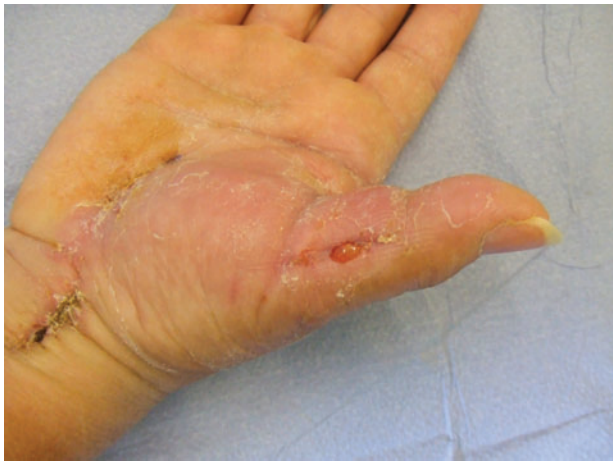
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Figure 16.28 Acute infections (3) (a) Flexor tenosynovitis of the middle finger following a cortisone injection. (b) Tuberculous synovitis of flexor pollicis longus. (c) Diffuse septic extensor tendinitis. (Courtesy of Professor S. Biddulph.) (d) Incisions after drainage of flexor sheath infection of the thumb.

of the joints. The flexor sheath catheter is left in place; using a syringe, the sheath is irrigated with 20 mL of saline three or four times a day for the next 2 days. The catheter and dressings are then removed and finger movements are started.

Stiffness is a very real risk and so early supervised hand therapy must be arranged.

Deep fascial space infection

The large thenar and mid-palmar fascial spaces may be infected directly by penetrating injuries or by secondary spread from a web space or an infected tendon sheath.

Clinical signs can be misleading; the hand is painful but, because of the tight deep fascia, there may be little or no swelling in the palm while the dorsum bulges like an inflated glove. There is extensive tenderness and the patient holds the hand as still as possible.

Treatment As with other infections, splintage and intravenous antibiotics are commenced as soon as the diagnosis is made. For drainage, an incision is made directly over the abscess (being careful not to cross the flexor creases) and sinus forceps inserted; if the web space is infected it, too, should be incised. A *thenar space abscess* can be approached through the first web space (but do not incise in the line of the skin-fold) or through separate dorsal and palmar incisions around the thenar eminence. Great care must be taken to avoid damage to the tendons, nerves and blood vessels. A thorough knowledge of anatomy is essential. The deep *mid-palmar space* (which lies between the flexor tendons and the metacarpals) can be drained through an incision in the web space between the middle and ring fingers, but wider exposure through a transverse or oblique palmar incision is preferable, taking care not to cross the flexor creases directly. Above all, do not be misled by the swelling on the back of the hand into attempting drainage through the dorsal aspect.

Occasionally, deep infection extends proximally across the wrist, causing symptoms of median nerve compression. Pus can be drained by anteromedial or anterolateral approaches; incisions directly over the flexor tendons and median nerve are avoided. Operation wounds are either loosely stitched or left open. Bulky dressings and saline irrigation are employed, more or less as described for tendon sheath infections.

Septic arthritis

Any of the MCP or finger joints may be infected, either directly by a penetrating injury or intra-articular injection, or indirectly from adjacent structures (and occasionally by haematogenous spread from a distant site). *Staphylococcus* and *Streptococcus* are the usual organisms; *Haemophilus influenzae* is a common pathogen in children. A 'fight-bite' is a common cause of infection of the MCP joints (see Figure 16.25).

Pain, swelling and redness are localized to a single joint, and all movement is resisted. The presence of lymphangitis and/or systemic features may help to

clinch the diagnosis; in their absence, the early symptoms and signs are indistinguishable from those of acute gout. Joint aspiration may give the answer.

Treatment Intravenous antibiotics are administered and the hand is splinted. If the inflammation does not subside within 24 hours, or if there are overt signs of pus, open drainage is needed. Dorsoulnar or dorsoradial incisions between the collateral ligaments and extensors are recommended for the finger IP joints; for the MCP joints, a mid-dorsal incision is needed. The capsule is closed with an absorbable suture but the skin wounds are left open, to heal by secondary intention. Copious dressings are applied and the hand is splinted in the 'position of safe mobilization' for 48 hours; thereafter, movement is encouraged.

Intravenous antibiotics are continued until all signs of sepsis have disappeared; it is prudent to follow this with another 2-week course of oral antibiotics.

Bites

ANIMAL BITES

Animal bites are usually inflicted by cats, dogs, farm animals or rodents. Many become infected and, although the common pathogens are staphylococci and streptococci, unusual organisms like *Pasteurella multocida* are often reported.

HUMAN BITES

Human bites are generally thought to be even more prone to infection. A wide variety of organisms (including anaerobes) are encountered, the commonest being *Staphylococcus aureus*, *Streptococcus* Group A and *Eikenella corrodens*.

Bites can involve any part of the hand, fingers or thumb; telltale signs of a human bite are lacerations on both volar and dorsal surfaces of the finger. Often, though, the 'bite' consists only of a dorsal wound over one of the MCP knuckles, sustained during a fist-fight. *All such wounds should be assumed to be infected*. Moreover, it should be remembered that a laceration of the clenched fist may have penetrated the extensor apparatus and entered the MCP joint; this will not be apparent if the wound is examined with the fingers in extension because the extensor hood and capsule will have retracted proximally.

X-rays should be obtained (to exclude a fracture, tooth fragment or foreign body) and swabs taken for bacterial culture and sensitivity.

Treatment

Fresh wounds should be carefully examined in the operating theatre and, if necessary, extended and debrided. Search for a fragment of tooth or – with a knuckle bite – for a *divot* of articular cartilage from the joint. The hand is splinted and elevated and antibiotics are given prophylactically until the laboratory results are obtained.

Infected bites will need debridement, wash-outs and intravenous antibiotic treatment. The common infecting organisms are all sensitive to broad-spectrum penicillins (e.g. amoxicillin with clavulanic acid) and cephalosporins. With animal bites one should also consider the possibility of rabies.

Postoperative treatment consists, as usual, of copious wound dressings, splintage in the 'position of safe mobilization' and encouragement of movement once the infection has resolved. Tendon lacerations can be dealt with when the tissues are completely healed.

Mycobacterial infections

Tuberculous tenosynovitis is uncommon even in countries where tuberculosis is still rife. The diagnosis should be considered in patients with chronic synovitis once the alternatives such as rheumatoid disease have been excluded; it can be confirmed by synovial biopsy. Treatment is by synovectomy and then prolonged chemotherapy.

'Fishmonger's infection' is a chronic infection of the hand caused by *Mycobacterium marinum* (Figure 16.29). The organism is introduced by prick injuries from fish spines or hard fins in people working with fish or around fishing boats. It may appear as no more than a superficial granuloma, but deep infection can give rise to an intractable synovitis of tendon or joint. Other causes of chronic synovitis must be excluded; definitive diagnosis usually requires biopsy for histological examination and special culture.

Superficial lesions often heal on their own; if not, they can be excised. Deep lesions usually require surgical synovectomy. Prolonged antibiotic treatment is needed to avoid recurrence; the recommended drug is a broad-spectrum tetracycline such as minocycline, or else chemotherapy with ethambutol and rifampicin.

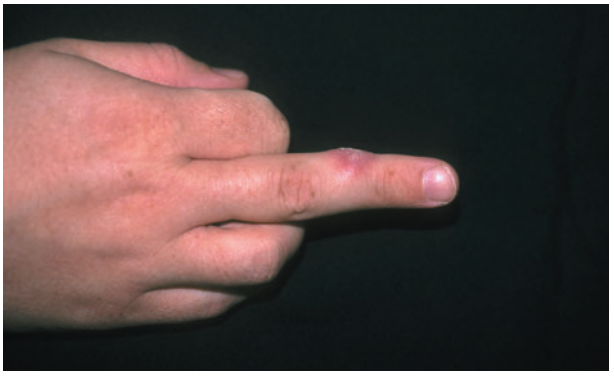


Figure 16.29 *Mycobacterium marinum* infection
Infection in an aquarium keeper.

Fungal infections

Superficial tinea infection of the palm and interdigital clefts (similar to 'athlete's foot') is fairly common and can be controlled by topical preparations. Tinea of the nails can be more difficult to eradicate and may require oral antifungal medication and complete removal of the nail.

Subcutaneous infection by *Sporothrix schenckii* (sporotrichosis) is rarely seen in the UK but is not uncommon in North America, where it is usually caused by a thorn prick. Chronic ulceration at the prick site, unresponsive to antibiotic treatment, may suggest the diagnosis, which can be confirmed by microbiological culture. The recommended treatment is oral potassium iodide.

Deep mycotic infection may involve tendons or joints. The diagnosis should be confirmed by microscopy and microbiological culture. Treatment is by local excision and administration of an intravenous antifungal agent. Resistant cases occasionally require limited amputation.

Opportunistic fungal infections are more likely to occur in debilitated and immunosuppressed patients.

VASCULAR DISORDERS OF THE HAND

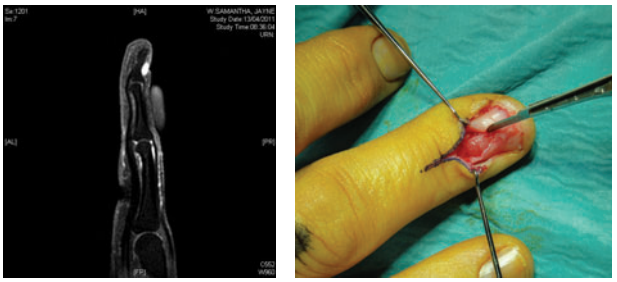
Emboli

Arising from the heart or from aneurysms in the arteries of the upper limb, emboli can lodge in distal vessels causing splinter haemorrhages, or in larger, more proximal vessels, causing ischaemia of the arm. A large embolus leads to the classic signs of pain, pulselessness, paraesthesia, pallor and paralysis. Untreated, gangrene or ischaemic contracture ensues.

Raynaud's disease

Raynaud's syndrome is produced by a vasospastic disorder which affects mainly the hands and fingers. Attacks are usually precipitated by cold; the fingers go pale and icy, then dusky blue (or cyanotic) and finally red. Between attacks the hands look normal. The condition is most commonly seen in young women who have no underlying or predisposing disease.

Raynaud's phenomenon is the term applied when these changes are associated with an underlying disease such as scleroderma or arteriosclerosis. Similar, though milder, changes are also seen in thoracic outlet syndrome. The hands must be kept warm. Calcium channel blockade, iloprost infusions, botulinum toxin or digital sympathectomy (surgical removal of the sympathetic plexus around the digital arteries) may be needed.



(a) (b)

Figure 16.30 Glomus tumour (a) MRI showing tumour beneath the nail; (b) removal under local anaesthetic.

A glomus tumour is a rare but very troublesome condition (Figure 16.30). Formed of small neural and vascular elements, it is very painful especially in colder weather. On examination there is a very localized and exquisitely tender spot, usually under or just alongside the nail bed. MRI sometimes shows the lesion. Treatment is by removal under local anaesthetic after very careful pinpoint marking before surgery.

Hand–arm vibration syndrome

Excessive and prolonged use of vibrating tools can damage the nerves and vessels in the fingers. The damage is proportional to the duration of exposure and amount of vibration. There are two components: vascular and neurological. The *vascular component* is similar to Raynaud's phenomenon, with the fingertips turning white in cold weather, then changing through blue and red as the circulation is restored. The *neurological component* involves numbness and tingling in the fingertips. In advanced cases there can be reduced dexterity. Some patients have carpal tunnel syndrome as well.

Treatment Treatment is generally unsatisfactory, but includes avoidance of cold weather and smoking as well as, of course, vibrating tools. Carpal tunnel syndrome associated with vibration, in the absence of a more diffuse neuropathy, responds fairly well to standard decompression.

ULNAR ARTERY THROMBOSIS

Repeated blows to the hand, especially using the hypothenar eminence as a hammer, can damage the intima of the ulnar artery, leading to either thrombosis or an aneurysm. The patient presents with cold intolerance in the little finger. Microvascular reconstruction of the ulnar artery may be needed.

OTHER GENERAL DISORDERS

A number of generalized disorders should always be borne in mind when considering the diagnosis of

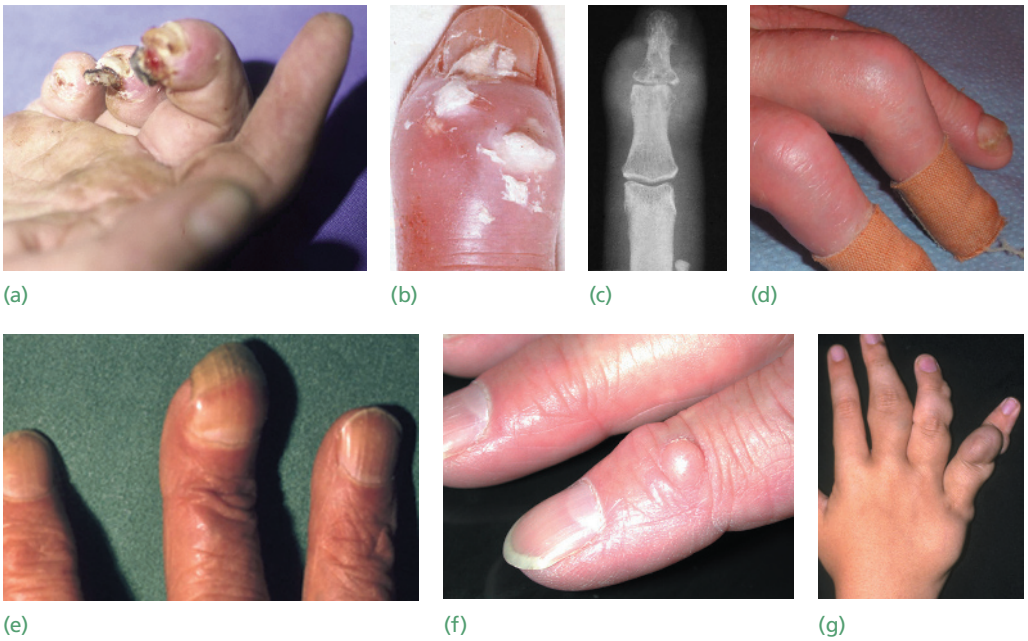


Figure 16.31 The hand in general disorders Some general conditions that may manifest with lesions in the hand: (a) scleroderma; (b,c) gouty tophi; (d) psoriasis; (e) implantation dermoid; (f) dermatofibroma; (g) Maffucci's syndrome; (h) Secretan's syndrome (hand oedema due to repetitive trauma, self-inflicted).

any unusual lesion that appears to be confined to the hand. It is beyond the scope of this text to enlarge on these conditions. The few examples shown in Figure 16.31 serve merely as a reminder that a general history and examination are as important as focused attention on the hand.

NOTES ON APPLIED ANATOMY

Function

The hand serves three basic functions: *sensory perception*, *precise manipulation* and *power grip* (see also Figure 16.1). The first two involve the thumb, index and middle fingers; without normal sensation and the ability to oppose these three digits, manipulative precision will be lost. The ring and little fingers provide power grip, for which they need full flexion though sensation is less important.

With the wrist flexed, the fingers and thumb fall naturally into extension. With the wrist extended, the fingers curl into flexion and the tips of the thumb, index and middle fingers form a functional tripod; this is the *position of function* (Figure 16.32b), because it is best suited to the actions of prehension.

Finger flexion is strongest when the wrist is powerfully extended; normal grasp is possible only with a painless, stable wrist. Spreading the fingers produces abduction to either side of the middle finger; bringing them together, adduction. Abduction and adduction of the thumb occur in a plane at right angles to the palm (i.e. with the hand lying palm upwards,

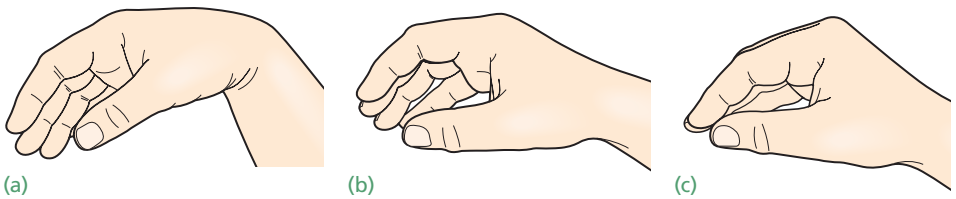


Figure 16.32 Three positions of the hand (a) The position of relaxation; (b) the position of function (ready for action); (c) the position of safe immobilization, with the ligaments taut.

abduction points the thumb to the ceiling). By a combination of movements the thumb can also be opposed to each of the other fingers. Functionally, the thumb is 40% of the hand.

Skin

The *palmar skin* is relatively tight and inelastic; skin loss can be ill-afforded and wounds sutured under tension are liable to break down. The acute sensibility of the digital palmar skin cannot be achieved by any skin graft. Although the *dorsal skin* seems lax and mobile with the fingers extended, flexion will show that there is very little spare skin. Loss of skin therefore often requires a graft or flap.

Just deep to the palmar skin is the palmar aponeurosis, the embryological remnant of a superficial layer of finger flexors; attachment to the bases of the proximal phalanges explains part of the deformity of Dupuytren's contracture. Incisions on the palmar surface are also liable to contracture unless they are placed in the line of the skin creases, along the midlateral borders of the fingers or obliquely across the creases.

Joints

The carpometacarpal joints

The second and third metacarpals have very little independent movement; the fourth and fifth have more, allowing greater closure of the ulnar part of the hand during power grip. The metacarpal of the thumb is the most mobile and the first CMC joint is a frequent target for degenerative arthritis.

The metacarpophalangeal joints

These flex to about 90 degrees. The range of extension increases progressively from the index to the little finger. The collateral ligaments are lax in extension (permitting abduction) and tight in flexion (preventing abduction). *If these joints are immobilized, they should always be in flexion, so that the ligaments are at full stretch and therefore less likely to shorten if they should fibrose.*

The interphalangeal joints

The IP joints are simple hinges, each flexing to about 90 degrees. Their collateral ligaments send attachments to the volar plate and these fibres are tight in extension and lax in flexion; *immobilization of the IP joints, therefore, should always be in extension.*

Muscles and tendons

Two sets of muscles control finger movements: the *long extrinsic muscles* (extensors, deep flexors and superficial flexors), and the *short intrinsic muscles* (interossei, lumbricals and the short thenar and hypothenar muscles). The extrinsics extend the MCP joints (long extensors) and flex the IP joints (long flexors). The intrinsics flex the MCP and extend the IP joints; the dorsal interossei also abduct and the palmar interossei adduct the fingers from the axis of the middle finger. Spasm or contracture of the intrinsics causes the *intrinsic-plus* posture – flexion at the MCP joints, extension at the IP joints and adduction of the thumb. Paralysis of the intrinsics produces the *intrinsic-minus* posture – hyperextension of the MCP and flexion of the IP joints ('claw hand').

Tough *fibrous sheaths* enclose the flexor tendons as they traverse the fingers; starting at the MCP joints (level with the distal palmar crease) they extend to the DIP joints. They serve as runners and pulleys, so preventing the tendons from bowstringing during flexion. Scarring within the fibro-osseous tunnel prevents normal excursion.

The long extensor tendons are prevented from bowstringing at the wrist by the extensor retinaculum; here they are liable to frictional trauma. Over the MCP joints each extensor tendon widens into an expansion which inserts into the proximal phalanx and then splits in three; a central slip inserts into the middle phalanx, the two lateral slips continue distally, join and end in the distal phalanx. Division of the middle slip causes a flexion deformity of the PIP joint (boutonnière); rupture of the distal conjoined slip causes flexion deformity of the DIP joint (mallet finger).

Nerves

The median nerve supplies the abductor pollicis brevis, opponens pollicis and lumbricals to the middle and index fingers; it also innervates the palmar skin of the thumb, index and middle fingers and the radial half of the ring finger.

The ulnar nerve supplies the hypothenar muscles, all the interossei, lumbricals to the little and ring fingers, flexor pollicis brevis and adductor pollicis. Sensory branches innervate the palmar and dorsal skin of the little finger and the ulnar half of the ring finger.

The radial nerve supplies skin over the dorsoradial aspect of the hand.

The neck

17

Jorge Mineiro & Nuno Lança

APPLIED ANATOMY

Anatomical considerations of the cervical spine

The neck has a gentle curvature with an anterior convexity. The bony structure of the neck is the cervical spine with *seven vertebrae*, arranged in a lordotic configuration of 16 to 25 degrees. This *physiologic lordosis* is never quite reversed, even in flexion, unless under pathologic conditions.

Important *palpable landmarks* of the neck are the hyoid bone, which lies at the level of C3, the thyroid cartilage, lying in front of C4, and the cricoid cartilage, at the level of C6 (Figure 17.1).

The seven cervical vertebrae are different in shape. The first two, the atlas (C1) and the axis (C2), are morphologically different from all the other five vertebrae (C3–C7) that have a similar morphology.

The *atlas* arises from three ossification centres. Without a vertebral body or spinous process, C1 has thick anterior and posterior arches merging laterally into large masses through which it articulates with the occipital condyles above and the axial facet joints below.

The *axis* originates from six ossification centres. The vertebral body has a characteristic superior peg, the *dens*, which articulates with the posterior surface of the anterior arch of the atlas. The dens can have a posterior angulation of up to 30 degrees. The transverse ligament of the atlas runs across the back of a narrowed waist of the odontoid process, stabilizing the joint, particularly in rotation. The *ossification of the dens* starts at 6 months of gestation, but fusion to the C2 vertebral body is only completed by the age of 5–6 years. However, ossification of the tip of the dens starts at 3–5 years of age and will only be completely fused at a later stage, during adolescence. The large spinous process of the axis allows for muscle insertion, namely the *rectus capitis* and the inferior oblique muscles.

The *subaxial cervical spine* extends from C3 to C7. With a smaller vertebral body, the subaxial cervical vertebrae, although similar in shape, differ from the vertebrae in other segments of the spine because these have two transverse *foramina* for the vertebral arteries, running from C6 (in 90% of cases) to C1, and two vertebral foramina for the nerve roots. The *vertebral body* is generally 17–20 mm wide, has two that are cranial projections on each side of the vertebral body, (uncal processes) that create a more concave shape to the superior end plate and participate in the motion pattern of the cervical spine, coupling bending and rotation.

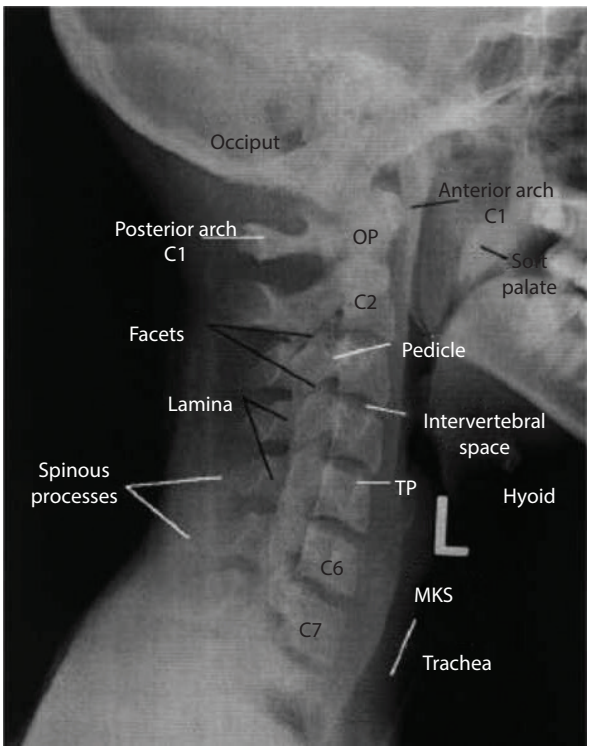


Figure 17.1 Radiological anatomy of the cervical region (Reproduced with permission from: Todd MM. Cervical spine anatomy and function for the anesthesiologist. *Can J Anaesth* 2001; **48**(Suppl 1): R1–R5.)

The short and medially oriented *pedicles* connect the vertebral body with the lateral masses. The diameter of the pedicles increases downwards, with C6 pedicles being the largest.

The cervical *articular facets* are oriented at 0 degrees in the coronal plane and 40–55 degrees in the sagittal plane, with the upper articulating surface oriented dorsosuperiorly and the inferior ventroinferiorly. *Spinous processes* are often bifid from C2 to C6, and the C7 spinous process is usually longer, the reason why it is called the *vertebra prominens*.

The primary function of the subaxial cervical spine is to resist compressive forces. The facets are part of a tripod of stable joints (two facets and one intervertebral disc) allowing flexion/extension, lateral bending and slight rotation. Under abnormal distractive forces they may also allow subluxation or dislocation to occur (even without fracture), a displacement that is usually prevented by the strong posterior ligaments.

The cervical *spinal canal* has a triangular shape in the axial plane and its diameter decreases from approximately 17 mm at C3 to 15 mm at C7. The *spinal cord* elongates and 'squeezes' in flexion and shortens and enlarges in extension. As much as 30% of cord compression can irreversibly damage the spinal cord.

The cervical spine contains eight pairs of *nerve roots*. They pass through relatively narrow *neural foramina*, above the similarly numbered vertebra, the first between the occiput and C1, and the eighth between C7 and the first thoracic (T1) vertebra. Hence, a lesion such as a disc prolapse between C5 and C6 might compress the sixth root.

Intervertebral discs lie between the vertebral bodies, with their posterior margin close to the nerve roots as they emerge through the foramina. Even a small herniation might compress or even stretch the nerve root exiting the spine, causing radicular symptoms (with radiating pain and paraesthesiae to the shoulder or upper limb) rather than neck pain.

Degenerative disc disease is associated with spur formation on both the posterior aspect of the vertebral body and the associated facet joints. Bone formation results in encroachment of the nerve root in the *intervertebral foramen*. Radiating pain can also be caused by facet joint degeneration or the soft surrounding structures. Facetary pain is typically aggravated with extension, lateral bending and rotation. Only radiculopathy (i.e. paraesthesiae and sensory or motor compromise) with shooting pain down the arm/forearm are unequivocal evidence of nerve root compression.

The *cervical spine motion* can be analysed in three different axes: flexion/extension, lateral bending and axial rotation. Head motion is a combination of all these movements.

The *occipitocervical junction* contributes to approximately 50% of the neck flexion-extension movement (the '*YES*' joint), with a C0–C1 range of motion of 21 degrees of flexion and 3.5 degrees of extension. At the *atlanto-occipital joint*, the movements that occur are nodding and tilting (lateral flexion).

The *atlantoaxial articulation* contributes to approximately 50% of neck rotation (the '*NO*' joint), with a C1–C2 range of motion of 47 degrees of axial rotation. The *vertebral artery* loop in this region allows the artery to adapt to the normal axial rotation.

In the subaxial cervical spine the main motion patterns are flexion-extension and lateral bending. The majority of the flexion-extension movement in the subaxial cervical spine occurs at the level of C4–C5 and C5–C6, the reason why these levels are more frequently affected in the degenerative process of the disc. The majority of lateral bending occurs from C2 to C4. The least mobile segment in the cervical spine is C7–T1 because it is usually deeply seated into the upper chest.

Surgical approaches to the cervical spine

The *Smith-Robinson-Cloward* approach is the most widely used for anterior cervical surgery. The spine is accessed through a slightly oblique skin incision on the side of the neck (right or left) in front of the sternocleidomastoid muscle (SCM). Deeper, soft-tissue dissection proceeds with incision of platysma and then the anterior cervical fascia on the medial border of the SCM. Progression medially to the carotid sheath, which is dorsolateral to the visceral space and ventrolateral to the prevertebral fascia, provides direct access to the midline of the anterior cervical spine. The cervical sympathetic chain is located posteromedially to the carotid sheath. The thoracic duct lies posterior to the carotid sheath on the left side. Sometimes crossing the operative field, the omohyoid muscle may be divided to facilitate the access. The anterior surface of the spine, just over the anterior longitudinal ligament, is separated from the pharynx by only a very thin layer of tissue with pharyngeal mucosa, constrictor muscles, buccopharyngeal fascia and prevertebral muscles.

The *oesophagus* at this level lies in front of the spine and behind the trachea. Due to its soft structure it can be easily injured if caution is not taken during the approach. *Dysphagia* is a common complication of anterior surgery of the cervical spine, although most frequently its aetiology is unclear.

The *recurrent laryngeal nerve* is another structure that is at risk in the cervical spine anterior approach. It supplies motor innervation to the intrinsic laryngeal muscles that control movement of the vocal cords and also supply sensory

innervation to the larynx below the vocal cords. Retraction of the recurrent laryngeal nerve during the anterior approach, mainly from the right side, where the nerve loops around the right subclavian artery and travels upwards being susceptible to injury by traction from the retractors, may cause *hoarseness* (or *aphonia*, if injured bilaterally). Disruption of the inferior *sympathetic cervical (stellate) ganglion*, which lies in front of the C7 transverse process, can result in *Horner syndrome*.

Anatomical variations of the course of the vertebral artery exist, such as medial loops of the vertebral artery or even the internal carotid artery, and these may increase the risk of surgical complications in anterior spine approaches (Figure 17.2). They are more likely in congenital and in certain degenerative conditions.

The *posterior midline approach* to the spine is also common in spine surgery. It is used to address different conditions such as trauma, certain degenerative diseases and pathology of other posterior elements. Longitudinal midline exposure through the ligamentum nuchae is done with dissection carried out detaching muscular insertions from the spinous processes and lamina, retracting the muscular layers laterally to access the canal/foramina.

In cervical *decompressive surgery*, from posterior or anterior, at the C5 level, *C5 nerve palsy* is a known complication. Its aetiology is not completely understood, but it might be associated with a traction phenomenon of the shorter C5 nerve root with dorsal translation of the cord.

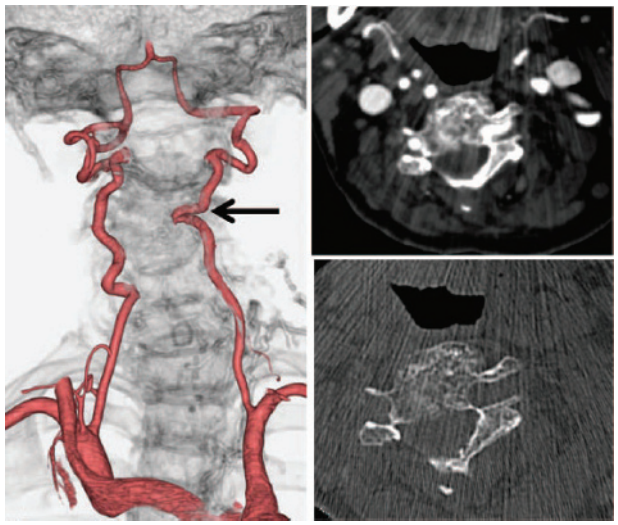


Figure 17.2 Anatomical variations of the course of the vertebral artery (Reproduced with permission from: Wakao N, *et al.* Risks for vascular injury during anterior cervical spine surgery: prevalence of a medial loop of vertebral artery and internal carotid artery. *Spine* 2016; **41**(4): 293–8.)

CLINICAL ASSESSMENT

Symptoms

Pain originating in the cervical spine can be due to pathology either at the disc, bone, articular or musculotendinous structures or at the neural structures (nerves or spinal cord). Pain is usually localized near the midline or around the shoulder girdle, but it can also radiate to the upper limb or the occipital region. A sudden onset of pain after exertion, exaggerated by coughing or straining and radiating down the arm/forearm is the typical clinical picture of a disc prolapse with cervical root irritation or compression, sometimes associated with paraesthesia in the same area of the upper limb. Pain in the cervical region can be direct, from an underlying condition, or referred, if caused by a pathologic condition at distance.

Referred neck pain can be muscular, developing secondarily as a result of postural adaptations to a primary pathology in the shoulder, the craniovertebral junction or at the temporomandibular joint.

Radiating pain down the arm/forearm can be caused by many pathologies besides herniated disc prolapse: peripheral entrapment syndromes, rotator cuff/shoulder pathology, brachial plexitis, *Herpes zoster*, thoracic outlet syndrome, sympathetic mediated pain syndrome, intraspinal or extraspinal tumours, epidural abscess and cardiac ischaemia.

Chronic or recurrent neck pain in older people is usually due to degenerative cervical spine pathology (i.e. *cervical spondylosis*), occurring as a result of ageing in the majority of the adult population. In this age group, the source of pain is multiple: from the degenerative disc itself, associated arthritis and synovitis of the facet joints and postural changes in the alignment of the cervical lordosis. It is crucial to define the *characteristics of pain* arising from the cervical region. Apart from the onset, type of pain, duration, precise localization and radiation, it is important to define the aggravating and alleviating factors, such as pain associated with any posture or movement.

Stiffness may be an associated symptom, either intermittent or continuous. The inability to move the neck, usually caused by pain and muscle spasm, can also be a spontaneous protective mechanism of the spine.

Numbness, tingling and weakness in the upper limbs may be due to irritation or pressure on a nerve root, but difficulty with hand *coordination*, cramping and weakness in the arms, hands and in the lower limbs, sometimes associated with an altered gait, may be the result of cord compression in the cervical spine.

Headache, especially occipital headache, sometimes originates from the cervical spine, but if this is the only symptom other causes should be ruled out.

Cervicogenic headache is a referred pain syndrome, usually unilateral in distribution, originating from various cervical structures innervated by the upper three cervical spinal nerves. They can be the atlanto-occipital joint, atlantoaxial joint, C2–3 facet joint, C2–3 intervertebral disc, myofascial trigger points and also the spinal nerves.

'Tension' is often mentioned as a cause of neck pain and occipital headache. The neck and back are common 'target zones' for psychosomatic illness and therefore cause undue tension (muscle spasm) in the posterior shoulder girdle or cervical spine.

Signs

Note the difference between the following:

- *radiculopathy* – a lower motor neuron lesion resulting from nerve root compression causing conduction impairment, expressing as sensory and motor deficits and diminished or absent reflexes at the involved level
- *radicular pain* – the result of nerve root irritation/inflammation and presents as a radiating pain down the upper limb
- *myelopathy* – an upper motor neuron lesion, expressing with hyperreflexia below the involved level.

Deformity in this region of the spine usually appears as a wry neck (or *torticollis*). The painful neck may be fixed in flexion or rotation or a combination of both.

The *clinical examination* of the neck is only complete with the examination of the upper trunk, upper limbs and shoulder girdle. The assessment of any anatomic region of the musculoskeletal system should have three phases – *look*, *feel* and *move* (Figure 17.4).

Look

Any *deformity* should be noted, assessing the neck from the front, from the side and from behind. Look for facial and shoulder *asymmetry*. Look for any scars or *lumps* in the supraclavicular fossa or on the midline. Note any asymmetry of the pupils, drooping eyelids and dry skin, characteristics of *Horner syndrome*.

Torticollis, due to muscle spasm, may suggest a disc lesion, an inflammatory disorder or cervical spine injury, but it also occurs with intracranial lesions and disorders of the eyes or semicircular canals. The '*cock robin*' posture describes the head tilted to the side. It is important to observe the prominence of the SCM, as it may give clues to the underlying cause. In congenital torticollis, the muscle bulk is tightened and shortened, prominent on the tilted side and in atlanto-axial subluxation it is prominent on the opposite side.

Neck stiffness is usually fairly obvious by the spine being 'splinted' due to muscle spasm.

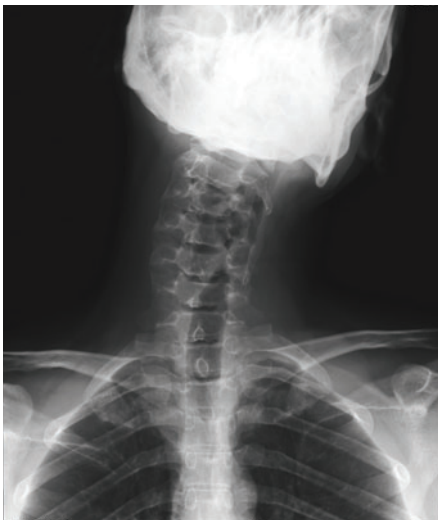


Figure 17.3 Disc prolapse A 39-year-old male with unremitting neck pain derived from cervical disc prolapse.

With the patient standing, look for unsteadiness and ask the patient to walk assessing the gait pattern.

Feel

The front of the neck is most easily palpated with the patient seated and the examiner standing behind. Always remember to *feel the neck from the four quadrants* – anterior, posterior and lateral (left and right). The best way to feel the back of the neck is with the patient lying prone and relaxed, allowing the *bony eminences* to be easily palpated. Feel for *tender spots* or *lumps* and note for paravertebral *muscle spasm*, particularly the posterolateral muscles and also assess the tension of the SCM.

Move

Start to assess active *range of motion* (Figure 17.5). Forward flexion, extension, lateral flexion and rotation are tested, and then shoulder movements. Range of motion normally diminishes with age, but even then movement should be smooth and pain-free. Remember that the *shoulder girdle* and the *cervical spine* are somehow synchronous in their movements – if one is injured and has a restricted range of motion, the other segment will have to compensate spontaneously. Very often we see patients who present with shoulder symptoms and subsequently develop neck pain and vice versa.

Enquire about any painful motion. Pain elicited by rotation and extension that is referred to the trapezium and shoulder blade area is very often due to facet joint pathology. Movement-induced pain and paraesthesia down the arm/forearm is particularly relevant for a herniated disc prolapse.

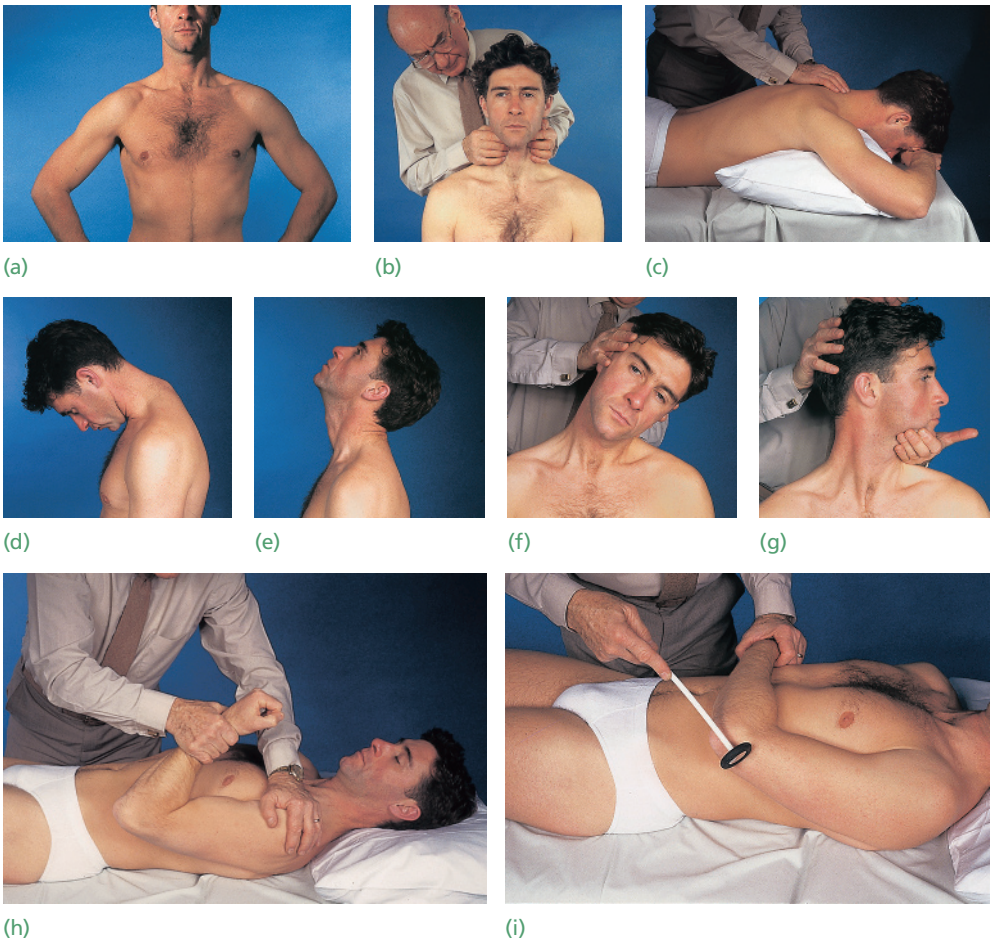


Figure 17.4 Examination (a) Look for any deformity or superficial blemish which might suggest a disorder affecting the cervical spine. (b) The front of the neck is felt with the patient seated and the examiner standing behind him. (c) The back of the neck is most easily and reliably felt with the patient lying prone over a pillow; this way muscle spasm is reduced and the neck is relaxed. (d–g) Movement: flexion ('chin on chest'); extension ('look up at the ceiling'); lateral flexion ('tilt your ear towards your shoulder') and rotation ('look over your shoulder'). (h,i) Neurological examination is mandatory.

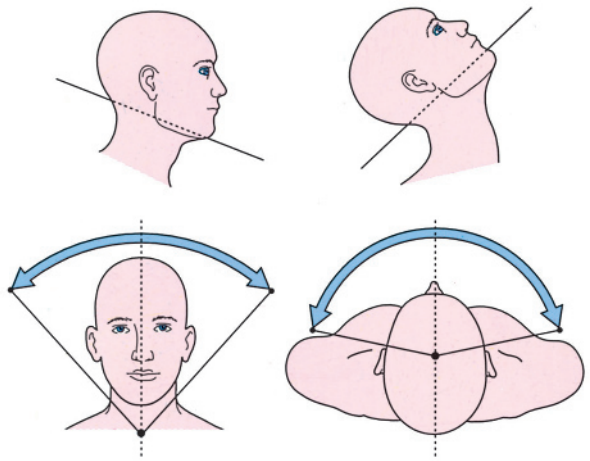


Figure 17.5 Normal range of motion Flexion and extension of the neck are best gauged by observing the angle of the occipitomental line – an imaginary line joining the tip of the chin and the occipital protuberance. In full flexion, the chin normally touches the chest; in full extension, the occipitomental line forms an angle of at least 45° with the horizontal, and more than 60° in young people. Lateral flexion is usually achieved up to 45° and rotation to 80° each way.

Neurological examination

Neurological examination of the upper limbs is mandatory in all cases. In some patients the lower limbs should also be examined. Muscle tone, power, sensation and reflexes should be carefully tested; even small degrees of asymmetry may be significant. Muscle power and sensation should be examined sequentially and bearing in mind the myotome and dermatome map. Test the C5 (biceps), C6 (brachioradialis) and C7 (triceps) reflexes.

Special tests

Tests for arterial compression If the thoracic outlet is tight, the radial pulse may disappear if, when the patient holds a deep breath, the neck is turned towards the affected side and extended (*Adson's test*), or if the shoulder is elevated and externally rotated (*Wright's test*).

The Spurling's test The patient is instructed to rotate the neck to one side with the chin elevated and laterally flexed, a position in which neural foramina are narrowed: if ipsilateral upper limb pain and paraesthesia are reproduced with axial compression of the head, the test is positive and that would increase the suspicion of a disc prolapse with cervical root compression. In these cases, pain may be relieved by the patient abducting the arm overhead (the abduction relief sign).

Tests for cervical myelopathy The following are physical findings suggestive of upper motor neuron compromise and cervical myelopathy:

- *Hoffmann's sign* – involuntary flexion of the thumb and index finger distal phalanx by flicking of the terminal phalanx of the middle finger
- *finger escape sign* – little finger abduction when the patient is asked to stretch his or her hands in front
- *finger fatigue test* – patient fatigues when asked to open and close his or her fists quickly
- *Lhermitte's sign* – an electric shock-like sensation along the spine if the spine is flexed
- *clonus* – rapid movements of the feet triggered by forceful passive motion of the ankle into dorsiflexion from a plantar position.

Assessment of peripheral nerve entrapments should also be carried out.

Imaging

Imaging examination should complement but never overcome the *clinical assessment* and should be directed at confirming or excluding a diagnosis.

X-rays

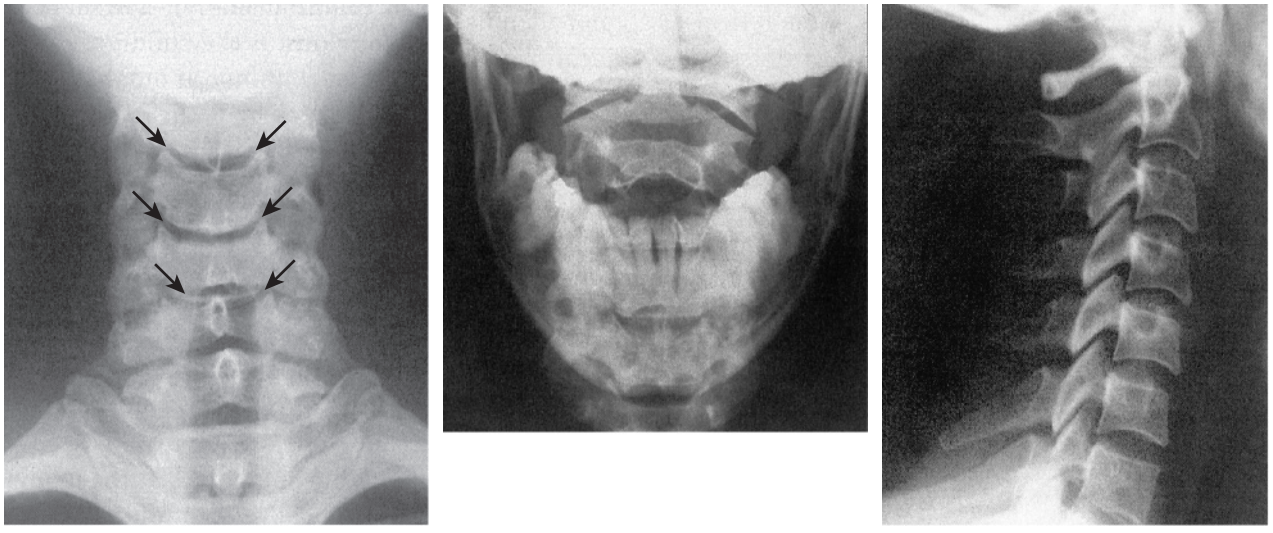
The *standard radiographic series* for the cervical spine comprises anteroposterior, lateral and open-mouth views (Figure 17.6). The lateral view should always include the base of the skull and the cervicothoracic junction, especially in the trauma case. Additional *lateral dynamic views* in flexion and extension can be obtained in the cooperative and neurologically intact patient. In the case of an acute neck injury, if needed, dynamic views should be obtained in the presence of the physician. *Oblique views* can also help, especially in the trauma scenario.

The *anteroposterior view* should show the regular, undulating outline of the lateral masses; destructive lesions or fractures may disturb its symmetry. The alignment of the spinous processes should be in a straight line.

An *open-mouth view* is required to show the axis and the atlantoaxial junction. The lateral margin of the atlas should align with the lateral margin of the axis and the space on each side of the dens should be equal, if the neck is not rotated.

The *lateral view* should include all seven vertebrae; there have been cases of serious spinal injuries because a fracture-dislocation at C6–C7 or C7–T1 was missed. The normal lateral view of the cervical spine shows *four parallel lines*: one along the anterior surfaces of the vertebral bodies, *one* along their posterior surfaces, *one* along the bases of the spinous processes, and *one* along the tips of the spinous processes; any malalignment suggests subluxation. The *disc spaces* are inspected; loss of disc height, the presence of osteophytic spurs at the margins of adjacent vertebral bodies and inversion of the natural lordosis suggest intervertebral disc degeneration. The *posterior interspinous spaces* are compared; if one is wider than the rest, this may signify chronic instability of that segment, possibly due to a previously undiagnosed subluxation. The direction of the *spinous processes* should be confluent in an imaginary point on the concave side of the spine. Flexion and extension views may be needed to demonstrate instability (Figure 17.7).

Children's X-rays have special particularities to be considered. Because the ligaments are relatively lax and the bones incompletely ossified, flexion views may show unexpectedly large shifts between adjacent vertebrae. The normal lateral X-ray of the child may show



(a) (b) (c)

Figure 17.6 Imaging – normal X-rays (a) Anteroposterior view – note the smooth, symmetrical outlines and the clear, wide uncovertebral joints (arrows). (b) Open-mouth view – to show the odontoid process and atlantoaxial joints. (c) Lateral view – showing all seven cervical vertebrae.

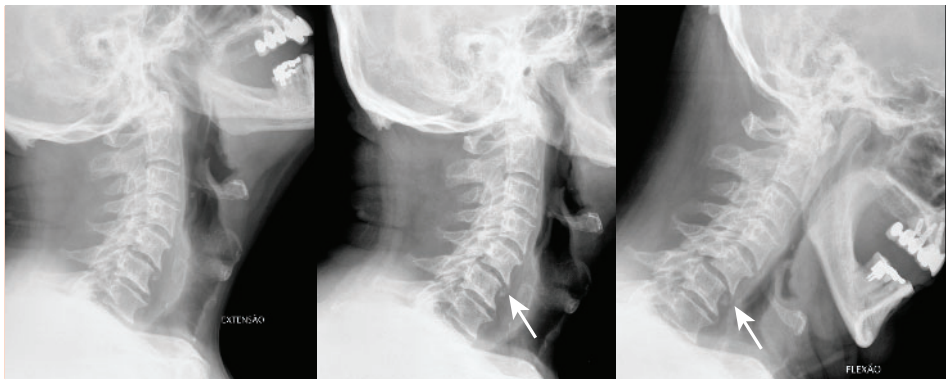


Figure 17.7 Imaging – dynamic X-rays Dynamic X-ray views of a 65-year-old male with a traumatic C5–C6 disc lesion. Note the instability at the disc level with anterolisthesis of C5 over C6 on hyperflexion.

an *atlantodental interval* of 4–5 mm (which in an adult would suggest rupture of the transverse ligament) or an *anterior pseudo subluxation at C2–C3 or C3–C4* of up to 3 mm. Note also that the *retropharyngeal space* between the cervical spine and pharynx at the level of C3 increases markedly on forced expiration (for example, when crying) and this can be misinterpreted as a soft-tissue mass. However, the increase in the prevertebral cervical space in the context of trauma should raise a red flag and demand further studies (CT or MRI) to rule out an underlying unstable traumatic lesion both in children and adults. Another error is to mistake the *normal synchondrosis between the dens and the body* of C2 (which only fuses at about 6 years) for an odontoid fracture. Finally, remember that normal radiographs in children do not exclude the possibility of a spinal cord injury.

Computed tomography

CT of the cervical spine provides excellent osseous detail. It is useful to demonstrate the shape and size of the spinal canal and intervertebral foramina, as well as the integrity of the bony structures. It is particularly helpful for the imagological assessment of axial and subaxial cervical spine trauma (Figure 17.8).

CT also has a high performance for the measurement of the anatomical features as part of routine preoperative workup and planning. However, the amount of radiation for a CT scan is not negligible and this should be taken into account when the decision is made to request such an examination in a child.

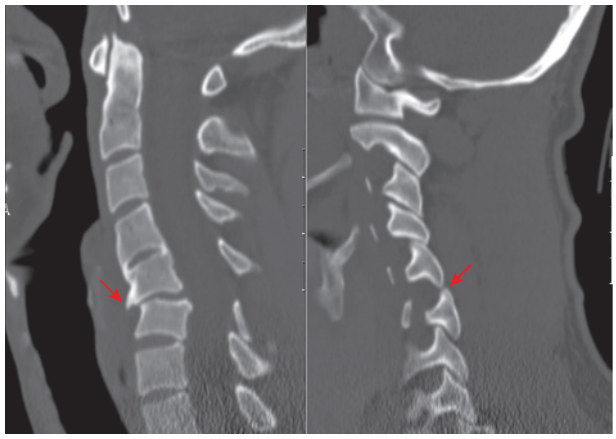


Figure 17.8 Imaging – CT scan A 39-year-old male with a C5–C6 unilateral locked facet well demonstrated in a sagittal CT scan frame.

Myelography

Changes in the contour of the contrast-filled thecal sac suggest intradural and extradural compression. However, this is an invasive investigation and fairly non-specific. Its usefulness is enhanced by performing a post-contrast CT scan. Due to its invasiveness and contrast side effects, it is seldom used routinely at present. Myelography can be substituted by modern CT scan techniques.

Magnetic resonance imaging

MRI is non-invasive, does not expose the patient to radiation and provides excellent resolution of the soft tissue, such as the intervertebral disc and neural structures (Figure 17.9). It is very sensitive for

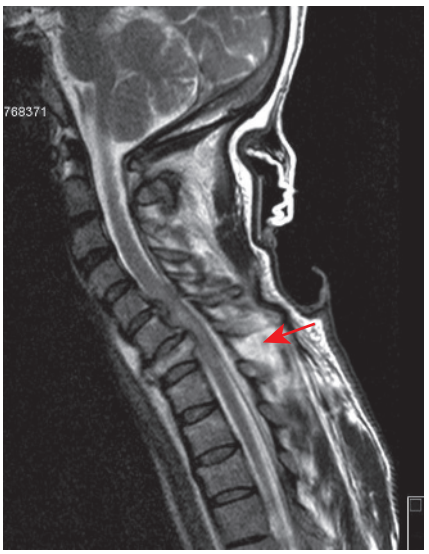


Figure 17.9 Imaging – MRI scan A 41-year-old female with a C7 fracture and associated C5–C6 disc prolapse. The role of MRI to assess the posterior ligamentous injuries is, nevertheless, associated with an important percentage of false positives.

demonstrating tumours and infection/inflammation. It also provides information on the size of the spinal canal and neural foramina. Its sensitivity can be a drawback: 20% of asymptomatic patients show significant abnormalities and the scans must therefore be interpreted in conjunction with the clinical picture. In the trauma scenario it can help to determine the compromise of the posterior ligamentous structures, acute lesions of the intervertebral disc and the presence of oedema in the spinal cord.

CERVICAL SPINE ABNORMALITIES IN CHILDREN

DEFORMITIES AND CONGENITAL ANOMALIES

A variety of deformities of the neck are encountered in children, some reflecting postural adjustments to underlying disorders and others a clinical manifestation of developmental anomalies.

Torticollis and related syndromes

TORTICOLLIS

Torticollis is a cervical deformity in which the head is rotated and tilted towards one side with some lateral flexion, the so-called '*cock-robin*' position. The SCM muscle is 'shortened' and may feel tight and hard.

It is often a presenting feature of a *congenital* osseous cervical spine anomaly, particularly of the atlas, but it can also be *acquired* and the presenting sign of a tumour (for example, eosinophilic granuloma), infection (for example, discitis, lymphadenitis or, rarely, caused by an ear or upper respiratory tract infection) or a cervicothoracic scoliosis. There is often a history of trauma, although it can be triggered by simple neck rotation. In up to 25% of cases, no underlying cause is identified.

In children and adolescents, acute torticollis is characterized by atlantoaxial rotational subluxation of sudden onset.

The correct workup of a child who presents with torticollis should include a cervical spine X-ray, and a CT scan should be considered on occasions.

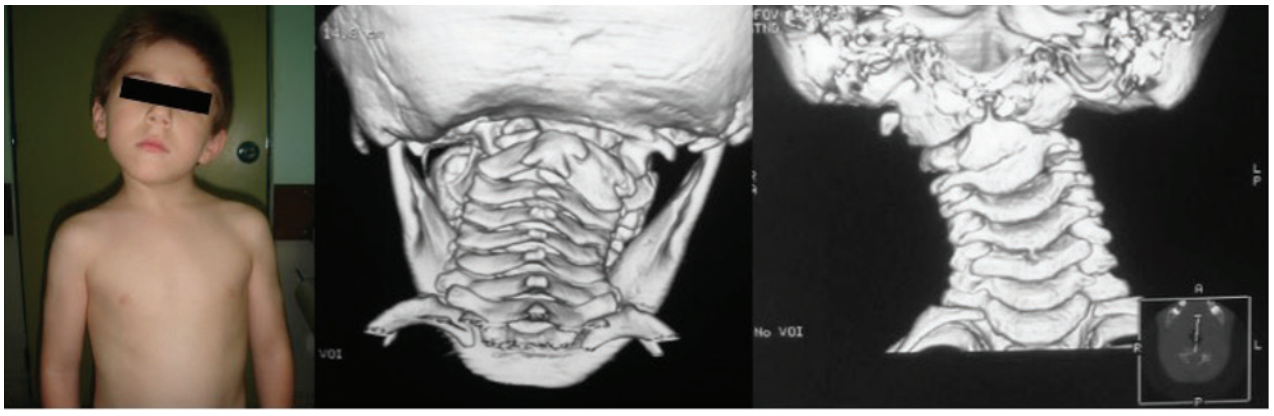
INFANTILE (CONGENITAL) TORTICOLLIS

This is a common disorder in neonates and infants in which one of the SCM muscles is fibrous and fails to elongate as the child grows, resulting in a progressive deformity with a reported incidence of approximately 1%. Although the aetiology is unclear, it may be associated with intrauterine packaging disorders or the result of a birth injury causing localized ischaemia. A history of difficult labour or breech delivery is common.

Clinically, a lump can be visible in the first few weeks after birth, disappearing within a few months. No deformity or obvious limitation of movement may be apparent until the child is 1–2 years old.

Along with the classical visible deformity of the neck, with the head tilted towards the affected side and the face rotated towards the contralateral shoulder so that the ear approaches the shoulder (Figures 17.10 and 17.11), an asymmetry of the face (hemihypoplasia) and plagiocephaly may be noticeable. These features can worsen and become more obvious as the child grows.

Treatment Most children have a complete spontaneous resolution with time, but some cases may require physiotherapy. If the diagnosis is made early, daily muscle stretching may prevent the incipient deformity.



(a) (b) (c)

Figure 17.10 Congenital torticollis (a) Clinical picture of a young child with congenital torticollis. Note the head tilting towards the left shoulder with slight rotation to the contralateral side. (b,c) 3D reconstruction of CT images of the same child showing atlantoaxial fusion.

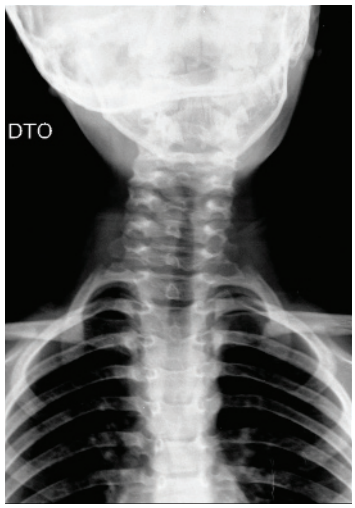


Figure 17.11 Congenital torticollis AP view of the cervical spine of a child with congenital torticollis. Note the head tilting towards the right shoulder with slight rotation to the contralateral side.

The benign SCM lump can completely disappear. However, the clinician should be aware of other causes such as tumours and cysts in the neck, which may need surgical excision. If the condition persists beyond 1 year, operative correction is required to avoid progressive facial deformity (Figure 17.12).

SECONDARY TORTICOLLIS

Childhood torticollis, as an acquired condition, has several aetiologies. It may be secondary to *infection* (lymphadenitis, retropharyngeal abscess, discitis, tuberculosis), *tumours* (posterior fossa, intraspinal tumours), *inflammatory disorders* (juvenile rheumatoid arthritis), *neurogenic causes* (benign paroxysmal torticollis) or *trauma* and can also be *idiopathic*.

Atlantoaxial rotatory subluxation Atlantoaxial rotatory fixation is a pathological displacement of the atlas on the axis in a position that is normally accomplished during head rotation. It can be associated with minor trauma or with a recent nasopharyngeal infection, tonsillectomy or even a retropharyngeal abscess (*Grisel's syndrome*). It can present with an acute onset or after a period of weeks. In the acute setting, there is pain and muscle spasm. In fixed deformities, pain subsides but motion is restricted and the child cannot correct the deformity.

The mechanism behind *Grisel's syndrome* is not completely understood, but anatomical factors permit that inflammation of the pharynx can lead to *attenuation of atlantoaxial ligaments* or the synovium. The chin is shifted laterally or laterocaudally and the head fixed in this position. Early diagnosis and therapy are crucial to prevent neurological complications caused by compression of the medulla oblongata by the dislocated odontoid.

Plain X-ray interpretation may be challenging. Open-mouth views should be obtained. *CT scans* in both neutral and maximum lateral rotation are the most helpful investigation.

Most cases are mild and can be managed expectantly with a soft collar and analgesics. If there is no resolution after a week, halter traction (Figure 17.13a), bed rest and analgesics should be prescribed. In this setting physiotherapy may be contraindicated. Attempts for manual reposition without general anaesthesia are not tolerated. In more resistant cases, halo traction may be required. Occasionally, if the articulation remains unstable, subluxation persists or recurs easily or if there is neurological compromise, then a C1–C2 fusion is recommended.

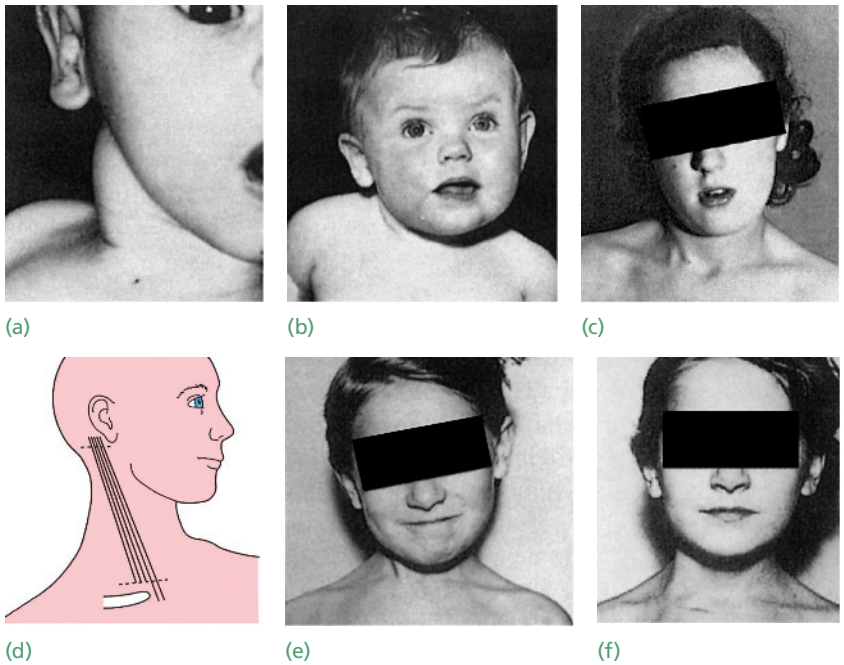
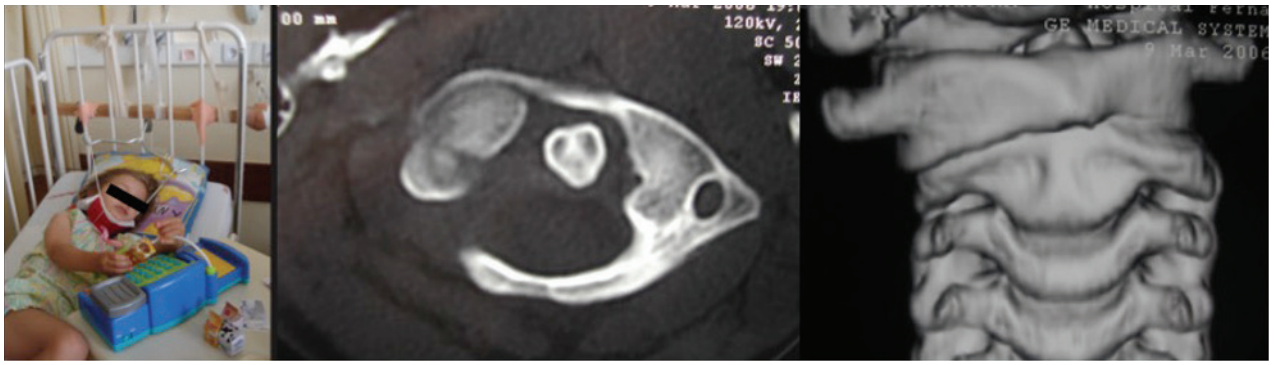


Figure 17.12 Torticollis Natural history: (a) sternomastoid tumour in a young baby; (b) early wry neck; (c) deformity with facial hemiatrophy in the adolescent. Surgical treatment: (d) two sites at which the sternomastoid may be divided; (e,f) before and a few months after operation.



(a) (b) (c)

Figure 17.13 Atlantoaxial rotary subluxation (a) A child with atlantoaxial subluxation on halter traction. Axial (b) and 3D reconstruction images (c) of C1–C2 rotatory subluxation.

Vertebral abnormalities

Congenital osseous cervical spine anomalies are rare, but their detection and complete diagnosis are needed in order to be able to establish a prognosis and treatment, as these deformities are often associated with instability and potential neurological injury associated with spinal cord encroachment. Most cases are innocuous and may go undetected throughout life, with some being diagnosed only when a serious complication occurs.

Mutations of *homeobox genes* may be responsible for congenital osseous anomalies of the cervical spine that probably arise during somatogenesis. The *occiput*, *atlas* and axis are formed by a separate mechanism from that responsible for the other vertebral bodies. The *remaining subaxial cervical vertebrae* develop in a manner similar to the rest of the spine. Failures of segmentation from the third to eighth weeks of fetal life can lead to several fusion defects, such as fusion of C1 to the occiput or C2–C3. These defects can be associated with congenital malformations of other organ systems, such as the kidneys and the heart.

Neurological signs and symptoms (head and neck pain, visual and hearing deficits, weakness and numbness in the extremities, long tract and posterior column signs, ataxia and nystagmus) can present with various anomalies including occipitalization of the atlas, basilar invagination, *os odontoideum* and chronic atlantoaxial dislocation.

Although imaging through conventional radiographs may be enough for the diagnosis, CT scan is the gold standard imaging for classifying this type of abnormalities. However, taking into account the high rate of associated underlying neural abnormalities, all these cases should also be screened with MRI.

Occipitoatlantal instability

Instability at the occipitoatlantal joint has been described after trauma to the cervical spine and in association with Down's syndrome, familial cervical dysplasia and hyperlaxity syndromes. Symptoms of non-traumatic occipitoatlantal instability can include neck pain, headache, torticollis and weakness as well as vertebrobasilar symptoms such as nausea, vomiting and vertigo.

Arthrodesis of the occiput to the atlas for all patients with non-traumatic occipitoatlantal instability is recommended.

Klippel-Feil syndrome

This rare developmental disorder is caused by a failure of segmentation of the cervical somites during the third to eighth week of embryogenesis, resulting in *fusion of at least two cervical segments.* Congenital fusion can occur at any level in the cervical spine, but approximately 75% occur in the upper cervical spine.

Klippel-Feil is often associated with other *skeletal and extraskeletal abnormalities* such as scoliosis (60%), renal abnormalities (35%, most commonly unilateral renal agenesis), Sprengel deformity (30%), deafness (30%) and congenital heart disease (14%, most commonly ventricular septal defect). There is also an important association with fetal alcohol syndrome. Other associated deformities include hand anomalies such as syndactyly, thumb hypoplasia and extra digits.

The classical clinical triad of children with synostosis is short neck with various degrees of neck webbing, low posterior hairline and limitation of neck mobility (Figure 17.14). Nevertheless, less than 50% have all these findings. Furthermore, there is often compensatory hypermobility on the mobile adjacent segments.

Symptoms tend to arise in the second or third decades, not from the fused segments but from the adjacent mobile segments, and they are related to the extension of involvement of the spine and the presence of other anomalies. The most consistent clinical finding is a limited range of motion of the neck, especially lateral bending. There may be pain due to joint hypermobility or neurological symptoms from instability.

(a) (b)

Figure 17.14 Klippel–Feil syndrome Clinical pictures of a young child with Klippel–Feil syndrome. Note the presence of the typical features: short neck, low posterior hairline and a wry neck.

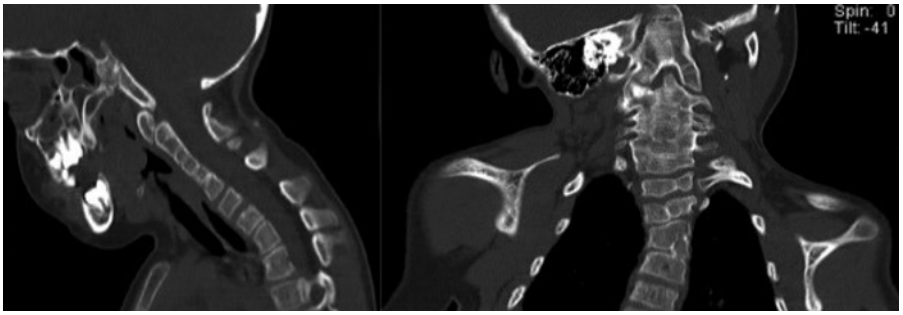


Figure 17.15 Klippel–Feil syndrome CT images of a child with congenital torticollis due to Klippel–Feil syndrome. Note the presence of several cervical fused vertebral bodies.

Imaging

X-rays and *CT scans* reveal fusion of two or more cervical vertebrae (Figures 17.15 and 17.16). The vertebrae are also often widened and flattened (so-called 'wasp-waist appearance', which is considered pathognomonic).

All patients with Klippel–Feil syndrome should have an *ultrasound evaluation of the renal system*.

Treatment

For asymptomatic patients, treatment is unnecessary but parents should be warned of the risks of contact sports. Sudden catastrophic neurological compromise can occur after minor trauma. Children with symptoms may need cervical fusion.

Basilar impression

Basilar impression or invagination is a disease of the atlantoaxial facet joints causing progressive *vertical instability* so that the floor of the skull is indented by the upper cervical spine, usually the odontoid, which may sit within the foramen magnum and impinge upon the brainstem (Figure 17.17).

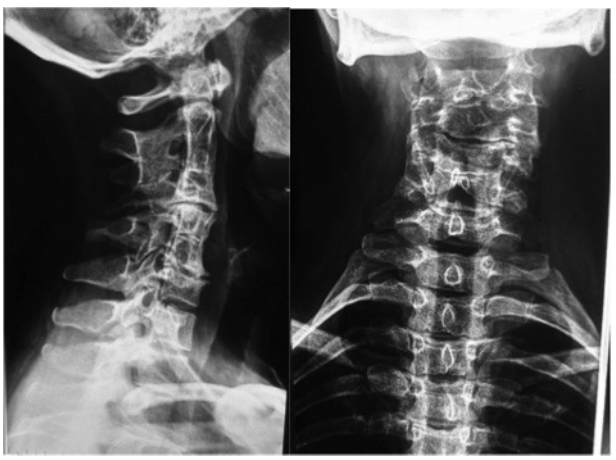


Figure 17.16 Klippel–Feil syndrome Lateral and AP X-ray views of a 55-year-old patient with Klippel–Feil syndrome. Note the presence of several cervical fused vertebral bodies and also the degenerative changes at adjacent levels.

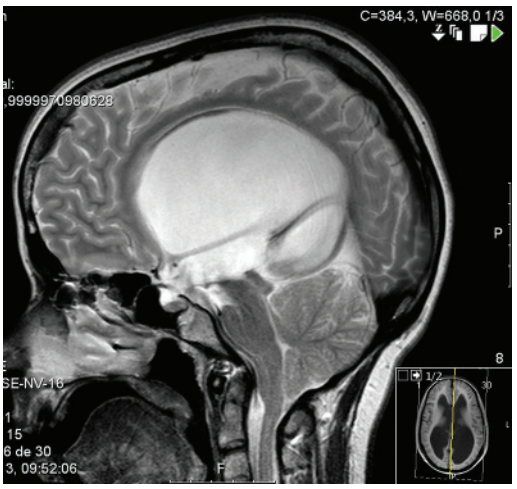


Figure 17.17 Basilar impression An example of a patient with basilar impression. Note the close relation between the tip of the odontoid and the *medulla oblongata*.

Basilar invagination can be congenital or acquired. More commonly, *primary invagination* occurs in association with occipitoatlantal fusion, hypoplasia of the atlas, a bifid posterior arch of the atlas, odontoid anomalies, *Morquio syndrome*, *Klippel–Feil syndrome* and *achondroplasia*. *Secondary basilar impression* is seen in association with conditions such as osteomalacia, rickets, *osteogenesis imperfecta*, *Paget's disease*, neurofibromatosis, skeletal dysplasia and degenerative destructive osteoarthropathies affecting the craniovertebral junction (for example, rheumatoid arthritis). Basilar impression is frequently associated with other congenital neurological anomalies, such as *Arnold-Chiari malformation* and syringomyelia.

Children usually present with a short neck, facial asymmetry and torticollis, which are not pathognomonic. Neurological signs and symptoms may not present until the second or third decade of life and may be precipitated by minor trauma. They are usually related to compression of the neural elements and the medulla oblongata at the level of the foramen magnum or can result from raised intracranial pressure (because the aqueduct of Sylvius becomes blocked). Patients may present with neck pain, headaches in the distribution of the greater occipital nerve, cranial-nerve involvement, ataxia, vertigo, nystagmus, weakness and paraesthesia of the limbs and even sexual dysfunction.

Imaging

Several *craniometric parameters* such as McRae's line, Chamberlain's line and McGregor's line have been described to quantify the relationship between the odontoid process and the foramen magnum. *Chamberlain's line* is defined from the posterior lip of the foramen magnum (the opisthion) to the dorsal margin of the hard palate. *McGregor's line*, the best for screening purposes as the landmarks are clearly seen on the lateral radiograph at all ages, is drawn from the upper surface of the posterior edge of the hard palate to the most caudal point of the occipital curve of the skull. *McRae's line* defines the opening of the foramen magnum. Patients in whom the odontoid is above this line will probably be symptomatic.

When these reference points are not well defined on radiographs, *CT and MRI scans* can be used to confirm the diagnosis.

Treatment

Treatment depends on the degree of neural compression and reducibility of the deformity and involves surgical decompression and stabilization with a posterior occipitocervical arthrodesis. If the symptoms are the result of a compressive aberrant odontoid that cannot be reduced, odontoidectomy may be indicated.

Odontoid abnormalities

Several odontoid abnormalities exist in which the odontoid may be absent, hypoplastic or a separate ossicle. *Ossiculum terminale persistens* is the term for an unfused apical dental segment. *Os odontoideum* is the term for an unfused basal odontoid to the axis body. *Os avis* is the term for a rare resegmentation error in which the apical dental segment is attached to the basion on the occiput and not to the dens. The C1–C2 joint has flat lateral articulations, weak posterior ligaments and the ligamentum flavum is replaced with a thin atlantoaxial membrane.

Anomalies of the odontoid are more common in patients with *Down's syndrome*, *Klippel–Feil syndrome*, multiple epiphyseal dysplasia and other skeletal dysplasia, and they should be suspected in this setting. This is especially important in patients undergoing operation, as the atlantoaxial joint may subluxate during general anaesthetic procedures.

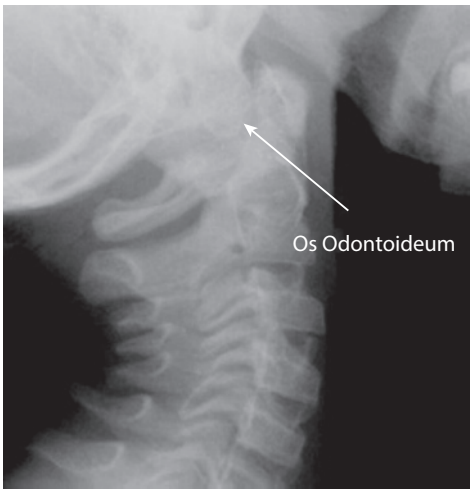
Os odontoideum

This condition refers to an independent osseous structure cephalad to the body of the axis. Its position can be in the normal location of the odontoid process (orthotopic) or rostrally displaced (dystopic). CT or MRI scans can confirm the diagnosis. The aetiology is not clearly defined, but it can be congenital or post-traumatic. Three types of os odontoideum have been described: round, cone and blunt tooth. The severity of myelopathy seems to be correlated with the round type.

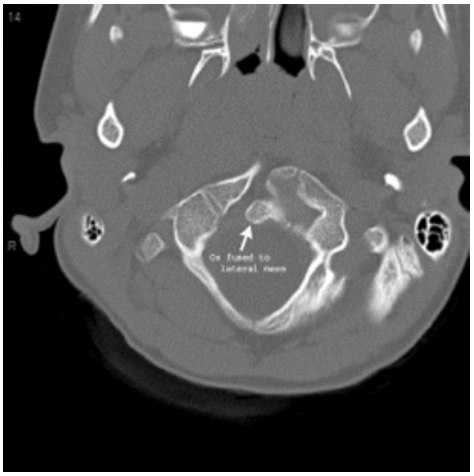
The os odontoideum accompanies the atlas during the normal flexion-extension motion and leads to biomechanical insufficiency of the apical odontoid and alar ligaments, which in turn can result in instability under physiological loads. *Translational instability and dislocation* result in posterior spinal cord compression. Vertical instability is also possible with invagination of the dens towards the skull with brainstem compression and subsequent neurological injury, including respiratory paralysis. Long-standing instability may become multidirectional, allowing the C1–C2 unit to become very unstable.

Signs and symptoms are the same as those described for other anomalies of the odontoid.

In the majority of cases the anomaly is discovered accidentally in a routine cervical spine X-ray, revealing as a wide radiolucent gap between the odontoid and the body of the axis (Figure 17.18). Note that the normal vestigial disc space between the dens and the body of the axis may be visible as a radiolucent line until 5 years of age. Open-mouth radiographs show the abnormality and lateral flexion-extension views may show C1–C2 instability with motion between the odontoid and the body of the axis. The degree of C1–C2 instability does not correlate with the severity of the neurological



(a)



(b)

Figure 17.18 Os odontoideum (a) Lateral view of the cervical spine showing the os odontoideum, and (b) an axial slice of a CT scan showing fusion of the os odontoideum to the left lateral mass of the atlas. (Reproduced with permission from: Hosalkar H, *et al.* Congenital osseous anomalies of the upper cervical spine. *J Bone Joint Surg* 2008; **90**: 337–48.)

deficits, but a space available for the cord in extension views of 13 mm is defined as critical.

The natural history of asymptomatic os odontoideum is unclear. Symptomatic patients should have surgical stabilization, which consists of C1–C2 posterior fusion with or without decompression. Prophylactic treatment of asymptomatic patients is controversial.

Atlantoaxial instability

The atlantoaxial unit contributes to the majority of the neck rotation movement and is the most mobile segment of the spine, although it is structurally weak. Simultaneously, it has specific stabilizing structures that prevent excessive motion and disarrangement. The articulation between the atlas and axis comprises one midline atlanto-odontoid joint and two lateral atlantoaxial facet joints. The articular capsules of the lateral facets provide stability and are reinforced by important ligaments, such as the alar ligaments and the transverse atlantal ligament, which is the thickest and the primary stabilizer of the atlas against anterior subluxation. The transverse ligament allows rotation, while the alar ligaments prevent excessive rotation. The apical ligament has an accessory or vestigial role.

Congenital osseous anomalies in this region, such as occipitalization of the atlas, os odontoideum and basilar invagination, can lead to an increased risk of segmental instability and neurological compromise. Isolated laxity of the transverse atlantal ligament is a diagnosis of exclusion in the setting of chronic atlantoaxial dislocation without a predisposing cause. The end result is spinal canal encroachment and neurological impingement.

Atlantoaxial instability is an uncommon disease in children and is significantly more prevalent in Down's syndrome, occurring in up to 40% of all patients, but it is rarely symptomatic. This abnormality is thought to be secondary to the laxity of the transverse ligament and to the bony anomalies encountered in these patients.

Patients rarely become symptomatic before the third decade of life. With age, atlantoaxial articulation becomes more vulnerable and the central nervous system becomes less tolerant of intermittent compression. Some patients may be misdiagnosed with other conditions that mimic the puzzling clinical picture, including multiple sclerosis and amyotrophic lateral sclerosis.

Imaging

Important *radiological parameters* are defined to characterize this condition. The small lucent space between the anterior aspect of the odontoid and the posterior surface of the anterior arch of the atlas is the *atlantodental interval (ADI)*. The space from the back of the odontoid to the anterior aspect of the posterior arch of C1 is defined as the *space available for spinal cord (SAC)*. Generally, more than 10 degrees of flexion at C1–C2 indicates subluxation. An ADI of >3 mm in adults suggests transverse ligament insufficiency. An ADI of >4 mm on lateral flexion-extension radiographs of the immature cervical spine indicates instability. In patients with Down's syndrome instability is defined as an ADI of >7 mm.

CT and MRI scans can help in defining the diagnosis.

Routine radiographic examination of the cervical spine of Down's syndrome patients is recommended, especially in the context of sports participation.

Treatment

The surgical treatment of paediatric patients with atlantoaxial instability is challenging, especially in children with Down's syndrome. C1–C2 fusions are indicated for patients showing >5 mm of instability on flexion-extension views and those with severe cervical cord compression.

CERVICAL SPINE ANOMALIES IN ADULTS

DEGENERATIVE PATHOLOGY

Cervical degenerative disc disease occurs with advancing age, leading to structural changes of the intervertebral discs, including herniation and disc space narrowing.

Intervertebral disc degeneration affects the majority of the population over 60 years of age, but it is predominately asymptomatic. The main symptom associated with cervical spine degenerative disease is neck pain, which has a reported incidence of 30% in the general population. Cervical degenerative disc disease can also present as radiculopathy or myelopathy, as a result of compression of nerve roots or the spinal cord.

Imaging

MRI is the most sensitive method for the assessment of disc pathology. T2-weighted images are more sensitive than T1-weighted images for detecting disc degeneration. Miyazaki and colleagues proposed a grading system for the severity of intervertebral disc degeneration consisting of five grades (Table 17.1).

Treatment

Most patients are managed conservatively. Surgical treatment is reserved for patients with persistent or worsening of symptoms and usually involves partial removal of the extruded disc or fusion. Anterior cervical discectomy and fusion (ACDF) remains the most common surgical strategy for single- or double-level disease. Non-fusion alternatives include posterior foraminotomy, cervical disc arthroplasty or replacement (CDR), laminectomy with fusion and laminoplasty. Motion preservation will theoretically lessen the incidence of adjacent segment degeneration.

ACUTE INTERVERTEBRAL DISC PROLAPSE

Acute disc prolapse is not as common in the neck as in the lower back. The mechanical environment in the cervical region is more favourable than in the lumbar region although the pathological features are similar.

The acute prolapse of the cervical intervertebral disc may be precipitated by local strain or injury, especially sudden unguarded flexion and rotation, and it usually occurs immediately above or below the sixth cervical vertebra. In many cases (perhaps in all) there is a predisposing abnormality of the disc with increased nuclear tension. The extruded disc material migrates posteriorly into the spinal canal and may press on the posterior longitudinal ligament or compress the dura or the nerve roots. Intradural disc herniation has also been reported although this is a rare type.

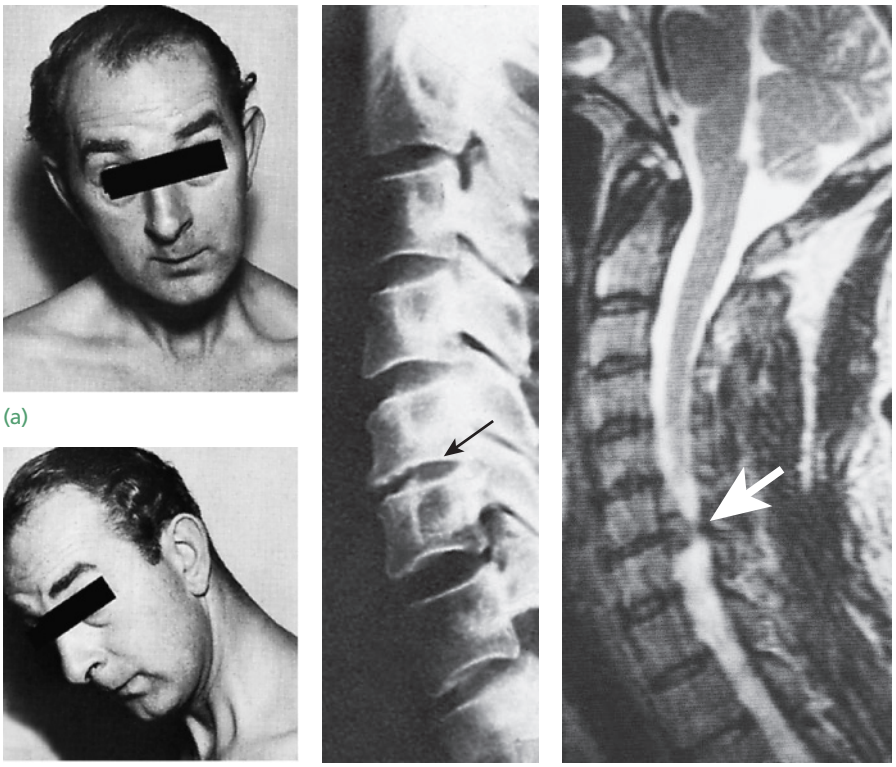
Clinical features

Unilateral or rarely bilateral arm pain is the main presentation symptom of cervical disc herniation, and it can be associated with variable degrees of neck pain and stiffness. The herniated nucleus pulposus in the spinal canal causes nerve irritation and pressure on the nerve roots. This can result in radiculopathy with paraesthesia or hypoesthesia, usually in the distribution of C6 or C7 (outer elbow, back of the wrist and the index and middle fingers), decreased reflexes and motor weakness, although this is a rare finding taking into account that the most commonly affected levels are C5–C6 and C6–C7 (Figure 17.19). Patients may sometimes complain of pain radiating to the scapular region or to the occiput, usually by compromise of the upper cervical nerve roots. On clinical examination there may be a painful wry neck (torticollis), muscle spasm and tenderness with restricted range of motion.

Table 17.1 Grading system for cervical intervertebral disc degeneration

Grade	Nucleus signal Intensity	Nucleus structure	Distinction of nuclear and annulus	Disc height
I	Hyperintense	Homogenous, white	Clear	Normal
II	Hyperintense	Inhomogenous with horizontal band, white	Clear	Normal
III	Intermediate	Inhomogenous, gray to black	Unclear	Normal to decreased
IV	Hypointense	Inhomogenous, gray to black	Lost	Normal to decreased
V	Hypointense	Inhomogenous, gray to black	Lost	Collapsed

Source: Miyazaki M, *et al*. Reliability of a magnetic resonance imaging-based grading system for cervical intervertebral disc degeneration. *J Spinal Disord Tech* 2008; **21**(4): 288–92.



(a) (b) (c) (d)

Figure 17.19 Acute disc prolapse (a,b) Acute wry neck due to a prolapsed disc. (c) The intervertebral disc space at C5–C6 is reduced. (d) MRI in another case showing a large disc prolapse at C6–C7.

Acute onset of symptoms can be related to a specific strain episode, such as acute flexion of the neck during intense physical exertion or a 'whiplash' injury. Subsequent attacks may be sudden or gradual in onset and triggered by trivial causes. Between attacks the pain subsides or alleviates, although residual axial pain may persist, along with slight neck stiffness.

Upper-limb neurological examination should be complete. The *C6 nerve root* innervates the biceps reflex, the biceps muscle and wrist dorsiflexion and sensation of the lateral forearm, thumb and index finger. *C7 nerve root* innervates the triceps and radial reflexes, the triceps muscle, wrist flexors and finger extensors and sensation in the middle finger. Rotation, tilting of the neck to the affected side and axial compression, as elicited by the Spurling manoeuvre, may trigger radicular symptoms, as does the Valsalva manoeuvre.

Imaging

X-rays may reveal loss of the normal cervical lordosis (due to muscle spasm) and disc space narrowing. The most sensitive imaging exam is *MRI*, which will reveal the extruded material or protruded disc and its relationship to the cord or nerve root in most cases (Figure 17.20).

Differential diagnosis

Acute soft-tissue strain Acute neck strain is often associated with pain, stiffness and vague 'tingling' sensation in the upper limbs. It is important to bear in mind that pain radiating into the arm is not necessarily due to nerve root compromise.

Neuralgic amyotrophy This condition can closely resemble an acute disc prolapse and should always be thought of if there is no definite history of a strain episode. Pain is sudden and severe, and localized over the shoulder girdle rather than in the neck itself. Careful examination will show that more than one neural level is affected – an extremely rare event in disc prolapse.

Cervical spine infections Pain is unrelenting and local spasm severe. X-rays may show erosion of the vertebral end plates and disc space narrowing.

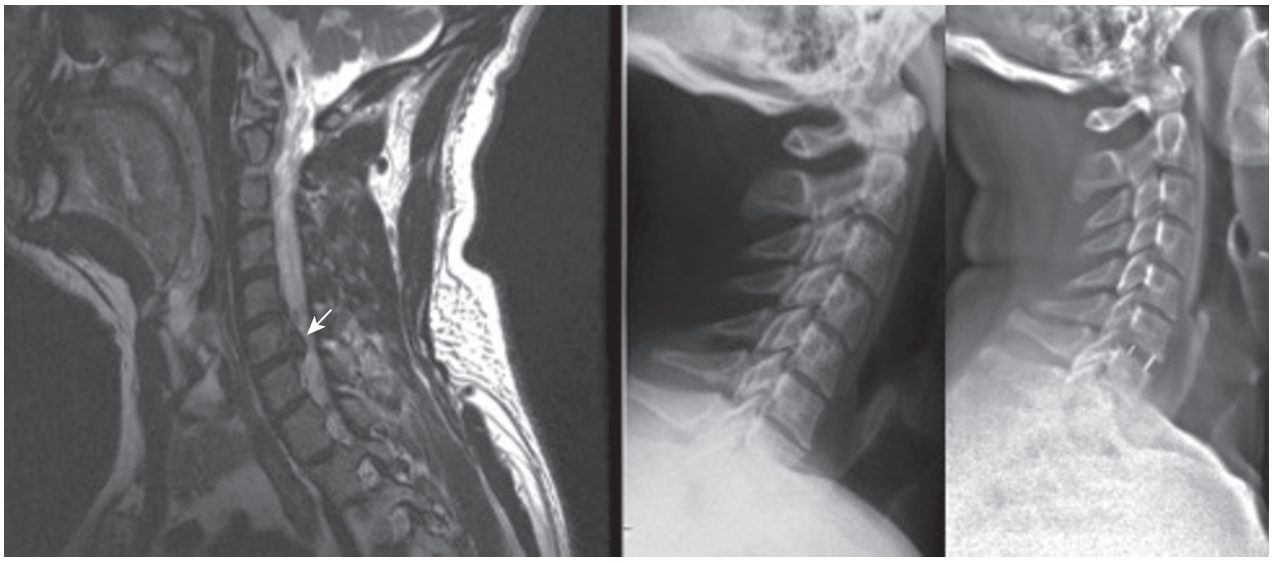
Cervical tumours Neurological signs may be progressive and unremitting and X-rays may reveal bone destruction. Increasing night pain is usually one of the alarming features.

Rotator cuff lesions Although the distribution of pain may resemble that of a prolapsed cervical disc, tenderness is localized to the lateral aspect of the shoulder and arm (typically never radiates below the elbow) and shoulder movements are abnormal.

Treatment

CONSERVATIVE TREATMENT

Conservative treatment often consists of patient education, heat, non-steroidal anti-inflammatory



(a) (b) (c)

Figure 17.20 Disc herniation – MRI (a) Sagittal T2 sequence MRI of the cervical spine of a patient with C6–C7 disc herniation. X-ray images of the patient showing (b) preop features with disc collapse and pain-causing flattening of the cervical lordosis and (c) postop lateral X-ray view with the physiological lordosis restored.

medication, oral corticosteroids, corticosteroid injections, rest, cervical collar (seldom needed for more than a week or two) and physical therapy.

Traction applied intermittently for no more than 30 minutes at a time may improve the radiating pain. A 'distraction' cervical collar can also be worn. Most patients recover completely with conservative treatment.

OPERATIVE TREATMENT

When conservative treatment fails to relieve the pain or there are severe progressive symptoms, including a progressive neurological deficit, then surgical treatment is indicated. There is no consensus on the duration of conservative therapy before surgery is indicated. Motor deficit and myelopathy caused by spinal cord compression are absolute indications for surgery. Prior changes on MRI signal intensity in the spinal cord seems to be an important risk factor for delayed recovery after surgical decompression. The main purpose of surgery is to relieve the pain, improve the clinical picture and stop progression of the neurological deficit by removing the compression on the nerve root.

ACDF has been the standard treatment, in which the disc is removed through an anterior approach and the affected segment fused (Figure 17.21). If only one level is affected and there is no bony encroachment on the intervertebral foramen, anterior decompression and fusion can be expected to give good long-term relief from radicular symptoms. Although associated with high success rates, fusion has been postulated as a major contributing factor to adjacent segment degeneration, with possible symptomatic adjacent-level

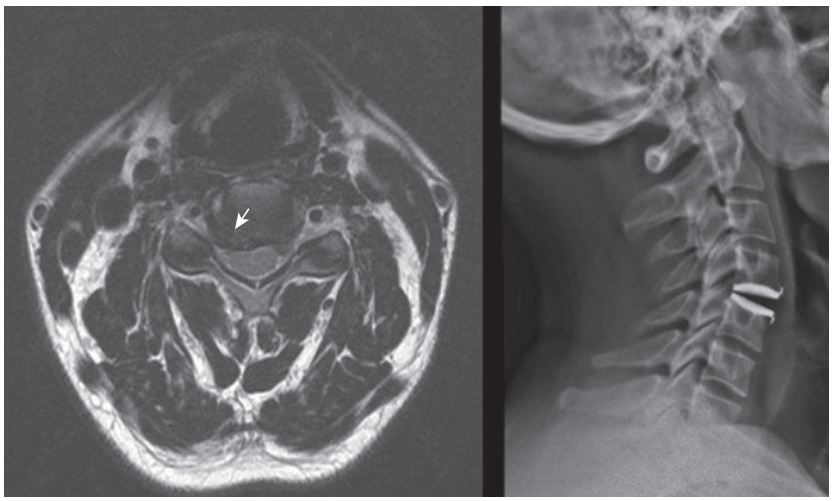


Figure 17.21 Disc prolapse – surgery An example of a patient with a C4–C5 right side disc herniation who underwent anterior discectomy and total disc arthroplasty.

spondylosis and stenosis. Furthermore, it can complicate with pseudarthrosis, although anterior plating may decrease the rate of this complication.

Cervical disc replacement (CDR) preserves motion at the implanted level and normal motion at the adjacent levels and is an alternative to ACDF. It is still debatable which treatment is superior to the other and both are cost-effective at 5 years. Nonetheless, heterotopic ossification can appear with time and interfere with the CDR success, especially in bi-level procedures.

Cervical spondylosis

This vague term refers to the cluster of abnormalities arising from the *ageing of the functional spinal unit* (two adjacent vertebrae and the disc in between), especially the *intervertebral (IV) disc*. Changes are most common in the C5–C6 and C6–C7 segments, the area that is more prone to intervertebral disc prolapse. As the discs degenerate, they lose their original biochemical and biomechanical properties. The ability of the disc to retain water is impaired, it desiccates, the amount of keratin sulfate increases and chondroitin sulfate decreases, which results in altered viscoelasticity. The disc loses its original height and becomes thinner and less elastic. Facet joints are progressively submitted to increased stresses and instability and the *uncovertebral joints* become arthritic, giving rise to pain and stiffness in the neck. Bony spurs, ridges or bars appear at the anterior and posterior margins of the vertebral end plates reducing the dimensions of the spinal canal and foramina. The disc collapses and protrudes and posterior bone spurs and infolded ligamentum flavum may encroach upon the spinal canal and foramina, causing pressure on the pain sensitive dura and the neural structures, resulting in a variably complex clinical picture.

Clinical features

Degenerative changes at the cervical spine are asymptomatic in most of the population. Nevertheless, patients, usually after 40 years of age, can present with *axial back pain* and *stiffness, radiating pain* to the upper extremity, altered *dermatomal sensation* or even signs of *myelopathy*. The onset of symptoms is usually insidious and they are often worse after a period of postural steadiness. The pain may radiate to different regions: to the occiput, the back of the shoulder girdle, the interscapular area and down to one or both upper limbs. *Paraesthesia* is often an associated symptom, as well as, *weakness* and clumsiness in the forearm and hand, although less frequently. The typical clinical course is characterized by exacerbations of acute discomfort, alternating with long periods of relative quiescence.

On clinical examination, the posterior and lateral neck and periscapular musculature may present with *spasm* and *tenderness*. Neck movements are limited and painful. *Decreased reflexes* of the upper limb may be present (Table 17.2).

Neck movements are limited and painful. Sometimes, features arising from narrowing of the intervertebral foramina and compression of the nerve roots (*radiculopathy*) dominate the clinical picture. These include pain referred to the interscapular area and upper limb, numbness and/or paraesthesia in the upper limb or the side of the face, muscle weakness and depressed reflexes in the arm or hand (Table 17.2). In advanced cases there may be narrowing of the spinal canal and changes due to pressure on the cord (*myelopathy* – see below).

Table 17.2 Cervical radiculopathy: clinical findings according to the level of involvement

Nerve root	Disc	Painful area	Areas of paraesthesia	Motor involvement	Reflexes
C3 Radiculopathy	C2–C3	Sub-occipital, back of the ear	Sub-occipital, back of the ear	–	–
C4 Radiculopathy	C3–C4	Lower neck, superior part of the shoulder	Lower neck, superior part of the shoulder	Diaphragm	–
C5 Radiculopathy	C4–C5	Superior part of the shoulder to the lateral mid-arm	Superior part of the shoulder to the lateral mid-arm	Deltoid (*biceps brachialis*)	*Biceps* reflex
C6 Radiculopathy	C5–C6	Lateral aspect of elbow, radial forearm and digits	Lateral aspect of elbow, radial forearm and digits	Wrist extensors, *biceps brachialis*	*Brachioradialis* reflex
C7 Radiculopathy (most common)	C6–C7	Dorsum of the forearm and middle finger	Dorsum of the forearm and middle finger	*Triceps*, wrist flexors, finger extensors	*Triceps reflex*
C8 Radiculopathy	C7–T1	Ulnar border of arm, forearm and digits	Ulnar border of arm, forearm and digits	Intrinsics, *flexor digitorum profundus* of index and long finger, *flexor pollicis longus* of thumb	–

Imaging

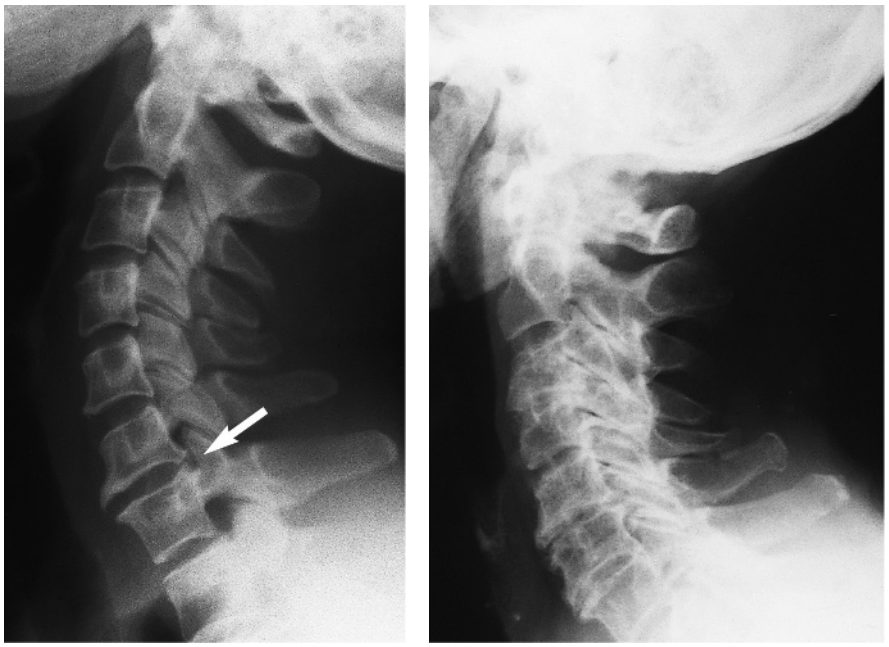
X-rays show narrowing of one or more intervertebral spaces, with bony spur formation (or lipping) at the anterior and posterior margins of the disc (at the end plates). These bony ridges (often referred to as 'osteophytes') may encroach upon the intervertebral foramina (Figure 17.22). Loss of the normal lordotic curve or even inversion might be found. The sagittal cervical spinal canal diameter can be measured: a canal less than 17 mm is often associated with symptomatic cervical spondylosis and less than 13 mm is usually associated with neurological compromise.

MRI is more sensitive to the whole degenerative process showing details of the discs, facets, vertebrae and ligamentum flavum, changes not otherwise visible (Figure 17.23). It is more reliable for the neural structures, showing the degree of compromise of the spinal cord or whether the clinical picture is due to nerve root compression.

Differential diagnosis

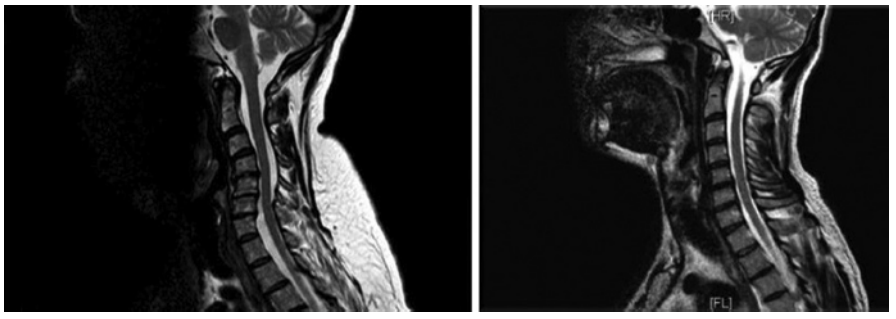
Around two-thirds of the adult population experience neck pain during their lifetime. Although very prevalent, neck pain is non-specific. Spondylosis is so common after the age of 40 years that it is likely to be seen in most middle-aged and elderly people who complain of neck pain. It is easily over-diagnosed as the cause of the patient's symptoms in this age group. Other disorders associated with neck and/or arm pain and sensory symptoms must be excluded.

Nerve entrapment syndromes Median or ulnar nerve entrapment may also give rise to intermittent symptoms of pain and paraesthesia in the hand. Typically, symptoms are worse at night and may be postural. Careful examination will show that the *changes follow a peripheral nerve* rather than a nerve root distribution. In doubtful cases, nerve conduction studies and electromyography will help to establish the diagnosis. Remember, though, that the patient



(a) (b)

Figure 17.22 Cervical spondylosis – X-rays
(a) Degenerative features at one level, C6–C7. Note the prominent 'osteophytes' at the anterior and posterior borders of these two vertebral bodies. (b) Marked degenerative changes at multiple levels.



(a) (b)

Figure 17.23 Cervical spine age
MRI of 46-year-old man (a) and of an 80-year-old man (b). The severity of imagiological features does not match the patient's chronological age, nor does it strictly correlate with the severity of symptoms. (Reproduced with permission from: Wierzbicki V, *et al.* How old is your cervical spine? Cervical spine biological age: a new evaluation scale. *Eur Spine J* 2015; **24**(12): 2763–70.)

may have symptoms from both a peripheral and a central neural structure compromise. At present, there is some evidence to suggest that long-standing cervical spondylosis may make the patient more vulnerable to the effects of peripheral nerve entrapment.

Rotator cuff lesions Pain may resemble that of cervical spondylosis because it radiates to the arm *above the elbow*. However, shoulder movements are abnormal, aggravate the pain and there may be X-ray and MRI features of rotator cuff degeneration.

Cervical tumours Metastatic deposits in the cervical spine can cause misleading symptoms, but sooner or later *bone destruction* produces diagnostic X-ray features. With tumours of the spinal cord or nerve roots, symptoms are usually unremitting and the lesion may be seen on MR imaging.

Thoracic outlet syndrome This condition is described in Chapter 11. Symptoms resemble those of cervical spondylosis. Pain and sensory abnormalities appear mainly in the ulnar border of the forearm and hand and may be *aggravated by upper limb traction or by elevation and external rotation of the shoulder*. It is due to compromise of the lower brachial plexus roots/trunk over a cervical or the first thoracic rib. In a thoracic outlet syndrome neck movements are neither painful nor restricted. X-rays may reveal a cervical rib, although the mere presence of this anomaly is not necessarily diagnostic.

Treatment

CONSERVATIVE TREATMENT

This is the mainstay of treatment. Analgesics and anti-inflammatory medication can be prescribed to control acute and exacerbating pain. Heat and massage are often soothing and restricting neck movements with a collar is an effective treatment during acute pain. Physiotherpay is a very important part of the treatment strategy, which includes exercises to optimize the range of motion and muscular control. Gentle passive manipulation and intermittent traction can be useful. Prolonged use of a cervical collar or brace may be detrimental.

OPERATIVE TREATMENT

If conservative measures fail to relieve the patient's symptoms and particularly if there is neurological compromise with radiculopathy arising from nerve root compression at one or two identifiable levels, surgical treatment may be indicated. There are several surgical strategies to address cervical spondylosis, depending on the pattern and the levels of involvement.

Anterior discectomy and fusion This operation has a 'track record' of more than 25 years and is particularly suitable if the problem is primarily one of unrelieved neck pain and stiffness, although it is also successful in relieving radicular symptoms. Through an anterior approach the intervertebral disc can be removed without disturbing the posteriorly placed neurological structures. After clearance of the intervertebral space, a suitably shaped spacer, autogenous bone graft or substitute (usually a *peek* or metallic implant filled with autogenous bone graft taken from the iliac crest) is inserted firmly between the adjacent vertebral bodies. An anterior plate may be added to improve the stability, particularly if several levels are fused. Complications such as graft dislodgement and failed fusion (with pseudarthrosis) are less likely to occur with intervertebral plating. Pseudarthrosis of cervical discectomy and fusion of more than three levels can be higher than 20%. Some surgeons recommend a combined anterior and posterior procedure in multilevel stenosis, especially if there is a kyphotic deformity. There is some concern about the possibility that fusion at one level may predispose to degeneration at an adjacent level.

Foraminotomy Foraminotomy (enlarging the IV foramen) through a posterior approach, may occasionally be indicated if there is isolated referred pain in the upper limb and/or radiculopathy, revealed on MRI as foraminal narrowing and nerve root compression. It is a very successful operation for pain relief, but only part of the facet joint is removed so as not to leave this segment unstable. Patients should be warned that pre-existing axial neck pain might not be eliminated and that further surgery may be required as the adjacent segments may go on to develop symptomatic disc degeneration in the future.

Intervertebral disc replacement Disc replacement has the theoretical advantages of preserving movement at the affected site and the stresses upon the adjacent discs. It has the drawback of time-related heterotopic ossification that can compromise these debatable advantages.

Laminoplasty This procedure (enlarging the spinal canal by lifting up the posterior elements of the vertebra – Figure 17.24) is indicated for spinal cord compression secondary to developmental spinal canal stenosis, continuous or mixed type of ossified posterior longitudinal ligament, multisegmental spondylosis associated with a narrow spinal canal and a distal type of cervical spondylotic amyotrophy with canal stenosis. Laminoplasty should be an option for younger patients. It is preferable to laminectomy because it can lessen postoperative kyphosis, instability and pain. With this procedure the central canal is decompressed but nerve root decompression can still easily be accomplished, addressing foraminal stenosis. However, the incidence of neck pain after laminoplasty is reported to be high, and this is one of the

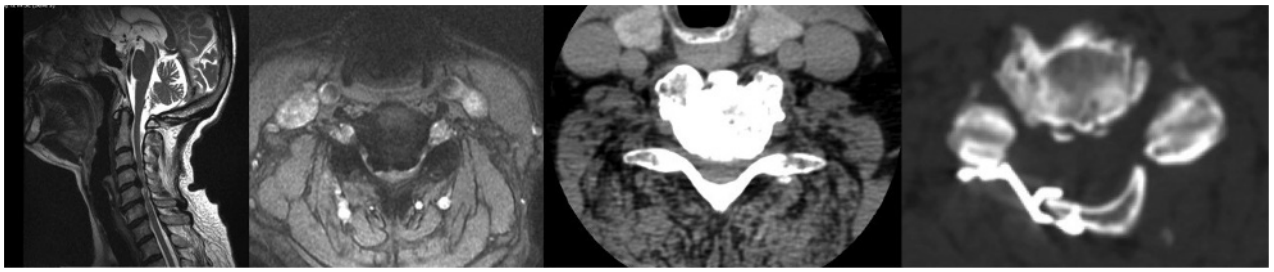


Figure 17.24 Laminoplasty A patient with cervical stenosis and myelopathy treated with laminoplasty: preop MRI and pre- and postop CT scan images.

most discouraging complications despite the advantages. Although preservation of spinal mobility is one of the aims of laminoplasty, the range of motion after this procedure usually decreases significantly.

Ossification of the posterior longitudinal ligament

Ossification of the posterior longitudinal ligament (OPLL) is a chronically progressive disease of ectopic enchondral and membranous ossification of the posterior longitudinal ligament, of unknown aetiology. Original reports appeared mainly from Japan, but it is recognized at present as a common and widespread condition elsewhere.

There is general consensus that it is a *multifactorial condition* representing a complex interaction of underlying genetic and environmental factors. Recent genetic analysis suggests the involvement of certain genes, such as *COL6A1*, *COL11A2* and *NPPS* in the origin of OPLL. The fibroblasts derived from OPLL patients exhibit osteoblast-like properties and PERK (a membrane protein kinase) is significantly upregulated in cells from these patients in contrast to those from non-OPLL patients.

The PLL is a two-layer structure: the superficial layer is located in close contact with the dura and bridges three or four vertebrae; the deep layer is located posterior to the vertebral body and connects two adjacent vertebrae.

OPLL is regarded as a rare disease in Western countries, in contrast to the Japanese population, where OPLL is one of the major causes of cervical myelopathy and was once called '*Japanese disease*'. The reported prevalence of OPLL in the Japanese is around 3%, whereas in Europeans or North Americans it is less than 1%.

It occurs mainly in the cervical spine, most often at the level of C5 or, less frequently, C4 and C6, and it may be associated with other bone-forming conditions such as diffuse idiopathic skeletal hyperostosis (DISH) and fluorosis. Coexisting ossification in the thoracic and/or lumbar spine has been reported in patients with cervical OPLL. OPLL is usually *associated with various metabolic disorders* such as obesity, diabetes mellitus, acromegaly and hypoparathyroidism.

This condition can cause spinal stenosis and myelopathy with varying degrees of severity as a result of cord compression. The dura mater may also become ossified and fuse with the posterior longitudinal ligament in a condition known as dural ossification.

The average age of onset of symptomatic disease is 50 years and patients may present with any combination of axial neck and upper-limb pain, sensory symptoms and muscle weakness in the arms and upper motor neuron symptoms and signs in the lower limbs. The most disturbing features are motor abnormalities such as *weakness*, *incoordination*, *clumsiness*, *muscle wasting* and *bladder-bowel dysfunction*. Ossification is often present for a long period before the onset of clinical symptoms. Only a small percentage of patients with typical imaging findings present symptomatic myelopathy and require surgical treatment. Cervical spinal cord injury (SCI) can be induced by minor cervical trauma in these patients.

Diagnosis

The diagnostic criteria for OPLL are:

- *radiological:* OPLL visible on lateral view X-ray (CT scan may be used to better assessment)
- *clinical:* cervical myelopathic symptoms, radicular symptoms and cervical spine range of movement abnormality.

Cervical OPLL is classified in four types:

a. *continuous* – a long lesion extending over several vertebral bodies
b. *segmental* – one or several separate lesions behind the vertebral bodies
c. *mixed* – a combination of the continuous and segmental types
d. *circumscribed* – a lesion mainly located posterior to a disc space

These are illustrated in Figure 17.25.

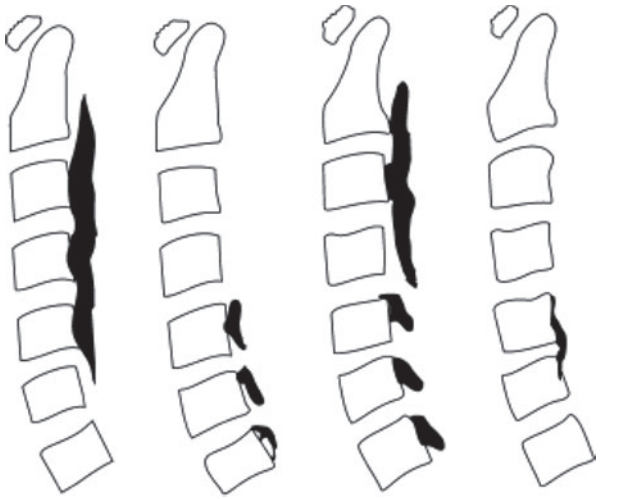


Figure 17.25 Types of OPLL (Reproduced with permission from: Izumi T, *et al.* Three-dimensional evaluation of volume change in ossification of the posterior longitudinal ligament of the cervical spine using computed tomography. *Eur Spine J* 2013; **22**: 2569–74.)

Imaging

X-rays show dense ossification along the back of the vertebral bodies (and sometimes also the ligamentum flavum) in the mid-cervical spine (Figure 17.26).

CT scan may show the double-layer sign on axial bone window, consisting of an anterior (ligamental) and a posterior (dural) rims of hyperdense ossification separated by a central hypodense mass (Figure 17.27). The double-layer sign is a sensitive factor to diagnose the dural ossification.

MRI scan is a sensitive examination for myelopathy. The signal intensity changes on MRI reflect the pathological changes in the spinal cord. Hypointensity on T1-weighted sequences and hyperintensity on T2 are primary changes seen in spinal cord lesions.

Treatment

Medical treatment is generally ineffective and mainly targeted for symptomatic relief. It consists of analgesics, anti-inflammatory drugs, antidepressants, anticonvulsants and opioids. However, the gold standard treatment for OPLL is surgical decompression, indicated in severe or progressive disease. The duration of symptoms prior to surgery is known to be one of the factors most significantly associated with a negative prognosis. Surgical treatment should be provided before the advent of intramedullary spinal cord changes in signal intensity on MRI.

Surgical decompression is performed through an anterior, a posterior or a combined approach.

Spinal stenosis and cervical myelopathy

The sagittal diameter of the mid-cervical spinal canal (the distance, on plain X-ray, from the posterior surface of the vertebral body to the base of the spinous process) varies considerably between individuals. The sagittal diameter of the adult spinal cord averages approximately 8 mm from C3 to C7. *A spinal canal with a diameter of less than 11 mm is suggestive of stenosis.*

Degenerative forms of cervical myelopathy as a result of spinal stenosis are the most common cause of spinal cord dysfunction in the adult population. Ageing of the cervical spine involves a range of anatomical changes that can result in spinal canal

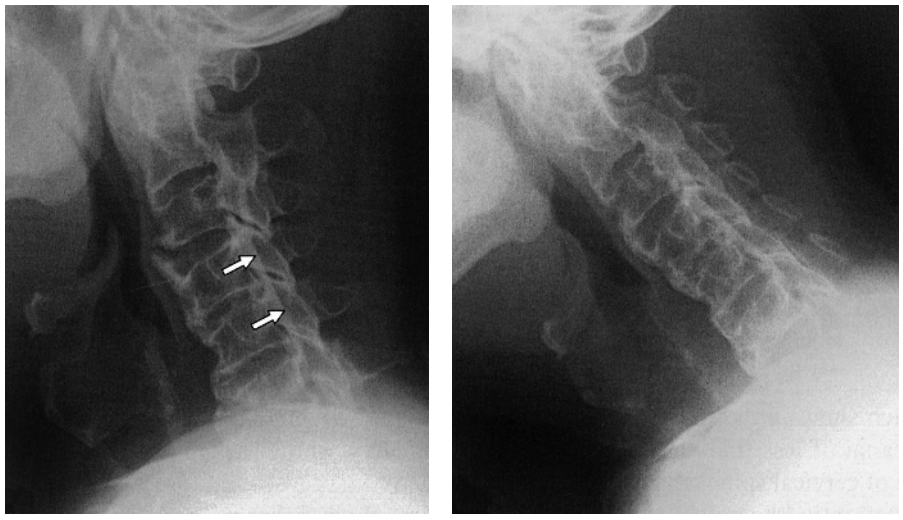


Figure 17.26 Ossification of the posterior longitudinal ligament (a) Lateral X-ray of the cervical spine showing the thin dense band running down the backs of the vertebral bodies (arrows); this appearance is typical of posterior longitudinal ligament ossification, which resulted in cervical spinal stenosis. (b) X-ray taken after posterior spinal decompression (laminoplasty); the spinous processes have been removed, the laminae split on one side of the midline and the posterior arch 'jacked' open. The sagittal diameter of the spinal canal is now considerably greater than before. (Courtesy of Mr H. K. Wong, Singapore.)

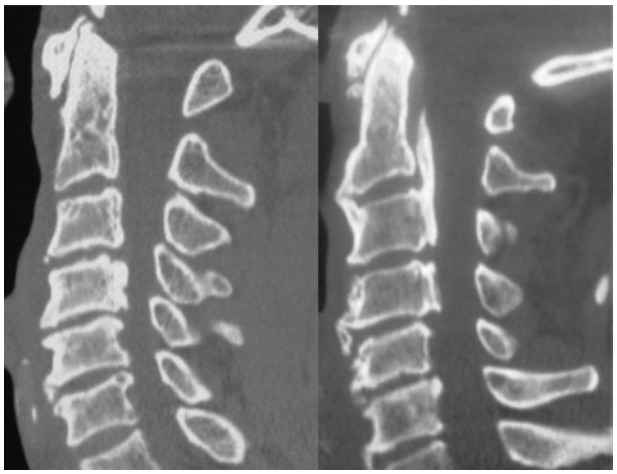


Figure 17.27 OPLL Sagittal view of a CT scan showing ossification of the PLL. (Reproduced with permission from: Fujimori T, *et al.* Ossification of the posterior longitudinal ligament of the cervical spine in 3161 patients: a CT-based study. *Spine* 2015; **40**: E394–E403).

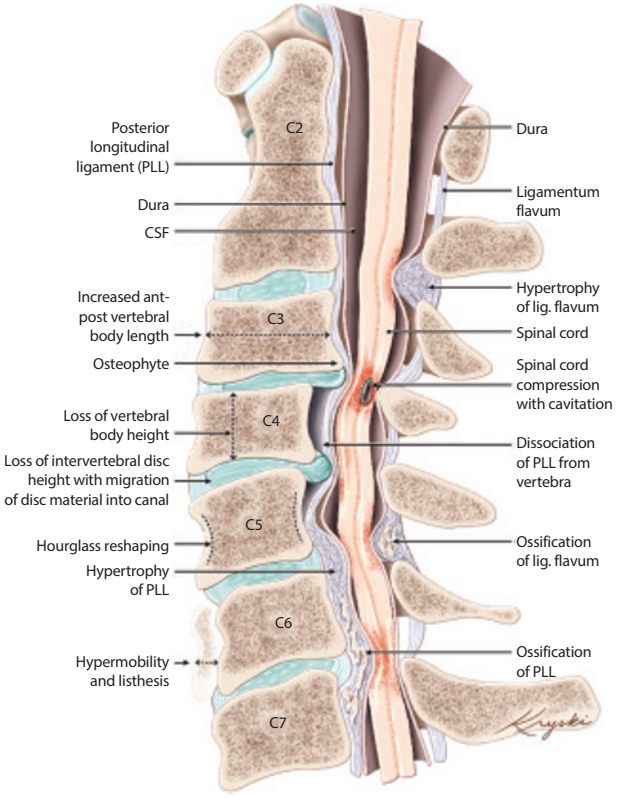


Figure 17.28 Cervical myelopathy The multiple features that characterize degenerative cervical myelopathy (Reproduced with permission from: Nouri A, *et al.* Degenerative cervical myelopathy: epidemiology, genetics, and pathogenesis. *Spine* 2015; **40**: E675–E693.)

stenosis, segment instability, sagittal malalignment and imbalance (Figure 17.28). If the changes are severe enough to compromise the spinal cord, the patient may develop neurological symptoms and signs of cord compression, which are thought to be due to both direct compression and ischaemia of the cord and nerve roots. Many asymptomatic and apparently normal people also have small canals and they are at risk of developing the clinical symptoms of spinal stenosis if there is any further encroachment due to intervertebral disc space narrowing, posterior osteophytes, wear of the facet joints, hypertrophy of the ligamentum flavum, ossification of the posterior longitudinal ligament or vertebral displacement.

Abnormally small canals are also seen in rare dysplasias, such as achondroplasia, and may give rise to cord compression. *Hirayama disease* or *cervical flexion myelopathy* is a rare form of cervical myelopathy in which segment instability and dynamic compression might play a role.

Cervical spondylotic myelopathy also results from dynamic factors leading to local spinal cord ischaemia, in combination with the static factors explained above, such as in cases of athetoid cerebral palsy or even in Gilles de la Tourette syndrome.

Clinical features

Patients usually have *neck pain* and *brachialgia* but also complain of paraesthesia, *numbness*, *weakness* and *clumsiness* in the arms and legs. *Gait might also be affected* with a broad-based unstable pattern, decreased velocity, decreased step and stride length, increased double support time, decreased plantar flexion at push-off and increased dorsiflexion of the ankle joint at swing phase, along with the onset of postural stability abnormalities. *Hand clumsiness* is one of the most common complaints in the setting of compressive cervical myelopathy. Exaggerated deep tendon reflexes, finger escape sign and difficulty in the finger grip and release test characterize this presentation pattern, also called *myelopathic hand*.

Symptoms may be precipitated by acutely hyperextending the neck, and some patients present for the first time after a hyperextension injury. They may experience involuntary spasms in the legs and, occasionally, episodes of spontaneous clonus. In severe cases there may be *bowel-bladder dysfunction* or incontinence. Patients with cervical spine-related headaches may report neck pain radiating to the low occipital and temporal regions.

The classical picture of *weakness* and *spasticity* in the legs and *numbness* in the hands is easy to recognize, but the signs are not always clear. Degenerative changes at the cervical spine with cord dysfunction result in the development of long tract signs, as a

result of atrophy and neuronal loss in the anterior horn and intermediate zone.

A detailed neurological examination is the current standard to the diagnosis of cervical myelopathy. Careful examination should reveal *upper motor neuron signs* in the lower limbs (increased muscle tone, brisk reflexes and clonus), while sensory signs depend on which part of the cord is compressed: there may be decreased sensibility to pain and temperature (spinothalamic tracts) or diminished vibration and position sense (posterior columns). The symptoms and signs reflect the degree to which the posterior, dorsolateral and ventrolateral columns, the ventral horns and the cervical nerve roots are involved.

The *Hoffmann sign* (elicited by flicking the terminal phalanx of the middle or ring finger to elicit the finger flexor response) and the *Trömner sign* (flexion of the thumb and index finger in response to tapping the volar surface of the distal phalanx of the middle finger held partially flexed between the examiner's finger and thumb) are established neurological signs for pyramidal response in the upper extremity and are commonly used as clinical neurological examinations for upper motor neuron lesions above the fifth or sixth cervical segments of the spinal cord. *Hyperreflexia* and the Hoffmann reflex have the highest sensitivity in patients with cervical myelopathy. These pathognomonic pyramidal signs may be absent in approximately one-fifth of myelopathic patients, but their prevalence is correlated with the severity of myelopathy.

Myelopathy is usually slowly progressive, but occasionally a patient with long-standing symptoms starts deteriorating rapidly and treatment becomes urgent. Nevertheless, the exact natural history of cervical spondylotic myelopathy is yet to be clarified.

The Japanese Orthopaedic Association Cervical Myelopathy Evaluation Questionnaire (JOACMEQ) and the Neck Disability Index (NDI) are recommended scores for the evaluation and outcomes measure of cervical spondylotic myelopathic patients (see Box 17.1).

BOX 17.1 THE JAPANESE ORTHOPAEDIC ASSOCIATION'S EVALUATION SYSTEM FOR CERVICAL MYELOPATHY (Total: 17 points)

I Upper extremity function
- 0 Impossible to eat with either chopsticks or spoon
- 1 Possible to eat with spoon, but not with chopsticks
- 2 Possible to eat with chopsticks, but inadequately
- 3 Possible to eat with chopsticks, but awkwardly
- 4 Normal

II Lower extremity function
- 0 Impossible to walk
- 1 Need cane or aid on flat ground
- 2 Need cane or aid on stairs
- 3 Possible to walk without cane or aid, but slowly
- 4 Normal

III Sensory disturbance

A Upper extremity
- 0 Apparent sensory loss
- 1 Minimal sensory loss
- 2 Normal

B Lower extremity
- 0 Apparent sensory loss
- 1 Minimal sensory loss
- 2 Normal

C Trunk
- 0 Apparent sensory loss
- 1 Minimal sensory loss
- 2 Normal

IV Bladder function
- 0 Complete retention
- 1 Severe disturbance (Inadequate evacuation of the bladder, straining, dribbling of urine)
- 2 Mild disturbance (Urinary frequency, urinary hesitancy)
- 3 Normal

Imaging

A *plain lateral radiograph* showing an anteroposterior diameter of the spinal canal of less than 11 mm strongly supports the diagnosis of cervical spinal stenosis. A better measure is the *Pavlov ratio* (the anteroposterior diameter of the canal divided by the diameter of the vertebral body at the same level) because this is not affected by magnification error. *A ratio of less than 0.8 is abnormal.*

MRI demonstrates the spinal cord and soft-tissue structures and helps to exclude other causes of similar neurological dysfunction. It is the gold standard method for evaluating these patients because it can determine the severity of degenerative changes, quantify the degree of cord compression due to canal stenosis, reveal intrinsic spinal cord abnormalities and effectively distinguish degenerative spine diseases from other etiologies (Figure 17.29). Nevertheless, increased signal intensity may, in fact, reflect various intramedullary pathologies such as oedema, gliosis, demyelination and myelomalacia. Furthermore, *T2 signal intensity abnormalities*, related to the cord compression, are not always present and correlate poorly with the disease severity and prognosis. Diffusion tensor imaging (an MRI technique that

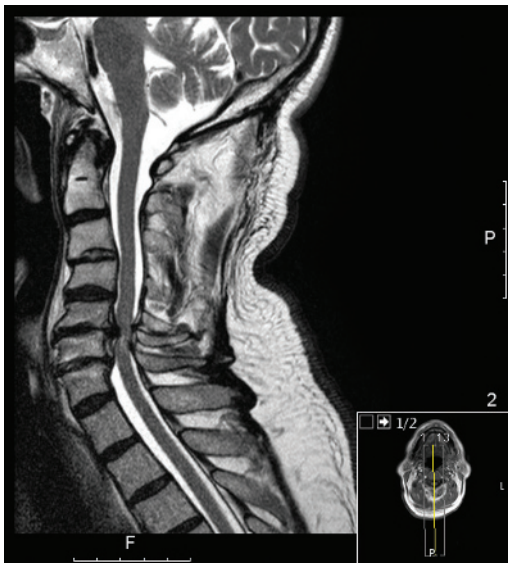


Figure 17.29 Spinal stenosis and myelopathy Sagittal T2 MRI view of a 69-year-old male patient with spinal stenosis and myelopathy. Note the multiple features of cervical spine degeneration and the hyperintense signal of the cord at the level of C5–C6.

allows evaluation of water molecule movement), however, provides relevant information about spinal cord integrity and impairment.

MRI has significant limitations in the evaluation of facet arthropathy.

CT myelography is superior to MRI in demonstrating osseous detail.

Differential diagnosis

Full neurological investigation is required to eliminate other diagnoses such as multiple sclerosis (episodic symptoms), amyotrophic lateral sclerosis (purely motor dysfunction), syringomyelia and spinal cord tumours.

Motor unit potentials analysis from nerve conduction studies may be useful for detecting axonal degeneration and reinnervation and the site of neurological compromise in the context of neuropathic disorders.

Treatment

Most patients can be treated conservatively with analgesics, a collar, isometric exercises and gait training. Manipulation and traction should be avoided. Epidural spinal injections are relatively safe and effective, although major complications such as spinal cord infarction have been reported. Given the unpredictably progressive nature of cervical myelopathy, the indications for non-operative management seem limited.

Patients with progressive myelopathy should be considered for surgery, as it usually prevents further deterioration and improves neurological outcomes and quality of life. The aim of surgical treatment is to decompress the spinal cord before permanent damage occurs. It is still a challenge to predict which patients will benefit the most from surgical treatment.

Acute, severe myelopathy is a surgical emergency, requiring immediate decompression. Surgical decompression of the cervical spinal cord can be performed by either an anterior or a posterior approach. A combined posterior decompression and reconstruction using pedicle or lateral mass screw instrumentation might be useful for patients with local kyphosis, segmental instability and in revision surgery.

INFECTIOUS PATHOLOGY

Spinal infections can be divided into two main types: *pyogenic* (caused by bacteria, mainly *Staphylococcus aureus*) or *non-pyogenic* (tuberculous, brucellar, aspergillar and fungal, which originate granulomatosis infections). Less frequently, parasites are the infecting agents.

Nowadays, the majority of spinal infection cases are pyogenic and only a quarter tuberculous.

These agents spread to the spine by a *haematogenous route*, *direct external inoculation* or from *contiguous tissues.* The direct external pathway is frequently associated with surgical spinal procedures and contiguous spread may result from adjacent infection (oesophageal ruptures, retropharyngeal abscesses or infections of aortic implants).

Vertebral infection may occur at any age. However, it affects primarily adult patients with a slight predominance of the male gender. It has a reported incidence between 0.2 and 2.4 per 100 000 per annum in developed countries and the age-adjusted incidence increases progressively after the fifth decade of life. There is a reported tendency for higher prevalence and more aggressiveness of cervical spondylodiscitis in the last decades.

In children, infection is located mainly within the *intervertebral disc* because of the rich anastomotic net between intraosseous arteries and vessels penetrating the disc, while *in adults spondylodiscitis predominates* because the disc is avascular (Figure 17.30). Infection can reach and collect inside the spinal canal, causing epidural or subdural abscesses. An uncontrolled infection can lead to spread to the surrounding tissues, causing *paravertebral abscesses.* The spread to the posterior structures is rare, being more common in the case of spinal tuberculosis.

Known predisposing risk factors for spinal infection include: previous spine surgery, septicemia, diabetes, protein malnutrition, intravenous drug use, HIV infection or another immunosuppressive states, chronic renal failure and liver cirrhosis.

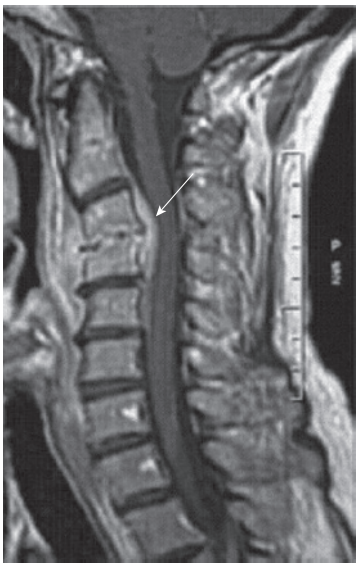


Figure 17.30 Spondylodiscitis Sagittal view of gadolinium-enhanced T1 image of a patient with C3–C4 spondylodiscitis with secondary epidural abscess. (Reproduced with permission from: Urrutia J, *et al*. Cervical pyogenic spinal infections: are they more severe diseases than infections in other vertebral locations? *Eur Spine J* 2013; **22**: 2815–20.)

Diagnosis

The diagnosis requires a high level of suspicion and is supported by clinical, laboratory and imaging findings. An *insidious onset of axial pain*, sometimes worsening at night, is frequently the first symptom. In adults, *fever* occurs in approximately half of the patients with pyogenic spondylodiscitis and in less than 20% of tuberculous cases. Dysphagia and torticollis may raise suspicion of cervical involvement. In children, the non-specific clinical picture may include irritability and refusal to walk. Neurological deficits are rare.

There is usually a delay between the onset of symptoms and the definitive diagnosis and treatment, mainly due to the low specificity of the clinical picture at presentation.

Erosion of vertebral end plates and osteolytic lesions, which can lead to instability, deformity and even cord compression, characterize the typical imagiological presentation, but these are late features.

Erythrocyte sedimentation rate is a sensitive marker of infection, although having a low specificity. C-reactive protein is also sensitive and considered the best monitor of treatment response. White blood cell count has the lowest sensitivity.

If there is clinical suspicion of a spinal infection, it is recommended to obtain blood and urine cultures. Aerobic cultures are performed routinely and anaerobic cultures are highly recommended, as anaerobic bacteremia is a re-emerging problem. Cultures should be obtained before antibiotic initiation. Following the rule of '*culture all tumours and biopsy all infections*', the histopathology adds an important value to microbiological culture in distinguishing pyogenic from granulomatous diseases. The definitive diagnosis of pyogenic spondylitis is only achieved by microbiological and histological examination of the infected tissues from biopsies. Percutaneous CT-guided needle and open biopsies can be used. The accuracy of *percutaneous vertebral biopsy* in patients with spondylodiscitis has been reported to be about 70%. The harvested tissue should be submitted to: *Gram and acid-fast bacilli smears*; *aerobic*, *anaerobic*, *fungal* and *tuberculosis cultures*; and *polymerase chain reaction*.

For a faster diagnosis in the case of *Mycobacterium tuberculosis*, which has a slow pattern of growth (up to 8 weeks), the use of interferon-gamma release assays (IGRAs), measured from whole blood plasma, is a valuable aid providing results in less than 24 hours. Tuberculosis has a recognized reputation as one of the great mimickers in medicine, making it difficult to diagnose.

Treatment

Spondylodiscitis is a life-threatening disease with a mortality rate of up to 20%.

The principles of treatment of spinal infections are: antibiotic therapy; neurological decompression in the setting of neurological deficits; preservation of stability and correction of deformity.

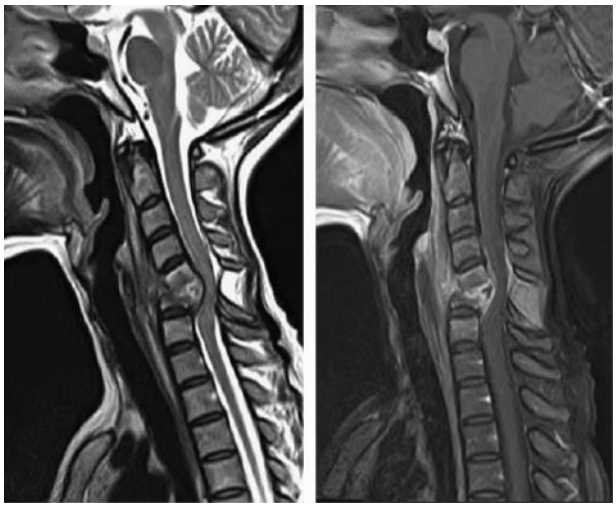
Although antibiotic therapy should be initiated only after a definitive etiological diagnosis in a stable patient, in the presence of sepsis or the impossibility of an etiological diagnosis, empirical antibiotic therapy should be considered. The antibiotic spectrum must generally cover *S. aureus* and *E. coli*, the commonest pathogens for pyogenic spondylodiscitis. In cases of methicillin-resistant *S. aureus*, vancomycin is usually chosen. In the setting of confirmed tuberculosis spondylitis, tuberculostatic therapy should be initiated.

The treatment of spinal infections is mainly non-surgical, although surgical intervention might be needed if conservative treatment fails or if there is significant bone destruction, mechanical instability, progressive deformity, neurological compromise and recurrent infection. Surgical treatment usually includes complete debridement of infected tissue, decompression of neural elements, reconstruction of the involved segments and spinal stabilization.

Since vertebral infections most commonly involve the anterior elements of the spine, the anterior operative approach is the main route for surgical treatment.

Pyogenic infection

Pyogenic infection involving the cervical spine is unusual and therefore often misdiagnosed in the early stages when antibiotic treatment is most effective. Only 11% of pyogenic spondylodiscitis caused



(a) (b)

Figure 17.31 MRI scan image showing a C5 and C6 pyogenic spondylitis with significant bone destruction and resulting kyphotic deformity. (a) Sagittal T2 image. (b) Sagittal gadolinium enhanced T1 image showing the involved area. (Reproduced with permission from: Miyazaki M, *et al.* Clinical features of cervical pyogenic spondylitis and intraspinal abscess. *J Spinal Disord Tech* 2011; **24**: E57–E61.)

by haematogenous spread affects the cervical spine (Figure 17.31). Nonetheless, this location has the highest risk of neurological compromise and the greatest potential for causing disability among spinal infections. Initially, destructive changes are limited to the intervertebral disc space and the adjacent parts of the vertebral bodies. Later, abscess formation occurs and pus may extend into the spinal canal or into the soft-tissue planes of the neck.

Post-procedural discitis represents up to 30% of all cases of pyogenic spondylodiscitis. There is a recognized association between pyogenic vertebral osteomyelitis and infectious endocarditis.

Pyogenic spondylodiscitis is difficult to diagnose at the initial stage, but early diagnosis and prompt treatment should be the goal in cervical spine pyogenic infections, considering the potentially high morbidity and high mortality.

According to the main monomicrobial pattern of pyogenic spondylodiscitis, up to about 59% of positive blood cultures identify the causative microorganism.

Clinical features

The patient complains of *neck pain*, usually with an insidious onset and a progressively severe course, associated with *muscle spasm* and *stiffness*. On examination, neck movements are usually severely restricted. Nevertheless, *systemic symptoms are often mild*, even in the presence of intraspinal abscess.

Imaging

X-rays at first show either no abnormality or only slight narrowing of the disc space. Later on, there may be more obvious signs of bone destruction (Figure 17.32).

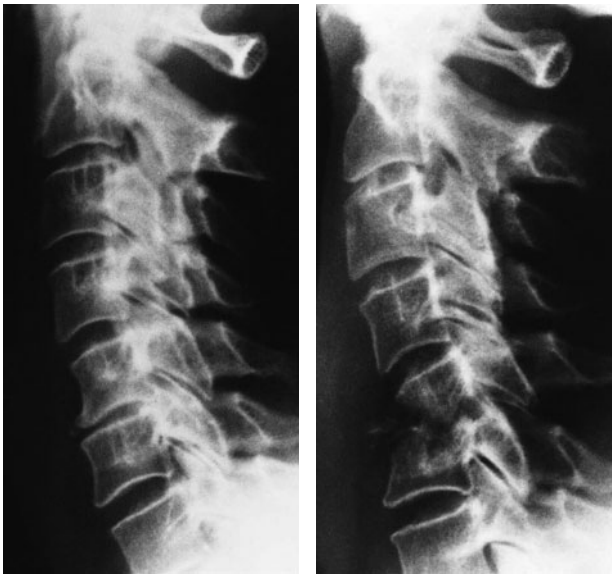
Treatment

The mainstay of treatment is *antibiotherapy* and *rest*. The cervical spine is 'immobilized' by traction and, once the acute phase subsides, a collar may suffice. The usual natural history is the remission of infection and intervertebral space obliteration and fusion.

In resistant cases or if there is significant abscess formation, debridement and drainage will be required. Autologous bone strut grafting combined with ventral instrumentation is considered the 'gold standard' of surgical treatment.

Tuberculosis

Skeletal tuberculosis is the most frequent type of extrapulmonary tuberculosis, and spinal affection comprises around 50% of the cases. Spinal tuberculosis, also known as *Pott's disease*, is preferentially located in the thoracic spine and often involves more than two levels, being the most frequent form of skeletal tuberculosis. Cervical spine tuberculosis, however,



(a) (b)

Figure 17.32 Pyogenic infection (a) The first X-ray, taken soon after the onset of symptoms, shows narrowing of the C5–C6 disc space but no other abnormality. (b) Three weeks later there is dramatic destruction and collapse; the speed at which these have occurred distinguishes pyogenic from tuberculous infection.

is rare, but the most dangerous, especially if it involves the upper cervical spine.

Tuberculosis is showing resurgence in developed countries and a significant increase in the developing regions. It usually affects the lung prior to spreading to the spine through the Batson's plexus or by lymphatic drainage. As with other types of infection, the *organism is blood-borne* and the infection localizes in the *intervertebral disc* and the anterior subchondral region of the adjacent vertebral bodies.

As the *bone is destroyed*, the *cervical spine collapses into kyphosis*. A retropharyngeal abscess forms and points behind the sternocleidomastoid muscle at the side of the neck (*scrofula*, the Latin word applied to describe tuberculosis of the neck). In late cases cord damage may cause neurological signs varying from mild weakness to tetraplegia. Because the tuberculosis of the spine is slowly developing, the spinal cord tolerates the gradually increasing extradural compression without immediate neurological deficits.

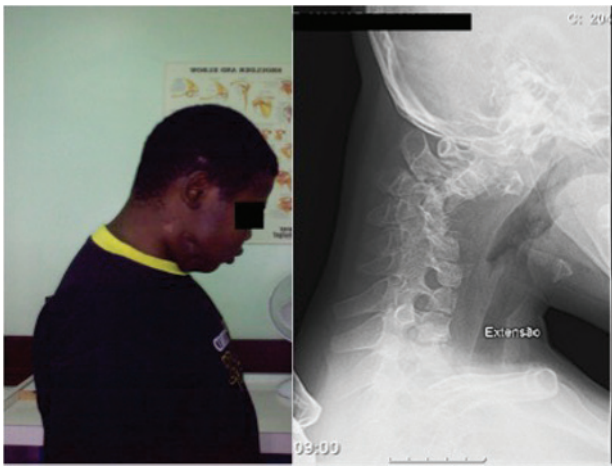


Figure 17.33 Tuberculosis Cervical kyphotic deformity due to destruction of vertebral bodies from C3 to C5 secondary to tuberculosis.

Clinical features

The patient, more commonly a child than an adult, complains of *neck pain* and *stiffness*. In neglected cases a retropharyngeal abscess may cause difficulty in swallowing or swelling at the side of the neck. On examination the neck is extremely tender and all movements are restricted. In late cases there may be obvious *kyphosis* (Figures 17.33 and 17.34), a fluctuant abscess in the neck (Figure 17.35) or a retropharyngeal swelling. The limbs should be examined for neurological defects.

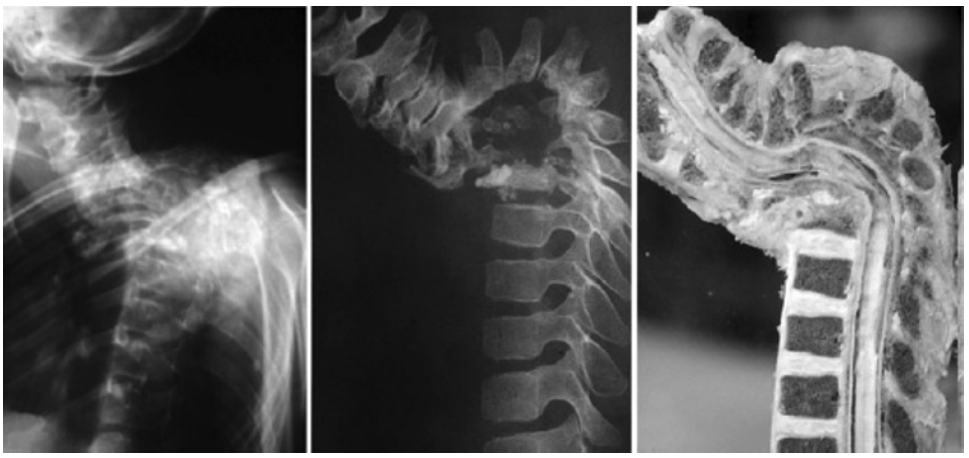
Imaging

X-rays show narrowing of the disc space and erosion of the adjacent vertebral bodies. *MRI* may show a cold abscess. Because of the possibility of *non-contiguous involvement* in spinal tuberculosis, it is recommended having an imagiological assessment of the entire spine in these patients (with X-ray or preferably MRI).

Treatment

Treatment is initially by anti-tuberculosis therapy and external immobilization of the neck in a cervical brace or plaster cast for 6 weeks up to 6 months, in neurologically intact patients and without signs of instability. Patients with instability or with neurological compromise can be placed on skull tongs traction.

Debridement of necrotic bone and anterior cervical vertebral fusion with bone grafts may be offered as an alternative to prolonged immobilization, in cases of bone destruction with severe kyphosis deformity. More urgent indications for operation are to drain a large retropharyngeal abscess (Figure 17.36), to decompress a threatened spinal cord, or to fuse an unstable spine.



(a) (b) (c)

Figure 17.34 Pott's disease X-ray (a), CT scan image (b) and pathological specimen (c) of a child with Pott's disease. Note the extent of anterior column deficit with multilevel involvement and the resulting kyphotic deformity. (Reproduced with permission from: Mak K, *et al.* Surgical treatment of acute TB spondylitis: indications and outcomes. *Eur Spine J* 2013; **22**(Suppl 4): S603–S611.)

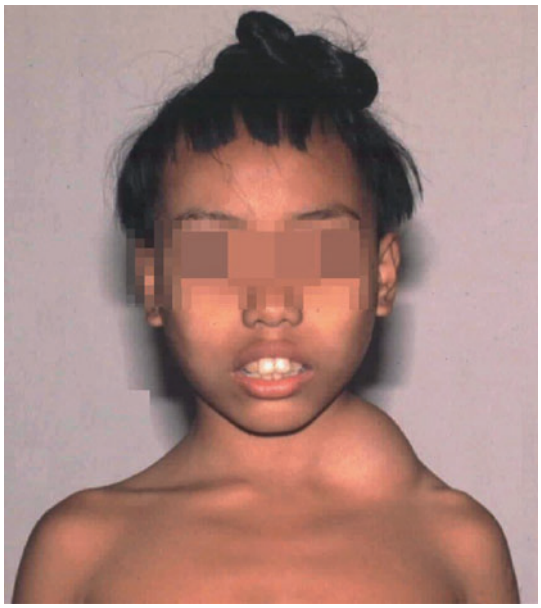


Figure 17.35 Tuberculosis This child had been complaining of neck pain and stiffness for several months. Eventually she was brought to the clinic with a lump at the side of her neck – a typical tuberculous abscess.

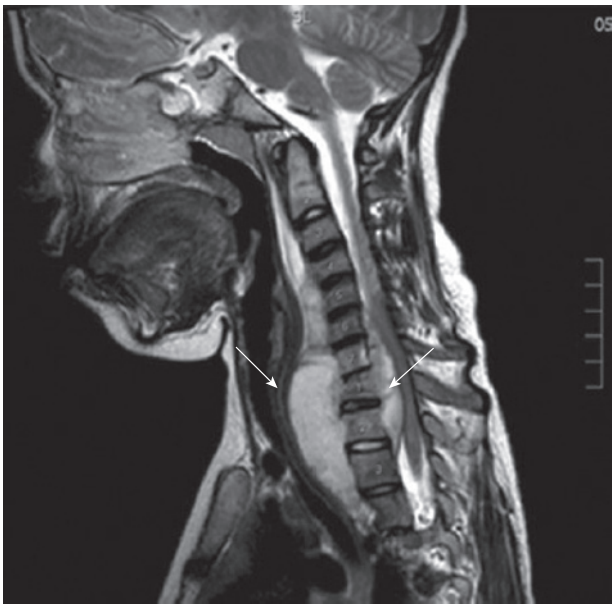


Figure 17.36 Tuberculosis – MRI Sagittal T2 MRI image of a young child with TB, showing a large anterior epidural and prevertebral abscess. (Reproduced with permission from: Manoharan S, *et al.* A large tuberculosis abscess causing spinal cord compression of the cervico-thoracic region in a young child. *Eur Spine J* 2013; **22**: 1459–63.)

INFLAMMATORY PATHOLOGY

The spine can be involved in most inflammatory disorders, such as rheumatoid arthritis (RA), seronegative spondyloarthritides (SpA), juvenile arthritides and, less frequently, disorders including pustulotic arthro-osteitis and SAPHO (synovitis, acne, pustulosis, hyperostosis, osteitis) syndrome. SpA includes ankylosing spondylitis (AS) and psoriatic arthritis (PsA).

Inflammatory changes at the sacroiliac joints always occur in AS and are part of most other forms of SpA. Spinal changes are also a feature of SpA, especially in the late stages of AS.

Damage of the periarticular bone and the *articular cartilage* are hallmarks of arthritis, symbolizing the destructive potential of chronic inflammation. RA, PsA and AS differ substantially in their patterns of bone and cartilage damage. These differences are at least partly based on the variable capability to form new bone, which may reflect a skeletal response to inflammation.

RHEUMATOID ARTHRITIS

The *atlantoaxial region* is the main target in cervical spine involvement of RA, the prototype of destructive arthritis that affects the synovial joints. The anatomy around the joint is illustrated in Figure 17.37.

Pannus formation (thickened synovium) can affect this joint, causing erosion of the articular cartilage and subchondral bone and destruction or attenuation of the surrounding ligaments (particularly the transverse ligament) and predisposing to instability. Subaxial involvement, the least common type, though, also occurs.

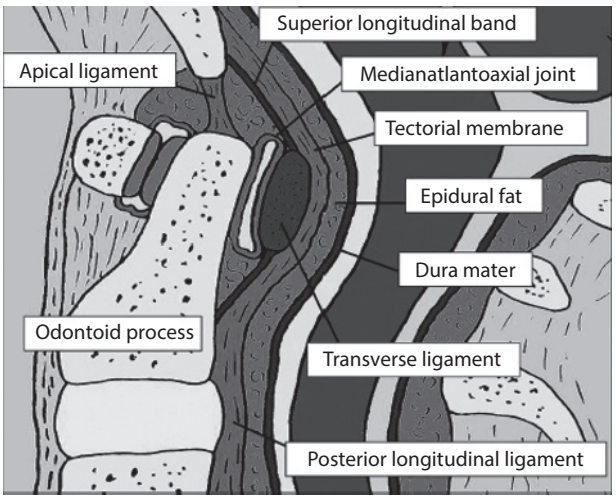


Figure 17.37 Anatomy around the atlantoaxial joint (Reproduced with permission from: Tojo S, *et al.* Factors influencing on retro-odontoid soft-tissue thickness: analysis by magnetic resonance imaging. *Spine* 2013; **38**: 401–6.)

The cervical spine is affected in approximately 17–86% of patients with RA and has a progressive course. There are three types of destructive and potentially unstable lesions:

1 *atlantoaxial subluxation (AAS)* – resulting from erosion of the atlantoaxial joints and the transverse ligament
2 *vertical subluxation (VS)* – also known as atlantoaxial impaction, resulting from erosion of the atlanto-occipital articulations allowing the odontoid peg to ride up into the foramen magnum; and
3 *subaxial subluxation (SAS)* – resulting from erosion of the facet joints in the mid-cervical region.

AAS is the most frequent form of instability in the occipitoatlantoaxial region (with a reported incidence of 27%), but lateral, rotatory and vertical subluxation also occurs. Although the amount of atlantoaxial displacement that occurs is often greater than 10 mm, neurological complications are uncommon and the majority of patients remain asymptomatic for years. However, complications do occur – especially in long-standing cases – and are produced by mechanical compression of the cord, by local granulation tissue formation or (very rarely) by thrombosis of the vertebral arteries. The prevalence of cervical myelopathy is 5% in rheumatoid patients.

Subaxial involvement can cause instability in the cervicothoracic transition and is usually seen in patients with severe chronic peripheral arthritis.

In addition, *vertebral osteoporosis* is common, due either to the disease or to the effect of corticosteroid therapy, or both.

Clinical features

The patient is usually a woman with advanced RA (Figure 17.38). There is often *neck pain* and *movements are markedly restricted*. Symptoms and signs of root compression may be present in the upper limbs. Less often there is lower-limb weakness and upper motor neuron signs due to cord compression. There may be *symptoms of vertebrobasilar insufficiency* or brainstem compression, such as vertigo, tinnitus and visual disturbance. Some patients with upper cervical RA develop thromboembolic stroke caused by positional and transient vertebral artery occlusions at the atlantoaxial junction.

General debility and peripheral joint involvement can mask the signs of myelopathy. A Lhermitte's sign (electric shock sensation down the spine on flexing the neck) may be present. Sudden death from catastrophic neurological compression is rare.

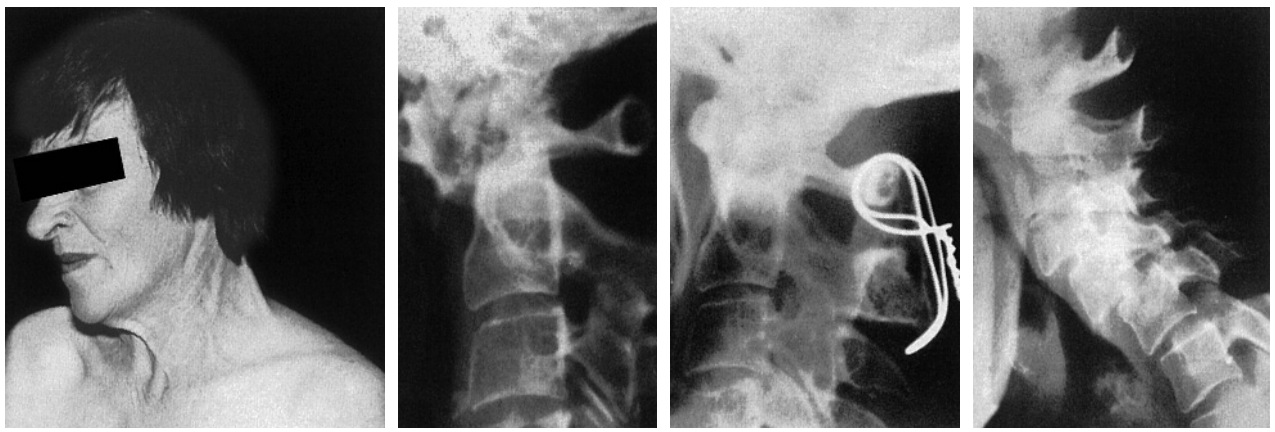
The presence of subluxation is not always associated with the presence of clinical symptoms and the radiographic severity of subluxation does not correspond to the development of neurological symptoms. Indeed, some patients, though completely unaware of any neurological deficit, are found to have mild sensory disturbance or pyramidal tract signs on careful examination.

Atlantoaxial subluxation also affects young patients with juvenile idiopathic arthritis.

Imaging

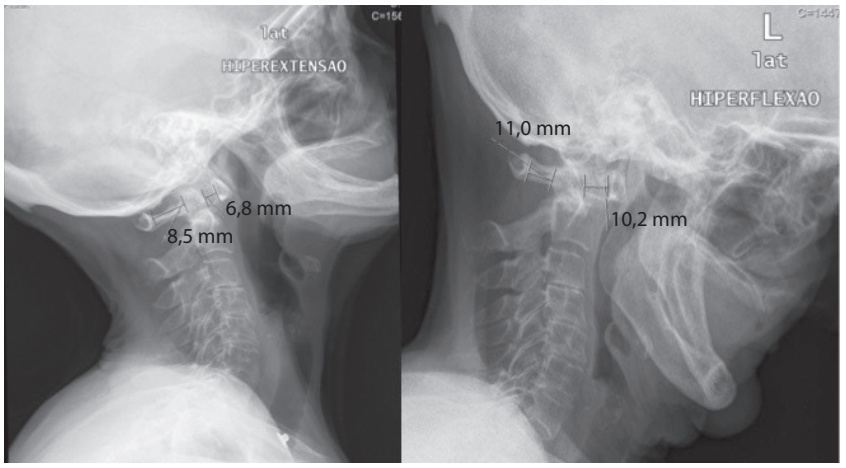
X-rays Radiography of the cervical spine is mandatory in rheumatoid patients with neck pain. Flexion-extension X-rays are an important part of any preoperative evaluation routine, especially in identifying C1–C2 abnormal motion (Figure 17.39).

The typical radiographic features of RA are *cortical bone erosions* (resulting from a continuous inflammatory attack of the synovial membrane on bone), *joint space narrowing* and *periarticular osteoporosis*.



(a) (b) (c) (d)

Figure 17.38 Rheumatoid arthritis (a) Movement is severely restricted; attempted rotation causes pain and muscle spasm. (b) Atlantoaxial subluxation is common; erosion of the joints and the transverse ligament has allowed the atlas to slip forward about 2 cm; (c) reduction and posterior fusion with wire fixation. (d) This patient has subluxation, not only at the atlantoaxial joint but also at two levels in the mid-cervical region.



(a) (b)

Figure 17.39 Rheumatoid arthritis – X-rays Lateral dynamic X-ray views of an 82-year-old female patient with RA with C1–C2 instability. Note the change in the measured ADI and SAC from hyperextension (a) to hyperflexion (b).

Atlantoaxial instability is visible in lateral films taken in flexion and extension. In flexion, the anterior arch of the atlas rides forwards, leaving a gap of 5 mm or more on the ADI (atlantodental interval). This subluxation is reduced on extension. Atlanto-occipital erosion is more difficult to see, but a lateral tomography shows the relationship of the odontoid to the foramen magnum. Normally the odontoid tip is less than 5 mm above *McGregor's line* (a line from the posterior edge of the hard palate to the lowest point on the occiput). In erosive arthritis the odontoid tip may be 10–12 mm above this line. The diagnosis and severity of vertical subluxation of the axis can be described using the *Redlund-Johnell* or *Ranawat criteria*.

Flexion views may also show anterior subluxation in the mid-cervical region. Erosive arthritis, usually at several levels, may cause subluxations producing a stepladder appearance on lateral views. If the subaxial canal diameter is <14 mm there is a possibility of spinal cord compression.

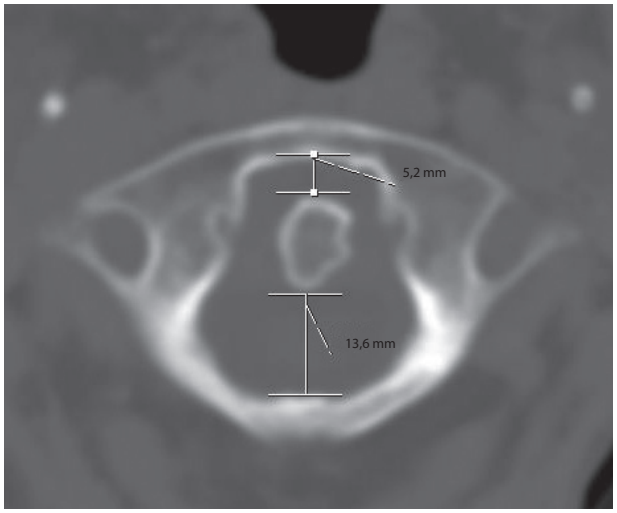


Figure 17.40 Rheumatoid arthritis – CT Axial CT scan image of the same patient as in Figure 17.39. Measurements of ADI and SAC.

CT and MRI These methods are useful for imaging grey areas such as the atlantoaxial and atlanto-occipital articulations and for viewing the soft-tissue structures (especially the cord) (Figures 17.40 and 17.41).

Treatment

Effective control of inflammation by conventional disease-modifying antirheumatic drugs (DMARDs), glucocorticoids, methotrexate, sulfasalazine and leflunomide retards structural damage in RA. Wearing a collar can usually relieve pain.

The indications for *operative stabilization* of the cervical spine are: (1) severe and unremitting pain and (2) neurological signs of root or cord compression. The presence of deformity and stenosis

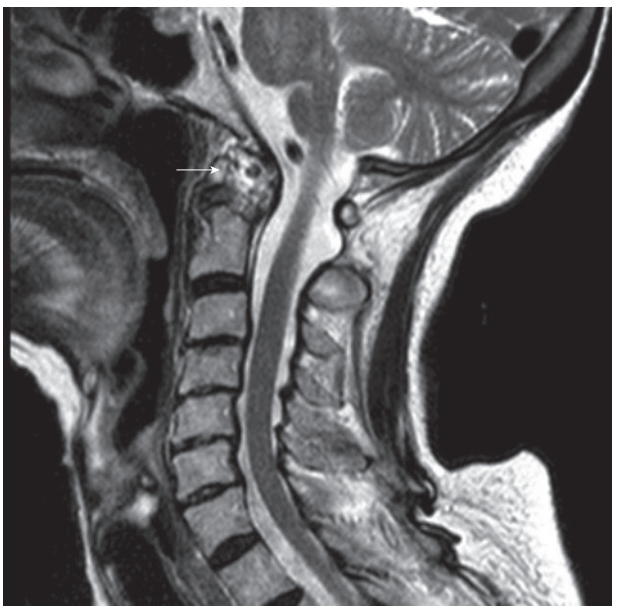
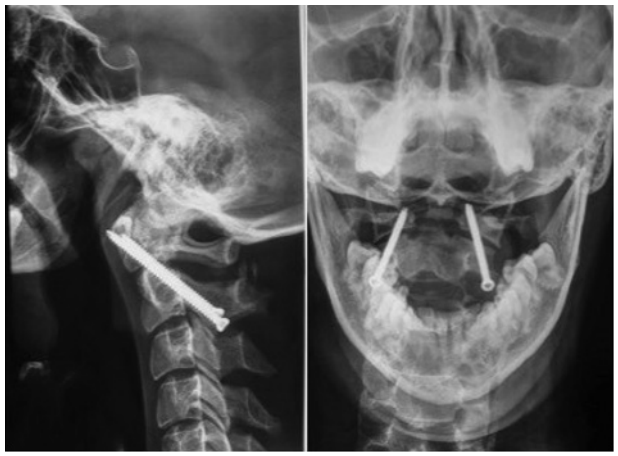


Figure 17.41 Rheumatoid arthritis – MRI Sagittal T2-weighted MRI image of the same patient as in Figures 17.39 and 17.40. Note the heterogeneous hyperintense *pannus* surrounding the tip of the dens.



(a) (b)

Figure 17.42 Rheumatoid arthritis – surgery Cervical C1–C2 fusion with atlantoaxial transarticular screw fixation (Magerl technique).

resulting in neurological symptoms and presumed presence of instability are indications for surgery.

Once neurological complications develop, 1-year mortality rate can be as high as 50% if the condition is left untreated. For those who need surgical intervention, early surgical intervention is therefore obviously desirable. Reducible AAS occurs first and irreducible AAS is a sign of VS, which has a high mortality risk.

Atlantoaxial surgical stabilization is usually accomplished through a posterior approach by transarticular screw fixation (*Magerl technique* – Figure 17.42) or a C1 lateral mass–C2 pedicle screw fixation construct (*Harms technique*). Postoperatively a cervical brace can be worn. However, if instability is marked and operative fixation insecure, a halo jacket may be necessary. In patients with very advanced disease and severe erosive changes, postoperative morbidity and mortality are high.

Severe deformities, very poor bone quality, erosive pedicles and abnormal vertebral arteries are recognized risks in rheumatoid cervical spine surgery.

Ankylosing spondylitis

Ankylosing spondylitis (AS) is the most common and usually the most disabling form of seronegative SpA to affect the cervical spine. According to the modified New York criteria, the diagnosis of definite AS requires the following: established *sacroiliitis* on radiographs and at least one of the following clinical criteria: (1) *low back pain* and *stiffness* for more than 3 months *improving with activity*, (2) *limited movement* of the lumbar spine, and (3) *reduced chest expansion*.

Clinical features

AS often starts in early adulthood, predominantly affecting men, and has a chronic progressive course. Neck pain and stiffness tend to occur some years after the onset of backache. The neck becomes progressively stiff and kyphotic, although some movement is usually preserved at the atlanto-occipital and atlantoaxial joints.

Severe functional limitation in the setting of an unacceptable *'chin-on-chest' deformity*, with significant compromise of the horizontal gaze, swallowing or jaw opening, is an indication for corrective cervical spine osteotomy. The thoracolumbar segment is the most affected by kyphotic deformities.

Progressive spinal deformity associated with stiffness and restricted spinal movements in a brittle and osteoporotic bone makes the AS spine *prone to fracture*, a life-threatening complication. A patient with ankylosing spondylitis and an increase in neck pain, even after minor trauma, must be assumed to have a fracture until proven otherwise. *CT or MRI scans must be included* in the workup if the plain radiographs are inconclusive. Disease-related chronic pain may mask the acute pain related to an acute fracture and late presentation of patients to hospital is not uncommon. Fractures often occur at intervertebral spaces but usually involve the ankylosed posterior structures and are thereby unstable three-column injuries, with commonly associated neurological compromise. The reported lifetime risk for fracture in AS is around 15%. Up to around 20% of patients have associated *spinal cord injury*. The lower cervical spine is the most commonly involved location and there is an increased risk of noncontiguous fractures in this population. AS patients complicated by vertebral fracture have also a high risk for inpatient complications and mortality.

X-rays

Many years may pass before AS has its full radiological expression, which explains why diagnosis is often delayed up to 10 years. Typical radiographic spinal changes include: *erosion of vertebral corners* (*Romanus lesions*), causing vertebral *squaring* and eliciting reactive *sclerosis* appearing as condensation of vertebral corners; *syndesmophytes* (slim ossifications in the annulus fibrosus); *bamboo spine* (syndesmophytes crossing the intervertebral spaces in addition to fusion of apophyseal joints); *supra and interspinous ligaments ossification* ('dagger sign' is the appearance of a single central radiodense band; 'trolley-track sign' is the result of ligamentous ossification together with the apophyseal joint capsules, appearing as three vertical radiodense lines on frontal views); *Andersson lesions* (erosive lesions affecting the three columns within intervertebral spaces); and *ossifying enthesopathy*. See Figures 17.43 and 17.44.

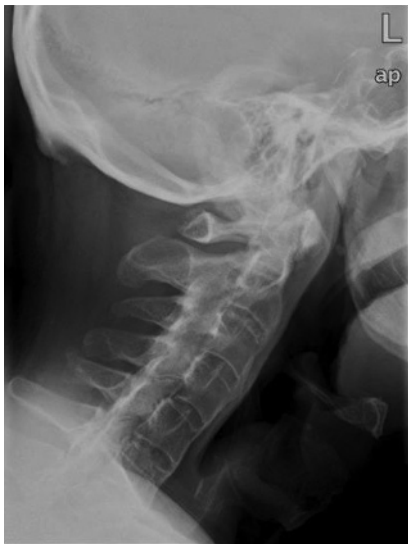


Figure 17.43 Ankylosing spondylitis Lateral X-ray view of the cervical spine of a 64-year-old patient with AS. Note the fusion pattern affecting the multiple cervical segments.

Treatment

Therapeutic strategies include non-pharmacological treatment, such as regular exercise and physiotherapy, and pharmacological treatment, including non-steroidal anti-inflammatory drugs (NSAIDs), analgesics, glucocorticoids, disease-modifying antirheumatic drugs and anti-tumor necrosis factor therapy. Patients on continuous NSAID therapy show reduced radiographic progression.

Surgical treatment may be considered for severely disabling deformities (Figure 17.45). Deformity corrective surgery in AS, usually extension osteotomies such as the *Simmons osteotomy* at C7–T1, is associated with a high risk of neurovascular compromise. Most of the fractures of the ankylosed spine are treated surgically, although surgical complications are common. The 3-month reported mortality associated with spinal fractures related to AS is about 20%.

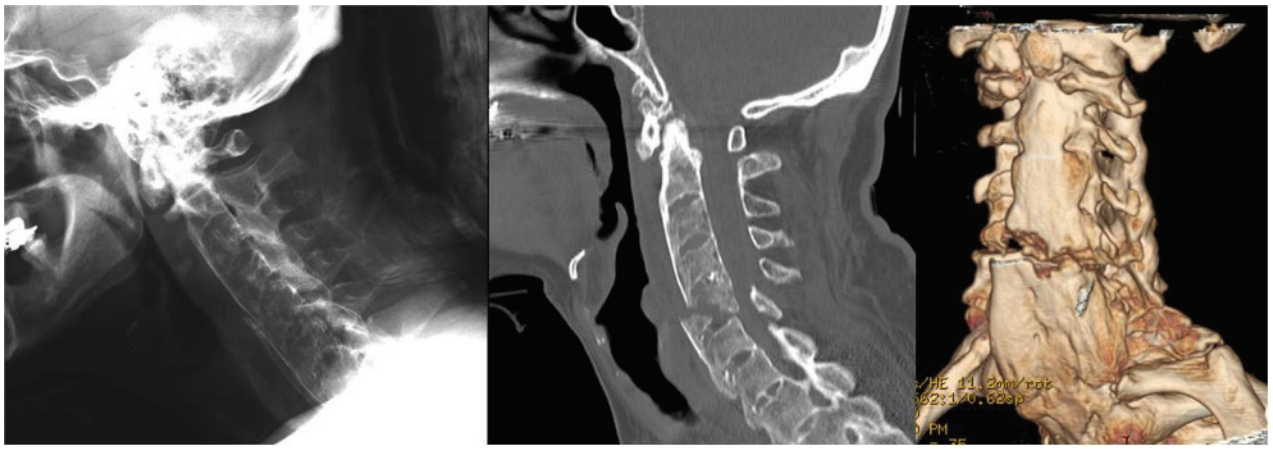
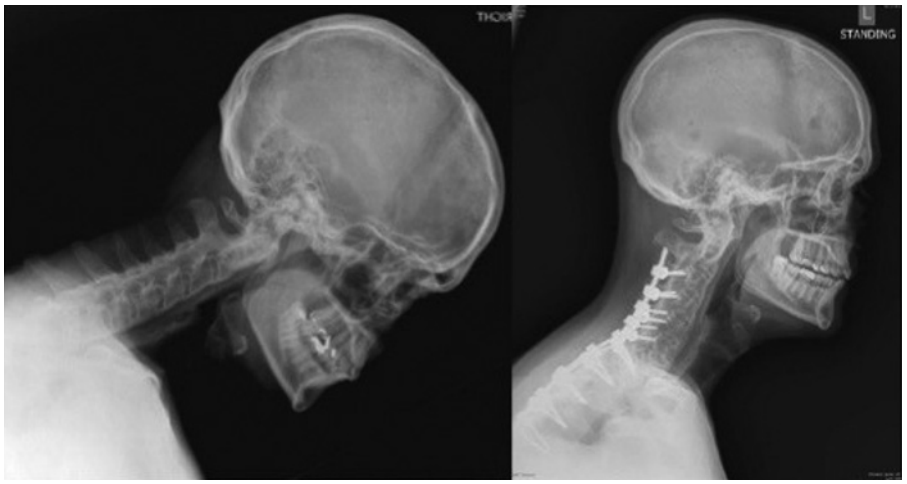


Figure 17.44 Ankylosing spondylitis A cervical spine fracture involving the C5–C6 level in a patient with AS. Note the low definition of X-ray to define the fracture on the left, further clarified by a CT scan that shows the instability pattern of this severe injury. Note also the fusion pattern affecting the multiple cervical segments.



(a) (b)

Figure 17.45 AS chin-on-chest Lateral X-ray views of an AS patient with chin-on-chest deformity (a) before and (b) after surgical correction. (Reproduced with permission from: Mehdian S, *et al*. Cervical osteotomy in ankylosing spondylitis. *Eur Spine J* 2012; **21**: 2713–17.)

Psoriatic arthritis

Axial involvement affects approximately half of patients with peripheral PsA. The involvement of the cervical spine is frequent, including atlantoaxial instability, new bone formation in the region of the dens and apophyseal joint changes.

Radiographically, parasyndesmophytes, osteitis and erosion of vertebral plates are typical features. The absence of sacroiliitis distinguishes psoriatic arthritis from AS. Erosive features with cortical bone resorption resemble RA, but enthesiophytes distinguish PsA from that condition.

MISCELLANEOUS PATHOLOGY

Spasmodic torticollis

Involuntary twisting or clonic movements of the neck characterize this condition, the most common form of focal dystonia. Spasms are sometimes triggered by emotional disturbance or attempts at correction. Even at rest the neck assumes an abnormal posture, the chin usually twisted to one side and upwards and the shoulder on that side often elevated. In some cases involuntary muscle contractions spread to other areas and the condition is revealed as a more generalized form of dystonia. The exact cause is unknown, but some cases are associated with lesions of the basal ganglia.

Correction is extremely difficult (Figure 17.46). Various drugs, including anticholinergics, have been used, although with little success. Some patients respond to local injections of botulin toxin into the sternocleidomastoid muscle.

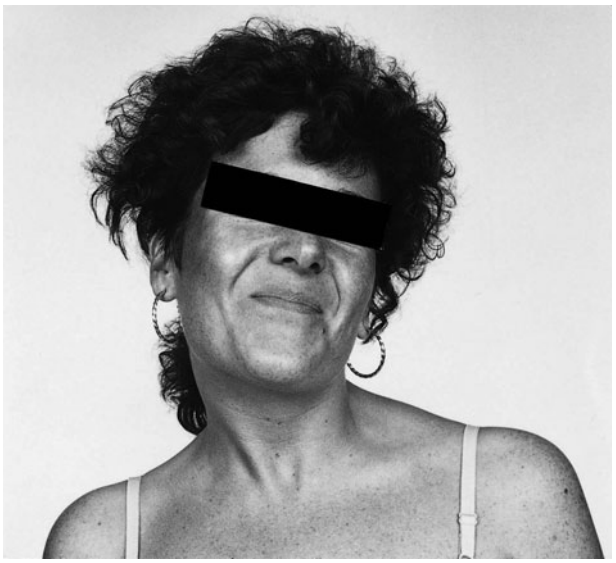


Figure 17.46 Spasmodic torticollis Attempted correction was forcibly resisted. The deformity can be very distressing.

Sarcoidosis of the cervical spine

Sarcoidosis is an *idiopathic multisystem disorder*. It most frequently occurs in Northern Europe, Japan and central USA, particularly in adults after the fourth decade of life, especially in women and Afro-Americans.

The most frequent form is the respiratory type. Bone involvement, when present, most often affects the small bones of the hands and feet, whereas spinal sarcoidosis is rare and usually involves the thoracolumbar region. Involvement of the vertebrae can present with an appearance similar to that of multifocal metastatic disease. The diagnosis for multiple enhancing vertebral lesions should include sarcoidosis, especially given typical lung findings or medical history of the disease.

Diagnosis is based on clinical and radiological findings and is confirmed by histological analysis of biopsy specimens, revealing the characteristic noncaseating granuloma.

Clinical features

The clinical course can be symptomless, but patients in the acute stage of the disease may present with symptoms and signs related to *Löfgren's syndrome* (i.e. *bilateral hilar lymphadenopathy*, *arthritis*, *erythema nodosum* and *fever*). In spinal sarcoidosis, patients may complain of axial pain that resolves spontaneously or after oral corticosteroids. Pathological fractures have been reported with associated neurological compromise.

The natural history of sarcoidosis is unpredictable as it can be progressive or resolve spontaneously.

Spinal cord sarcoidosis most commonly occurs at the cervical level, presenting with subacute or chronic myelopathy and focal weakness, potentially progressing to paraplegia.

Imaging

Chest X-ray, CT and gadolinium-enhanced MRI scans and PET with fluorodeoxyglucose are helpful methods for the diagnosis.

The reported radiographic appearance of vertebral sarcoid lesions is generally lytic with well-defined borders, but there may be a mixed pattern. The MRI may show multifocal lesions with T2 hyperintense signal and hypointense to isointense on T1-weighted images. The posterior spinal elements and the intervertebral discs are usually spared.

Spinal cord sarcoidosis should be borne in mind in the differential diagnosis of a high signal intensity area observed within the spinal cord on T2-weighted MRI images, in patients with spondylotic changes. For patients over the age of 40, differential diagnosis with malignancy is mandatory.

Treatment

Management of spinal sarcoidosis with steroids may be effective in the presence of neurological symptoms without spinal instability. The preferred treatment of spinal cord sarcoidosis is high-dose corticosteroid therapy.

The surgical management of spinal sarcoidosis should be reserved for cases of progressive vertebral destruction, spinal cord compression or instability secondary to fracture.

FURTHER READING

Cheung WY, Luk KDK. Pyogenic spondylitis. *International Orthopaedics (SICOT)* 2012; **36**: 397–404.

Dong F, Shen C, Jiang S, Zhang R, Song P, Yu Y, Wang S, Li X, Zhao G, Ding, C. Measurement of volume-occupying rate of cervical spinal canal and its role in cervical spondylotic myelopathy. *Eur Spine J* 2013; **22**: 1152–7.

Duarte RM, Vaccaro AR. Spinal infection: state of the art and management algorithm. *Eur Spine J* 2013; **22**: 2787–99.

Fujimori T, Le H, Hu SS, Chin C, Peckmezci M, Schairer W, Tay BK, Hamasaki T, Yoshikawa H, Iwasaki M. Ossification of the posterior longitudinal ligament of the cervical spine in 3161 patients: a CT-based study. *Spine* 2015; **40**: E394–E403.

Harrop JS, Naroji S, Maltenfort M, Anderson DG, Albert T, Ratliff JK, Ponnappan RK, Rihn JA, Smith HE, Hilibrand A, Sharan AD, Vacarro A. Cervical myelopathy: A clinical and radiographic evaluation and correlation to cervical spondylotic myelopathy. *Spine* 2010; **35**: 620–4.

Jurik AG. Imaging the spine in arthritis—a pictorial review. *Insights Imaging* 2011; **2**: 177–91.

Kato S. Fehlings MG. Prevalence of cervical spondylotic myelopathy. *Eur Spine J* 2015; **24**(Suppl 2): S139–S141.

Lukasiewicz AM, Bohl DD, Varthi AG, Basques BA, Webb ML, Samuel AM, Grauer JN. Spinal fracture in patients with ankylosing spondylitis – Cohort definition, distribution of injuries, and hospital outcomes. *Spine* 2016; **41**: 191–6.

Mak KC, Cheung KM. Surgical treatment of acute TB spondylitis: indications and outcomes. *Eur Spine J* 2013; **22**(Suppl 4): S603–S611.

Matsunaga S, Sakou T. Ossification of the posterior longitudinal ligament of the cervical spine etiology and natural history. *Spine* 2012; **37**: E309–E314.

Mori K, Imai S, Omura K, Saruhashi Y, Matsusue Y, Hukuda S. Clinical output of the rheumatoid cervical spine in patients with mutilating-type joint involvement for better activities of daily living and longer survival. *Spine* 2010; **35**: 1279–84.

Nouri A, Tetreaul L, Singh A, Karadimas SK, Fehlings, MG. Degenerative cervical myelopathy: Epidemiology, genetics, and pathogenesis. *Spine* 2015; **40**: E675–E693.

Oda T, Yonenobu K, Fujimura Y, Ishii Y, Nakahara S, Matsunaga S, Shimizu T, Matsumoto M. Diagnostic validity of space available for the spinal cord at C1 level for cervical myelopathy in patients with rheumatoid arthritis. *Spine* 2009; **34**: 1395–8.

Rao R. Neck pain, cervical radiculopathy, and cervical myelopathy: Pathophysiology, natural history and clinical evaluation. *J Bone Joint Surg Am* 2002; **84A**(10): 1871–81.

Rhee JM, Heflin JA, Hamasaki T, Freedman B. Prevalence of physical signs in cervical myelopathy – A prospective, controlled study. *Spine* 2009; **34**: 890–5.

Wierzbicki V, Pesce A, Marrocco L, Piccione E, Colonnese C, Caruso R. How old is your cervical spine? Cervical spine biological age: a new evaluation scale. *Eur Spine J* 2015; **24**: 2763–70.

Yoshikawa M, Doita M, Okamoto K, Manabe M, Sha N, Kurosaka M. Impaired postural stability in patients with cervical myelopathy: Evaluation by computerized static stabilometry. *Spine* 2008; **33**: E460–E464.

Yurube T, Sumi M, Nishida K, Takabatake M, Kohyama K, Matsubara T, Ozaki T, Maeno KR, Kakutani K, Zhang Z, Doita M. Progression of cervical spine instabilities in rheumatoid arthritis – A prospective cohort study of outpatients over 5 years. *Spine* 2011; **36**: 647–53.

The back

18

Robert Dunn & Nicholas Kruger

CLINICAL ASSESSMENT

Symptoms

The usual symptoms of back disorders are pain, stiffness and deformity in the back, and pain, paraesthesia or weakness in the lower limbs. The mode of onset is very important: did it start suddenly, perhaps after a lifting strain; or gradually without any antecedent event? Are the symptoms constant, or are there periods of remission? Are they related to any particular posture? Has there been any associated illness or malaise?

Pain, either sharp and localized or chronic and diffuse, is the commonest presenting symptom. Backache is usually felt low down and on either side of the midline, often extending into the upper part of the buttock and even into the lower limbs. Mechanical back pain is aggravated by activity and relieved by rest. It may originate from the disc, facets and ligamentous structures.

Sciatica, most commonly due to a prolapsed intervertebral disc pressing on a nerve root, is characteristically more intense than referred low back pain, is aggravated by coughing and straining and is often accompanied by symptoms of root pressure such as numbness and paraesthesiae, especially in the foot. It radiates from the buttock along the distribution of the affected nerve root.

Stiffness may be sudden in onset from muscular spasm ('locked back' attack, or a disc prolapse) or continuous and predictably worse in the mornings from inflamed spinal joints (suggesting arthritis or ankylosing spondylitis).

Deformity is usually noticed by others, or the patient may become aware of shoulder or breast asymmetry or poorly fitting clothes.

Numbness or *paraesthesia* is felt anywhere in the lower limb, or it may follow a dermatomal distribution. It is important to ask if it is aggravated by standing or walking and relieved by sitting down – the classic symptom of spinal stenosis.

Urinary retention or *incontinence* can be due to pressure on the cauda equina. *Faecal incontinence* or urgency, and *impotence*, may also occur.

Signs with the patient standing

Adequate exposure is essential; patients should strip to their underclothes. Refer to Figures 18.1 and 18.2

Look

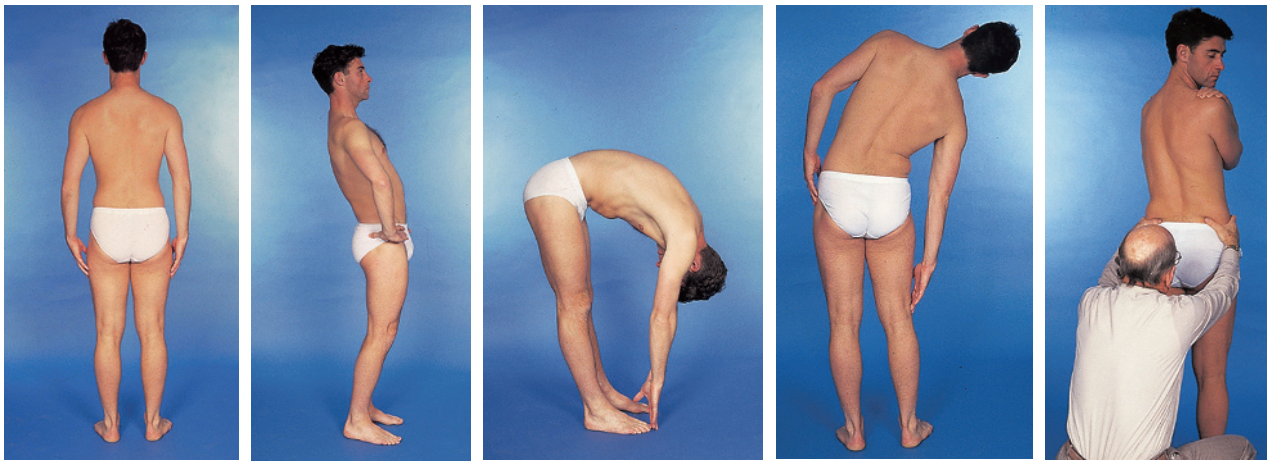
Start by examining the skin. Scars (previous surgery or injury), pigmentation (neurofibromatosis?) or abnormal tufts of hair (spina bifida?) are important clues to underlying spinal disorders. Look carefully at the patient's shape and posture, from both the front and behind. Asymmetry of the chest, trunk or pelvis may be obvious or may appear only when the patient bends forward. Lateral deviation of the spinal column is described as a *list* to one or other side; lateral curvature is *scoliosis*.

Seen from the side, the back normally has a slight forward curve, or *kyphosis*, in the thoracic region and a shorter backward curve, or *lordosis*, in the lumbar segment (the 'hollow' of the back). Excessive thoracic kyphosis is sometimes called *hyperkyphosis*, to distinguish it from the normal; if the spine is sharply angulated the prominence is called a *kyphos* or *gibbus*. The lumbar spine may be excessively lordosed (hyperlordosis) or unusually flat (effectively a lumbar kyphosis).

If the patient consistently stands with one knee bent (even though his legs are equal in length), this suggests nerve root tension on that side; flexing the knee relaxes the sciatic nerve and reduces the pull on the nerve root.

Feel

Palpate the spinous processes and interspinous ligaments, noting any unusual prominence or 'steps'. Tenderness should be localized to: (1) bony structures; (2) intervertebral tissues; (3) paravertebral



(a) (b) (c) (d) (e)

Figure 18.1 Examination With the patient standing upright (a), look at his general posture and note particularly the presence of any asymmetry or frank deformity of the spine. Then ask him to lean backwards (extension) (b), forwards to touch his toes (flexion) (c) and then sideways as far as possible (d), comparing his level of reach on the two sides. Finally, hold the pelvis stable and ask the patient to twist first to one side and then to the other (rotation). Note that rotation occurs almost entirely in the thoracic spine (e) and not in the lumbar spine.

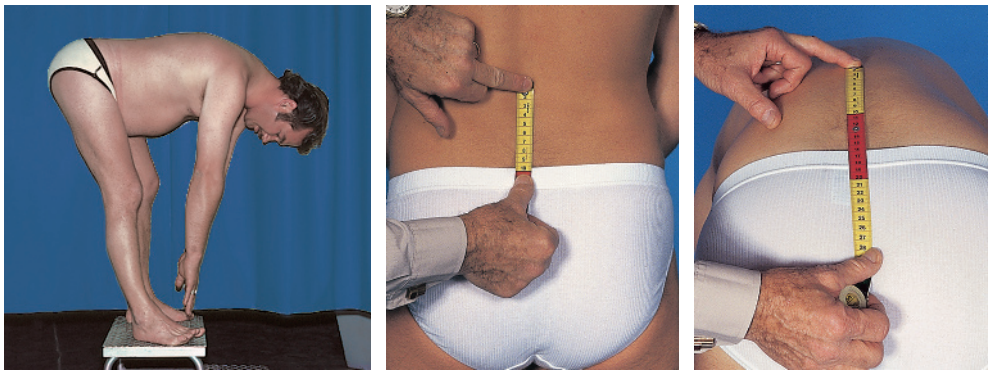
muscles and ligaments, especially where they insert into the iliac crest.

Move

Flexion is tested by asking the patient to try to touch his toes (Figure 18.2). With a stiff back the movement occurs at the hips and there may be no spinal excursion.

The mode of flexion (whether it is smooth or hesitant) and the way in which the patient comes back to the upright position are also important. With mechanical back pain, the patient tends to regain the upright position by pushing on the front of their thighs. To test *extension*, ask the patient to lean backwards, without bending their knees. Poor extension may be due to facet pathology or spinal stenosis. The '*wall test*' will unmask a minor flexion deformity (kyphosis, as in ankylosing spondylitis or Scheuermann's osteochondrosis); standing with the back flush against a wall, the heels, buttocks, shoulders and occiput should all make contact with the vertical surface.

Lateral flexion is tested by asking the patient to bend sideways, sliding their hand down the outer side of the leg; the two sides are compared. *Rotation* is examined by asking them to twist the trunk to each side in turn while the pelvis is anchored by the



(a) (b) (c)

Figure 18.2 Measuring the range of flexion Bending down and touching the toes may look like lumbar flexion but this is not always the case. The patient in (a) has ankylosing spondylitis and a rigid lumbar spine, but he is able to reach his toes because he has good flexibility at the hips. Compare his flat back with the rounded back of the model in Figure 18.1c. You can measure the lumbar excursion. With the patient upright, select two bony points 10 cm apart and mark the skin (b); as the patient bends forward, the two points should separate by at least a further 5 cm (c).

examiner's hands; this is essentially a thoracic movement and is not limited in lumbosacral disease.

Rib-cage excursion is assessed by measuring the *chest circumference* in full expiration and then in full inspiration; the normal difference is about 7 cm. A reduced excursion may be the earliest sign of ankylosing spondylitis.

Ask the patient to stand on their toes (plantarflexion) and on their heels (dorsiflexion) as a useful screen for motor power in the legs; small differences between the two sides are easily spotted.

Signs with the patient lying prone

Make sure that the patient is lying comfortably on the examination couch, and remove the pillow so that they are not forced to arch their back. Again, look for localized deformities and muscle spasm, and examine the buttocks for *gluteal wasting*. Feel the *bony outlines* (is there an unexpected 'step' or prominence?) and check for consistently localized lumbosacral *tenderness* or soft-tissue 'trigger' points. The popliteal and posterior tibial *pulses* are felt, hamstring *power* is tested and *sensation* on the back of the limbs assessed.

The *femoral nerve stretch test* (for lumbar 3rd or 4th nerve root irritation) is performed by flexing the patient's knee and lifting the hip into extension; pain may be elicited down the front of the thigh (Figure 18.3).

Signs with the patient lying supine

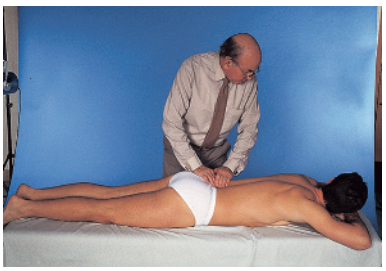
The patient is observed as they turn – is there pain or stiffness? A rapid appraisal of the thyroid, chest (and breasts), and abdomen (and scrotum) is advisable, and essential if there is even a hint of generalized disease. Hip and knee mobility are assessed before testing for spinal cord or root involvement.

The *straight-leg raising test* discloses lumbosacral root tension (Figure 18.4a,b). With a straight knee, the leg is slowly raised until pain is produced – not merely in the lower back (which is common and not significant) but also in the buttock, thigh and calf. The angle at which pain occurs is noted. Lasègue's test reproduces pain by extending the knee in a leg which has already been lifted. Normally it should be possible to raise the limb to 80–90 degrees; people with hypermobility can go even further. In disc prolapse with nerve root compression, straight-leg raising may be restricted to less than 30 degrees because of severe pain. At the point where the patient experiences discomfort, passive dorsiflexion of the foot may cause an additional stab of sciatic pain.

The *'bowstring sign'* is even more specific (Figure 18.4c). Raise the patient's leg gently to the point where they experience sciatic pain; now, without reducing the amount of lift, bend the knee so as to relax the sciatic nerve. Buttock pain is immediately relieved; pain may then be re-induced without extending the knee by simply pressing on the lateral popliteal nerve behind the lateral tibial condyle, to tighten it like a bowstring.

Occasionally straight-leg raising on the unaffected side produces pain on the affected side. This *crossed straight-leg raise test* is highly specific for a disc prolapse, often a large central prolapse. Cauda equina syndrome should be excluded.

A *full neurological examination* of the lower limbs is carried out, systematically assessing each dermatome, myotome and reflex. The Babinski sign and ankle clonus should always be assessed, especially in elderly patients. Circulatory assessment includes abdominal palpation for aortic aneurysm, femoral, popliteal and foot pulses and the presence of trophic changes. Hip and sacroiliac joints should be routinely screened for pain and stiffness and any leg length discrepancies documented.



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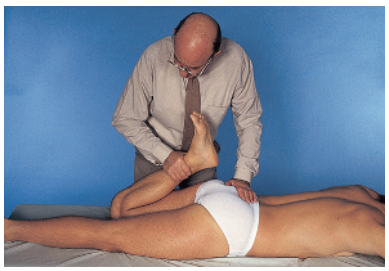

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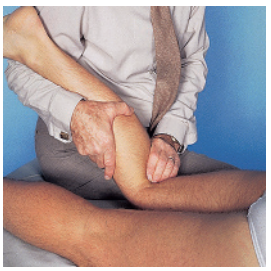
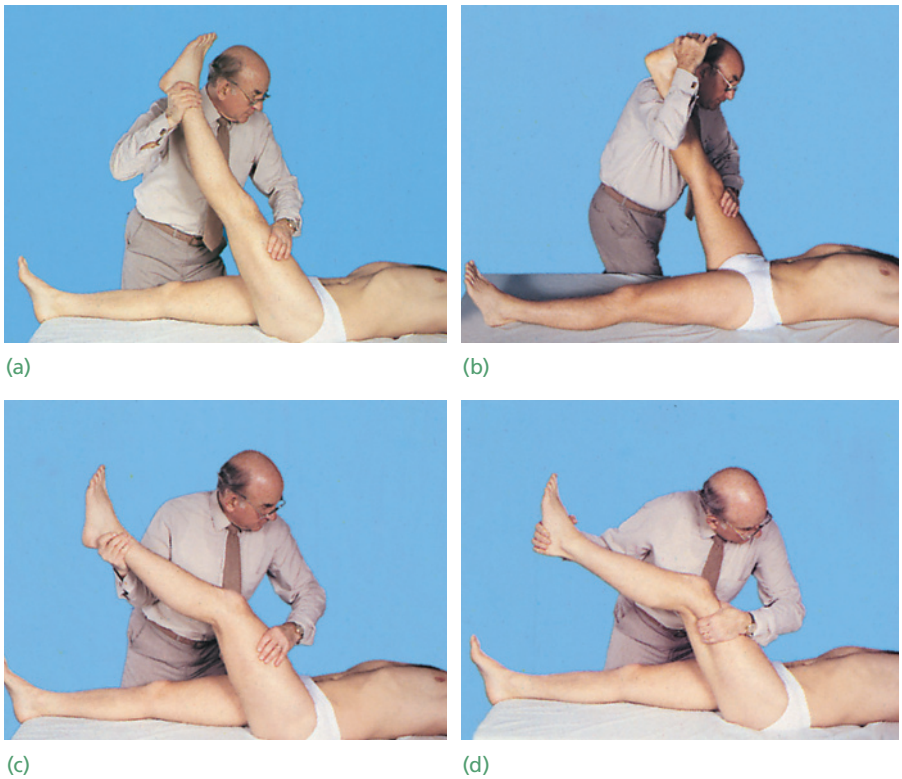


(c)

Figure 18.3 Examination with the patient prone (a) Feel for tenderness, watching the patient's face for any reaction. (b) Performing the femoral stretch test. You can test for lumbar root sensitivity either by hyperextending the hip or by acutely flexing the knee with the patient lying prone. Note the point at which the patient feels pain and compare the two sides. (c) While the patient is lying prone, take the opportunity to feel the pulses. The popliteal pulse is easily felt if the tissues at the back of the knee are relaxed by slightly flexing the knee.



(a) (b) (c) (d)

Figure 18.4 Sciatic stretch tests (a) Straight-leg raising. The knee is kept absolutely straight while the leg is slowly lifted (or raised by the patient); note where the patient complains of tightness and pain in the buttock – this normally occurs around 80–90°. (b) At that point a more acute stretch can be applied by passively dorsiflexing the foot – this may cause an added stab of pain. (c) The 'bowstring sign' is a confirmatory test for sciatic tension. At the point where the patient experiences pain, relax the tension by bending the knee slightly; the pain should disappear. Then apply firm pressure behind the lateral hamstrings to tighten the common peroneal nerve (d); the pain recurs with renewed intensity.

Imaging

Plain X-rays

For the lower back, standing anteroposterior and lateral X-rays of the lumbar spine (Figure 18.5) and anteroposterior pelvis X-rays are required; occasionally lumbar oblique and sacroiliac joint views are useful.

In the anteroposterior view the spine should be perfectly straight and the soft-tissue shadows should outline the normal muscle planes. Curvature (scoliosis) is obvious, and best shown in standing views. Bulging of the psoas muscle or loss of the psoas shadow may indicate a paravertebral abscess. Individual vertebrae may show alterations in structure, such as asymmetry or collapse. Identify the pedicles: a missing or misshapen pedicle could be due to erosion by infection, a neurofibroma or metastatic disease.

In the lateral view the normal thoracic kyphosis (up to 40 degrees) and lumbar lordosis should be regular and uninterrupted. Vertebral anterior shift (spondylolisthesis) may be associated with defects of the posterior arch, best illustrated with oblique views. Vertebral bodies, which should be rectangular, may be wedged or biconcave, deformities typical of osteoporosis or old injury. Bone density and trabecular markings also are best seen in lateral films. Lateral views in flexion and extension may reveal excessive intervertebral movement, a possible cause of back pain.

The intervertebral spaces may be edged by bony spurs (suggesting long-standing disc degeneration) or bridged by fine bony syndesmophytes (a cardinal feature of ankylosing spondylitis). The sacroiliac joints may show erosion or ankylosis, as in tuberculosis (TB) or ankylosing spondylitis, and the hip joints may show arthrosis, not to be missed in the older patient with backache.

Radioisotope scanning

Isotope scans may pick up areas of increased activity, suggesting a fracture, a local inflammatory lesion or a 'silent' metastasis. Bone scans may include whole-body, three-phase, or regional imaging and single-photon emission computed tomography (SPECT).

Computed tomography

Computed tomography (CT) is helpful in the diagnosis of structural bone changes (e.g. vertebral fracture) and intervertebral disc prolapse although this has largely been superseded by MRI. CT scan is very useful to assess spinal implant placement but requires *myelography* to demonstrate the dural contents.

Discography and facet joint arthrography

These investigations for chronic back pain are largely obsolete due to low specificity and poor correlation with symptoms. There are also concerns that discography can accelerate degeneration in the lumbar discs.

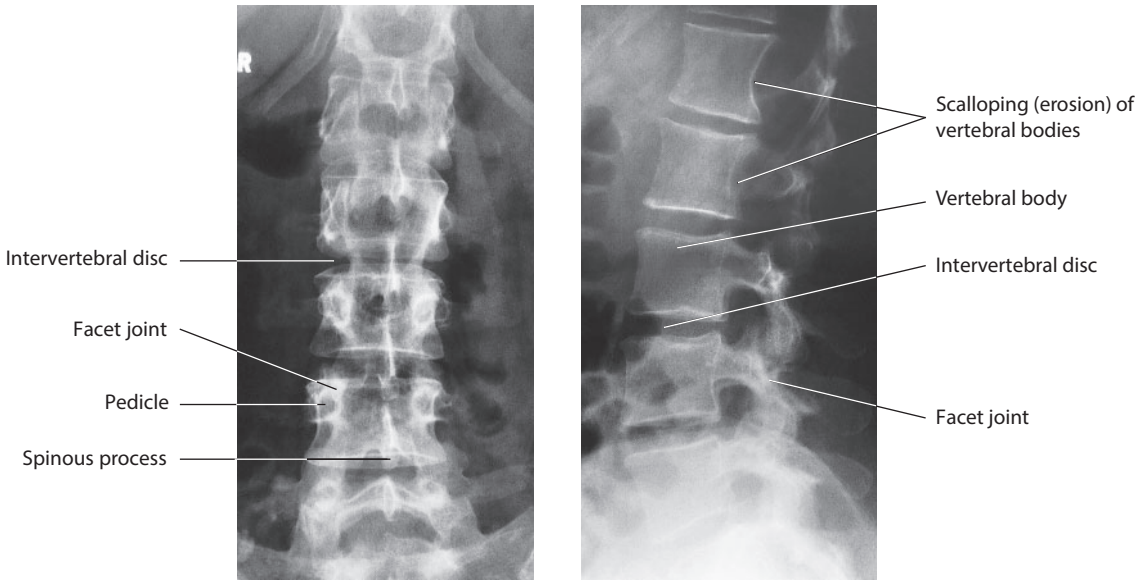


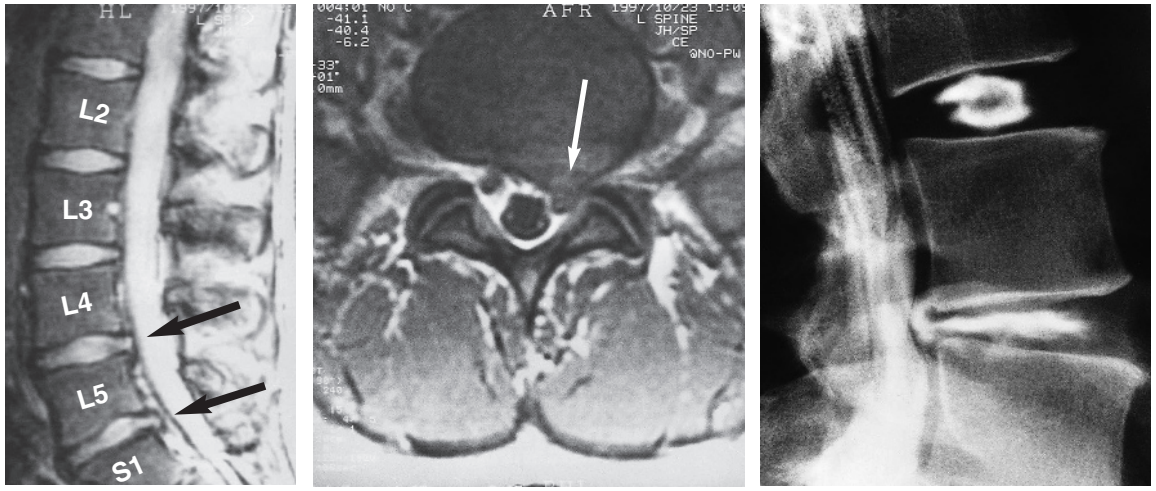
Figure 18.5 Lumbar spine X-rays The most important normal features are demonstrated in the lower lumbar spine. In this particular case there are also signs of marked posterior vertebral body and facet joint erosions at L1 and L2, features that are strongly suggestive of an expanding neurofibroma.

Magnetic resonance imaging

MRI has virtually done away with the need for myelography, discography, facet arthrography, and much of CT scanning. The spinal canal and disc spaces are clearly outlined in various planes (Figure 18.6). Scans can reveal the physiological state of the disc as regards dehydration, as well as the effect of disc degeneration on bone marrow in adjacent vertebral bodies.

SPINAL DEFORMITIES

'Spinal deformity' refers to the loss of normal alignment of a straight spine in the coronal plane or the cervical lordosis, thoracic kyphosis and lumbar lordosis in the sagittal plane. It may be due to a congenital or developmental malformation. The latter may be idiopathic, associated with neuromuscular conditions or the consequence of degenerative processes.



(a) (b) (c)

Figure 18.6 MRI and discography (a) The lateral T2-weighted MRI shows a small posterior disc bulge at L4/5 and a larger protrusion at L5/S1. (b) The axial MRI shows the disc prolapse encroaching on the intervertebral canal and the nerve root on the left side. (c) Discography, showing normal appearance at the upper level and a degenerate disc with prolapse at the level below.

SCOLIOSIS

Scoliosis is a complex rotational deformity which may manifest with a thoracic or lumbar prominence, shoulder imbalance, coronal shift and infrequently pain.

Two broad types of deformity are defined: *postural* and *structural*.

Postural scoliosis

In postural scoliosis the deformity is secondary, or compensatory, to some condition outside the spine, such as a short leg, or pelvic tilt due to contracture of the hip. When the patient sits (thereby cancelling leg length asymmetry), the curve disappears. Local muscle spasm associated with a prolapsed lumbar disc may cause a skew back; although sometimes called 'sciatic scoliosis' this, too, is a spurious deformity. The scoliosis is usually mild and has minimal rotation (Figure 18.7).

Structural scoliosis

In structural scoliosis there is a non-correctable deformity of the affected spinal segment, an essential component of which is vertebral rotation (Figure 18.8). The spinous processes point towards the concavity of the curve and the transverse processes on the convexity rotate posteriorly. In the thoracic region the ribs on the convex side stand out prominently, producing the rib hump, which is a characteristic part of the overall deformity. Dickson and co-workers pointed out in the 1980s that this is really a lordoscoliosis associated with rotational buckling of the spine. The initial deformity is probably correctable but, once it exceeds a certain point of mechanical stability, the spine buckles and rotates into a fixed deformity that does not disappear with changes in posture. *Secondary (compensatory) curves* nearly always develop to counterbalance the primary deformity; they are usually less marked and more flexible but with time they, too, become fixed.

The deformity tends to progress throughout the growth period. Thereafter, further deterioration is slight, although curves greater than 50 degrees may go on increasing by 0.5–1 degree per year. With very severe curves, chest deformity is marked and cardiopulmonary function is usually affected.

The largest group is termed *idiopathic scoliosis* with no obvious cause but appears to have a genetic basis and a predictable course. Other causes are *congenital* failure of vertebral formation or segmentation, within the *neuromuscular group* ranging from cerebral palsy, spinal muscular atrophy to muscle dystrophies, and a miscellaneous group of connective-tissue disorders.

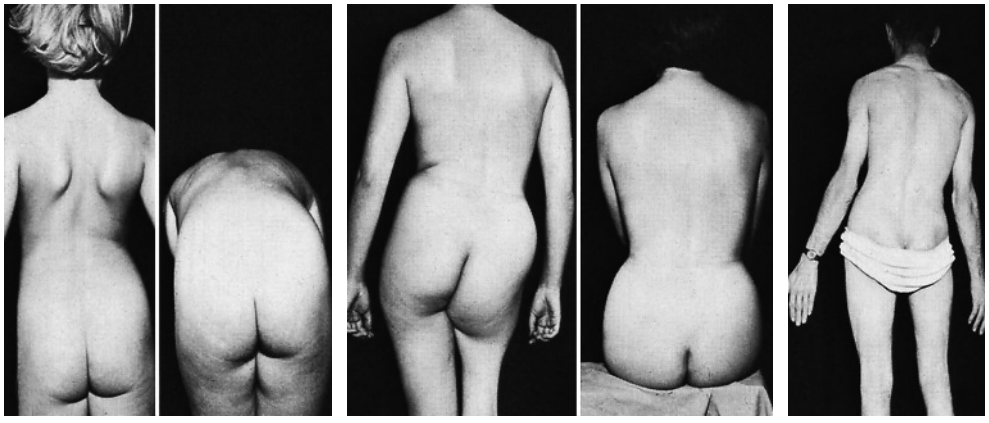
Clinical features

Deformity is usually the presenting symptom: an obvious skew back or rib hump in thoracic curves, and asymmetrical prominence of one hip or flank crease in thoracolumbar curves. Balanced curves sometimes pass unnoticed until quite severe. Where school screening programmes are conducted, children will be referred with very minor deformities and the recent evidence suggests that screening is not justified.

Pain is a rare complaint and should alert the clinician to the possibility of an unusual underlying cause and the need for investigation. There may be a *family history* of scoliosis or a record of some abnormality *during pregnancy* or *childbirth*; the *early developmental milestones* should be noted.

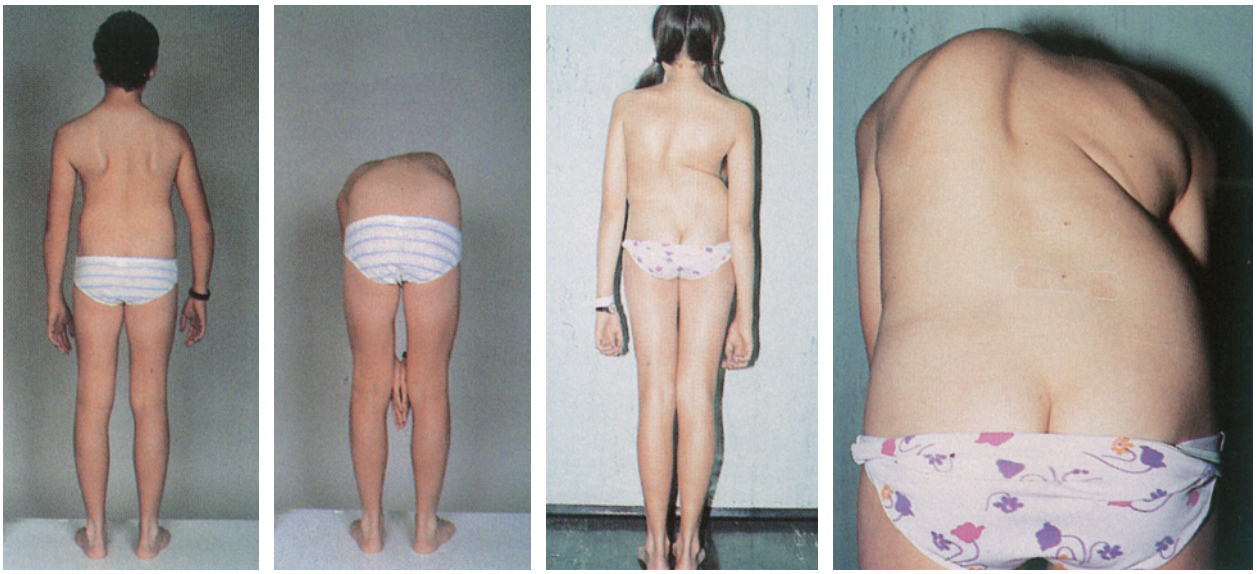
The trunk should be completely exposed and the patient examined from in front, the back and the side. *Skin* pigmentation and congenital anomalies such as sacral dimples or hair tufts are sought.

The *spine* may be obviously deviated from the midline, or a prominence may become apparent when the patient bends forward (the *Adams test*). The level and direction of the major curve convexity are noted (e.g. 'right thoracic' means a curve in the thoracic spine and convex to the right). The hip (pelvis) sticks out on the concave side and the scapula on the convex. The breasts and shoulders may be asymmetrical.



(a) (b) (c)

Figure 18.7 Postural scoliosis (a) This young girl presented with thoracolumbar 'curvature'. When she bends forwards, the deformity disappears; this is typical of a postural or mobile scoliosis. (b) Short-leg scoliosis disappears when the patient sits. (c) Sciatic scoliosis disappears when the prolapsed disc settles down or is removed.



(a) (b) (c) (d)

Figure 18.8 Structural scoliosis (a) Slight curves are often missed on casual inspection but the deformity becomes apparent when the spine is flexed (b). The young girl in (c) has a much more obvious scoliosis and asymmetry of the hips but what really worries her is the prominent rib hump, seen best when she bends over (d).

With thoracic scoliosis, rotation causes the rib angles to protrude, thus producing an asymmetrical rib hump on the convex side of the curve. In balanced deformities the occiput is over the midline; in unbalanced (or decompensated) curves it is not. This can be determined more accurately by dropping a plumbline from the prominent spinous process of C7 and noting whether it falls along the gluteal cleft. Spinal mobility should be assessed and the effect of lateral bending on the curve noted; is there some flexibility in the curve and can it be passively corrected?

Side-on posture should also be observed. There may appear to be excessive kyphosis or lordosis.

Neurological examination is important including abdominal reflexes which may identify syringomyelia. Any neurological abnormality calls for MRI investigation.

Leg length is measured. If one side is short, the pelvis is levelled by standing the patient on wooden blocks and the spine is re-examined.

General examination includes a search for the possible cause and an assessment of cardiopulmonary function (which is reduced in severe curves).

Imaging

PLAIN X-RAYS

Full-length posteroanterior (PA) and lateral X-rays of the spine and iliac crests must be taken with the patient standing erect. Structural curves show vertebral rotation: in the PA X-ray, vertebrae towards the apex of the curve appear to be asymmetrical and the spinous processes are deviated towards the concavity. Remember that PA in relation to the patient is not PA in relation to the rotated vertebrae!

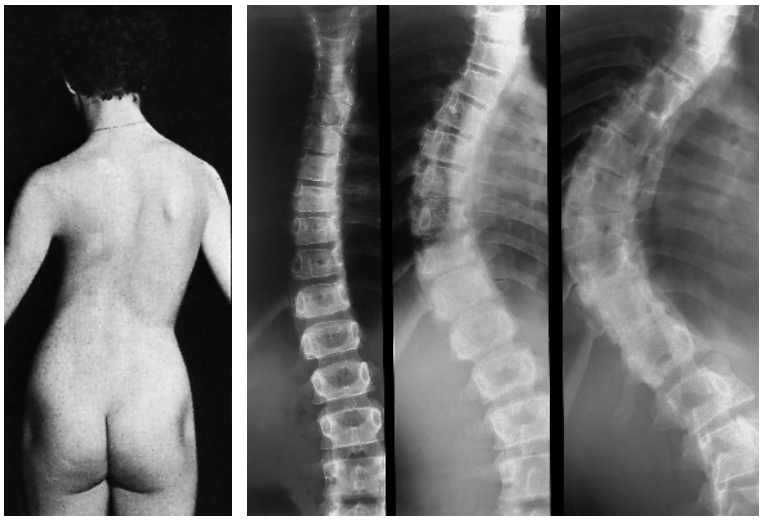
The upper and lower ends of the curve are identified as the levels where vertebrae start to angle away from the curve. The degree of curvature is measured by drawing lines on the X-ray at the upper border of the uppermost vertebra and the lower border of the lowermost vertebra of the curve; the angle subtended by these lines is the *angle of curvature* (*Cobb's angle*).

The site of the curve apex should be noted. Right thoracic curves are the commonest, the great majority in girls in adolescent idiopathic scoliosis (Figure 18.9). Left thoracic curves are unusual and have a higher incidence of underlying neurological pathology in the adolescent group justifying MRI investigation. The primary structural curve is usually balanced by compensatory curves above and below, or by a second 'primary' curve also with vertebral rotation (sometimes there are multiple 'primary' curves).

What is not readily appreciated from these films is the degree of lordosis in the primary curve(s) and kyphosis in the compensatory curves; indeed it is postulated that flattening or reversal of the normal thoracic kyphosis superimposed on coronal plane asymmetry leads, with growth, to progressive idiopathic scoliosis. Lateral bending views are taken to assess the degree of curve correctability.

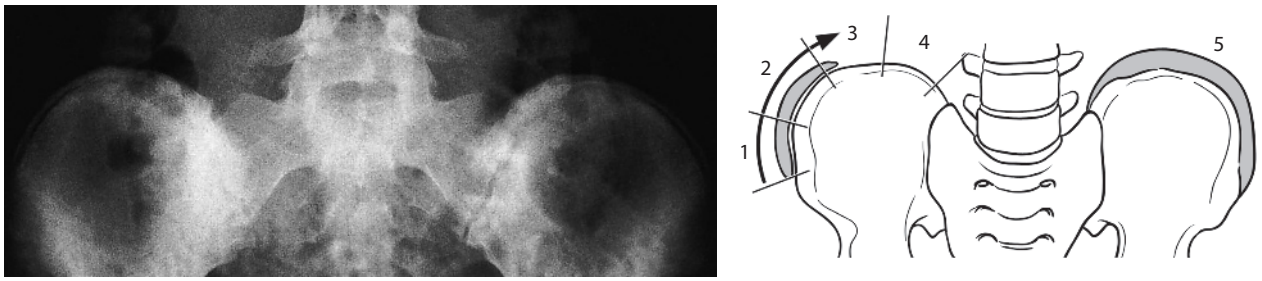
SKELETAL MATURITY – RISSER'S SIGN

Skeletal maturity is assessed in several ways. This is important because the curve often progresses most during the period of rapid skeletal growth and maturation. The iliac apophyses start ossifying shortly after



(a) (b) (c)

Figure 18.9 Adolescent idiopathic scoliosis (a) Typical thoracic deformity. (b) Serial X-rays show how this curve increased over a period of 4 years. (c) The angle of curvature is measured on the X-ray by Cobb's method: lines projected from the top of the uppermost and the bottom of the lowermost vertebral bodies in the primary curve define Cobb's angle.



(a) (b)

Figure 18.10 Risser's sign The iliac apophyses normally appear progressively from lateral to medial (stages 1–4). When fusion is complete, spinal maturity has been reached and further increase of curvature is negligible (stage 5).

puberty; ossification extends medially and, once the iliac crests are completely ossified, further progression of the scoliosis is minimal (*Risser's sign*: Figure 18.10). This stage of development usually coincides with fusion of the vertebral ring apophyses. 'Skeletal age' may also be estimated from X-rays of the wrist and hand.

SPECIAL IMAGING

CT and MRI may be necessary to define a vertebral abnormality or cord compression.

Special investigations

Pulmonary function tests are performed in all cases of severe chest deformity. A marked reduction in vital capacity is associated with diminished life expectancy and carries obvious risks for surgery. Patients with muscular dystrophies or connective-tissue disorders require full *biochemical and neuromuscular investigation* of the underlying condition.

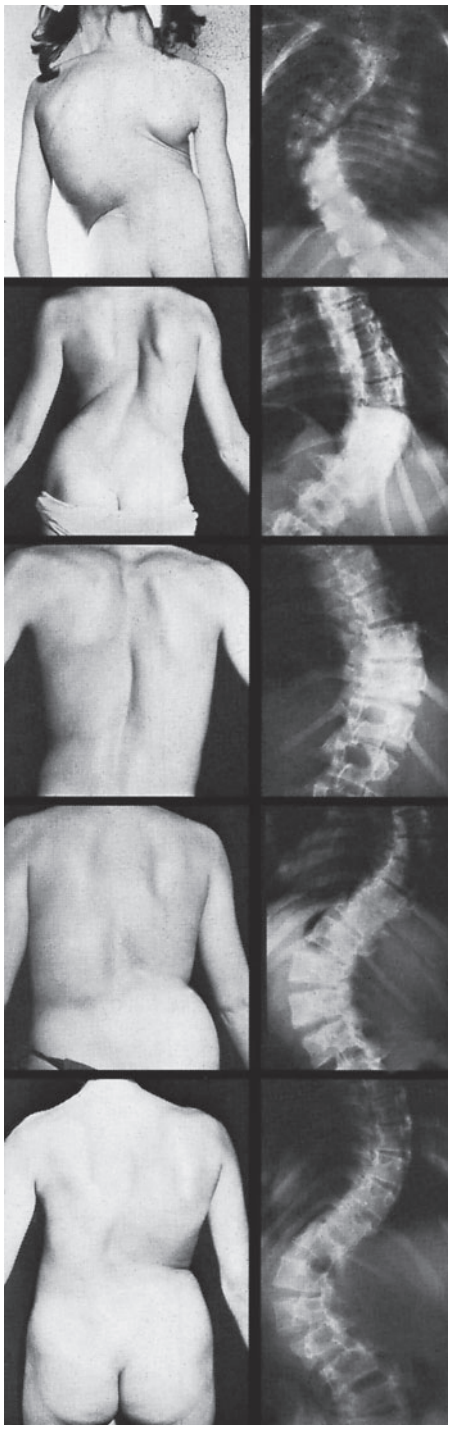
Prognosis and treatment

Prognosis is the key to treatment: the aim is to prevent severe deformity. Generally speaking, the younger the child and the higher the curve magnitude the worse is the prognosis. Management differs for the different types of scoliosis, which are considered later.

IDIOPATHIC SCOLIOSIS

This group constitutes about 80% of all cases of scoliosis. The deformity is often familial and the population incidence of serious curves (over 30 degrees and therefore needing treatment) is 3 per 1000; trivial curves are very much more common. The age at onset has been used to define three groups: *adolescent*, *juvenile* and *infantile*. A simpler division now in general use is *early-onset scoliosis* (before puberty) and *late-onset scoliosis* (after puberty).

Patterns of idiopathic scoliosis are summarized in Figure 18.11.



Infantile thoracic

60% male.
90% convex to left.
Associated with ipsilateral plagiocephaly.
May be resolving or progressive.
Progressive variety becomes severe.

Adolescent thoracic

90% female.
90% convex to right.
Rib rotation exaggerates the deformity.
50% develop curves of greater than 70°.

Thoracolumbar

Slightly more common in females.
Slightly more common to right.
Features mid-way between adolescent thoracic and lumbar.

Lumbar

More common in females.
80% convex to left.
One hip prominent but no ribs to accentuate deformity.
Therefore not noticed early, but backache in adult life.

Combined

Two primary curves, one in each direction.
Even when radiologically severe, clinical deformity relatively slight because always well balanced.

Figure 18.11 Patterns of idiopathic scoliosis Bracing is used far less than previously because of serious doubts as to its effectiveness beyond natural history.

Late-onset (adolescent) idiopathic scoliosis (aged 10 or over)

Late-onset idiopathic scoliosis is the commonest type, occurring in 90% of cases, mostly in girls. Primary thoracic curves are usually convex to the right, lumbar curves to the left; intermediate (thoracolumbar) and combined (double primary) curves also occur. Progression is not inevitable; indeed, most curves less than 20 degrees either resolve spontaneously or remain unchanged. However, once a curve starts to progress, it usually goes on doing so throughout the remaining growth period (and, to a much lesser degree, beyond that). Reliable predictors of progression are: (1) a very young age; (2) marked curvature; and (3) an incomplete Risser sign at presentation.

In prepubertal children, rapid progression is liable to occur during the growth spurt.

Treatment

The aims of treatment are: (1) to prevent a mild deformity from becoming severe; and (2) to correct an existing deformity that is unacceptable to the patient. A period of preliminary observation may be needed before deciding between conservative and operative treatment. At 4 to 9-monthly intervals the patient is examined, photographed and X-rayed so that curves can be measured and checked for progression.

NON-OPERATIVE TREATMENT

If the patient is approaching skeletal maturity and the deformity is acceptable (which usually means it is less than 30 degrees and well balanced), treatment is probably unnecessary unless sequential X-rays show definitive evidence of progression.

Exercises are often prescribed; they have no effect on the curve but they do maintain muscle tone and may inspire confidence in a favourable outcome.

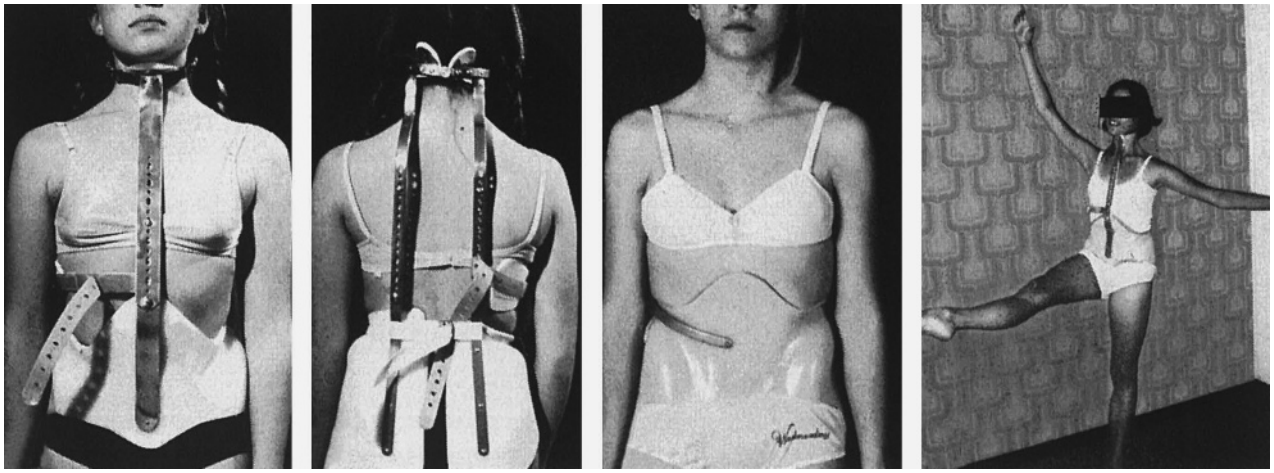
Bracing has been used for many years in the treatment of progressive scoliotic curves between 20 and 30 degrees (Figure 18.12). A variety of braces are available, some cast and others prefabricated. They generally include the thoracic and lumbar spine to the pelvis with pads to push the spine into a more normal alignment. Compliance determines success and they need to be worn for more than 20 hours a day – only to be removed for sport and bathing. If successful, the final curve magnitude will be the same as when the bracing was initiated, not straight. For these reasons, bracing is not popular, especially in warmer regions where compliance is especially poor.

OPERATIVE TREATMENT

Surgery is indicated for curves that are predicted to be more than 50 degrees at maturity in growing patients or those with established large, cosmetically unacceptable curves. The objectives are: (1) to halt progression of the deformity; (2) to restore the normal spinal contours with instrumentation; and (3) to arthrodese the entire primary curve by bone grafting.

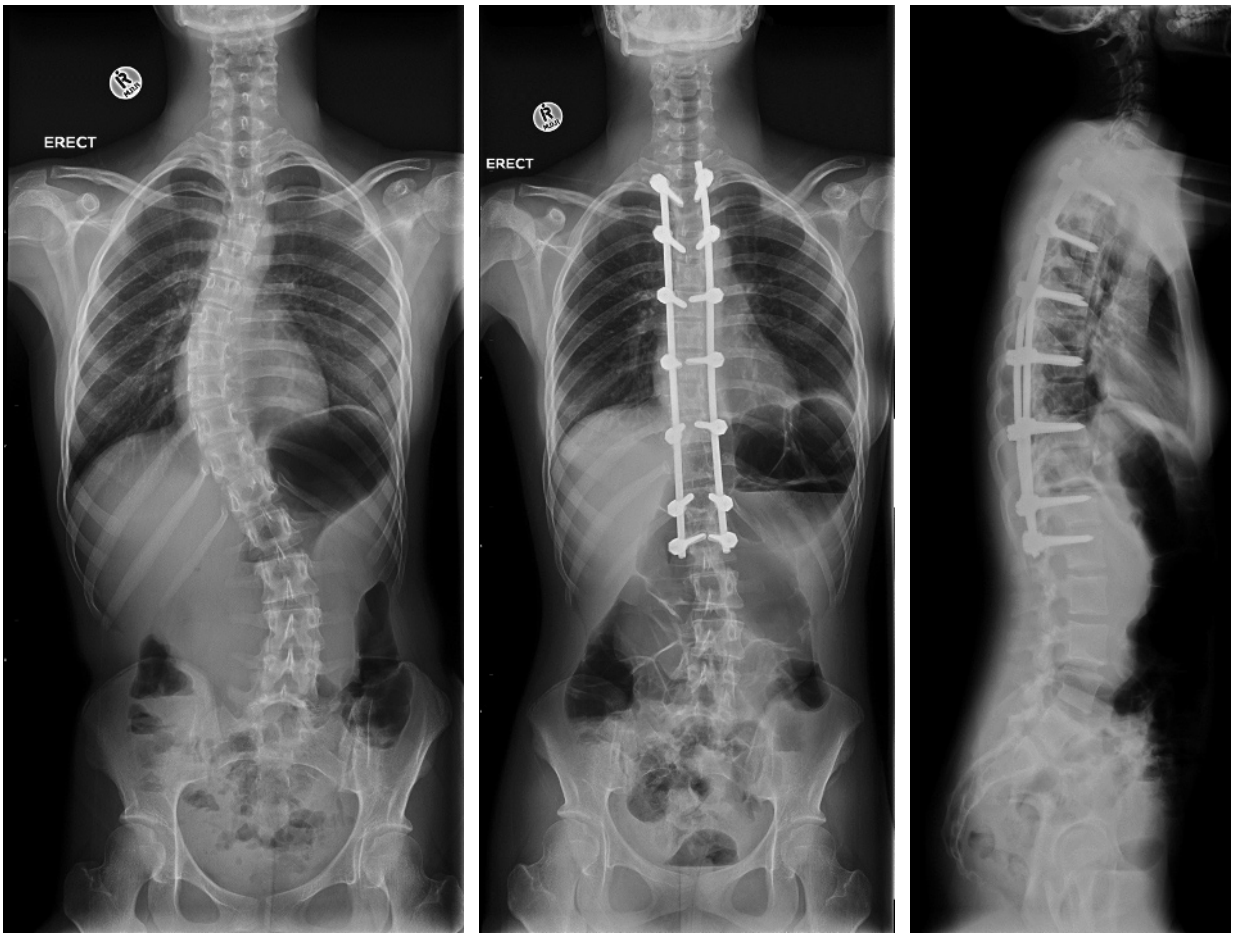
In *posterior instrumentation* the spine is instrumented segmentally from posterior with pedicle screws and hooks which are connected to pre-contoured rods to correct the deformity via the mobile discs (Figure 18.13). Older sub-laminar wiring techniques may still be used if pedicles are too small for screws or to reduce cost in long neuromuscular scoliosis. If the deformity is rigid, it may require resection of the facets (ponte osteotomies) and even concave rib resections to allow correction.

Anterior surgery is another option where the discs are resected and screws placed into the vertebral bodies, straightened with the addition of the rod. The advantage may be a shorter construct but increased morbidity of a transthoracic approach. It is useful in thoracolumbar curves where excellent rotational correction can be induced with the disc release.



(a) (b) (c) (d)

Figure 18.12 Structural scoliosis – bracing (a,b) The Milwaukee brace fits snugly over the pelvis below; chin and head pads promote active postural correction and a thoracic pad presses on the ribs at the apex of the curve. (c) The Boston brace is used for low curves. All braces are cumbersome, but (d) if well-made they need not interfere much with activity. Nowadays bracing is used far less often than before because of doubts about its ability to alter the natural progress of structural scoliosis.



(a) (b) (c)

Figure 18.13 Scoliosis – posterior instrumentation (a) Preoperative anteroposterior X-ray; (b) postoperative anteroposterior X-ray; (c) postoperative lateral X-ray.

WARNING: During correction, spinal cord injury may occur due to cord traction with column lengthening. Spinal cord electrophysiological monitoring should be performed, ideally both somatosensory and motor-evoked potential monitoring, during spinal correction. If these facilities are not available or there is an electrophysiological alert, the 'wake-up test' is used. Anaesthesia is reduced to bring the patient to a semi-awake state and he or she is then instructed to move their feet. If there are signs of cord compromise, the instrumentation is relaxed or removed and reapplied with a lesser degree of correction.

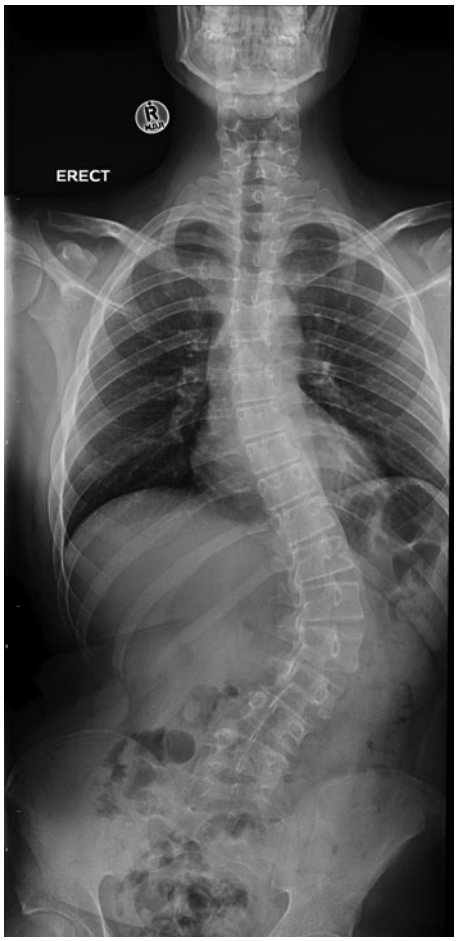
Rib hump In severe, rigid curves, even the best of the instrumentation systems cannot completely eliminate the rib hump – and it is often this that troubles the patient most of all. If the deformity is marked, it can be reduced significantly by performing a costoplasty, where short sections of rib are excised at multiple levels on the rib hump (convex) side, close to the vertebral articulation.

Complications of surgery

Neurological compromise With modern techniques the incidence of permanent paralysis has been reduced to less than 1%. From the patient's point of view this is small comfort. Every effort should be made to provide adequate safeguards.

Pseudarthrosis Incomplete fusion occurs in about 2% of cases and may require further operation and grafting.

Implant failure Implants may dislodge and rods fracture especially in delayed/non-union.



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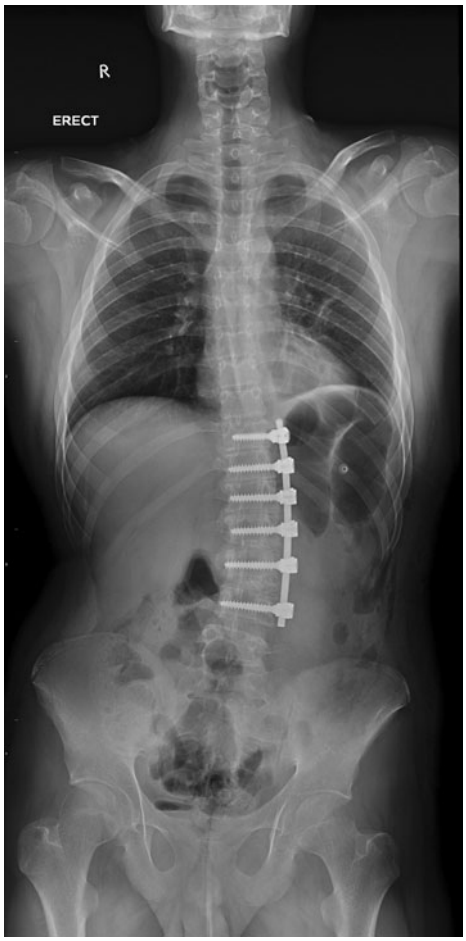


(b)

Figure 18.14 Scoliosis – anterior instrumentation (a) Preoperative anteroposterior X-ray; (b) postoperative anteroposterior X-ray.

Early-onset (juvenile) idiopathic scoliosis

Presenting in children aged 4–9 years, this type of idiopathic scoliosis is uncommon. The characteristics of this group are similar to those of the adolescent group, but the prognosis is worse and surgical correction may be necessary before puberty. However, if the child is very young, a brace may control the curve until the age of 10 years, when fusion can be performed without severe pulmonary effects.

Early-onset (infantile) idiopathic scoliosis

This variety is rare. Boys predominate and most curves are thoracic with convexity to the left (Figure 18.15). Although 90% of infantile curves resolve spontaneously, progressive curves can become very severe; those in which the rib–vertebra angle at the apex of the curve differs on the two sides by more than 20 degrees are likely to deteriorate. Because this also influences the development of the lungs, there is a high incidence of cardiopulmonary dysfunction.

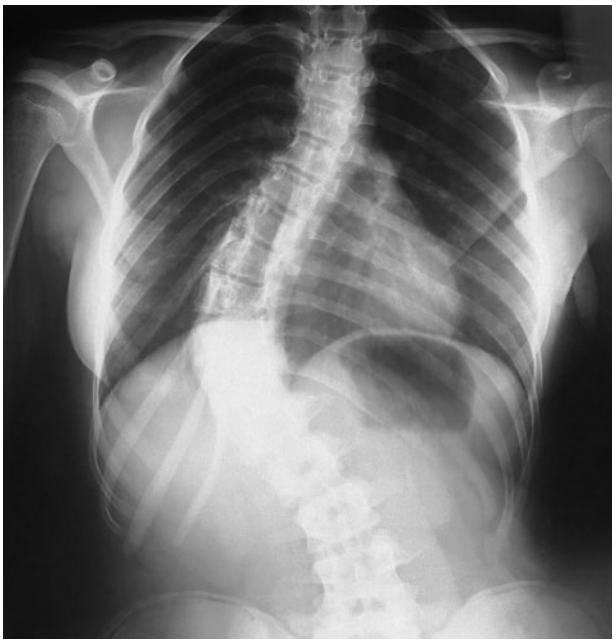


Figure 18.15 Early-onset scoliosis 'Idiopathic' curves in young children usually resolve, but some increase progressively and become very severe. Measurement of the rib–vertebra angles at the curve apex in the early stages of the deformity is a good prognostic indicator.

Curves assessed as being potentially progressive should be treated by applying serial elongation–derotation–flexion (EDF) plaster casts under general anaesthesia, until the deformity resolves or until the child is big enough to manage in a brace. From about the age of 4 years onwards curve progression slows down or ceases and the child may not need further treatment.

If the deformity continues to deteriorate, surgical correction may be required. Growth rods may be employed where the curve is controlled with instrumentation without fusion. This requires 6-monthly surgical correction. Newer technology offers non-invasive magnetically induced lengthening but at great cost.

Congenital scoliosis

This includes the more common *failure of formation* (hemivertebrae) and less common *failure of segmentation* (bar) (Figure 18.16). Clinically there may be hair, dimples or a pad of fat over the spine. There is a common association with renal, cardiac and neurological anomalies requiring investigation by abdominal ultrasound and MRI.

While congenital scoliosis is often mild, some cases progress to severe deformity, particularly those with unilateral fusion of vertebrae with contralateral hemivertebrae. Progression can be predicted by the nature of the hemivertebrae – i.e. whether it is non-, partially or fully segmented – suggesting the presence of growth plates and growth potential.

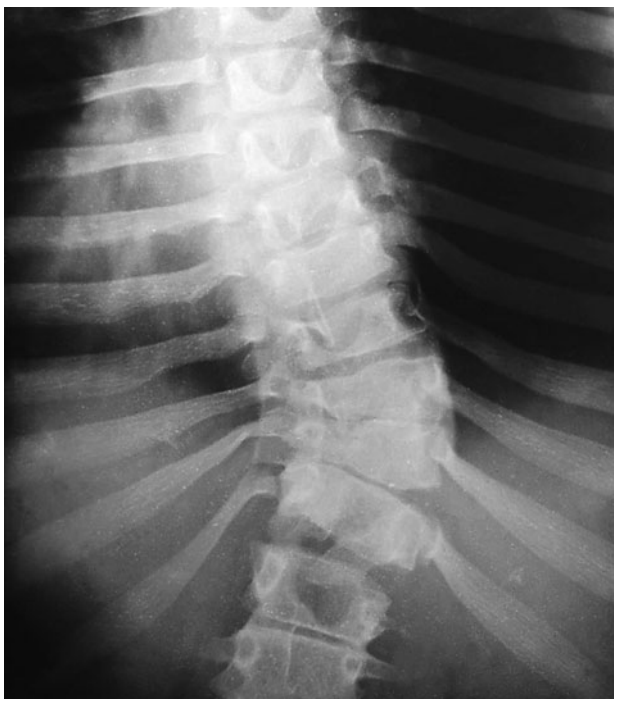


Figure 18.16 Congenital scoliosis Failure of segmentation and formation of the vertebrae at T10, 11 and 12 has resulted in a localized scoliosis.

Treatment

Progressive deformities (usually involving rigid curves) will not respond to bracing alone, and surgical correction is technically challenging in these small patients. Management involves recognizing the progression potential and impairing growth with fusion over the small involved segment. If there is an existing deformity, hemivertebral resection and fusion may be required.

Neuromuscular scoliosis

Neuromuscular conditions associated with scoliosis include poliomyelitis, cerebral palsy, syringomyelia, Friedreich's ataxia and the rarer lower motor neuron disorders and muscle dystrophies; the curve may take some years to develop. The typical paralytic curve is a long, C-shaped curve; initially it is flexible but it becomes rigid with time. As the curve progresses, pelvic obliquity will ensue with sitting balance problems and pressure areas.

X-ray with traction applied shows the extent to which the deformity is correctable.

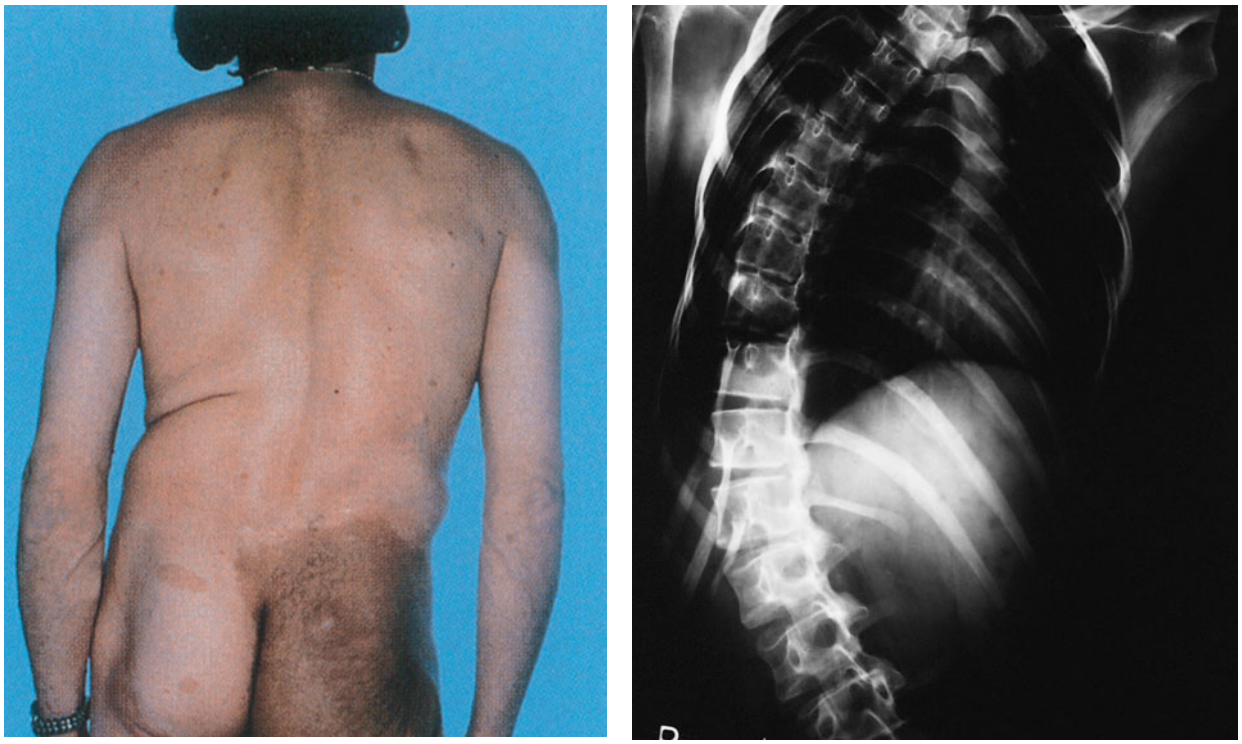
Treatment

Treatment depends upon the degree of functional disability. Mild curves may require no treatment at all. Moderate curves with spinal stability are managed as for idiopathic scoliosis. Severe curves, associated with pelvic obliquity and loss of sitting balance, can often be managed by fitting a suitable sitting support. If this does not suffice, operative treatment may be indicated. This involves a long instrumented fusion of the spine to the pelvis.

Scoliosis and neurofibromatosis

About one-third of patients with neurofibromatosis develop spinal deformity, the severity of which varies from very mild (and not requiring any form of treatment) to the most marked manifestations accompanied by skin lesions, multiple neurofibromata and bony dystrophy affecting the vertebrae and ribs. The scoliotic curve is typically 'short and sharp'. Other clues to the diagnosis lie in the appearance of the skin lesions and any associated skeletal abnormalities (Figure 18.17).

Mild cases are treated as for idiopathic scoliosis. More severe deformities will usually need combined anterior and posterior instrumentation and fusion. As with other forms of skeletal neurofibromatosis, graft dissolution and pseudarthrosis are not uncommon.



(a) (b)

Figure 18.17 Other types of scoliosis (a) This patient has a short structural curve plus multiple skin lesions – features suggesting neurofibromatosis. (b) By contrast, the typical post-poliomyelitis 'paralytic' scoliosis shown in this X-ray is characterized by a long C-shaped curve.

KYPHOSIS

Rather confusingly, the term 'kyphosis' is used to describe both the normal (gentle rounding of the thoracic spine) and the abnormal (excessive thoracic curvature or straightening out of the cervical or lumbar lordotic curves). Excessive thoracic curvature might be better described as 'hyperkyphosis'. *Kyphos*, or gibbus, is a sharp posterior angulation due to localized collapse or wedging of one or more vertebrae. This may be the result of a congenital defect, a fracture (sometimes pathological) or spinal tuberculosis (see Figure 18.24).

Postural kyphosis

Postural kyphosis is usually associated with other postural defects such as flat feet. It is voluntarily correctable. If treatment is needed, this consists of posture training and exercises. Compensatory kyphosis is secondary to some other deformity, usually increased lumbosacral lordosis (Figure 18.18). This deformity, too, is correctable.

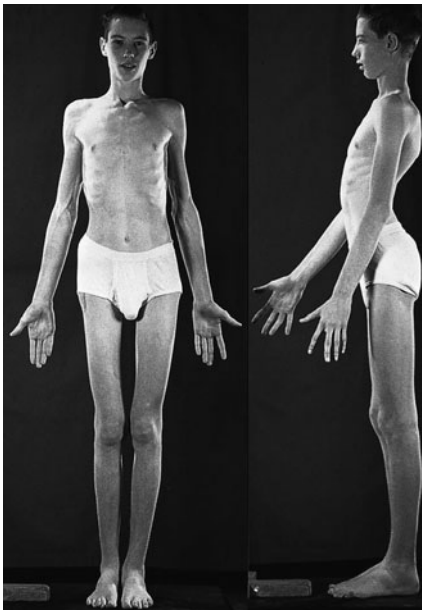


Figure 18.18 Postural kyphosis This tall teenager has Marfan syndrome and ligamentous laxity. He has also developed a postural thoracic hyperkyphosis and lumbar hyperlordosis.

Structural kyphosis

Structural kyphosis is fixed and associated with changes in the shape of the vertebrae. In *children* this may be due to congenital vertebral defects; it is also seen in skeletal dysplasias such as achondroplasia and in osteogenesis imperfecta. Older children may develop severe deformity secondary to

tuberculous spondylitis. In *adolescents* the commonest cause is Scheuermann's disease (see later).

In *adults* kyphosis could be due to an old childhood disorder; tuberculous spondylitis, ankylosing spondylitis or spinal trauma. In *elderly people*, osteoporosis may result in vertebral compression and an increase in a previously mild, asymptomatic deformity.

CONGENITAL KYPHOSIS

Vertebral anomalies leading to kyphosis may be due to failure of formation (type I), failure of segmentation (type II) or a combination of the two.

Type I (failure of formation) This is the commonest (and the worst) type. If the anterior part of the vertebral body fails to develop, progressive kyphosis and posterior displacement of the hemivertebra may lead to cord compression. In children younger than 6 years with curves of less than 40 degrees, posterior spinal fusion alone may prevent further progression. Older children or more severe curves may need combined anterior and posterior fusion, and those with neurological complications will require cord decompression as well as fusion.

Type II (failure of segmentation) Type II usually takes the form of an anterior intervertebral bar; as the posterior elements continue to grow, that segment of the spine gradually becomes kyphotic. The risk of neurological compression is much less but, if the curve is progressive, a posterior fusion will be needed.

ADOLESCENT KYPHOSIS (JUVENILE OSTEOCHONDROSIS; SCHEUERMANN'S DISEASE)

Scheuermann, in 1920, described a condition that he called 'juvenile dorsal kyphosis', distinguishing it from the more common postural (correctable) kyphosis. The characteristic feature was a fixed round-back deformity associated with wedging of several thoracic vertebrae. The term 'vertebral osteochondritis' was adopted because the primary defect appeared to be in the ossification of the ring epiphyses that define the peripheral rims on the upper and lower surfaces of each vertebral body. The true nature of the disorder is still not known; the cartilaginous end plates may be weaker than normal (perhaps due to a collagen defect) and are then damaged by pressure of the adjacent intervertebral discs during strenuous activity. The normal curve of the thoracic spine ensures that the anterior edges of the vertebrae are subjected to the greatest stress and this is where the damage is greatest. Similar changes may occur in the lumbar spine, but here wedging is unusual.

Clinical features

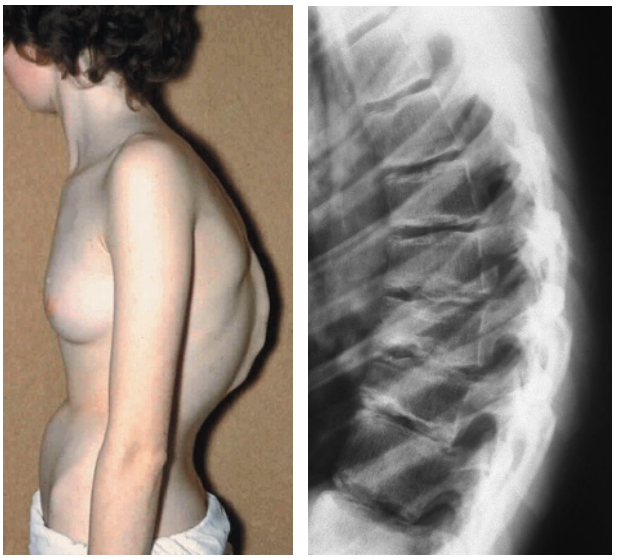
The condition starts at puberty and affects boys more often than girls. The parents notice that the child, an otherwise fit teenager, is becoming increasingly round-shouldered. The patient may complain of backache and fatigue; this sometimes increases after the end of growth and may become severe.

A smooth thoracic kyphosis is seen; it may produce a marked hump and below this is a compensatory lumbar lordosis (Figure 18.19). The deformity cannot be corrected by changes in posture. Movements are normal but tight hamstrings often limit straight leg raising. A mild scoliosis is not uncommon. Rare complications are spastic paresis of the lower limbs and – with severe deformity of the thorax – cardiopulmonary dysfunction.

In later life patients with thoracic kyphosis may develop lumbar backache. This has been attributed to chronic low back strain or facet joint dysfunction due to compensatory hyperextension of the lumbar spine. In some cases, however, lumbar Scheuermann's disease itself may cause pain (see below).

X-rays

In lateral radiographs of the spine the vertebral endplates of several adjacent vertebrae (usually T6–T10)



(a) (b)

Figure 18.19 Scheuermann's disease (a) A young girl with marked exaggeration of the usual thoracic kyphosis. (b) X-ray examination showed the typical indentations in the vertebral end plates and wedging of vertebral bodies.

appear irregular or fragmented. The changes are more marked anteriorly and one or more vertebral bodies may become wedge-shaped. There may also be small radiolucent defects in the subchondral bone (Schmorl's nodes), which are thought to be due to central (axial) disc protrusions.

The angle of deformity is measured in the same way as for scoliosis, except that here the lateral X-ray is used and the lines mark the uppermost and lowermost affected vertebrae. Wedging of more than 5 degrees in three adjacent vertebrae and an overall kyphosis angle of more than 40 degrees are abnormal. Mild scoliosis is not uncommon.

Differential diagnosis

Postural kyphosis Postural 'round back' is common in adolescence. It is painless, and the patient can correct the deformity voluntarily. The curve is a long one and other postural defects are common. The X-ray appearance is normal.

Discitis, osteomyelitis and tuberculous spondylitis If the changes are restricted to one intervertebral level, they can be mistaken for an infective lesion. However, infection causes more severe pain, may be associated with systemic symptoms and signs and produces more marked X-ray changes, including signs of bone erosion and paravertebral soft-tissue swelling.

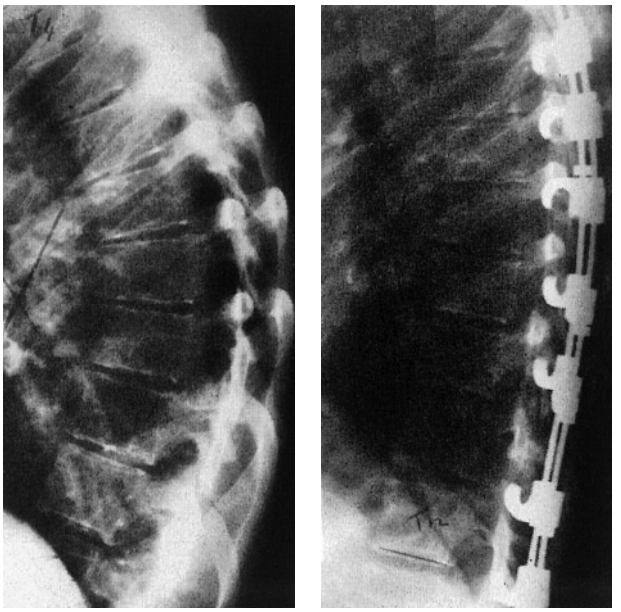
Spondyloepiphyseal dysplasia In mild cases this can produce changes at multiple levels resembling those of Scheuermann's disease. Look for the characteristic defects in other joints.

Outcome

The condition is often quite painful during adolescence, but (except in the most severe cases) symptoms subside after a few years. There may be a recurrence of backache in later life, though overall disability is seldom marked.

Treatment

Curves of 40 degrees or less require only back-strengthening exercises and postural training. More severe curvature in a child who still has some years of growth ahead responds well to a period of 12–24 months in a brace that holds the lumbar spine flat and the thoracic spine in 'extension' (decreased kyphosis). Check the position by X-ray to ensure that the brace is effective. The older adolescent or young adult with a rigid curve of more than 60 degrees may need operative release, correction and instrumented fusion (Figure 18.20).



(a) (b)

Figure 18.20 Scheuermann's disease – operative treatment A severe curve may need operation especially if, as in this girl (a), it is associated with chronic pain. (b) The same girl after operative correction and fixation with Wisconsin rods; bone grafts were added and can be expected to produce fusion after a year or two.

Thoracolumbar Scheuermann's disease

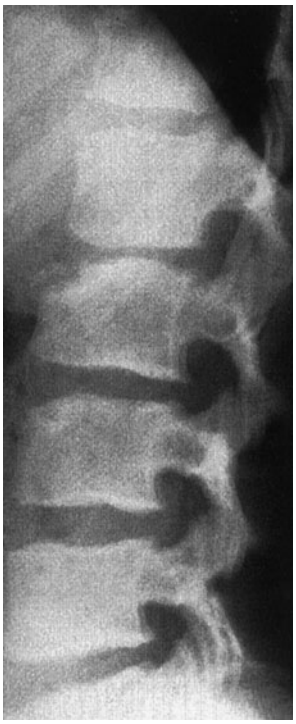
Vertebral end-plate defects are sometimes limited to the lower thoracic and/or the lumbar spine. In mild cases the condition is usually asymptomatic and discovered only incidentally when X-rays are obtained for other reasons (see Figure 18.21). In some cases, however, the patient (usually a teenager at the end of growth or a young adult) complains of back pain and inability to undertake sustained bending, lifting and carrying activities. There is nothing striking to see on clinical examination and it may be difficult to determine whether the backache is due to the Scheuermann's disorder or to some other condition such as spondylolysis or facet joint dysfunction.

Treatment

Treatment consists of muscle strengthening exercises and avoidance of excessive bending and lifting.

KYPHOSIS IN THE ELDERLY

Degeneration of intervertebral discs probably produces the gradually increasing stoop characteristic of the aged. The disc spaces become narrowed and the vertebrae slightly wedged. There is little pain unless osteoarthritis of the facet joints is also present.



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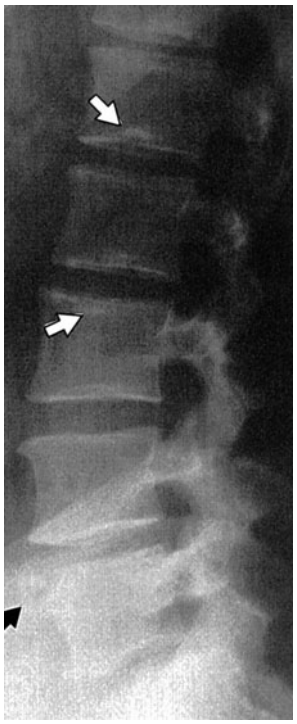
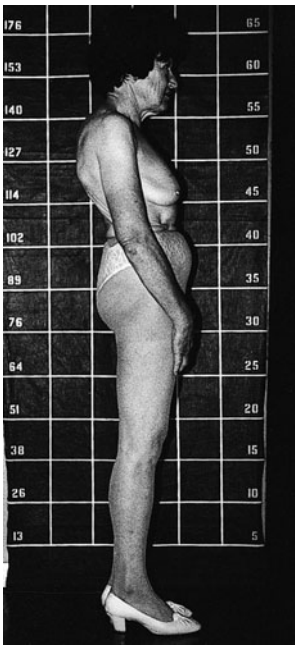


(b)

Figure 18.21 Lumbar Scheuermann's disease (a) The X-ray appearances of lumbar Scheuermann's disease are often mistaken for a fracture (or worse). The 'fragmentation' anteriorly is due to abnormal ossification of the ring epiphysis. (b) Schmorl's nodes (arrows) may also be seen.



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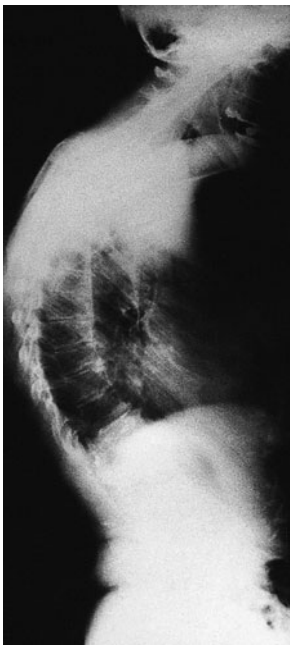


(b)

Figure 18.22 Osteoporotic kyphosis Postmenopausal osteoporosis often results in compressive wedging of the thoracic vertebral bodies and a gradual increase in the natural thoracic kyphosis.

Osteoporotic kyphosis

Postmenopausal osteoporosis may result in one or more compression fractures of the thoracic spine. Patients are usually in their sixties or seventies and may complain of pain. Kyphosis is seldom marked (Figure 18.22). Often the main complaint is of lumbosacral pain, which results from the compensatory lumbar lordosis in an ageing, osteoarthritic spine. Treatment is directed at the underlying condition and may include hormone and bone mineral replacement therapy.

Senile osteoporosis affects both men and women. Patients are usually over 75 years of age, often incapacitated by some other illness, and lacking exercise. They complain of back pain, and spinal deformity may be marked. X-rays reveal multiple vertebral fractures. It is important to exclude other conditions such as *metastatic disease* or *myelomatosis.*

Treatment

Treatment is symptomatic. Bed rest and spinal bracing merely aggravate the osteoporosis. More recently, fresh compression fractures are being treated by the transpedicular injection of methacrylate or bone graft substitute paste in order to stop further deformity and control pain ('vertebroplasty') or to correct the wedge deformity and maintain correction ('kyphoplasty'). The authors believe it is too early to recommend this treatment as the long-term outcome and potential complications have yet to be fully assessed.

SPINAL INFECTION

The axial skeleton accounts for 2–7% of all cases of osteomyelitis. Predisposing factors include diabetes mellitus, malnutrition, substance abuse, human immunodeficiency virus (HIV) infection, malignancy, long-term use of steroids, renal failure and septicaemia.

PYOGENIC DISCITIS/OSTEOMYELITIS

Acute pyogenic infection of the spine is uncommon and diagnosis and treatment are often unnecessarily delayed. The elderly, chronically ill and immunodeficient patients are at greatest risk.

Pathology

Staphylococcus aureus is responsible in 50–60% of all cases, but in immunosuppressed patients

Gram-negative organisms such as *Escherichia coli* and *Pseudomonas* are the most common. The usual sources of infection are:

- haematogenous spread from a distant focus of infection
- inoculation during invasive procedures (spinal injections and disc operations).

The infection usually begins in the vertebral end plates with secondary spread to the disc and adjacent vertebra. It may also spread along the anterior longitudinal ligament to an adjacent vertebra, or outwards into the paravertebral soft tissues: from the thoracic spine along the psoas to the groin; from the lumbar region to the buttock, the sacroiliac joint or the hip.

The spinal canal is rarely involved but, when it is, in the form of an epidural abscess, it is a surgical emergency! Despite rapid surgical decompression, the patient is often left with some degree of permanent paralysis.

Clinical features

Localized pain – the cardinal symptom – is often intense, unremitting and associated with muscle spasm and restricted movement. There may also be point tenderness over the affected vertebra. Intercostal neuralgia is a frequent symptom with thoracic spine involvement.

The patient may give a history of some invasive spinal procedure or a distant infection during the preceding few weeks. A careful history and general examination are essential to exclude a focus of infection (skin, ENT, chest, pelvis).

Systemic signs such as pyrexia and tachycardia are often present but not particularly marked. In children

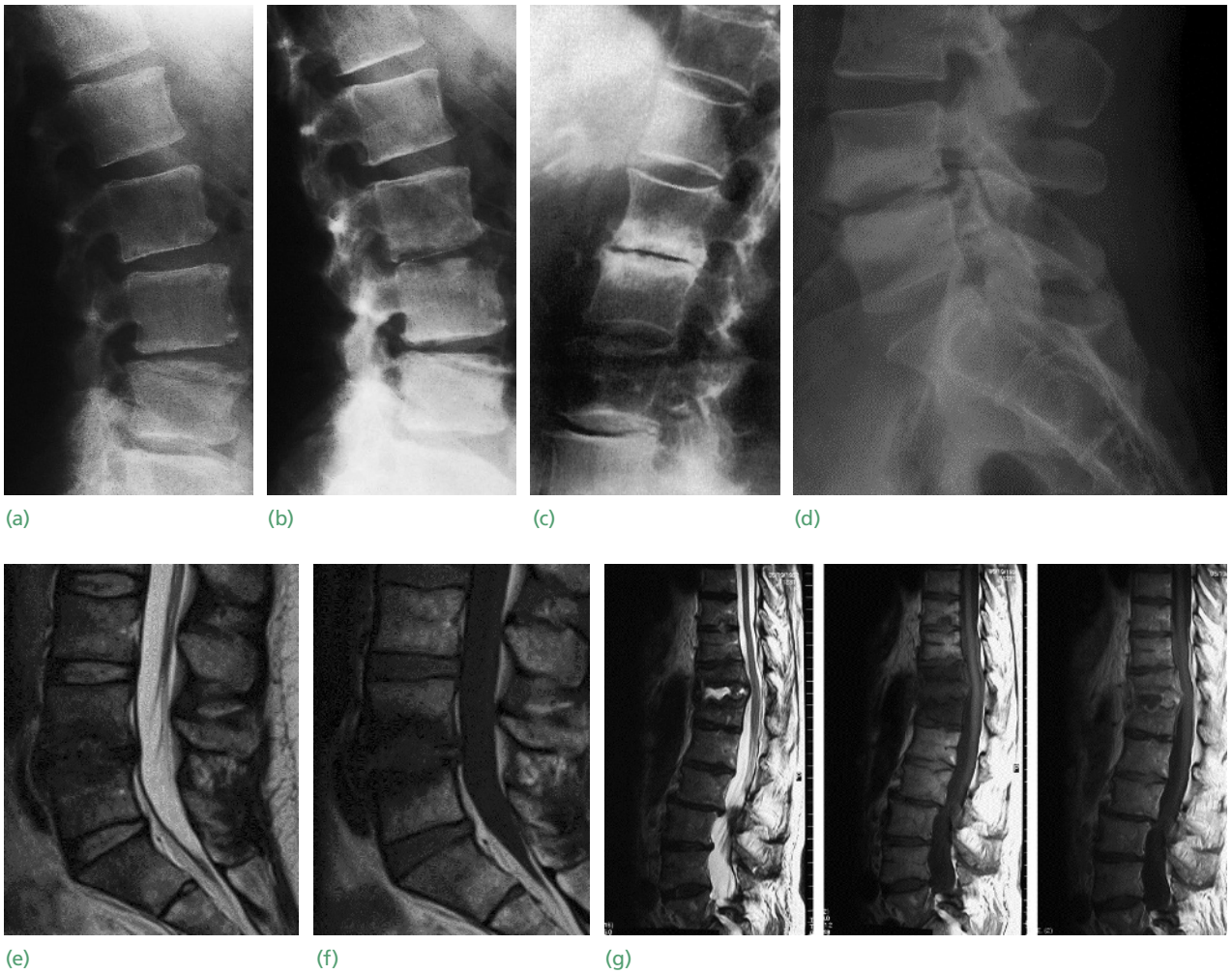


Figure 18.23 Pyogenic osteomyelitis and discitis Typical X-ray features are loss of disc height, irregularity of the disc 'space', end-plate erosion and reactive sclerosis. Progressive changes are shown in (a) and (b). Reactive bone changes, shown in (c), may end with ankylosis at the affected level; (d) X-ray shows L4/5 loss of disc height, end-plate erosion and sclerosis; (e–f) MRI shows the disc and end-plate involvement without pus collection, typical of a low-grade discitis commonly from skin commensals such as *Proprionibacterium acnes*; (g) MRI of T12/L1 spondylodiscitis with T2, T1 and with gadolinium contrast enhancement showing pus in the disc space.

the diagnosis can be particularly difficult; often they have an awkward gait with a stiff spine or, if the lumbar spine is involved, they can present with abdominal symptoms and signs.

Imaging

X-rays may show no change for several weeks; if the diagnosis is delayed, the examination should be repeated. Early signs are loss of disc height, irregularity of the disc space, erosion of the vertebral end plate and reactive new bone formation. Soft-tissue swelling may be visible. The early loss of disc height distinguishes vertebral osteomyelitis from metastatic disease, where the disc can remain intact despite advanced bony destruction.

Radionuclide scanning will show increased activity at the site but this is non-specific. *MRI* may show characteristic changes in the vertebral end-plates, intervertebral disc and paravertebral tissues; this investigation is highly sensitive but not specific.

Similar features may be seen in discitis.

Other investigations

The white cell count, C-reactive protein (CRP) level and erythrocyte sedimentation rate (ESR) are usually elevated. Agglutination tests for *Salmonella* and *Brucella* can be considered in endemic regions and in patients who have recently visited these areas. Blood culture is essential in patients who are febrile though it is often negative in the early stages of infection. Biopsy is definitive. Needle biopsies have variable yields with open biopsies and larger cores of tissue usually identifying the organism.

Treatment

NON-OPERATIVE TREATMENT

Ideally, the organism should be identified on biopsy and appropriated intravenous antibiotics initiated. If not yet known, both Gram-positive and Gram-negative cover should be used in consultation with a microbiologist according to the likely local microbiological flora.

Intravenous antibiotics are continued for 1–2 weeks. If there is a good response (clinical improvement, a falling CRP and ESR and a normal white cell count), oral antibiotics can be used for a total treatment period of 6 weeks. Some centres treat for 12 weeks but with little evidence for this increased period. During this period nutritional support and management of comorbidities are essential in ensuring a successful outcome.

OPERATIVE TREATMENT

Operative treatment is seldom needed. Should the CRP or pain not be settled, surgical debridement may be indicated. An anterior approach is preferred; necrotic and infected material is removed and, if necessary, the cord is decompressed. The anterior column defect is reconstructed with bone grafts. If the spine is unstable, posterior instrumentation may be necessary. For a primary epidural abscess with neurological symptoms, laminectomy and drainage is indicated.

Outcome

The outcome (with prompt and effective treatment) is usually favourable. Spontaneous fusion of infected vertebrae is a common radiological feature of healed staphylococcal osteomyelitis.

TUBERCULOSIS

The spine is the most common site of skeletal tuberculosis (TB), and it accounts for 50% of all musculoskeletal TB. It is thought that there are approximately 2 million people with spinal tuberculosis worldwide.

Pathology

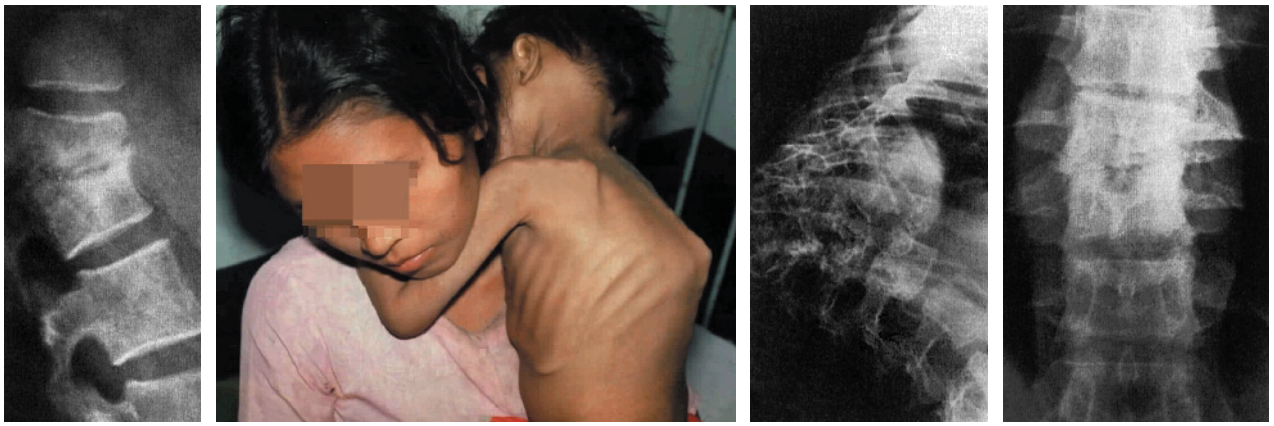
Spread is haematogenous and the infection usually settles in a vertebral body adjacent to the intervertebral disc. Bone destruction and caseation follow, with infection spreading to the disc space and the adjacent vertebrae. In the lumbar area the paravertebral abscess may track along muscle planes to involve the sacroiliac or hip joint, or along the psoas muscle to the thigh. As the vertebral bodies collapse, a sharp angulation (gibbus or kyphos) develops. There is a major risk of cord damage due to pressure by the abscess, granulation tissue, sequestra or displaced bone, or (occasionally) ischaemia from spinal artery thrombosis.

Spontaneous bony fusion of the involved levels may occur with healing but the persistent infection may cause spinal cord attenuation and late-onset paraplegia. Reactivation of healed disease may also occur.

Clinical features

There is usually a long history of ill health and backache; in late cases a gibbus deformity is the dominant feature. Concurrent pulmonary TB is a feature in most children under 10 years with thoracic spine involvement. Occasionally the patient may present with a cold abscess pointing in the groin, or with paraesthesiae and weakness of the legs. There is local tenderness in the back and spinal movements are restricted.

In cervical spine disease, dyspnoea and dysphagia are features of advanced infection, especially in children; these patients present with a stiff painful neck. Children under 10 years of age with thoracic spine TB usually develop a *pectus carinatum* ('pigeon chest') deformity (Figure 18.24).



(a) (b) (c) (d)

Figure 18.24 Tuberculosis of the spine (a) Early X-ray changes with loss of disc space. (b) Young patient with advanced tuberculous deformity. (c) X-ray showing vertebral collapse and severe kyphosis. (d) X-ray appearance of a psoas abscess in the paravertebral tissues.

Neurological examination may show motor and/or sensory changes in the lower limbs. As spinal tuberculosis is found mostly in the thoracic spine, spastic paraparesis is a common presentation in adults.

ATYPICAL FEATURES

Even in areas where tuberculosis is no longer as common as it was in the past, it is important to be alert to the possibility of this diagnosis. The task is made harder when the patient presents with atypical features:

- Lack of deformity, e.g. a patient with a primary epidural abscess
- Involvement of only the posterior vertebral elements
- Infection confined to a single vertebral body
- Involvement of multiple vertebral bodies and posterior elements (especially in HIV-positive patients) resulting in a kyphoscoliosis

POTT'S PARAPLEGIA

Paraplegia is the most feared complication of spinal tuberculosis.

Early-onset paresis Paresis is usually within 2 years of disease onset and is due to pressure by inflammatory oedema, an abscess, caseous material, granulation tissue or sequestra. The patient presents with lower-limb weakness, upper motor neuron signs, sensory dysfunction and incontinence. CT and MRI may reveal cord compression. In these cases the prognosis for neurological recovery following surgery is good.

Late-onset paresis This is due to direct cord compression from increasing deformity, or (occasionally) vascular insufficiency of the cord; recovery following decompression is poor.

Imaging

The entire spine should be X-rayed, because vertebrae distant from the obvious site may also be affected without any obvious deformity. The earliest signs of infection are local osteoporosis of two adjacent vertebrae and loss of crispness of the end plates. Progressive disease is associated with signs of bone destruction and collapse of adjacent vertebral bodies into kyphosis. Paraspinal soft-tissue shadows may be due either to oedema, swelling or a paravertebral abscess. The radiological picture may mimic those of other infections including fungal infections and parasitic infestations. A chest X-ray is essential to exclude associated pulmonary TB.

With healing, bone density increases, the ragged appearance disappears and paravertebral abscesses may undergo resolution or fibrosis or calcification.

MRI and CT scans are invaluable to assess neurological compression, posterior vertebral element involvement, paravertebral abscesses, an epidural abscess and cord compression (Figure 18.25).

Special investigations

Skin tests with attenuated mycobacterium (Mantoux and Tine) are sensitive but non-specific as they will react in vaccinated patients. ESR can also be useful but it is a non-specific marker. The white cell count is usually normal but the differential may have a lymphocytosis and platelets raised. HIV status should be confirmed due to their association. Retroviral therapy may be required depending on the CD4 count and the local hospital protocols.

Biopsy is necessary to confirm the diagnosis and exclude drug-resistant strains which are increasingly common. Microscopy, culture, polymerase chain reaction (PCR) and histological investigation should be requested. PCR is both sensitive and specific and

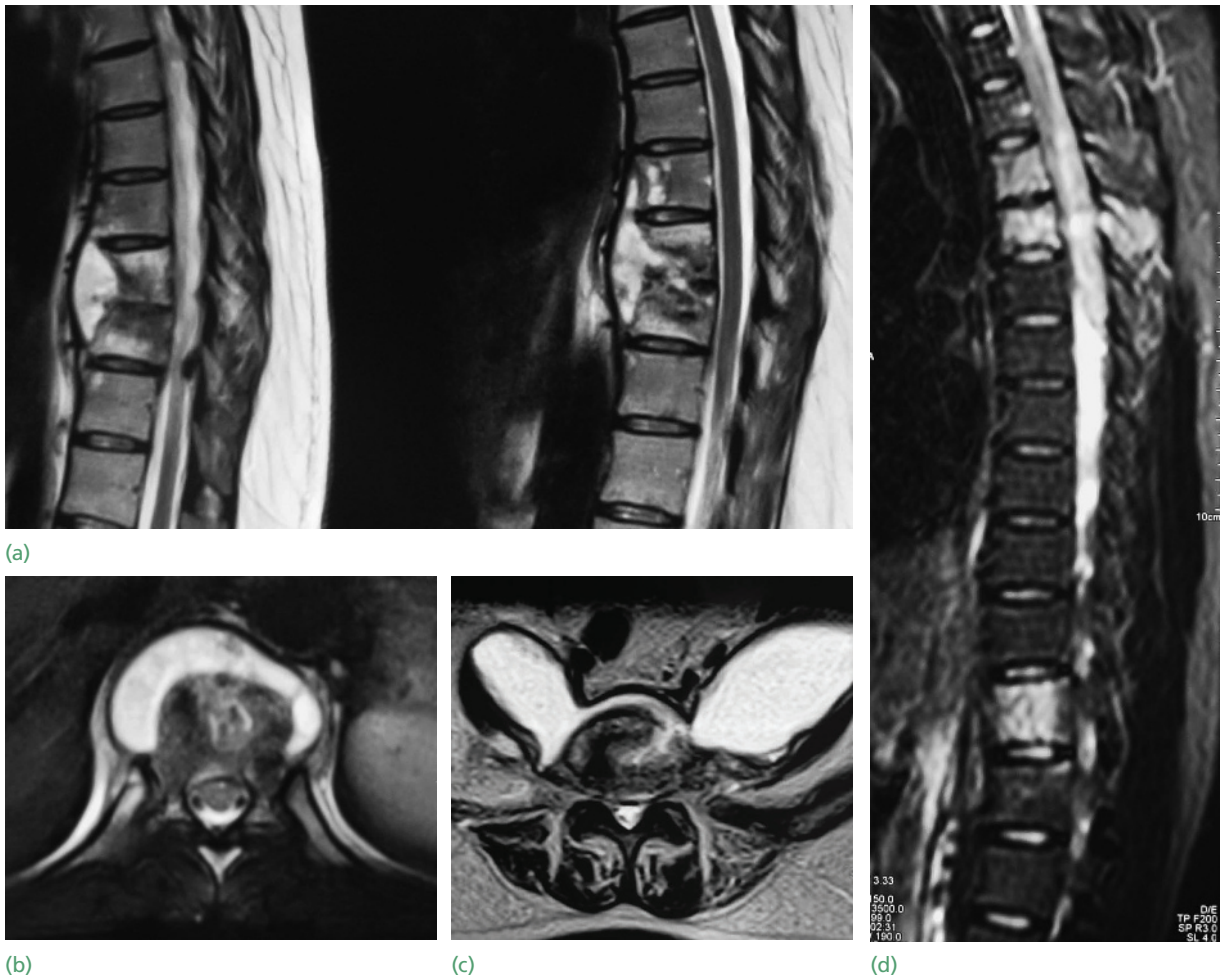


Figure 18.25 Spinal tuberculosis – MRI features Scanning in several planes shows details that cannot be seen in plain X-rays. (a) Sagittal MR images of advanced tuberculous infection with abscess formation beneath the anterior longitudinal ligament. (b,c) Axial images showing psoas abscesses communicating across the front of the spine. (d) In countries where TB is endemic, additional active lesions can be detected by MRI in almost 40% of patients presenting with 'local' lesions.

provides a result in 1–2 days as opposed to culture which takes up to 6 weeks.

Differential diagnosis

Spinal tuberculosis must be distinguished from other causes of vertebral pathology, particularly pyogenic and fungal infections, malignant disease and parasitic infestations such as hydatid disease. Disc-space collapse is typical of pyogenic infection, involvement of vertebrae either side of a preserved disc typical of TB and involvement of a single vertebral body typical of a tumour. However, TB is a great mimic and the presentation and appearances can be similar to all of the above.

Treatment

The objectives are to: (1) eradicate or at least arrest the disease; (2) prevent or correct deformity; and (3) prevent or treat associated neurological deficit.

There is little consensus on antituberculous chemotherapy in terms of either drugs or duration. Most high incidence regions use a combination of drugs, usually three or four (rifampicin, isoniazid, ethambutol and pyrazinamide), to minimize resistance and act against both the easily accessible mycobacteria and the latent intracellular group. They are available in a combination single tablet. Many use full drug treatment for the full duration of 9–12 months, whereas others follow the pulmonary strategy of a two-drug continuation phase after 2 months of three/four drug therapy. Dosages are weight-dependent and need to be adjusted as the patient improves in health status and gains weight. The patient must be monitored for the drugs' side effects of hepatitis, depression and loss of visual acuity. If resistant mycobacteria are confirmed on PCR or culture, second-line agents are required, usually involving a fluoroquinolone and aminoglycoside.

In patients with minimal deformity and normal or mild neurological impairment, medical therapy is adequate. With progressive neurological deficit or marked kyphosis, surgical debridement and corrective instrumented fusion should be considered. Anterior resection of diseased tissue and anterior spinal fusion with a strut graft offers the double advantage of early and complete eradication of the infection and prevention of spinal deformity (Figure 18.26).

The following approach to treatment is advocated:

- *Ambulant chemotherapy alone* is suitable for early or limited disease with no abscess formation or neurological deficit. Treatment is continued for 6–12 months, or until the X-ray shows resolution of the bone changes, the patient is constitutionally well and ESR has returned to normal. Therapeutic compliance is sometimes a problem.
- *Continuous bed rest and chemotherapy* may be used for more advanced disease when the necessary skills and facilities for radical anterior spinal surgery are not available, or where the technical problems are too daunting (e.g. in lumbosacral tuberculosis) – provided there is no abscess that needs to be drained.
- *Operative treatment* is indicated:
  - when there is an abscess that can readily be drained
  - for advanced disease with marked bone destruction and threatened or actual severe kyphosis
  - neurological deficit including paraparesis that has not responded to drug therapy.

All infected and necrotic material is evacuated or excised and reconstructed with autogenous rib/fibula/iliac crest or allograft humerus shafts or titanium cages. This can be done from an anterior- or

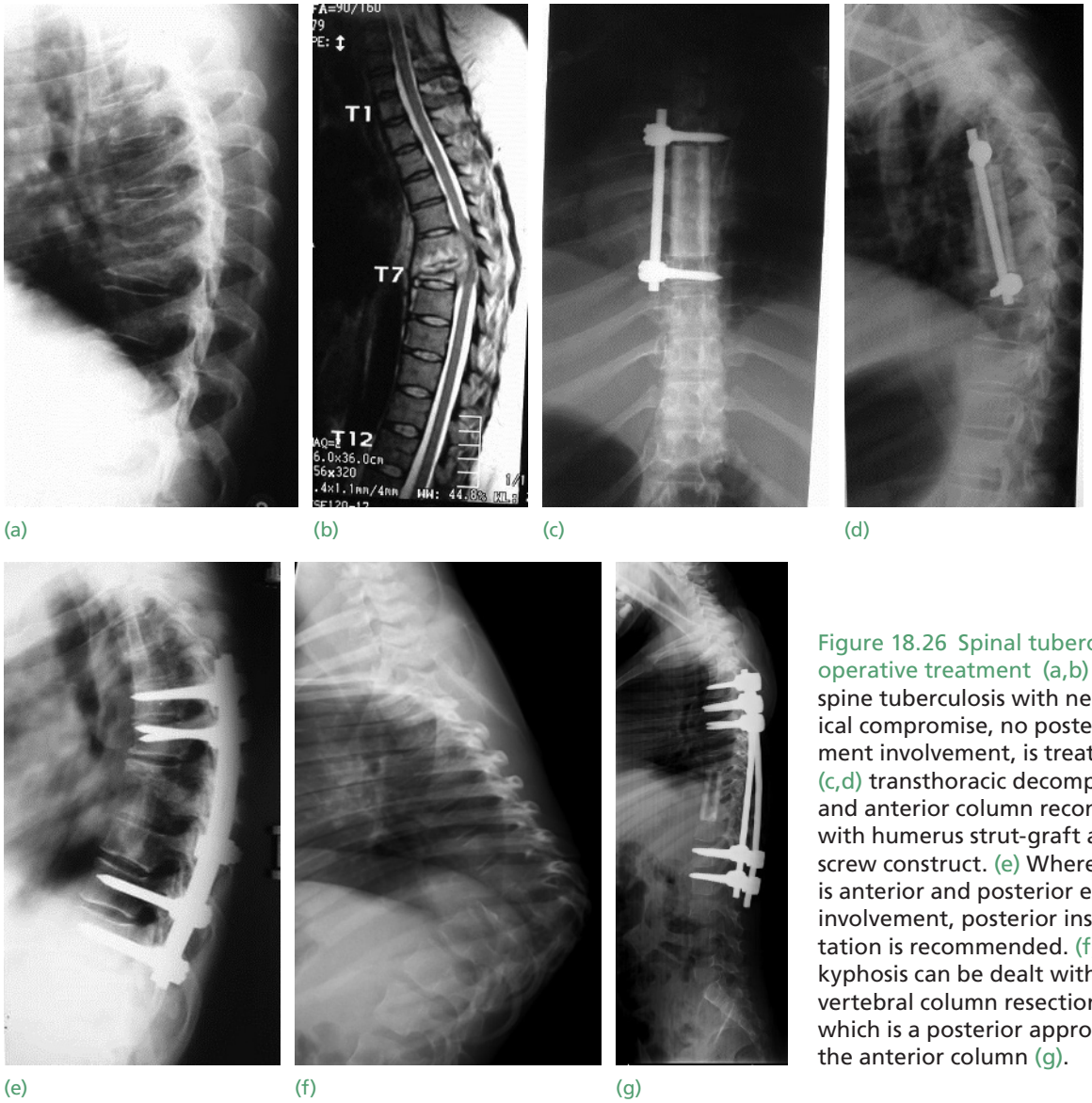


Figure 18.26 Spinal tuberculosis – operative treatment (a,b) Thoracic spine tuberculosis with neurological compromise, no posterior element involvement, is treated with (c,d) transthoracic decompression and anterior column reconstruction with humerus strut-graft and rod-screw construct. (e) Where there is anterior and posterior element involvement, posterior instrumentation is recommended. (f) Severe kyphosis can be dealt with by a vertebral column resection (VCR) which is a posterior approach to the anterior column (g).

posterior-based approach using a vertebral column resection technique. Posterior instrumentation may be required for increased stability. Children are at risk of developing severe kyphosis if they have extensive growth remaining and may need fusion of the posterior elements to minimize the expected deformity.

Human immunodeficiency virus and spinal tuberculosis

HIV is one of the main reasons for the resurgence of TB, especially in the developing world. Initially spinal TB, which is an extrapulmonary focus, was considered as AIDS-defining but, due to the high incidence of both diseases in some regions, this is not always the case. HIV patients are not homogeneous in their immune state: some still have relatively good CD4 counts whereas others do not. Patients with significantly impaired immunity are prone to developing opportunistic infections and atypical mycobacterial infections (*Mycobacterium intracellulare*, *M. avium*, *M. fortuitum)*. There may be increasing atypical TB presentation in HIV patients.

Medical and surgical management is essentially the same but, in those patients with advanced disease, the patient's ability to survive a surgical insult must be considered. Often they have large paraspinal abscesses with little kyphosis, and a simple procedure such as a costotransversectomy is all that is required. Initiation of anti-retrovirals should be considered but it may result in increased TB disease due to a recovering immune system (immune reconstitution inflammatory syndrome, IRIS). This can be life-threatening and needs to be managed by an infectious diseases specialist.

FUNGAL INFECTION

These are opportunistic infections occurring in an immunocompromised host (e.g. due to HIV, malignancy, steroid therapy or chronic granulomatous disease) and a patient with extensive burns; however, they may also affect a normal host. *Aspergillosis* and *Cryptococcus* are airborne fungi that initially affect the lungs; the spine is involved by haematogenous spread.

In children with chronic granulomatous disease, thoracic spine involvement is due to contiguous spread from the lungs. The presentation, clinical findings and radiographic features may mimic those of TB. The chest X-ray may show a fungal ball or pneumonia. The diagnosis is made by sputum examination and bronchoscopy. The immunodiffusion test is specific for *Aspergillosis* and the latex agglutination test for *Cryptococcus*. A biopsy is performed to confirm the diagnosis.

Treatment

Neurological deficit is an indication for operative decompression. Specific treatment includes 5-flucytosine and amphotericin B, which act synergistically. Synthetic oral antifungals (ketoconazole, fluconazole, itraconazole) are well absorbed and the serum and cerebrospinal fluid (CSF) concentrations are high. Concurrent treatment of the underlying immunocompromised state is essential.

PARASITIC INFESTATION

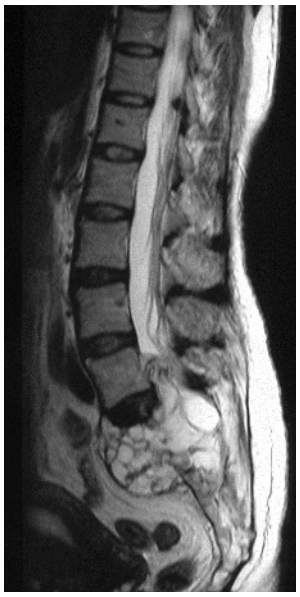
The commonest parasitic infestation affecting the spine is due to the cestode worm *Echinococcus granulosis*, which causes hydatid disease. It is encountered mainly in areas where sheep are raised: Australasia, South America, parts of Africa, Wales and Iceland. The definitive host is the dog as well as other canine animals. The sheep is the intermediate host and humans are affected by the ingestion of ova that are usually carried in the dog's excreta or fur. The embryo worm enters the human host either by being ingested through faecal contamination or by inhalation of desiccated particles in dust. In that way the embryos come to lodge in the liver and the lungs, but in about 10% of cases there is dissemination to other sites, including the bones (mainly the spine, skull and long bones) where hydatid cysts develop in about 1% of cases.

Hydatid disease is usually picked up in childhood but it may be many years before the diagnosis is made. The presentation and clinical features are similar to those of other forms of spondylitis. X-rays may reveal a translucent area with a sclerotic margin in the affected vertebral body (Figure 18.27). In untreated cases this can lead to bone destruction. Neurological deficit, the difficulty in eradicating the disease and the tendency to recurrence make for significant morbidity and mortality.

Systemic treatment is with albendazole, which is active against the larvae and the cysts; three cycles of 25 days each is the usual recommendation. Operative treatment to achieve spinal decompression may be called for; spillage of cyst contents must be avoided. The prognosis is generally poor when the liver and lungs are affected.

NON-INFECTIVE INFLAMMATORY DISEASE

Ankylosing spondylitis and seronegative spondyloarthropathies are dealt with in Chapter 3.



(a)

(b)

Figure 18.27 Hydatid disease (a) MRI showing hydatid lumbosacral disease; (b) hydatid cysts removed during decompression.

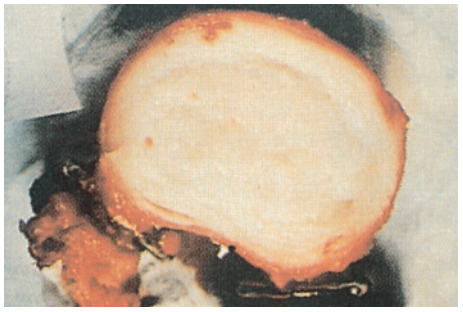
DEGENERATIVE DISORDERS OF THE SPINE

INTERVERTEBRAL DISC DEGENERATION

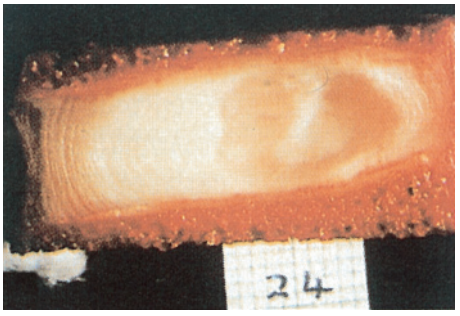
Lumbar backache is one of the most common causes of chronic disability in Western societies, and in the majority of cases the backache is associated with degeneration of the intervertebral discs in the lower lumbar spine. This is an age-related phenomenon that occurs in over 80% of people who live for more than 50 years and in most cases it is asymptomatic.

Pathology

With normal ageing, glycosaminoglycan production diminishes leading to gradual desiccation of the disc. The annulus fibrosus develops fissures and disc nuclear material may prolapse through. The discs lose height and bulge beyond the margins of the vertebral bodies (Figure 18.28).



(a)



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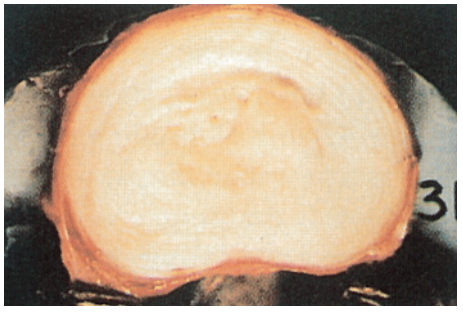

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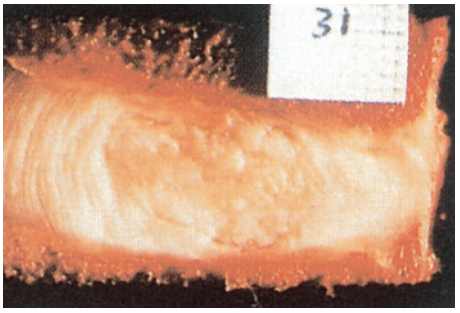

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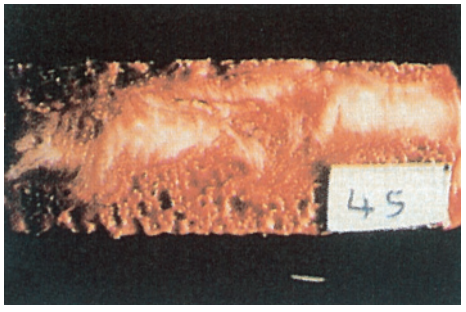
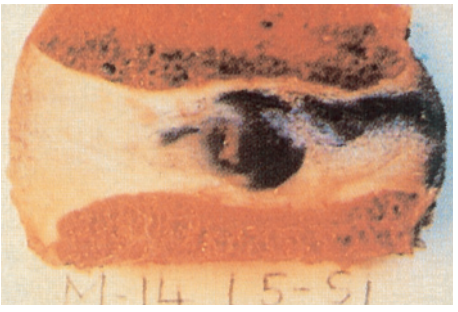
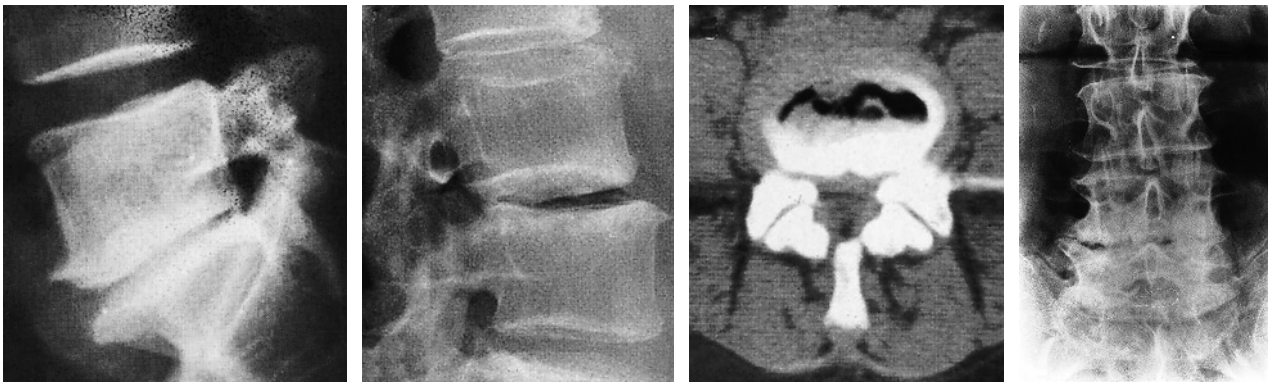


(e)



(f)

Figure 18.28 Disc lesions – pathology (a,b) Transverse and sagittal sections through a young (teenage) intervertebral disc. The nucleus is soft, homogeneous and almost translucent. The annulus is composed of regular lamellae of fibrocartilage. (c,d) Mature (50-year-old) normal disc. The nucleus is more fibrous and less homogeneous. The annulus is thickened and the vertebral body and end plates are intact. (e) Degenerating disc, which is markedly flattened with break-up of the nucleus and disruption of the vertebral body end plates. (f) Young disc stained with analine blue dye to demonstrate a fissure extending posteriorly through the annulus fibrosis.



(a) (b) (c) (d)

Figure 18.29 Spondylosis and osteoarthritis Typical X-ray features are (a) narrowing of the intervertebral space and anterior 'osteophytes'. (b) Other features are slight retrolisthesis and a dark (vacant) area in the disc space – the 'vacuum sign' – better demonstrated in the axial CT (c), which also shows the hypertrophic osteoarthritis of the facet joints. (d) In advanced cases there are well-marked signs of osteoarthritis.

Disc protrusion against the ligaments causes formation of marginal osteophytes. Adjacent vertebral end plates ossify and become sclerotic while fatty change occurs in the subchondral bone marrow. This process is termed *spondylosis* (Figure 18.29). (For a classification of the age-related changes in lumbar discs, see Boos N, *et al.* Classification of age-related changes in lumbar intervertebral discs. *Spine* 2002; **27**: 2631–44.)

Secondary effects

Disc degeneration results in altered biomechanics and increased loading forces on the facet joints (which are intrinsically linked). Facet joint osteoarthritis may develop with osteophyte encroachment into the canal causing spinal lateral recess stenosis. The ligamentum flavum thickens, which contributes to the stenosis, while the disc bulges from anteriorly into the spinal canal. Occasionally the facets become incompetent and segmental instability arises.

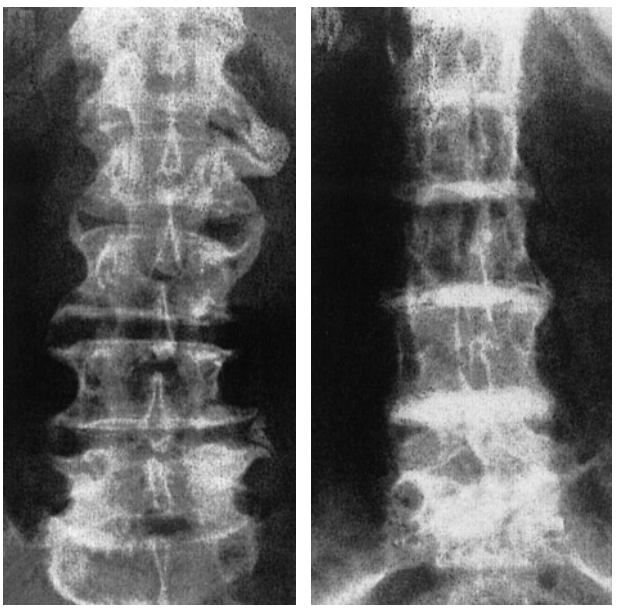
Clinical features

Disc degeneration is inherent with ageing and causes the normal age-related back pain endemic in humans. When symptoms occur, they are usually due to the secondary effects of disc degeneration and are described later.

Imaging

X-ray features of intervertebral disc degeneration have little correlation with symptoms and it is not possible to prognosticate whether an asymptomatic individual with X-ray signs of disc degeneration will develop disabling backache in the future. More than 50% of asymptomatic individuals of 30–39 years of age have disc degeneration.

MRI findings include annular fissures, Modic changes (subchondral changes thought to be degenerative – Figure 18.31), disc degeneration and disc herniation. Modic changes are found in 46% of patients with non-specific low back pain (LBP) and 6% in the asymptomatic general population. Type 1 Modic changes have been linked to *Propionibacterium acnes* and *Corynebacterium propinquum* low-grade infections. This remains contentious and research continues into the condition. Most patients with proven disc pathology will not have Modic changes. As diagnostic markers, Modic signs have relatively low sensitivity.



(a) (b)

Figure 18.30 Conditions resembling spondylosis (a) *Forestier's disease*: at first sight this looks like osteoarthritic spondylosis; there are large spurs at multiple levels, but the disc spaces are usually preserved. (b) *Ochronosis*: intervertebral calcification is characteristic.

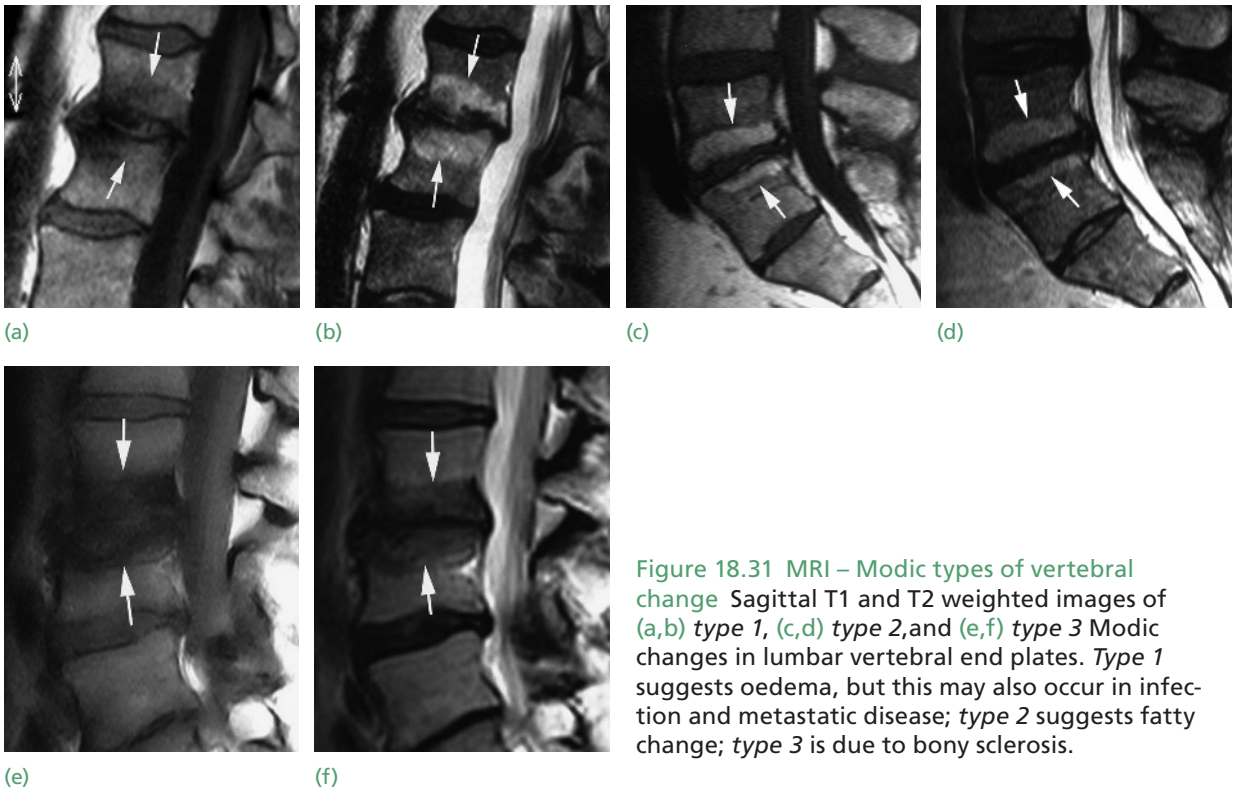


Figure 18.31 MRI – Modic types of vertebral change Sagittal T1 and T2 weighted images of (a,b) *type 1*, (c,d) *type 2*,and (e,f) *type 3* Modic changes in lumbar vertebral end plates. *Type 1* suggests oedema, but this may also occur in infection and metastatic disease; *type 2* suggests fatty change; *type 3* is due to bony sclerosis.

Treatment

Asymptomatic lumbar disc degeneration does not require any treatment besides reassuring the patient that they are normal. Symptomatic conditions related to disc degeneration are discussed below.

ACUTE INTERVERTEBRAL DISC PROLAPSE

Acute disc herniation (prolapse, rupture) is a result of underlying disc degeneration. It occurs most commonly in the fourth to fifth decades of life, is more common in men than women (3:1 ratio) and occurs mostly at L4/5 and L5/S1 disc levels. Risk factors include smoking, heavy lifting especially with torsional stress, strenuous physical activity, and occupational driving.

A *'protrusion'* is a posteriorly bulging disc with the outer annulus intact. When *rupture* occurs, fibrocartilaginous disc material is extruded posteriorly (*extrusion*) through the posterior longitudinal ligament; when disc material breaks free to lie in the canal, it is termed *sequestration* (Figure 18.32). A large central rupture may cause compression of the cauda equina. A posterolateral rupture presses on the nerve root proximal to its point of exit through the intervertebral foramen; thus a herniation at L4/5 will compress the fifth lumbar nerve root, and a herniation at L5/S1, the first sacral root. Local inflammatory response with oedema aggravates the symptoms.

Acute back pain at the onset of disc herniation probably arises from disruption of the outermost layers of the annulus fibrosus and stretching or tearing of the posterior longitudinal ligament. Nerve root irritation causes pain in the buttock which may be referred or radiate down the posterior thigh and calf (*sciatica*).

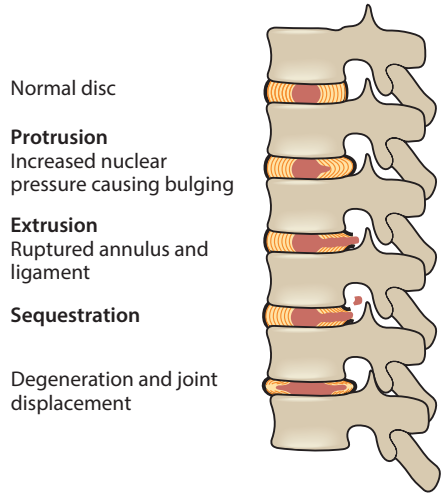


Figure 18.32 Disc lesions – pathology From above, downwards: an abnormal increase in pressure within the nucleus causes splitting and bulging of the annulus; the posterior segment may rupture, allowing disc material to extrude into the spinal canal; with chronic degeneration (lowest level) the disc space narrows and the posterior facet joints are displaced, giving rise to osteoarthritis.

Pressure on the nerve root itself causes *paraesthesia* and/or *numbness* in the corresponding dermatome, as well as *weakness* and *depressed reflexes* in the muscles supplied by that nerve root.

Clinical features

Acute disc prolapse may occur at any age but it is uncommon in the very young and the very old. A common presentation is acute severe back pain which improves, followed by the development of buttock and leg pain (sciatica) a few days later. Both backache and sciatica are made worse by coughing or straining. Paraesthesia or numbness in the leg or foot, and occasionally muscle weakness may occur. Cauda equina compression is rare but may cause urinary retention and perineal numbness (Box 18.1).

The patient usually stands with a list to one side (sciatic scoliosis – Figure 18.33). Sometimes the knee on the painful side is held slightly flexed to relax tension on the sciatic nerve; straightening the knee makes the skew back more obvious. All back movements are restricted, and during forward flexion the list may increase.

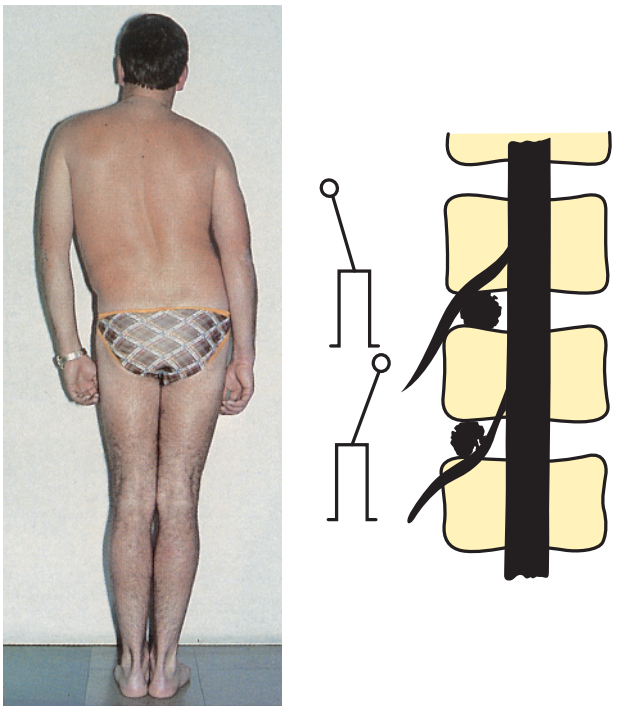
Palpation may find tenderness in the midline and paravertebral muscle spasm. Straight-leg raising is restricted and painful on the affected side; dorsiflexion of the foot and bowstringing of the lateral popliteal nerve may accentuate the pain, and a crossed straight-leg raise test, if present, is highly specific for a disc prolapse. With a high or mid-lumbar prolapse the femoral stretch test may be positive.

Neurological examination may show muscle weakness (and, later, wasting), diminished reflexes and sensory loss corresponding to the affected level. L5 impairment causes weakness of knee flexion and big toe extension as well as sensory loss on the outer side of the leg and the dorsum of the foot. Normal reflexes at the knee and ankle are characteristic of L5 root compression. Paradoxically, the knee reflex may appear to be *increased*, because of weakness of the antagonists (which are supplied by L5). S1 impairment causes weak plantarflexion and eversion of the foot, a depressed ankle jerk and sensory loss along the lateral border of the foot. Occasionally an L4/5 disc prolapse compresses both L5 and S1. Cauda equina compression causes urinary retention and sensory loss over the sacrum.

BOX 18.1 FEATURES OF CAUDA EQUINA SYNDROME

Bladder and bowel incontinence
Perineal numbness
Bilateral sciatica
Lower limb weakness
Crossed straight-leg raising sign

Note: Scan urgently and operate urgently if a large central disc is revealed.



(b) (b)

Figure 18.33 Lumbar disc – signs (a) The patient has a sideways list or tilt. (b) If the disc protrudes medial to the nerve root, the tilt is towards the painful side (to relieve pressure on the root); with a far lateral prolapse (lower level) the tilt is away from the painful side.

Imaging

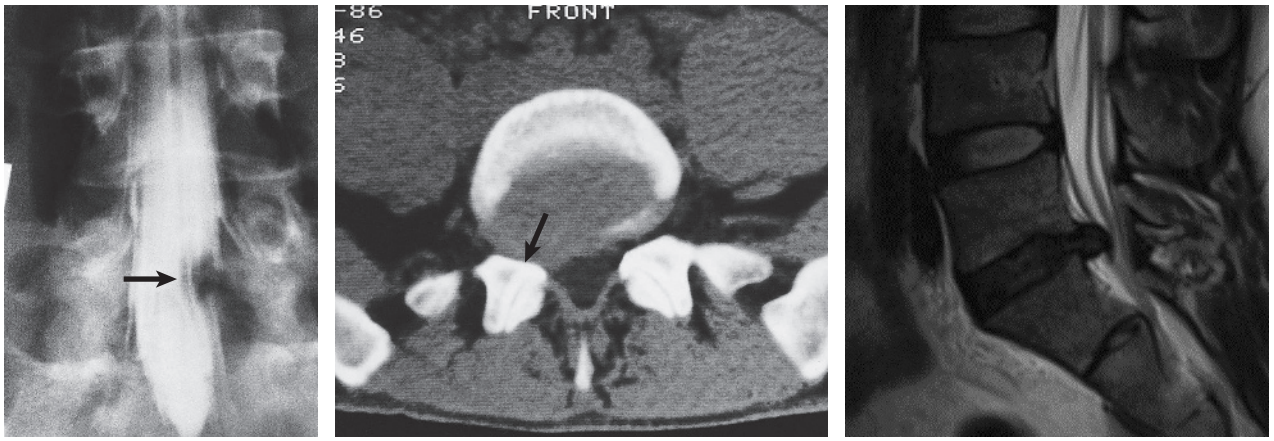
X-rays are helpful to exclude bony pathology and reassure the clinician and patient.

MRI is the default investigation for spinal pathology and has replaced other imaging modalities (Figure 18.34). Where there are contraindications to MRI (such as MRI incompatible pacemaker) a CT myelogram is indicated.

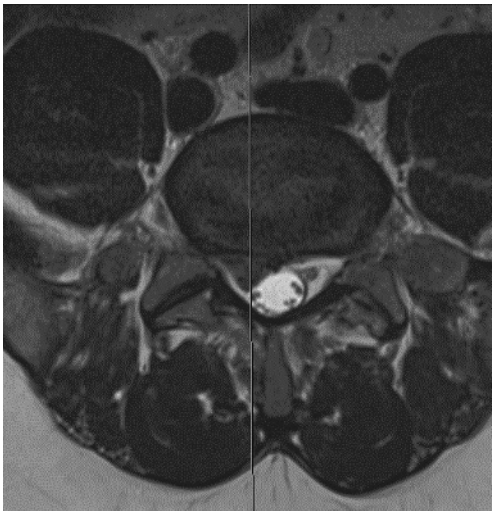
Differential diagnosis

Space-occupying lesions, epidural abscess, tumours, epidural haematoma, stenosis and intradural pathology may present with sciatic symptoms. 'Red flags' (see 'Lower back pain' below) require further investigation.

Direct sciatic nerve compression in the pelvis and upper thigh may occur with piriformis syndrome (compression neuropathy).



(a) (b) (c)



(d)

Figure 18.34 Disc prolapse – imaging (a) Radiculogram in which the gap in the contrast medium (arrow) shows where a disc has protruded. (b) CT scan showing how disc protrusion can obstruct the intervertebral foramen. (c,d) MRI: T2 sagittal and axial view of L5/S1 right-sided disc protrusion. MRI is the default investigation for spine pathology.

Treatment

Around 90% of symptomatic lumbar disc herniations will improve over 6 weeks irrespective of the advice or treatment given. All the usual conservative treatment modalities are symptomatic and have not been shown to change the natural history.

NON-INTERVENTIONAL TREATMENTS

Commonly prescribed non-interventional treatments are:

- Bedrest, heat, ice, short course of analgesics, non-steroidal anti-inflammatory drugs (NSAIDs), antidepressants and muscle relaxants
- Massage, spinal manipulation, spinal traction, acupuncture, advice to stay active, exercise therapy

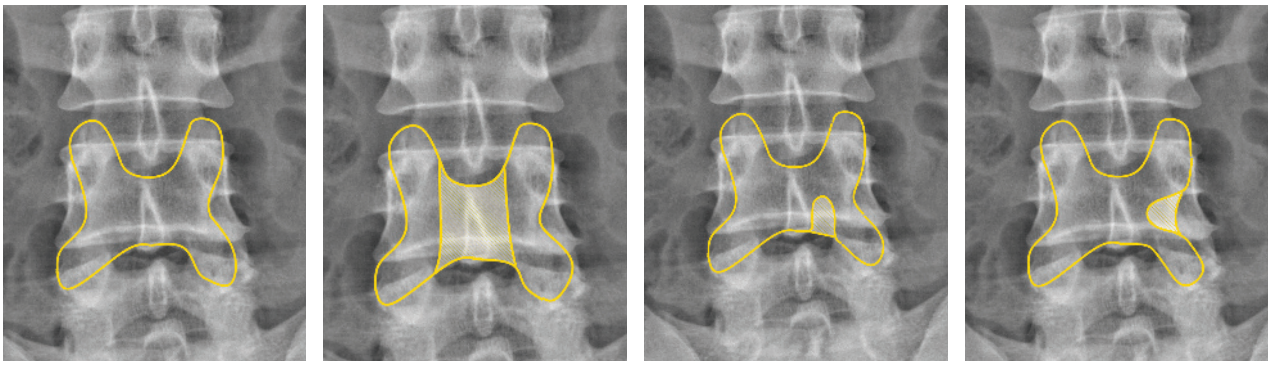
INTERVENTIONAL TREATMENTS

The following modalities of treatment are available but cannot be recommended on the current evidence:

- Epidural corticosteroids
- Automated percutaneous discectomy
- Laser discectomy
- Percutaneous disc decompression
- Chemonucleolysis dissolution of the nucleus pulposus by percutaneous injection of a proteolytic enzyme (chymopapain). Controlled studies have shown that this is less effective (and potentially more dangerous) than surgical removal of the disc material.

An absolute indication for operative removal of a prolapse is a cauda equina compression syndrome – this is an emergency. Relative indications are neurological deterioration and persistent pain and failed conservative treatment.

The presence of a prolapsed disc, and the level, must be confirmed by imaging and the anatomical location of the disc prolapse needs to correlate with the symptoms. Surgery in the absence of a clear preoperative diagnosis is usually unrewarding. Surgical terms used are *laminectomy*, *laminotomy*, *discectomy* and *microdiscectomy* (Figure 18.35). These are largely historical divisions and most procedures are now done with magnification through a unilateral hemilaminotomy approach.



(a) (b) (c) (d)

Figure 18.35 Decompression options (a) Schematic lamina superimposed on X-ray; (b) laminectomy; (c) laminotomy; (d) extraforaminal approach.

The ligamentum flavum is removed on the relevant side, if necessary, with some margin of the bordering laminae and medial third of the facet joint. The dura and nerve root are retracted towards the midline and the disc bulge or extrusion/sequestration is displayed. With an intact annulus an annulotomy is performed and the disc material removed. A far lateral disc protrusion cannot be visualized through the interlaminar approach and usually needs an extraforaminal approach.

Complications include epidural bleeding, dural tears, nerve root injury and incomplete removal of prolapsed disc fragments. Epidural bleeding is minimized by positioning the patient prone with the abdomen free thus minimizing venous pressure. The major postoperative complication is disc space infection, but fortunately this is rare. Recurrent prolapse with sciatica is more common (5–11% incidence) and may require revision decompression surgery.

REHABILITATION

After recovery from an acute disc rupture, or disc removal, the patient is mobilized and needs to limit physical exertion until 6–8 weeks to allow the annular defect to scar up and minimize re-prolapse. Non-impact exercises are usually safe.

OUTCOMES

The recent Spine Patient Outcomes Research Trial (SPORT) showed clinical and economic superiority of surgical versus non-operative care for lumbar disc herniation after 4 years.

PERSISTENT POSTOPERATIVE BACKACHE AND SCIATICA

Persistent symptoms after operation may be due to: (1) recurrent disc prolapse (typified by recurrence of sciatica after successful discectomy); (2) residual disc material in the spinal canal; (3) disc prolapse at another level; (4) postoperative discitis; (5) nerve root compression by a hypertrophic facet joint or a narrow lateral recess ('root canal stenosis'). After careful investigation, any of these may call for re-operation; but second procedures do not have a high success rate – third and fourth procedures still less.

ARACHNOIDITIS

Diffuse back pain and vague lower limb symptoms such as 'cramps', 'burning' or 'irritability' sometimes appear after myelography, epidural injections or disc operations. This diagnosis is now rarely made and is believed to have been a complication of oil-based contrast media used in myelography 30 years ago. There may also be sphincter dysfunction and male impotence. Patients complain bitterly and many are labelled neurotic. However, in some cases there are electromyographic abnormalities, and dural scarring with obliteration of the subarachnoid space can be demonstrated by MRI or at operation.

Treatment is generally unrewarding. Corticosteroid injections at best give only temporary relief, and surgical 'neurolysis' may actually make matters worse. Sympathetic management in a pain clinic, psychological support and a graduated activity programme are the best that can be offered.

LOWER BACK PAIN

An estimated 80% of the normal population will experience lower back pain (LBP) in their lifetime and this has enormous socioeconomic consequences. It is the leading cause of occupational disability worldwide. It is defined as pain and discomfort between the costal margin and inferior gluteal folds with or without leg pain.

BOX 18.2 CAUSES OF LOWER BACK PAIN (LBP)

Mechanical (80%)

- Muscular strains or from ligamentous injury
- Degenerative disc disease
- Facet joint disease (facet joint dysfunction)
- Spondylolysis
- Osteoporotic compression fractures
- Instability
- Sacroiliac joint pathology

Neurogenic (15%)

- Herniated disc
- Spinal stenosis
- Foraminal stenosis
- Disc annular tear and neuritis

Non-mechanical back pain (1–2%)

- Infections
- Neoplasms
- Inflammatory conditions

Referred pain (1–2%)

- Gastrointestinal disease
- Renal disease
- Aortic aneurysm

Other (2–4%)

- Somatization disorder
- Fibromyalgia
- Malingering

Pain may be acute (less than 6 weeks' duration), subacute (6–12 weeks) and chronic (more than 12 weeks).

Clinical features

Patient history is relevant. Mechanical pain is aggravated with movement and relieved by rest. Pain is described as dull, aching and similar to toothache and does not radiate down the leg. Patients may complain of symptoms on standing up from supine or seated positions, and pain on turning over in bed. An acute flare-up of pain often occurs on a background of chronic back pain and an increase in frequency may interfere with activities of daily living (ADLs). It is usually not possible to clinically distinguish the source of pain between the disc, facet joints, muscles, ligaments and the sacroiliac joints. Pain on flexion may be related to discogenic pain and facetogenic pain may be aggravated by hyperextension.

It is not clear whether *facet joint dysfunction* is a separate entity from degenerative disc disease. MRI studies have found facet joint osteoarthritis (OA) was most prominent at spinal levels with advanced disc degeneration and facet joint OA is rarely found in the absence of disc degeneration. In most people *degenerative disc disease (DDD)* precedes facet osteoarthritis. Facet joints, however, are true synovial joints and prone to all the normal synovial joint pathological processes.

Patients with pain radiating down to the buttocks and posterior thigh may have neurogenic pain such as *spinal stenosis*. Poor walking distance is a typical feature, stenotic symptoms are typically relieved by flexion and the 'shopping-cart sign' is almost pathognomonic of spinal stenosis. When other symptoms occur, such as pins and needles, sensation changes and weakness, a radiculopathy is likely.

Box 18.3 lists the red and yellow flag conditions to look for. *Red flags* are conditions in addition to back pain and warrant investigation to exclude serious pathology. *Yellow flags* are psychosocial factors that increase the risk of chronicity and disability from back pain.

BOX 18.3 LOWER BACK PAIN – RED AND YELLOW FLAGS

Red flags

- Age of onset less than 20 years or more than 55 years
- Recent history of violent trauma
- Constant progressive, non-mechanical pain (no relief with bed rest)
- Thoracic pain
- Past medical history of malignant tumour
- Prolonged use of corticosteroids
- Drug abuse, immunosuppression, HIV
- Systemically unwell
- Unexplained weight loss
- Widespread neurological symptoms (including cauda equina syndrome)
- Structural deformity
- Fever

Yellow flags

- Belief that back pain is harmful or potentially severely disabling
- Fear and avoidance of activity or movement
- Tendency to low mood and withdrawal from social interaction
- Expectation of passive treatment(s) rather than a belief that active participation will help

Examination

Spine examination may reveal muscle spasm, local tenderness and restriction of back movements. Pain on flexion may indicate disc pathology and with extension facet joint pain and spinal stenosis may predominate, but these tests have low specificity.

The *hips* should be examined to exclude hip joint pathology and the *sacroiliac joints* are routinely assessed (e.g. with the FABER (Flexion, ABduction, and External Rotation)/Patrick's test).

Neurological assessment includes eliciting nerve root irritation with the straight-leg raise test (L4–S1) and the femoral stretch test (L2–L4 nerve roots). Motor power, sensation and reflexes should be documented.

Investigations

X-RAYS

X-rays may be normal. However, in many cases there are mild to moderately severe features of intervertebral disc degeneration, mainly flattening of the 'disc space' and marginal osteophytes. The appearance of a 'traction spur' – a bony projection anteriorly a little distance from the upper or lower rim of the vertebral body – was held to be indicative of segmental instability (abnormal translational movement), but this has not been found to be statistically significant. In the lateral view, there may be slight displacement of one vertebra upon another, either forwards (spondylolisthesis) or backwards (retrolisthesis); this may become apparent only during flexion or extension. X-rays of the pelvis help assess the hip joints and sacroiliac joints for pathology.

Discography and facetography may reveal disc abnormalities, but they are unreliable in determining the source of pain.

BLOOD TESTS

FBC and ESR help screen for non-mechanical causes of lower back pain such as infections, inflammatory conditions and neoplasms. In elderly patients a serum protein electrophoresis and prostate-specific antigen in males should be part of the workup.

CT AND MRI

These investigations may reveal signs of disc degeneration as well as early features of OA in the facet joints (loss of articular cartilage space and curling over of the joint surfaces). MRI findings of high intensity zones (annular tear), disc degeneration and Modic end-plate changes are suggestive as causes of lower back pain, however their diagnostic potential is low. Many asymptomatic individuals have similar findings. Facet joint effusions on MRI have been shown to correlate with the dynamic instability shown on flexion/extension X-rays.

BONE SCAN AND SPECT SCAN

Active inflammatory conditions will show increased uptake in the facets and sacroiliac joints (as found in ankylosing spondylitis). Vertebral fractures and metastatic neoplasms can also be appreciated on bone scan.

Diagnosis

The diagnostic goals are to categorize the back pain into three categories: *serious spinal pathology*, *neural pain* and non-specific *back pain*. The history and clinical examination are thus first directed at distinguishing between non-spinal pathology and musculoskeletal back pain.

The second priority is to decide if there is a neural element of pain such as spinal stenosis or radiculopathy. If a neural pain source is absent from the clinical findings, the problem can be characterized as non-specific lower back pain and further into acute, sub-acute and chronic.

The clinical assessment should be supported with X-rays, FBC and an ESR. Where there are 'red flags' and/or abnormal specific findings, further imaging such as CT, MRI and bone scan may be required.

Treatment

Whatever the pattern in which the back pain may present, the pain may be sufficiently distressing or disabling to justify treatment in increasing degrees of invasiveness.

CONSERVATIVE TREATMENT

Initially if the symptoms are neither severe nor disabling; conservative measures should be encouraged for as long as possible:

Reassurance Many patients are concerned about possible serious disease or have a family history of malignancy. A clear explanation of the cause for their symptoms and counselling about the benign nature of the condition is often half the battle won. Patients can be reassured that most cases of acute back pain are self-limiting and resolve over a few weeks. A short course of analgesics and advice to stay active and continue normal daily activities including work will help. Smoking is associated with back pain and patients need to be made aware of this.

Medication Acute LBP treatment includes paracetamol and non-steroidal anti-inflammatory drugs (NSAIDs), short courses of opioids, or non-benzodiazepine muscle relaxants (cyclobenzaprine). Tricyclic antidepressants are more useful for chronic LBP and gabapentin tends to be used in radiculopathy.

Activity modification One of the most important aspects of treatment is modification of daily activities (bending, lifting, climbing, etc.) and specific activities relating to work. The patient may need retraining for a different job. The cooperation of employers is essential.

Physical therapy Conventional physiotherapy and spinal manipulation for patients may be of benefit. In the longer term, weight control and strengthening of the vertebral and abdominal muscles (core muscles) may prevent recurrences.

Spinal support A simple corset may provide symptomatic relief in some patients but in general they have no proven benefit and are costly.

Psychological support Chronic back pain can be psychologically as well as physically debilitating. Counselling and support are often welcomed by the patient. Perhaps the most successful treatment is the reassurance that the surgeon can provide for the vast majority of patients, to the effect that the patient has no serious spinal disease.

Injection therapy In chronic radiculopathy nerve root blocks may give short-term symptomatic relief and diagnostic information. Epidural steroids can be considered for spinal stenosis with some short-term benefit.

Injections are ineffective in non-specific lower back pain. No evidence supports the use of epidural steroids, facet blocks have little proven efficacy and denervation procedures (radiofrequency) even less evidence.

SURGERY

Only after all of the above measures have been tried and found to be ineffectual should a spinal fusion be considered. Even then, very strict guidelines should be followed if embarking on a road already crowded with patients labelled 'failed back surgery' is to be avoided:

1 Repeated examination should ensure that there is no other treatable pathology.
2 There should have been at least some response to conservative treatment; patients who 'benefit from nothing' will not benefit from spinal fusion either.
3 There should be unequivocal evidence of pathology at a specific level.
4 The patient should be emotionally stable and should not exaggerate his symptoms or display inappropriate physical signs (see later).
5 Patient expectations need to be in line with the surgeon's expectations. Discussion needs to had around the nature of the procedure, potential complications and the expected outcomes.
6 Surgery is effective for relief of pain and deformity in infections, tumours and fractures. Surgery is also cost effective and superior to non-operative treatment for degenerative conditions with neural pain (prolapsed disc, spinal stenosis and spondylolisthesis).
7 Surgery for non-specific back pain is far less effective. The recommendations are for at least 2 years of failed conservative treatment and a maximum of two levels of degenerative disc disease before it is even contemplated.

SURGICAL PROCEDURES FOR NON-SPECIFIC BACK PAIN

'Segmental instability' as an entity has long been thought to be a cause of back pain, either from disc degeneration or facet joint incompetence. No clinical findings or investigations have been shown to have diagnostic validity for accurate pain localization and this is partly a cause for poor clinical outcomes.

Surgery is traditionally aimed at stabilizing the painful segment with fusion; instrumentation increases the fusion rates but does not correlate with clinical outcomes. Options are posterior fusion, anterior fusion or combinations of both. Posterior fusions are instrumented with pedicle screws and anterior approaches allow insertion of cages to assist fusion and maintain sagittal alignment. Minimal access techniques have been popularized but have not shown clear benefit over traditional 'open' approaches, and there are steep learning curves.

Adjacent level degeneration is occasionally noted alongside fusion segments, possibly as a result of altered spinal biomechanics in addition to normal ageing processes. Non-fusion stabilization technologies such as total disc replacement (TDR) were developed to address this. Although these initially held great promise, a high revision rate and devastating approach-related complications resulted in loss of favour of these implants for use in the lumbar spine.

NOTE: As a rule of thumb, surgical outcomes for non-specific lower back pain do worse than surgery for neural pain, deformity, infection and tumours. Probably the best determinant of surgical outcome is patient selection. The presence of *yellow flags*, *Waddell signs* and *secondary gain issues* such as Workman's compensation are significant indicators of poor outcomes with surgical intervention, and these patients should rather be managed conservatively.

Surgical options are shown in Figure 18.36.

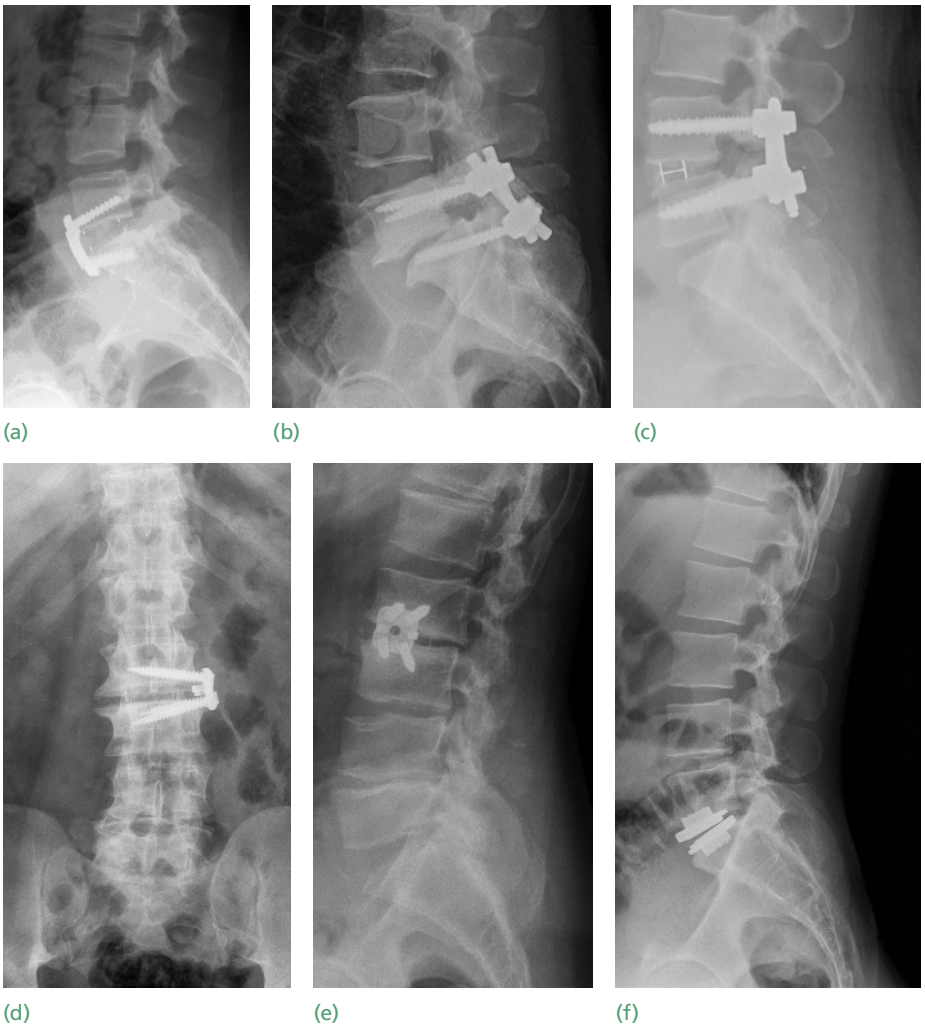


Figure 18.36 Surgical options (a) Anterior lumbar interbody fusion (ALIF); (b) posterior instrumented fusion; (c) transforaminal lumbar interbody fusion (TLIF); (d,e) direct lateral interbody fusion (DLIF); (f) total disc replacement (TDR).

SPONDYLOLISTHESIS

'Spondylolisthesis' means forward translation of one segment of the spine upon another. The shift is nearly always between L4 and L5, or between L5 and the sacrum (11% occur at L4/5 and 82% occur at L5/S1). Normal discs, laminae and facets constitute a locking mechanism that prevents each vertebra from moving forwards on the one below. Forward shift (or slip) occurs only when this mechanism has failed. Causes of spondylolisthesis are multifactorial but a large proportion are degenerative and the topic is therefore discussed here.

Classification

The *Wiltse–Newman classification of Spondylolisthesis* is most commonly used.

I Dysplastic
II Isthmic
  IIA Disruption of pars as a result of stress fracture
  IIB Elongation of pars without disruption related to repeated, healed microfractures
  IIC Acute fracture through pars
III Degenerative
IV Traumatic
V Pathologic
VI Iatrogenic

The first two types present in childhood and adolescence. Note that type VI (iatrogenic) is not part of the original Wiltse–Newman classification but, with injudicious facetectomy and pars fracture during laminectomies, iatrogenic instability can occur.

Dysplastic (4–8% incidence but accounts for 20% of all spondylolisthesis) Type I includes congenital abnormalities of the lumbosacral junction. The superior sacral facets are deficient or malorientated and the sacrum is dome-shaped or hypoplastic. The pars may be poorly developed. Slow and relentless forward slip leads to severe displacement. Associated anomalies (usually spina bifida occulta) are common.

Lytic or isthmic (5% incidence) In type II, the commonest variety, there are defects in the pars interarticularis (spondylolysis), or repeated breaking and healing may lead to elongation of the pars. The defect (which occurs in about 6% of people) is usually present

by the age of 7, but the slip may appear only some years later. It is difficult to exclude a genetic factor because spondylolisthesis often runs in families, and is more common in certain races, notably Eskimos; but the incidence increases with age up to the late teenage years, although clinical presentation with pain can continue into late middle age.

Type IIA is more common and is essentially a stress fracture from repetitive loading especially in competitive athletes (11% incidence in female gymnasts and 21% in college football players). This results in a radiolucent defect in the pars (non-union). Type IIB is characterized by repeated microfractures which heal with pars elongation and is occasionally confused with dysplastic type. Type IIC is a pars fracture caused by an acute injury.

Spondylolysis has a benign course. The general incidence of 6% does not change with increasing age from 20 to 80 years and the overwhelming majority of cases are asymptomatic. Only about 4% of pars defects tend to progress to significant slips of more than 20% over several years.

Degenerative (25% incidence) Degenerative (type III) changes in the disc and facet joint incompetence permit forward slip (nearly always at L4/5 and mainly in women of middle age). L4–L5 facets have a sagittal orientation which allows forward slippage (as opposed to the L5/S1 joints which are have a coronal orientation). Degenerative spondylolisthesis is commonly seen above a sacralized L5 vertebra due to increased mechanical stresses. These slips rarely progress more than 30% of the body width.

Post-traumatic Posterior arch fractures (not including the pars) may result in destabilization of the lumbar spine and allow vertebral slip.

Pathological Bone destruction (e.g. due to tuberculosis or neoplasm) may lead to vertebral slipping.

Iatrogenic As noted above, iatrogenic is not part of the original Wiltse–Newman classification but, with injudicious facetectomy and pars fracture during decompression, iatrogenic instability can occur leading to spondylolisthesis.

Pathology

Type I dysplastic spondylolisthesis will progress in 32% of cases. They are more likely to become high-grade slips with significant chance of neurological injury and more commonly require surgery. Anterior vertebral translation results in a sagittal deformity with compensatory pelvic rotation. This results in a vertical sacrum and loss of lumbar lordosis. With forward slipping there is compression on the cauda equina and the exiting foraminal nerve roots (L5). The degree of slip is measured by the amount of overlap of vertebral bodies and is expressed as a percentage. High-grade slips have more than 50% translation.

With *Type II pars isthmic* stress fractures, healing can occur with immobilization especially with unilateral defects. When non-union occurs, the fracture becomes corticalized and filled with fibrous tissue. A 'lytic' defect is visible on X-ray. The loss of the posterior facet support results in increased disc loads with subsequent degeneration and a small risk of spondylolisthesis (4%).

Type III is characterized by segmental 'instability' due to disc or facet incompetence with osteophytes and facet effusions. *Lateral recess stenosis* occurs due to facet osteophytes and ligamentum flavum hypertrophy which encroaches on the traversing nerve roots. Occasionally there is *foraminal stenosis* which compresses the exiting nerve root.

Clinical features

Typically a child or adolescent with spondylolysis presents with low back pain or pain that radiates to the buttock or posterior thighs. Onset is usually insidious and related to sporting activities; occasionally an acute injury may precipitate events. Neurological deficit is rare. There is a slightly higher incidence of spondylolysis in idiopathic scoliosis.

With a *dysplastic spondylolisthesis*, the child may have typical flat buttocks, a vertically oriented sacrum and a lumbosacral step palpable. Hamstring tightness is common and may result in flexed hips and knees. Neurological examination may reveal nerve root tension signs (commonly L5 root).

Degenerative spondylolistheses presents in middle age with chronic lower back pain, spinal stenosis or radicular pain (Figure 18.37). Walking distance is restricted and symptoms are relieved by forward flexion.

X-rays

Oblique films may demonstrate the classic 'Scotty dog neck' which is pathognomonic of a pars fracture with a broken neck or collar. About 20% of pars defects are only shown on oblique films.

Lateral views show the forward shift (spondylolisthesis) of the upper part of the spinal column on the stable vertebra below (Figure 18.38); elongation of the arch or defective facets may be seen.

When there is no gap, the pars interarticularis is elongated or the facets are defective. The degree of slip is measured by the amount of overlap of adjacent vertebral bodies and is usually expressed as a percentage.

Prognosis

Dysplastic spondylolisthesis appears at an early age, often goes on to a severe slip and carries a significant risk of neurological complications. If progression is predicted, early surgery is recommended.

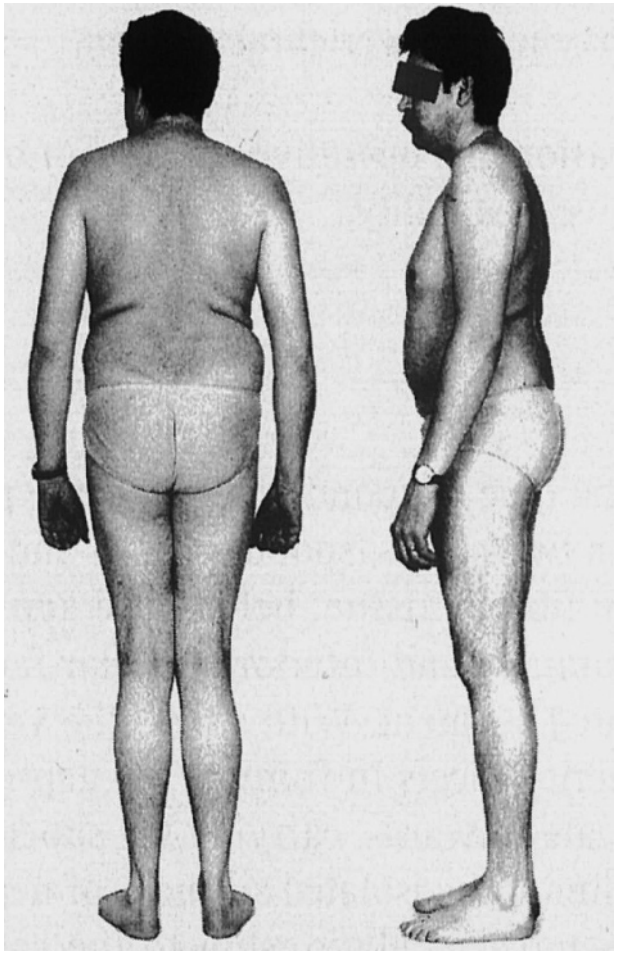


Figure 18.37 Spondylolisthesis – clinical appearance
The transverse loin creases, forward tilting of the pelvis and flattening of the lumbar spine are characteristic.

Lytic (isthmic) spondylolisthesis with less than 10% displacement does not progress after adulthood, but it may predispose the patient to later back problems. It is not a contraindication to strenuous work unless severe pain supervenes. With slips of more than 25% there is an increased risk of backache in later life.

Degenerative spondylolisthesis is uncommon before the age of 50, progresses slowly and seldom exceeds 30%.

Treatment

Conservative treatment, similar to that for other types of back pain, is suitable for most patients and is based on symptom management. Short-term bed rest, activity restriction, pain medication, NSAIDs, muscle relaxants, steroid injections, physical therapy and bracing are all common treatment modalities.

Operative treatment is indicated if: (1) the symptoms are disabling and interfere significantly with work and recreational activities (loss of activities of daily living); (2) the slip is more than 50% and progressing; (3) neurological compression is significant.

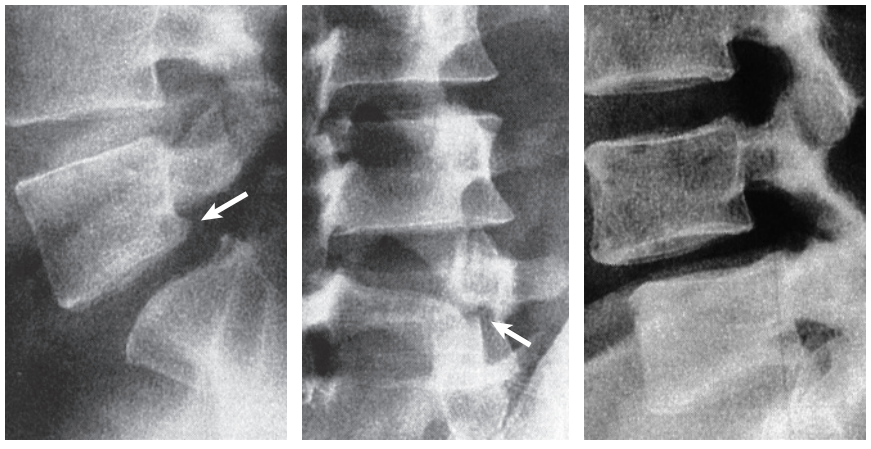
CHILDREN

In *dysplastic spondylolisthesis*, there is commonly progression to high grade slips (>50% translation) at L5/S1 and the child presents with lower back pain, hamstring tightness and sciatica. Surgical treatment is fusion of the dysplastic listhesed segment. Some controversy exists over the need for reduction of the slip, the extent of reduction and the surgical technique.

Historically, posterolateral *in situ* fusion has been the procedure of choice for paediatric spondylolisthesis. However, for high-grade spondylolisthesis, the small transverse processes result in a high pseudarthrosis rate and sagittal balance is not addressed. Instrumented reduction and circumferential fusion improves fusion rates and the posture, but the surgery is technically demanding and has a higher complication rate.

Spondylolysis The mainstay of conservative management is activity restriction. NSAIDs are used for analgesia as required. The pain-producing sporting activities need to be restricted and active competition stopped for 4–12 weeks. Bracing is controversial with no consensus.

With failed conservative treatment after 9–12 months, surgery is indicated. Posterior uninstrumented fusion is the default procedure but this



(a) (b) (c)

Figure 18.38 Spondylolisthesis – X-rays (a) There is a break in the pars interarticularis of L5, allowing the anterior part of the vertebra to slip forwards. In this case the gap is easily seen in the lateral X-ray, but usually it is better seen in the oblique view (b). In degenerative spondylolisthesis there is no break in the pars – the degenerate disc and eroded facet joints permit one vertebra to slide forwards on the other (c). There is no pars defect; the dehydrated disc permits slipping, usually at L4/5.

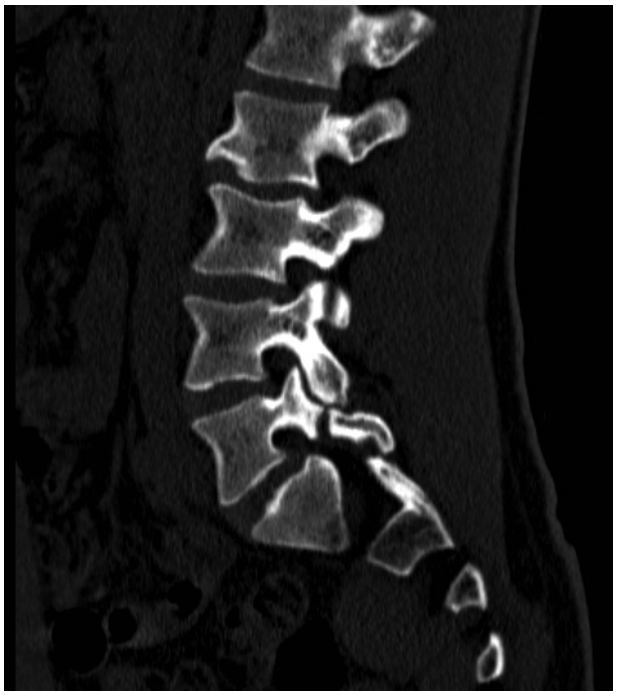


Figure 18.39 Spondylolysis CT scan shows corticalization and a fibrous non-union.

sacrifices a motion segment. Where there is no disc degeneration or spondylolisthesis in a young patient (<20), pars repair is possible, but the pain should be isolated to the lysis, which can be confirmed with local anaesthetic injections. When there are relative contraindications to pars repair, posterior fusion remains the gold standard.

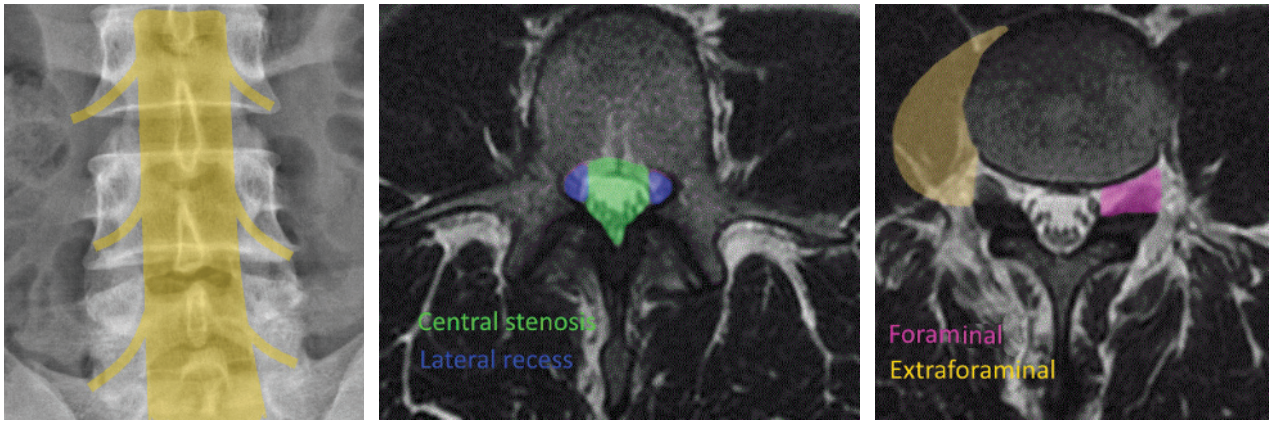
Degenerative spondylolisthesis Most patients respond to conservative treatment but 10–15% of patients with degenerative spondylolisthesis will require surgery for lower back pain, spinal stenosis and/or radiculopathy. Progressive weakness, cauda equina symptoms and loss of activities of daily living (ADLs) with poor quality of life are indications for surgery.

When symptoms are mainly those of spinal claudication, decompression alone is successful. However, mechanical back pain, abnormal motion on dynamic X-rays and large facet effusions on MRI are features which may require fusion in addition to decompression. Posterolateral fusion is the standard, and pedicle screw instrumentation produces higher fusion rates. Modern segmental pedicle screw fixation allows spondylolisthesis reduction and restoration of foraminal height for nerve root decompression. Posterior instrumentation may be augmented with anterior interbody fusion (circumferential fusion) either from posterior or a separate anterior approach. This allows improved lordosis correction and fusion rates (especially in smokers).

SPINAL STENOSIS

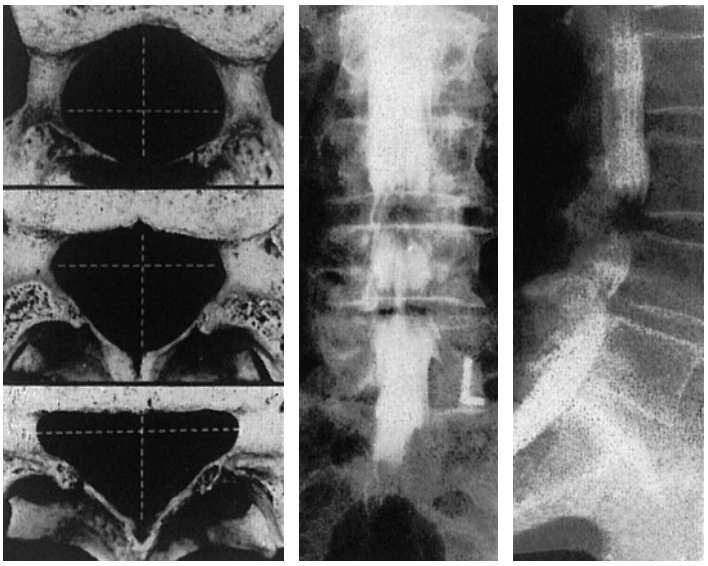
Spinal stenosis refers to reduced spinal canal dimensions and neural compression and may be congenital or acquired. *Congenital* vertebral dysplasia occurs in achondroplasia or hypochondroplasia. *Acquired* stenosis occurs mostly with degeneration but also in spondylolysis and spondylolisthesis, iatrogenic, post-traumatic, local infection and metabolic stenosis.

Anatomically, spinal stenosis is categorized as central, lateral or foraminal. *Central spinal stenosis* involves the area between the facet joints and contains the dural sac and nerve roots. The lateral border of the dura to the medial border of the pedicle defines the *lateral recess* containing the traversing nerve root. The *foraminal region* is situated under the pars and contains the exiting nerve root. Lateral to the pars and facet joints lies the *extraforaminal region* where a nerve root may be compressed by a 'far lateral' disc protrusion (Figure 18.40).



(a) (b) (c)

Figure 18.40 Spinal stenosis – anatomical regions (a) X-ray relationships of the dural sac and exiting nerve roots – note how the roots closely exit under the pedicles; (b) central spinal canal and lateral recess; (c) the foraminal region and extraforaminal region.



(a) (b) (c)

Figure 18.41 Spinal stenosis (a) The shape of the lumbar spinal canal varies from oval (with a large capacity) to trefoil (with narrow lateral recesses); further encroachment on an already narrow canal can cause an ischaemic neuropathy and 'spinal claudication'. (b,c) Myelogram showing marked narrowing of the radio-opaque column at the level of stenosis.

Degenerative stenosis is the most common and involves changes in the discs and facet joints. Facet joints develop synovitic changes, cartilage thinning and capsular laxity. This allows increased segmental motion and accelerated disc degeneration with loss of disc height and bulging or prolapse into the canal. Vertebral end-plate osteophytes contribute to the stenosis. Facet osteophytes enlarge and encroach on the spinal canal along with the ligamentum flavum, which thickens due to fibrosis. The normally oval canal becomes trefoil-shaped (Figure 18.41). The increasing canal narrowing, neural compression, vascular compromise and possibly neural demyelination cumulate in symptoms of spinal stenosis. In addition, there are also dynamic changes; with axial loading and extension the canal space decreases, while with flexion the canal space increases. This is the basis for symptom relief with flexion in spinal stenosis.

In *spondylolysis* fibrocartilage may proliferate in the pars defect causing foraminal stenosis. *Spondylolisthesis* may also result in foraminal stenosis as well as lateral recess stenosis. Occasionally foraminal stenosis may result from a foraminal disc herniation or a facet joint cyst.

Clinical features

Usually more common in women, the symptoms are complaints of aching, heaviness, numbness and paraesthesia in the thighs and legs. Pain occurs after standing upright or walking for 5–10 minutes, and it is relieved by sitting, squatting or leaning against a wall to flex the spine (hence the term 'spinal claudication'). The patient may prefer walking uphill, which flexes the spine (and maximizes the spinal canal capacity), to downhill, which extends it. With foraminal stenosis the symptoms may be unilateral. The patient sometimes has a previous history of disc prolapse, chronic backache or spinal surgery.

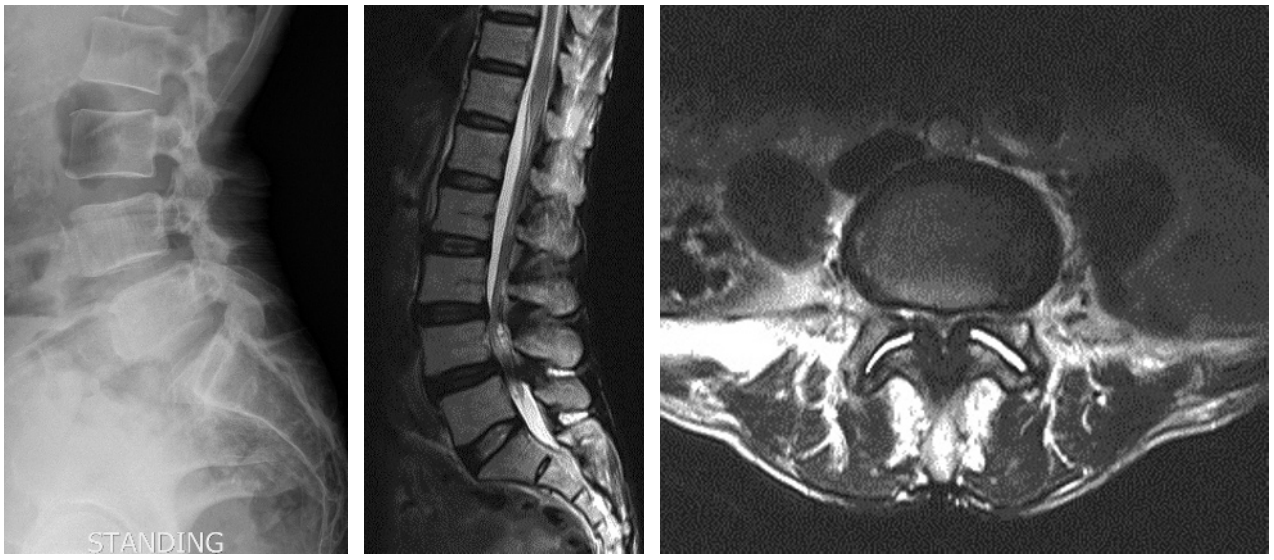
Examination, especially after getting the patient to reproduce the symptoms by walking, may (rarely) show neurological deficit in the lower limbs. Often the neurological examination is normal. Intact pedal pulses and absence of trophic skin changes would confirm the claudication as spinal rather than arterial, but beware of the older patient who could have both spinal and arterial claudication. In general, vascular claudication symptoms are relieved after a short rest (5 minutes or so) or while still standing whereas spinal stenosis requires flexion for relief. Vascular claudication is aggravated while walking uphill or riding a stationary bicycle.

It is imperative to exclude vascular claudication, hip joint pathology and peripheral neuropathy as a differential diagnosis.

Imaging

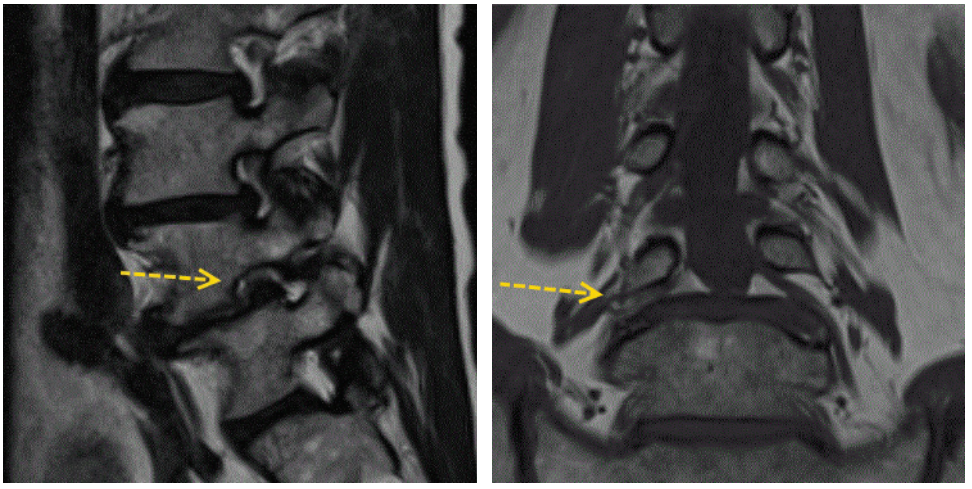
Absolute stenosis is defined when the mid-sagittal diameter of the canal is less than 10 mm and *relative stenosis* with a mid-sagittal canal diameter of 10–13 mm. These are useful guidelines but they do not have reliable clinical correlation.

AP, lateral lumbar and AP pelvis *X-rays* are mandatory as part of first-line investigations. Spinal alignment, disc degeneration, spondylolisthesis and fractures can be appreciated. Dynamic lateral X-rays may show instability with more than 5 mm translation and oblique views may demonstrate pars defects. Pelvis X-rays will help to evaluate the hip and sacroiliac joints.



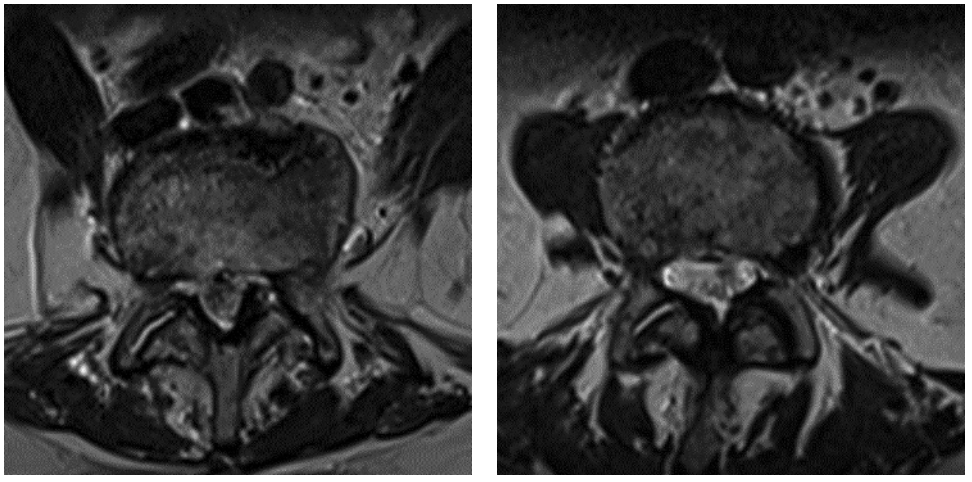
(a) (b) (c)

Figure 18.42 Spinal stenosis (a) L4/5 Degenerative spondylolisthesis; (b) MRI confirms stenosis at L4/5 level; note the listhesis has reduced since the MRI is taken supine; (c) axial MRI shows the lateral recess stenosis from facet hypertrophy; facet effusions are found with facet incompetence and instability.



(a) (b)

Figure 18.43 Foraminal stenosis (a) Sagittal MRI shows the compressed nerve root; (b) coronal MRI of foraminal stenosis.



(a) (b)

Figure 18.44 Lateral recess stenosis (a) Sub-facetal stenosis as opposed to a relatively normal level in the same patient (b).

MRI is the imaging of choice for investigation of spinal stenosis. It is non-invasive, safe and can differentiate various tissue types. Disc degeneration and disc prolapse can be appreciated as well as facet arthrosis and effusions. Central, lateral recess and foraminal stenosis are illustrated as well as extra-foraminal stenosis, which is not well shown on CT myelography.

Myelography with CT scan is indicated where there are contraindications to MRI or metallic implants causing artefact on MRI. CT myelography may better demonstrate canal narrowing with dynamic stenosis or severe scoliosis.

Treatment

CONSERVATIVE MANAGEMENT

With a short duration of symptoms and moderate pain, conservative management is the first line of treatment. Patient reassurance and education, analgesics (paracetamol/acetaminophen) and NSAIDs, non-impact exercise programme, aerobic fitness, activity modification and epidural steroid injections are relatively effective.

When there is failure of conservative treatment, loss of activities of daily living and severe neurological symptoms, surgery is indicated.

SURGICAL TREATMENT

Posterior decompression is the mainstay of surgical treatment. For central stenosis, laminectomy is performed. The lateral recess stenosis is decompressed with undercutting facetectomies and removal of ligamentum flavum. Care should be taken to excise less than 50% of the facet joints and avoid damage to the pars interarticularis to prevent iatrogenic instability, which would necessitate fusion. Foraminal stenosis requires fusion to decompress and maintain foraminal patency.

Spinal stenosis with spondylolysis, spondylolisthesis, scoliosis and kyphosis are indications for fusion with decompression.

The recent SPORT trial (Spine Patient Outcomes Research Trial) confirmed the cost-effectiveness of surgery over non-operative treatment over a 4-year period for spinal stenosis.

APPROACH TO DIAGNOSIS IN PATIENTS WITH LOW BACK PAIN

Chronic backache is such a frequent cause of disability in the community that it has become almost a disease in itself. The following is a suggested approach to more specific diagnosis. Careful history-taking and examination will uncover one of five pain patterns.

Transient backache following muscular activity This suggests a simple back strain that will respond to a short period of rest followed by gradually increasing exercise. People with thoracic kyphosis (of whatever origin), or fixed flexion of the hip, are particularly prone to back strain because they tend to compensate for the deformity by holding the lumbosacral spine in hyperlordosis.

Sudden, acute pain and sciatica In young people (those under the age of 20), it is important to exclude infection and spondylolisthesis; both produce recognizable X-ray changes. Patients aged 20–40 years are more likely to have an acute disc prolapse: diagnostic features are: (1) a history of a lifting strain; (2) unequivocal sciatic tension; and (3) neurological symptoms and signs. Elderly patients may have osteoporotic compression fractures, but metastatic disease and myeloma must be excluded.

Intermittent low back pain after exertion Patients of almost any age may complain of recurrent backache following exertion or lifting activities and this is relieved by rest. Features of disc prolapse are absent but there may be a history of acute sciatica in the past. In early cases, X-rays usually show no abnormality; later there may be signs of lumbar spondylosis in those over 50 years and osteoarthritis of the facet joints is common.

These patients need painstaking examination to: (1) uncover any features of radiological segmental instability or facet joint osteoarthritis; and (2) determine whether those features are incidental or are likely to account for the patient's symptoms. In the process, disorders such as ankylosing spondylitis, chronic infection, myelomatosis and other bone diseases must be excluded by appropriate imaging and blood investigations.

Back pain plus pseudoclaudication These patients are usually aged over 50 and may give a history of previous, long-standing back trouble. The diagnosis of spinal stenosis should be confirmed by CT and/or MRI.

Severe and constant pain localized to a particular site This suggests local bone pathology, such as a compression fracture, Paget's disease, a tumour or infection. Spinal osteoporosis in middle-aged men is pathological and calls for a full battery of tests to exclude primary disorders such as *myelomatosis*, *carcinomatosis*, *hyperthyroidism*, *gonadal insufficiency*, *alcoholism* or *corticosteroid* usage.

CHRONIC BACK PAIN SYNDROME

Patients with chronic backache may despair of finding a cure for their trouble (or, indeed, even a diagnosis that everyone agrees on), and they often develop

affective and psychosomatic ailments that subsequently become the chief focus of attention. This 'illness behaviour' is both self-perpetuating and self-justifying. It is usually accompanied by *'non-organic' (inappropriate) physical signs* such as: (1) pain and tenderness of bizarre degree or distribution; (2) pain on performing impressive but non-stressful manoeuvres such as pressing vertically on the spine or passively rotating the entire trunk; (3) variations in response to tests such as straight-leg raising while distracting the patient's attention; (4) sensory and/or motor abnormalities that do not fit the known anatomical and physiological patterns; and (5) overdetermined behaviour during physical examination (trembling, sweating, hyperventilating, inability to move, a tendency to fall and exaggerated withdrawal) – usually accompanied by loud groaning and exclamations of discomfort.

Patients with these features are unlikely to respond to surgery and they may require prolonged support and management in a special pain clinic – but only after every effort has been made to exclude organic pathology (Figure 18.45).

NOTES ON APPLIED ANATOMY

The spine as a whole

The spine has to move, to transmit weight and to protect the spinal cord. In upright man, the lumbar segment is lordotic and the column acts like a crane (Figure 18.46); the paravertebral muscles are the cables that counterbalance any weight carried anteriorly. The resultant force, which passes through the nucleus pulposus of the lowest lumbar disc, is therefore much greater than if the column were loaded directly over its centre. Even at rest, tonic contraction of the posterior muscles balances the trunk, so the lumbar spine is always loaded. When the intradiscal pressure in volunteers during various activities was measured, it was found to be as high as 10–15 kg/cm^2 while sitting, about 30% less on standing upright, and 50% less on lying down. Leaning forward or carrying a weight produces much higher pressures, although when a heavy weight is lifted breathing stops and the abdominal muscles contract, turning the trunk into a tightly inflated bag that cushions the force anteriorly against the pelvis. (Could it be that champion weightlifters benefit in this way from having voluminous bodies?)

Seen from the side, the dorsal spine is convex backwards (kyphosis); the cervical and lumbar regions are convex forwards (lordosis). In forward flexion the lordotic curves straighten out. Lying supine with the legs straight tilts the pelvic brim forwards; the lumbar spine compensates by increasing its lordosis. If the hips are unable to extend fully (fixed flexion deformity), the lumbar lordosis increases still more until the lower limbs lie flat and the flexion deformity is masked.

Vertebral components

Each segment of the vertebral column transmits weight through the vertebral body anteriorly and the facet joints posteriorly. Between adjacent bodies (and firmly attached to them) lie the intervertebral discs. These compressible 'cushions', and the surrounding ligaments and muscles, act as shock-absorbers; if they are degenerate or weak, their ability to absorb some of the force is diminished and the bones and joints suffer the consequences.

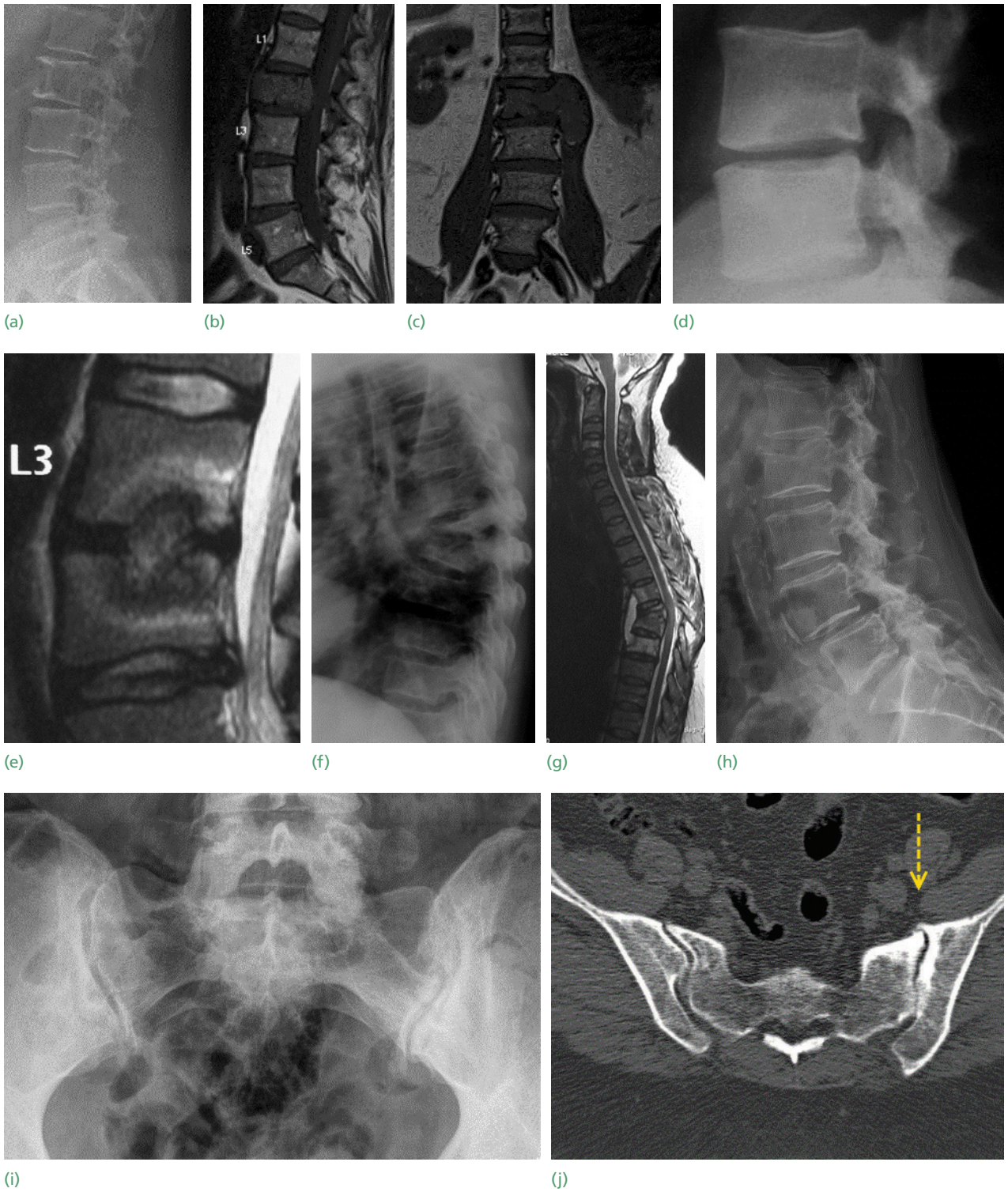
The vertebral body is cancellous, but the upper and lower surfaces are condensed to form sclerotic endplates. In childhood these are covered by cartilage, which contributes to vertebral growth. Later the peripheral rim ossifies and fuses with the body, but the central area remains as a thin layer of cartilage adherent to the intervertebral disc. The epiphyseal end plates may be damaged by disc pressure during childhood, giving rise to irregular ossification and abnormal vertebral growth (Scheuermann's disease).

Intervertebral disc

The disc consists of a central *avascular nucleus pulposus* – a hydrophilic gel made of protein–polysaccharide, collagen fibres, sparse chondroid cells and water (88%), surrounded by concentric layers of fibrous tissue – the *annulus fibrosus*. If the physicochemical state of the nucleus pulposus is normal, the disc can withstand almost any load that the muscles can support; if it is abnormal, even small increases in force can produce sufficient stress to rupture the annulus.

Movements

The axis of movements in the thoracolumbar spine is the nucleus pulposus; the disposition of the facet joints determines which movements occur. In the lumbar spine these joints are in the anteroposterior plane, so flexion, extension and sideways tilting are free but there is virtually no rotation. In the thoracic spine the facet joints face backwards and laterally, so rotation is relatively free; flexion, extension and tilting are possible but are grossly restricted by the ribs. The costovertebral joints are involved in respiration and their limitation is an early feature of ankylosing spondylitis.



(a) (b) (c) (d)

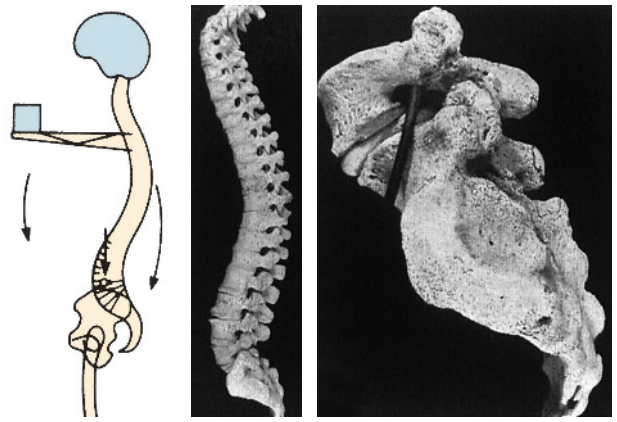
(e) (f) (g) (h)

(i) (j)

Figure 18.45 Other causes of chronic back pain (a,b,c) Metastatic disease; (d,e) spondylodiscitis; (f,g) tuberculosis; (h) L2 compression fracture; (i,j) sacroiliitis.

Spinal canal

The shape of the canal changes from ovoid in the upper part of the lumbar spine to triangular in the lower. Variations are common and include the trefoil canal, whose shape is mainly due to thickening of the laminae. This shape is harmless in itself, but further encroachment on the canal (e.g. by a bulging disc or hypertrophic facet joints) may cause compression of the spinal contents (spinal stenosis).



(a) (b) (c)

Figure 18.46 Anatomy (a,b) The vertebral column has a series of gentle curves that produce lordosis in the cervical and lumbar regions and kyphosis in the dorsal segment. The column functions like a crane, the weight in front of the spine being counterbalanced by contraction of the posterior muscles. (c) Relationship of nerve root to disc and facet joint.

Spinal cord

The spinal cord ends at about L1 in the conus medullaris, but lumbosacral nerve roots continue in the spinal canal as the cauda equina and leave at appropriate levels lower down. The dural sac continues as far as S2, and whenever a nerve root leaves the spine it takes with it a dural sleeve as far as the exit from the intervertebral foramen. These dural sleeves can be outlined by contrast-medium radiography (radiculography).

Intervertebral foramina and nerve roots

Each intervertebral foramen is bounded anteriorly by the disc and adjoining vertebral bodies, posteriorly by the facet joint, and superiorly and inferiorly by the pedicles of adjacent vertebrae. It can therefore be narrowed by a bulging disc or by joint osteophytes. The segmental nerve roots leave the spinal canal through the intervertebral foramina, each pair below the vertebra of the same number (thus, the fourth lumbar root runs between L4 and L5). The segmental blood vessels to and from the cord also pass through the intervertebral foramen. Occlusion of this little passage may occasionally compress the nerve root directly or may cause nerve root ischaemia (especially when the spine is held in extension).

Nerve supply of the spine

The spine and its contents (including the dural sleeves of the nerve roots themselves) are supplied by small branches from the anterior and posterior primary rami of the segmental nerve roots. Lesions of different structures (e.g. the posterior longitudinal ligament, the dural sleeve or the facet joint) may therefore cause pain of similar distribution. Pain down the thigh and leg ('sciatica') does not necessarily signify root pressure; it may equally well be referred from a facet joint or any painful spinal tissue.

Blood supply

In addition to the spinal arteries, which run the length of the cord, segmental arteries from the aorta send branches through the intervertebral foramina at each level. Accompanying veins drain into the azygos system and inferior vena cava and anastomose profusely with the extradural plexus, which extends throughout the length of the spinal canal (Batson's plexus).

FURTHER READING

Albert H, Sorensen J, Christensen B, *et al.* Antibiotic treatment in patients with chronic low back pain and vertebral bone edema (Modic type 1 changes): a double-blind randomized clinical controlled trial of efficacy. *Eur Spine J* 2013; **22**: 697–707.

Boos N, Weissbach S, Rohrbach H, *et al.* Classification of age-related changes in lumbar intervertebral discs. *Spine* 2002; **27**: 2631–44.

Brinjikji W, Luetmer P, Comstock B, *et al.* Systematic literature review of imaging features of spinal degeneration in asymptomatic populations. *Am J Neuroradiol* 2014; **36**(4): 811–16.

Guo JB, Zhu Y, Chen BL, *et al.* Surgical versus non-surgical treatment for vertebral compression fracture with osteopenia: a systematic review and meta-analysis. *PLoS One* 2015; **10**(5): e0127145.

Jensen TS, Karppinen J, Sorensen JS, *et al.* Prevalence of vertebral endplate signal (Modic) changes and their association with non-specific low back pain – A systematic literature review. *Eur Spine J* 2008; **17**:1407–22.

Tosteson A, Tosteson T, Lurie J, *et al.* Comparative effectiveness evidence from the Spine Patient Outcomes Research Trial. *Spine* 2011; **36**(24): 2061–8.

Waddell G, McCulloch JA, Kummel E, *et al.* Nonorganic physical signs in low-back pain. *Spine* 1980; **5**: 117–25.

Waddell G, Bircher M, Finlayson D, *et al.* Symptoms and signs: physical disease or illness behaviour. *BMJ* 1984; **289**: 739–41.

Weinstein JN, Lurie JD, Tosteson TD, *et al.* Surgical versus nonoperative treatment for lumbar disc herniation: four-year results for the Spine Patient Outcomes Research Trial (SPORT). *Spine* 2008; **33**(25): 2789–800.

The hip

19

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THE IMMATURE HIP

DEVELOPMENTAL DYSPLASIA OF THE HIP

The term developmental dysplasia of the hip (DDH), coined by Klisic in the late 1980s, has replaced the term congenital dislocation of the hip (CDH) in order to reflect a spectrum of abnormalities in the development of the hip joint, ranging from mild acetabular dysplasia to irreducible dislocation. The true incidence is not known precisely although there is probably some, at least transient, abnormality of the neonatal hip in up to 10% of females, but equipoise exists in terms of what needs to be treated and what can be observed and which patients, or their parents, can be reassured and discharged.

Broadly, DDH can be classed into four groups on the basis of a combination of clinical and sonographic examination.

1 *Reduced and stable but dysplastic*
The hip may be clinically normal on examination or may have subtle laxity without frank dislocation despite manipulation. When imaged ultrasonographically, the hips demonstrate a shallow acetabular cup.
2 *Reduced but dislocatable*
The femoral head normally sits within the acetabulum but can be dislocated by clinical manipulation using the *Barlow manoeuvre* to push the femoral head superior and posterior over the edge of the shallow acetabulum (Figure 19.1).
3 *Dislocated but reducible*
In this situation the femoral head naturally sits outside of the acetabulum (usually superior and posterior). However, by gentle manipulation using the *Ortolani manoeuvre* the femoral head can be reduced into the acetabulum (Figure 19.2).

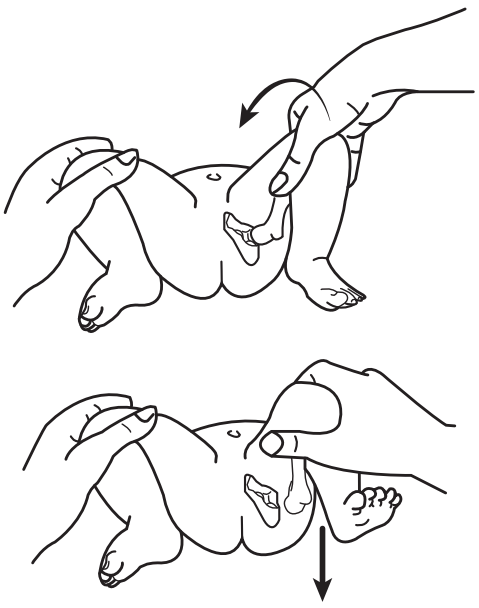


Figure 19.1 Barlow manouevre

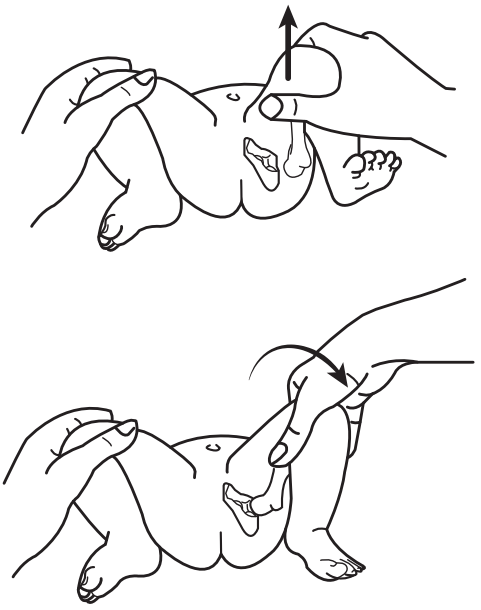


Figure 19.2 Ortolani manouevre

4 *Dislocated and irreducibile*
This is the most severe class of DDH and may be identified neonatally due to asymmetrical abduction of the hip. The hip may be stiff, with tightness of the adductor longus tendon, and the femoral head cannot be clinically manipulated into the acetabulum. Ultrasound imaging will confirm that the femoral head is located outside the acetabulum.

Pathophysiology

Embryonically the femoral head and acetabulum develop from a single cleft of primitive mesenchymal cells. The femoral head begins to separate between 7 and 8 weeks' gestation and the process is completed by 11–12 weeks, at which point the femoral head is almost completely contained within the acetabulum on axial cross section. The femoral head remains enlocated normally but relative growth results in least coverage at around term. Postnatal development of the labrum further deepens the socket to increase stability of the hip joint and walking (at around the child's first birthday) is a potent stimulus to further acetabular development.

Dislocation (or *dysplasia*) of the hip can occur at specific points of time during development. The first time at which dislocation may occur is 10 weeks when the lower extremity limb bud rotates medially. Neuromuscular disorders may cause muscular imbalance leading to dislocation around 18 weeks' gestation. Dislocation at these early stages leads to abnormal development of all parts of the hip joint and is thus termed a *teratological dislocation*. Children with teratological dislocations commonly have other developmental abnormalities such as Ehlers Danlos, Down's syndrome or arthrogryposis, and the principles of treatment are different from DDH.

During the final 4 weeks of gestation, mechanical forces play a large part in the positioning of the hip joint. In the left occiput anterior position, the commonest prenatal fetal position, the left hip is adducted against the maternal sacrum, increasing the risk of dislocation. Other factors at this stage are known to increase the risk of DDH, including breech position, oligohydramnios, multiple fetuses, circulating maternal relaxin combined with the relatively low percentage acetabular coverage of the femoral head. Postnatally, swaddling of the lower extremities holds the hips in extension and adduction and can be an additional contributing factor to subluxation or dislocation in some countries.

In the subluxed position, the labrum is flattened under the pressure of the femoral head and becomes flattened or everted. In addition, the acetabulum develops non-concentrically and does not deepen around the femoral head, which leads to a shallow acetabulum and unstable hip joint. Dislocation of the femoral head leads to stretching of the inferior capsule and adductors which, if untreated, may lead to contractures and limited range of abduction.

In addition to oligohydramnios and breech positioning, the main *risk factors* for DDH are female gender, firstborn children and family history. Three percent of all births are breech position and of these 23% will have some degree of DDH. Other skeletal abnormalities commonly associated with DDH include torticollis and postural foot abnormality such as metatarsus adductus.

Diagnosis

All neonates should undergo clinical examination of their hips, and it seems sensible that 'at-risk' hips might merit ultrasound examination, but there is still debate as to its optimal role.

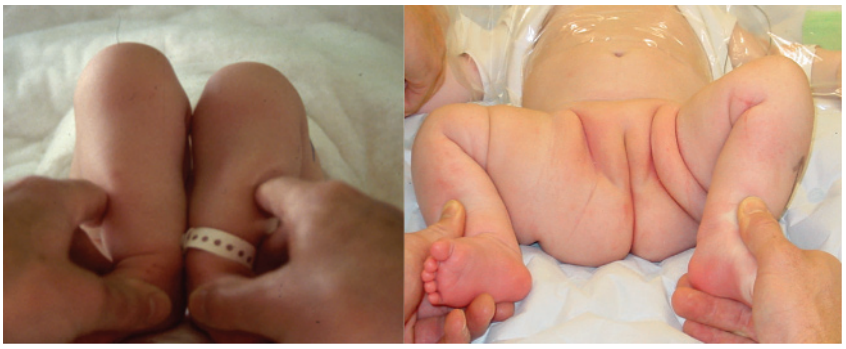
The Barlow and Ortolani tests are the mainstays in examination of the newborn hip. The *Barlow manoeuvre* is a provocation test conducted by adducting the flexed hip and applying gentle anterior to posterior pressure in order to push the femoral head superior and posterior over the edge of a shallow acetabulum. The *Ortolani test* is a relocation manoeuvre conducted by gently manipulating the flexed hip from adduction to abduction to bring the femoral head anteriorly back into the acetabulum from a dislocated position. These manoeuvres are best performed with the clinician's palms over the infant's knees and the middle fingertip placed over the greater trochanter.

At a later stage, clinical examination findings may include asymmetrical skin folds, although most experts feel that these are generally not significant. Once there is an established dislocation, one can detect a leg-length discrepancy with a shorter affected limb, positive *Galeazzi test* and reduced range of abduction (Figure 19.3). Bilateral DDH is always more difficult to diagnose as symmetrical changes are more difficult to pick up.

Imaging

The introduction of *ultrasound imaging*, in the 1980s, allowed visualization of the soft-tissue components of the infant hip including the cartilage of the femoral head and acetabulum, the capsule and the labrum. The use of multiplanar and dynamic ultrasound enables visualization of the femoral head within the acetabulum and assessment of the shape and depth of the acetabular cup. Ultrasound is best used for children before 6 months of age, after which ossification of structures makes plain radiographs increasingly more helpful.

The most widely used ultrasonographic technique was popularized by Graf. This technique uses static coronal images to determine the depth and shape of



(a) (b)

Figure 19.3 Galeazzi test
(a) Positive Galeazzi test, indicating limb-length discrepancy and (b) reduced abduction in flexion of the left hip.

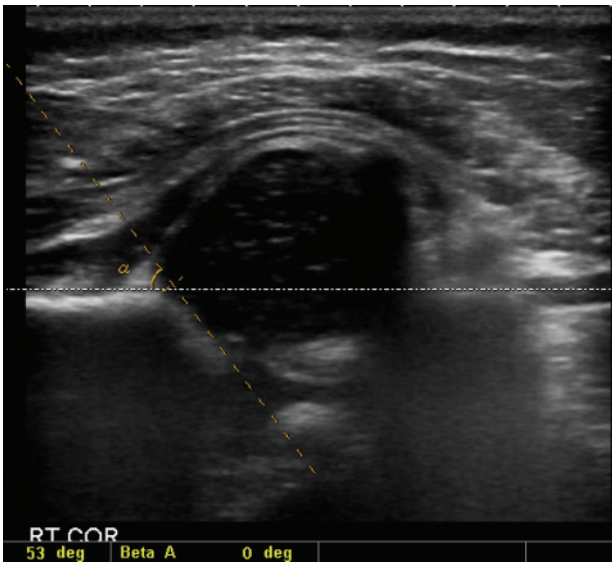


Figure 19.4 DDH Coronal static ultrasound of moderately dysplastic right hip with alpha angle of 53°.

the acetabulum by measuring the alpha angle, an angle formed between the straight edge of the ilium and the acetabular roof. A normal alpha angle measurement is 60 degrees or greater (Figure 19.4). In addition, Graf described the beta angle to measure cartilaginous coverage of the femoral head, with a normal angle measurement of 55 degrees, but this is less helpful in clinical practice. Table 19.1 demonstrates ultrasonographic classification of DDH according to Graf.

Harcke later described the use of dynamic imaging and transverse sequences to better understand the three-dimensional relationship between the femoral head and acetabulum. Consensus was reached at a meeting involving both Graf and Harcke that described the Dynamic Standard Minimum Examination combining a static coronal image and a transverse stress image for optimal assessment of the infant hip.

Management and treatment

It is clear that treating DDH is more complicated with a less favourable outcome the later the diagnosis is made and so significant resources have been applied to achieve early diagnosis by screening. *Clinical screening* is clearly sensible, but debate persists on the specific benefits, compared to cost and risk of overtreatment, of selective or universal ultrasound screening. Universal screening of infants using ultrasound is not recommended in the UK.

There is good evidence to support *management in the neonate*. The treatment of Ortolani-positive hips, dislocated at rest but reducible, should begin as soon as practical. Conversely, it is reasonable to delay treatment of Barlow-positive hips for up to 2 weeks without compromising the final outcome, as a large percentage of such hips will stabilize within that period. At 2 weeks, failure to improve either clinically or on the basis of an ultrasound is an indication to commence treatment.

Table 19.1 Classification of ultrasonographic hip types (Graf)

Type	α-angle (degrees)	β-angle (degrees)	Age (months)	Acetabular roof (shape)	Femoral head (location)
I	>60	<55	Any	Normal	Central
IIa	43–60	55–77	0–3	Normal	Central
IIb	43–60	55–77	>3	Normal, delayed ossification	Central
IIIa	<43	>77	Any	Upward displacement	Lateralized
IIIb	<43	>77	Any	Upward displacement and bending	Lateralized
IV			Any	Stretched	Dislocated

Treatment is generally accomplished by maintaining hip abduction through the use of a brace. The most commonly used abduction brace internationally is the *Pavlik harness (PH)*. Although readily used and widely accepted, PH therapy is not without risk of complication, and successful outcome is not universal. Poor positioning of the harness, prolonged treatment and lack of parental compliance can be associated with treatment failure. Significant complications that have been reported include avascular necrosis (AVN) of the femoral head, femoral nerve palsy, and brachial plexus palsy. PH therapy should be discontinued at least until resolution of the complication and often is abandoned completely. It is also widely accepted that, if the PH fails to reduce a dislocated hip within 3–4 weeks of treatment, the PH should be discontinued. The PH is worn 24 hours per day. Total treatment duration varies between clinicians but is generally agreed to be around 12 weeks for most patients and should not exceed 20 weeks. Few clinicians treat with PH for less than 6 weeks.

Success rates following PH are excellent if treatment is commenced before 8 weeks of age, but success reduces after that.

The management of hips that are clinically stable but dysplastic or unstable on dynamic ultrasound is complex and controversial, and decisions to treat are probably best made on the basis of multiple data points with ongoing clinical and ultrasound monitoring.

CLOSED REDUCTION

If a hip cannot be reduced and held reduced with an abduction device then the next step in the treatment algorithm is an examination under general anaesthetic and a *hip arthrogram* with a view to a closed reduction. The use of *preoperative traction* to gently stretch soft tissue and neurovascular structures is controversial but there is good evidence from some countries of its efficacy.

Closed reduction is performed by longitudinal traction, flexion and abduction of the hip while lifting the greater trochanter anteriorly towards the acetabulum. Reduction of the hip is confirmed with an intraoperative dynamic arthrogram. Medial pooling of dye between the femoral head and medial acetabulum indicates non-concentric and inadequate reduction. If more than 7 mm of medial dye pooling is seen, there is a high likelihood that closed reduction may fail and open reduction will be necessary.

Following reduction, the hip is immobilized in a *hip spica cast* within the *'safe zone of Ramsey'*. This safe zone is the arc of motion through which the hip remains reduced without forced abduction. Adductor tenotomy is often performed at the time of closed reduction in order to increase the abduction range and therefore increase the safe zone.

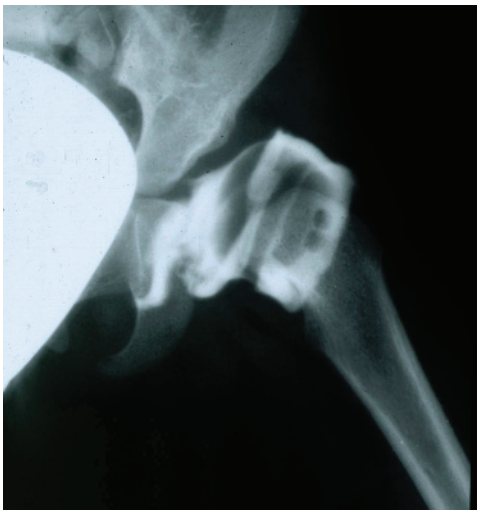


Figure 19.5 DDH Arthrogram with subluxed hip and hourglass constriction of joint caused by psoas tendon.

Postoperatively the position of the hip is confirmed by three-dimensional imaging, and the hip remains immobilized for approximately 12 weeks.

Closed reduction may fail for several reasons, most commonly due to an anatomical mechanical block to reduction or the inability of the femoral head to be held against a shallow acetabulum by a force applied some distance away. Extra-articular blocks to reduction include tight adductors and a tight iliopsoas tendon, often seen as an 'hourglass' constriction (Figure 19.5) on the arthrogram. These contractures limit the range of motion of the hip and reduce the stability of a reduced joint. The capsule of the joint may become contracted medially, particularly following a long period of dislocation, and it can also impede reduction with the femoral head lying in the 'baggy' stretched capsule supralaterally. The transverse acetabular ligament is placed under tension by the ligamentum teres in a dislocated position and may also hypertrophy and migrate superiorly, further limiting reduction of the femoral head into the acetabulum.

OPEN REDUCTION

The *timing of open reduction* is controversial. Some authorities feel that, if closed methods have failed, then the hip should be allowed to 'loosen' for a period of time before proceeding to open techniques, even if the ossific nucleus has not appeared, because the hip is under pressure and is more likely to have a better outcome the sooner it is reduced. In contrast, others feel that the presence of an ossific nucleus is protective and surgery should be deferred until it appears.

In the younger age group, often less than 1 year old, some surgeons prefer a medial approach to the hip, but others feel that this approach is associated with a poorer outcome in the longer term in terms of late AVN.

Open reduction allows direct visualization of the joint in order to assess and deal with blocks to reduction. Open reduction is usually performed through a transverse skin crease incision just inferior to the iliac crest. Following dissection in the gap between tensor fascia lata and sartorius and splitting of the iliac apophysis, the conjoined head of rectus femoris is divided and a psoas tenotomy performed at the pelvic brim. A T-shaped capsulotomy is performed such that the joint can be inspected. The ligamentum teres and transverse acetabular ligament are generally excised but the acetabular labrum, historically removed, is now recognized as being very important for acetabular development and is preserved if possible. An assessment can then be made of the best method to achieve and maintain concentric reduction of the femoral head. In younger children a capsulorraphy may suffice but with increasing age it is more likely that pelvic and/or femoral osteotomies may be required. Following reduction and stabilization of the joint the capsule is repaired.

Postoperatively the hip is immobilized in a spica cast for up to 12 weeks. It is imperative to maintain control of the operated limb during wound closure until the spica cast is applied to prevent redislocation and 3D imaging should be performed postoperatively to confirm reduction.

OSTEOTOMIES

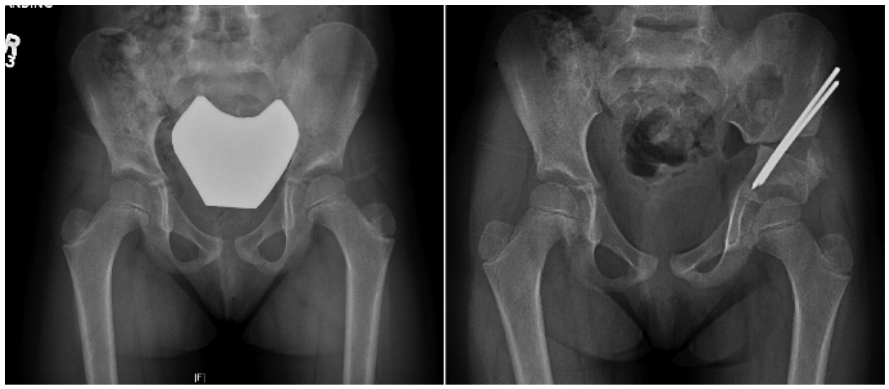
Osteotomies in DDH can be used at the time of open surgery to correct residual dysplasia on the acetabular side (rarely before the age of 18 months) and on the femoral side to correct femoral version or to reduce wound tension, or on both sides of the joint. They are also used commonly to treat residual or late-presenting dysplasia.

The Salter osteotomy (Figure 19.6)

In this redirectional osteotomy the pubic symphysis is used as a rotational hinge around which to rotate the position of the acetabulum. This procedure does not alter the overall shape or volume of the acetabulum but does increase the stability of the joint as well as the load-bearing portion of the acetabulum in a weight-bearing position.

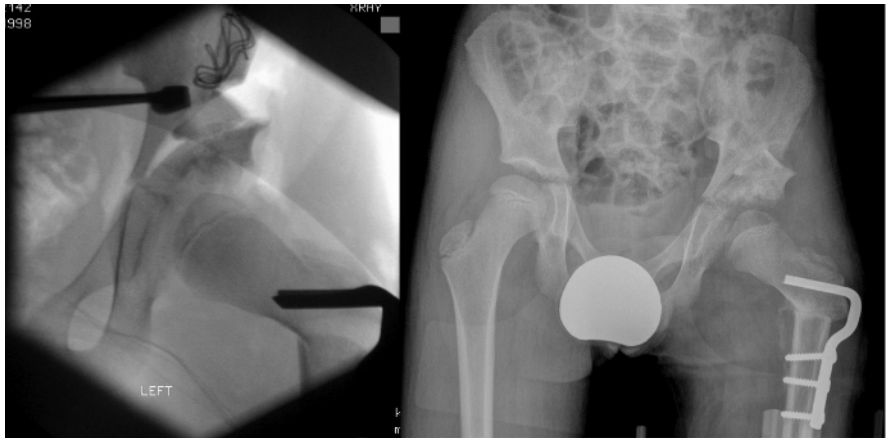
The Pemberton osteotomy (Figure 19.7)

This is a pericapsular pelvic osteotomy that achieves rotation of the acetabulum by exploiting the plasticity of the horizontal arms of the triradiate cartilage, which must remain open to perform this procedure. The Pemberton osteotomy is a volume-changing osteotomy commonly used in children with shallow acetabulum with anterolateral wall deficiency.



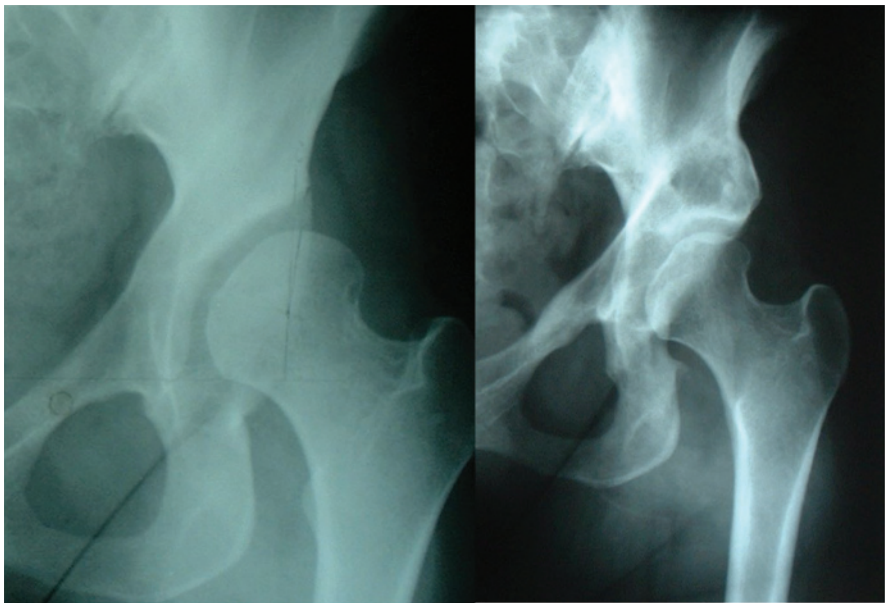
(a) (b)

Figure 19.6 Salter osteotomy This patient with bilateral DDH was treated with Pavlik therapy at birth and underwent Salter innominate osteotomy for persistent left-sided dysplasia at age 5. (a) Preoperative; (b) postoperative.



(a) (b)

Figure 19.7 Pemberton osteotomy (a) Intraoperative radiograph of Pemberton acetabular and femoral varising osteotomy. (b) 6 weeks postoperative.



(a) (b)

Figure 19.8 Periacetabular osteotomy (PAO)
(a) Preoperative; (b) 2 years postoperative PAO.

Proximal femoral varus derotation osteotomy In DDH the proximal femur is typically in a position of valgus and anteversion, contributing to instability of the hip joint. If the hip is stable in internal rotation and abduction but subluxes in flexion, then a proximal femoral osteotomy to increase the varus angle and reduce the excessive anteversion may increase stability of the reduction.

Femoral shortening osteotomy Older children who have a long-standing, high hip dislocation may require significant traction to achieve a reduction that is tight and under significant tension. This places increased pressure on the femoral head within the acetabulum and femoral shortening osteotomy may be required to reduce the risk of AVN.

Periacetabular osteotomy (PAO) Studies in adult populations undergoing total hip arthroplasty often reveal a history of dysplasia. In teenagers with closed growth plates and young adults who present with dysplastic hips it is reasonable to offer corrective surgery to relieve symptoms and reduce the rate of progression to degenerative change, such as the Ganz periacetabular osteotomy (Figure 19.8).

IRRITABLE HIP (TRANSIENT SYNOVITIS)

Aetiology

The irritable hip is a common childhood condition affecting up to 1 in 1000 children. Prospective and retrospective studies have identified multiple possible aetiologies of the condition. *Systemic infection*, viral or bacterial, is one of the most common sources, with recent upper respiratory tract infection present in up to 80% of patients. *Allergic reaction* is also a well described pathological process, with hypersensitivity of the synovial membrane in children with atopy. Finally, *trauma* can cause irritation of the hip capsule or bony contusion. The pathophysiology of transient synovitis includes non-pyogenic inflammation and hypertrophy of the synovial membrane. Incidence rates for the described aetiologies of this condition vary greatly throughout the literature.

Irritable hip usually occurs in children between 3 and 9 years of age, with a peak incidence around 6 years. Notably, this is the same time as the peak in Legg-Calvé-Perthes disease presentation. The condition is twice as common in boys as in girls and more common in Caucasians than other racial groups. The left and right hips are affected roughly equally, but bilateral disease is rare. Up to 10% of children will have a recurrent episode.

Presentation and examination

PRESENTATION

Children most commonly present with acute onset of a painful limp. Pain may be referred to the thigh, knee or buttock. Presentation commonly occurs after 2 days of symptoms. The natural history is a 5–10-day course of self-limiting symptoms although mild symptoms may persist beyond this.

PHYSICAL EXAMINATION

Physical examination often demonstrates refusal or limitations to weight-bearing on the affected limb with a marked limp. At rest the hip is held in flexion and external rotation to maximize capsular volume (and hence minimize intracapsular pressure), often

with associated muscle spasm to limit movement. Range of motion is commonly reduced, especially in internal rotation. Children remain systemically well but may have a low-grade fever, below 38 °C.

Investigation

Transient synovitis is a diagnosis of exclusion, after more sinister diagnoses have been ruled out. Following full history and physical examination, diagnostic investigation begins with laboratory blood work and plain radiographs. *Blood tests* should include full blood count (FBC), erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP). Initial *radiographs* should be AP and frog-lateral images of both hips. Based on results of these initial investigations, advance imaging such as *ultrasound* (with or without guided needle aspiration) or *MRI* may be required.

Septic hip arthritis is the most important diagnosis to rule out acutely. Diagnostic criteria were initially described by Kocher (Table 19.2). Studies assessing the initial Kocher criteria (fever, refusal to weight-bear, ESR and WBC) found that the presence of two criteria indicated a 40% risk of septic arthritis, increasing to 99.6% when all four criteria were present. If intraarticular sepsis is suspected, ultrasound can be confirmatory in demonstration of joint effusion. Ultrasound-guided needle aspiration of the hip joint is the gold-standard of diagnosis but may not be indicated in cases of high clinical suspicion when it may delay treatment. Treatment includes thorough irrigation and debridement of the joint followed by antibiotics until normalization of all inflammatory lab values and return of ability to weight-bear.

Osteomyelitis of the proximal femur may present with symptoms similar to the irritable hip. However, children with osteomyelitis are likely to have positive inflammatory markers, and symptoms will not resolve independently. MRI can be helpful in identifying bony changes and sub-periosteal collections to confirm the diagnosis.

Approximately 1.5% of children who present with transient synovitis will later be diagnosed with Legg-Calvé-Perthes disease.

Table 19.2 Modified Kocher criteria for septic hip arthritis

Criterion	Value
Fever	>38.5 °C
WBC	>12.0 × 10° cells/L
ESR	>40 mm/h
CRP	>2.0 mg/L
Weight-bearing	Complete inability

Treatment

Only after exclusion of important differentials can transient synovitis truly be diagnosed. Treatment is then largely focused on *symptom management* of this self-limiting condition. Bed rest is helpful in decreasing recovery time, but it is poorly tolerated by children. Non-steroidal anti-inflammatory medications are helpful for analgesia but do not alter the disease course. Weight-bearing aids, such as crutches, may be necessary until return of full range of hip motion. If symptoms do not completely resolved in 2–3 weeks, further investigation is warranted.

Complications

Transient synovitis has been associated with several sequelae. Most significantly, temporary local over-stimulation of growth may lead to coxa magna and over-lengthening of the affected limb. The long-term consequences and implications of this are poorly understood but seem to be relatively benign.

LEGG-CALVÉ-PERTHES DISEASE

Background

Legg-Calvé-Perthes disease (LCPD) describes idiopathic avascular necrosis of the proximal femoral epiphysis that affects approximately 1 in 1000 children. The condition most commonly presents between 4 and 8 years of age with reduced incidence near the equator and increased incidence in northern regions, particularly within Northern Europe. Caucasians and Chinese are significantly more affected than those of African descent, and boys are five times more commonly affected than girls. Bilaterally affected hips are seen in approximately 10% of cases and this is more common in girls. In bilateral disease hips do not follow the same time course and are often in different stages of the disease process.

LCPD has been associated with a number of other congenital conditions including genitourinary malformations, undescended testes, inguinal hernia, Down's syndrome and some coagulopathies. Risk factors include low birth weight, exposure to secondhand cigarette smoke, short body length at birth, family history, low socioeconomic status and attention deficit hyperactivity disorder type 1 (ADHD-1).

Pathophysiology

The pathophysiology of LCPD is not fully understood but it may include a vascular insult and uncoupling of the bone metabolic process. Initial bony destruction is exacerbated by prolonged, repetitive loading of the hip joint. Thrombophilic conditions such as thrombophilia and Protein C and S deficiencies have been

linked to LCPD. Additionally, bone age in children with LCPD often lags behind chronological age, resulting in a relatively small ossific nucleus within the femoral head with a much larger cartilage component. Vessels that traverse the cartilage cap are more susceptible to mechanical compression and are therefore vulnerable over a greater length in these children. Furthermore, studies have shown that in up to 68% of patients with LCPD there is an interruption at the origin of the lateral epiphyseal artery, a branch of the medial circumflex femoral artery via the posterior circumflex artery. Venous stasis may also contribute to the vascular insult. Abduction and internal rotation stretch the posterior circumflex artery and may completely interrupt flow to the lateral epiphyseal artery as it traverses the capsule.

Following initial insult, normal bone remodelling does not occur in LCPD. Rather, increased osteoclast activity driving bony resorption is followed by persistent fibroblastic proliferation and fibrovascular replacement of trabeculae instead of new bone formation. Prolonged repetitive loading of the hip joint leads to compression of this weak epiphyseal bone.

Presentation and examination

PRESENTATION

The presentation of LCPD is variable, ranging from a painless limp to acute transient synovitis. If children do have pain it is often referred to the knee, as with slipped capital femoral epiphysis (SCFE). Pain tends to be worsened by activity and relieved by rest. Children aged 4–9 years with asymptomatic limp or symptomatic synovitis lasting longer than 10 days should raise suspicion of LCPD and warrant investigation. This should include AP and frog-lateral radiographs of both hips and examination of blood inflammatory markers including FBC, ESR and CRP.

PHYSICAL EXAMINATION

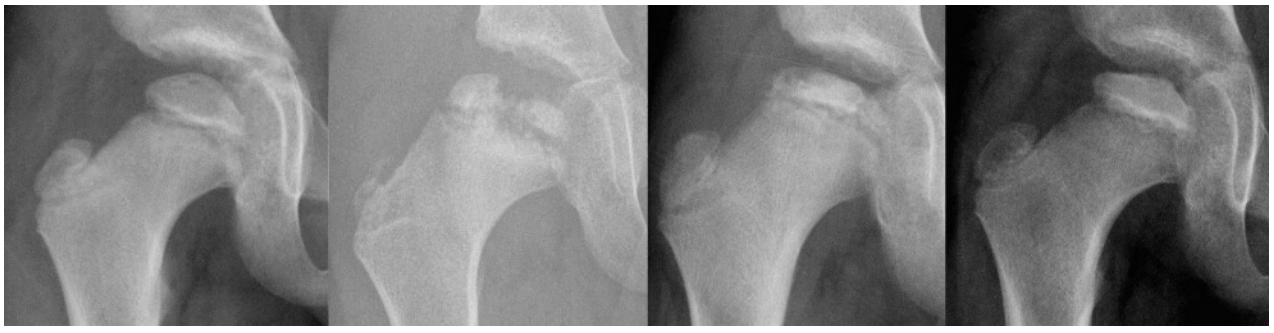
Physical examination often demonstrates hip pain with passive range of motion including log-roll of the leg. Range of motion may be reduced, especially in abduction and internal rotation, and hip flexion contracture may be seen in cases of long-standing disease. Prolonged disease leads to loss of epiphyseal height and proximal femoral deformity resulting in weakening of musculature around the hip. It can present with Trendelenburg gait and leg-length discrepancy.

Imaging

Radiographic changes can be seen after 3–6 months of disease process, beginning with medial joint space widening, cartilage thickening and joint effusion. The ossific nucleus is typically small and hyperdense. Lateral subluxation of the femoral head leads to further increases in inferomedial joint space. A fracture line is often present within the femoral head, identifying the zone of resorption with an osteoporotic segment of the lateral epiphysis. With continued collapse and near-total head involvement the physis becomes increasingly horizontal. Severe disease may progress to changes in the acetabulum, early closure of the triradiate cartilage, and bi-compartmentalization of the acetabulum and ischium varum.

WALDENSTROM STAGING (Figure 19.9)

- *Initial stage* – describes the first 3–6 months of the disease, which may be clinically and radiographically silent. If radiographic changes are present, they include medial joint space widening and a small, sclerotic epiphysis with increased density in the ossific nucleus (Figure 19.9a).
- *Fragmentation* – this stage is present from approximately 6–12 months and is often associated with clinical symptoms. Necrotic bone is irregularly resorbed and replaced with vascular fibrous tissue as revascularization begins. Radiographically the epiphysis demonstrates fragmentation with alternating areas of sclerosis and fibrosis and it may begin to collapse in height (Figure 19.9b).



(a) (b) (c) (d)

Figure 19.9 Waldenstrom staging of LCPD (a) Initial stage; (b) fragmentation; (c) reossification; (d) remodelling.

- *Reossification* – begins at around 12 months and lasts for up to 18 months. During this time reossification of the nucleus begins peripherally and progresses centrally as necrotic bone is fully removed. Gradually the epiphysis regains normal strength and density (Figure 19.9c).
- *Remodelling* – begins once the ossific nucleus is completely reossified and continues until skeletal maturity. Trabecular pattern is reformed throughout epiphysis although the femoral head may remain flattened and ovoid (Figure 19.9d).

CATERALL CLASSIFICATION

This grading system is applied during the *fragmentation stage* of disease as described by Waldenstrom and is based on the amount of epiphyseal involvement.

- *Grade 1* – represents less than 25% involvement, usually involving the anterior portion of the epiphysis with no metaphyseal reaction.
- *Grade 2* – involves 25–50% and progresses towards lateral epiphysis. A sequestrum is often present along with anterior and lateral metaphyseal reaction and a subchondral fracture line in the anterior half of the epiphysis
- *Grade 3* – involves 50–75% with sequestrum and posterior subchondral fracture line.
- *Grade 4* – involves 100% of the epiphysis with diffuse metaphyseal involvement.

This grading system has demonstrated poor inter-observer reliability; however, a grade of 3 or 4 has been shown highly predictive of poor outcomes.

Caterall also described several 'head-at-risk' signs on radiography that correlated with a more severe disease course and poorer outcomes. These include lateral subluxation of the femoral head, speckled calcification in the lateral epiphysis, a horizontal physis, metaphyseal cyst formation and *Gage's sign*. Gage's sign, described by Courtney Gage in 1933, is a 'V'-shaped osteoporotic segment in the lateral epiphysis and adjacent metaphysis (Figure 19.10).

SALTER AND THOMPSON CLASSIFICATION

The classification developed by Salter and Thompson in 1984 is focused on the extent of the superolateral dome of the femoral head that is affected by subchondral fracture (Figure 19.11). This system can be applied once a fracture line is present, often *up to 8 months before full fragmentation* when the Caterall classification can be applied.

- *Type A disease* – involves less than 50% of the femoral head and has a good prognosis.
- *Type B disease* – involves over 50%, often with associated lateral pillar collapse and suggests poor outcomes.

The main drawback of this classification is that up to two-thirds of patients do not demonstrate a clear subchondral fracture line.

HERRING CLASSIFICATION

This is the *most commonly used classification system* and it can be applied *early in the fragmentation stage*, usually within the first 6 months of clinical symptoms. The Herring classification is based on involvement of the lateral pillar of the epiphysis, leading to loss of pillar height on AP radiograph.

- *Group A* – the hips have no lateral pillar involvement.
- *Group B* – over 50% of lateral pillar height is maintained.

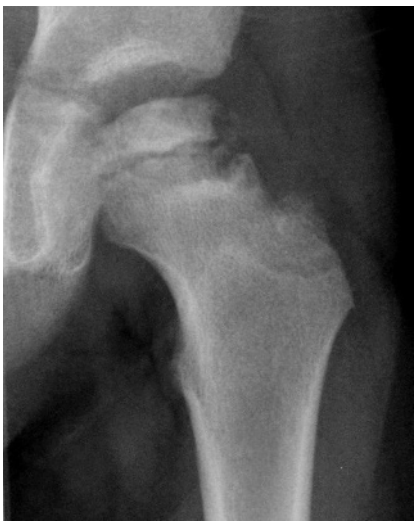


Figure 19.10 Gage's sign Note the 'V'-shaped osteoporotic segment.

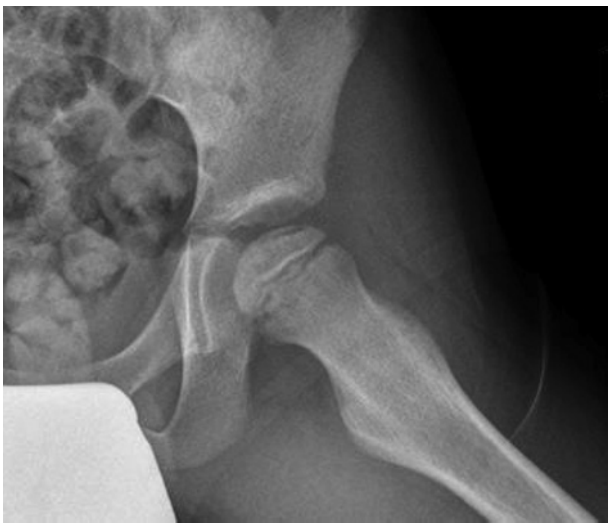


Figure 19.11 LCPD Frog-lateral radiograph demonstrating a subchondral fracture in the superolateral dome of the femoral head.

- *Group B/C* – 50% loss of height
  - *B/C-1*: the lateral pillar is less than 2–3 mm wide
  - *B/C-2*: there is minimal ossification
  - *B/C-3*: the lateral pillar is depressed in relation to the central pillar of the femoral head.
- *Group C* – there is collapse and loss of over 50% of lateral pillar height.

This classification has demonstrated strong prognostic value and inter-observer reliability. Group A is associated with universally good outcomes; Group B hips generally have poor outcomes in children older than 6 years and Group C hips have universally poor outcomes.

STULBERG GRADING

Stulberg grading is based on congruency and sphericity of the femoral head at skeletal maturity.

- *Grade I* – hips have a near-normal, round femoral head.
- *Grade II* – hips also have a spherical head but additionally demonstrate coxa-magna, coxa-breva or an abnormally steep acetabulum. Grade II is associated with approximately a 15% increase in risk of osteoarthritis.
- *Grade III* – hips have an ovoid femoral head and demonstrate a difference of greater that 2 mm between AP and frog-lateral radiographs. The aspherical congruity is associated with a 60% risk of osteoarthritis (OA) in adulthood.
- *Grade IV* – hips demonstrate a flat head with a shallow, steep acetabulum and are associated with a 75% risk of adult OA.
- *Grade V* – hips have a femoral head that is flat and aspherically incongruous with the relatively normal acetabulum leading to an 80% risk of OA before the age of 50 years.

Treatment

As described above, the LCPD disease process is defined by destruction followed by regeneration. The primary aim of treatment is to prevent deformity to the femoral head before the remodelling phase and many experts feel that 'containment' of the femoral head is important. Containment alters joint mechanics to distribute forces more evenly across the epiphysis thereby protecting the weak and fragmented femoral head until reossification can occur.

NON-OPERATIVE TREATMENT

Non-operative treatment is generally reserved for younger children, specifically those under 6 years of age with Herring A or B hips. These children typically have a good outcome regardless of treatment modality. Therapies include protected weight-bearing and activity restriction to reduce mechanical loading of the hip until reossification. Reduction in weight-bearing has proven significantly beneficial, but additional human studies are required. Physiotherapy is also beneficial to restore and maintain range of motion in the hip joint. Abduction bracing, using the Newington brace or Petri casting, was used historically to stretch adductors and increase hip range of motion to a position of better containment. Patients should be monitored regularly with radiographs over the 24–36-month natural history course of the disease. Ultimate shape of the femoral head after reossification will dictate long-term outcomes.

Bisphosphonates have increasingly been studied as a way to stop destruction by delaying resorption of necrotic bone and preventing collapse of the femoral head. Bone morphogenetic proteins (BMPs) are also being investigated as a possible treatment, by way of promoting osteoclastic bone resorption and thereby stimulating the healing process. Routine use of both these therapies remains controversial.

OPERATIVE TREATMENT

Operative intervention may be indicated in children with persistent losses in range of motion or unresolving clinical symptoms. This usually includes children over the age of 6 years with Herring B hips and all children with Herring B/C or C hips. Surgery is most beneficial if performed early in the fragmentation stage. Prerequisites for surgical containment include near-normal abduction under general anaesthetic and arthrogram demonstrating a containable congruent hip joint.

Under the age of 8 years the most common procedure is *proximal femoral varus osteotomy*. This corrects lateral subluxation of the femoral head and reduces point pressure of the articular surface by increasing head coverage and correcting excessive anteversion caused by metaphyseal involvement. Correction is performed through a medial wedge closing osteotomy and held with a fixed angle device.

Over the age of 8 years, or in children with more advanced disease, *pelvic osteotomy* is often required for adequate containment. Techniques include acetabular shelf osteotomy, Dega- and Salter-innominate osteotomies.

Failure of containment and significant head collapse can result in hinged abduction (Figure 19.12) through a short femoral neck and high greater trochanter leading to loss of abduction and gluteal insufficiency syndrome. In this case the hip joint is often congruent in extreme adduction. Hip joint salvage may be achieved through a *valgus subtrochanteric osteotomy* to redirect the head towards the acetabulum and improve joint congruity, the abductor lever arm and limb-length discrepancy.

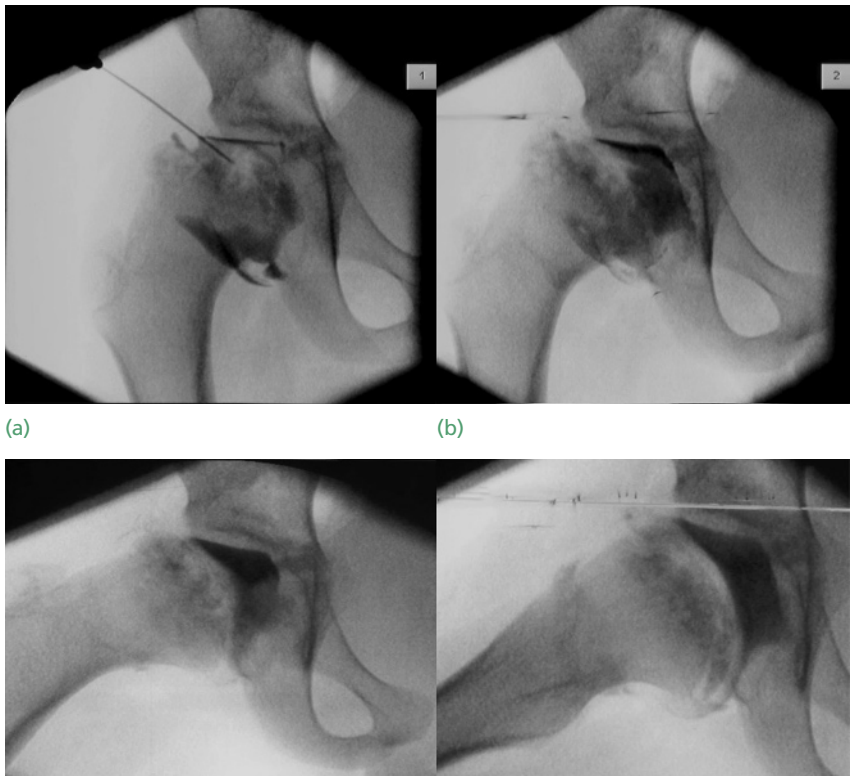


Figure 19.12 LCPD (a) Arthrogram image with dye injected into the joint showing some incongruity of the hip joint. (b) Increasing incongruity as abduction starts. (c) Radiograph showing 'hinge' on superolateral aspect of acetabulum as abduction increases. (d) 'Hinging' seen on frog-lateral radiograph.

SLIPPED CAPITAL FEMORAL EPIPHYSIS

The slipped capital femoral epiphysis (SCFE) (also known as slipped upper femoral epiphysis, SUFE) is the most common hip condition among adolescents from age 10 to 15 with an incidence of 1 per 10 000 worldwide.

Pathophysiology

Mechanical overloading leads to displacement through the proximal femoral physis with translation of the metaphysis anteriorly and superiorly in relation of the epiphysis. Slipping of the epiphysis typically occurs through the hypertrophic zone of the physis. Anatomically the hypertrophic zone often contains an anastomosis of the metaphyseal epiphyseal blood supplies. During adolescence, prior to growth-plate closure the extracapsular arterial ring supplying the metaphysis increases substantially and invests the subphyseal region, terminating at the hypertrophic zone. The epiphyseal side of the physis is supplied by the artery of the ligamentum teres, a branch of the obturator artery.

The physis is reinforced externally by a thick periosteal ring. During adolescence the periosteum begins to thin and the force required for displacement to occur is reduced. Internally, the physis is stabilized primarily by mammillary processes. Hips that develop SCFE often display widening of the physis, particularly through the hypertrophic zone, subjecting these mammillary processes to the risk of unlocking and leading to potential displacement.

Histologically the physis of hips that undergo SCFE are likely to demonstrate physeal widening up to 12 mm (normal width is 2–6 mm), larger chondrocytes, altered physeal column height and organization, and disruption of chondrocyte differentiation and ossification.

Biomechanically several factors predispose to SCFE. Increased retroversion of the proximal femur increases torsional stress and therefore rotational instability through the physis. Changes in the shape of the proximal femur during growth also contribute to reduced stability of the physis. During the growth spurt, significant lengthening of the femoral neck leads to increase varus as the neck shaft angle reduces from 160 degrees in infants towards 125 degrees in the adult skeleton. This leads to a more vertically oriented physis, with increased shear forces across the physis. Larger body habitus and additional weight further increase these shear forces.

Several endocrine disorders have been highly associated with the development of SCFE, particularly *hypothyroidism*, *osteodystrophy of chronic renal failure*, and *excessive growth hormone*. These children usually have an atypical SCFE with presentation under the age of 10 or over the age of 16. In addition bone age is

often found to lag behind chronological age in these children. Children who present with SCFE under the age of 10 or whose weight is below the 50th percentile for their age should have an endocrine workup beginning with thyroid-stimulating hormone (TSH), blood urea nitrogen (BUN) and creatinine.

Presentation and examination

PRESENTATION

Children with SCFE typically present in early adolescence with pain in the thigh, groin or knee. Some patients will present with isolated knee pain due to the reflex sensory arc of the leg. This can lead to detrimental delays in diagnosis and treatment. Therefore, in adolescents presenting with knee pain of unclear aetiology, plain radiographs of the hip should always be performed. Patients often present following an acute traumatic event, but may report prodromal symptoms for several weeks or months preceding the event.

The average age of diagnosis is 12 years. Children under 10 or over 16 years are more likely to have atypical SCFE with underlying endocrinopathy. Race plays a significant role in the risk of the condition, with highest incidences seen in the black, Hispanic and Pacific Islands populations. Approximately 20% of patients will have some bilateral symptoms at the time of presentation and this is significantly increased in the case of endocrine disorders. Between 15% and 35% of patients with initially unilateral symptoms will develop a slip of the contralateral side within 18 months of presentation. This may be termed a metachronous slip.

PHYSICAL EXAMINATION

Physical examination will often demonstrate a leg that is shortened and held in external rotation (Figure 19.13). There may be significant pain with hip range of motion including log-roll of the leg. Passive range of motion demonstrates limitations to abduction, flexion and internal rotation compared to the unaffected side. Additionally, hip flexion often leads to obligate external rotation and abduction; this indicates a positive *Drehmann's sign*. If they are able to weight-bear, patients often have an *antalgic* or *Trendelenburg gait* with notable external rotation of the foot on the affected side.

Imaging

Plain radiographs remain the pillar diagnostic imaging in children with symptoms of SCFE. Initial radiographs should always include AP and frog-lateral images of both hips. Lateral radiographs are more sensitive for early signs of slip. Early signs include widening or irregularity of the physis, loss of the anterior head-neck concavity, sharpening of the metaphyseal border of the head and loss of total epiphyseal height.

- *Klein's line* – a line drawn on the AP radiograph along the superior border of the neck (Figure 19.14). In a normal hip the line should intersect the epiphysis; failure to do so results in a positive *Trethowan sign* and may indicate slip. A difference of greater than 2 mm in the maximal width of epiphysis lateral to Klein's line between the affected and unaffected hips should increase suspicion of SCFE. Klein's line has relatively low sensitivity and poorly identifies pathological hips in the pre-slip stage. This line can also be used on the frog-lateral view.
- *Steel sign* – a double-density or metaphyseal blanch sign seen on the AP radiograph caused by overlapping of the posteriorly displaced epiphysis over the metaphysis.
- *Capener's sign* – reduction in the double-density crescent created by overlap of the posterior acetabulum and proximal metaphysis as the metaphysis is displaced laterally by worsening slip on the AP radiograph.

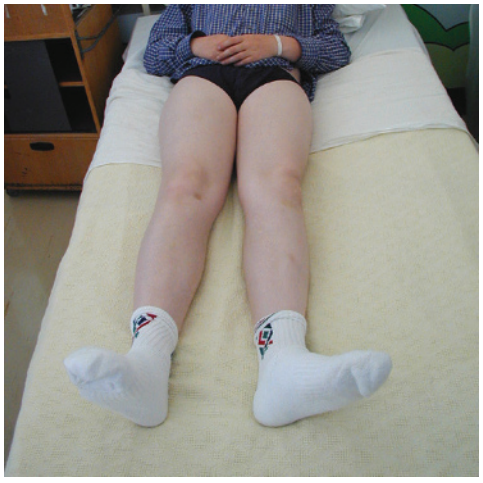


Figure 19.13 Slipped capital femoral epiphysis (SCFE) Child presenting with stable left-sided SCFE.

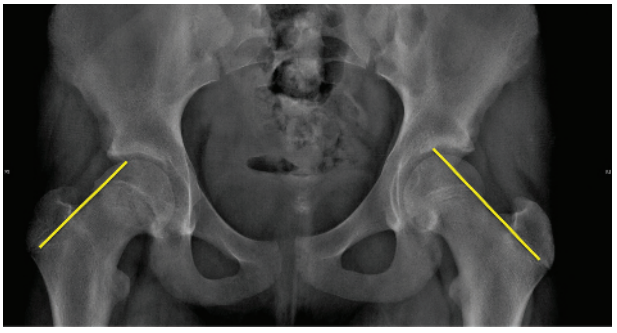


Figure 19.14 SCFE – Klein's line Line drawn bilaterally with positive Trethowan sign in the left hip.

Historically, Steel and Capener's signs have been identified as evidence of SCFE on plain radiographs. In modern practice, however, three-dimensional imaging has become increasingly available to aid in diagnosis. Specifically, *MRI* is useful in cases of high clinical suspicion with negative radiographic findings. MRI is more sensitive than plain radiographs and may identify hips in the pre-slip stage with physeal oedema and very small degrees of slip.

- *Southwick angle* – measured on the frog-lateral view, the angle created by a line drawn perpendicular to the physis and a line parallel to the femoral shaft (Figure 19.15). This angle can then be compared to the contralateral side. In the case of bilateral slip, 12 degrees can be used as a reference for normal. As slip worsens, the Southwick angle will increase.

Classification

TEMPORAL

Historically SCFEs were classified based on presentation as *acute* (symptoms for less than 3 weeks), *chronic* (symptoms for more than 3 weeks) or *acute-on-chronic* (an abrupt increase in symptoms with a preceding prodrome of non-specific pain). In recent decades this classification system has been found to have low prognostic value and therefore is less clinically relevant.

CLINICAL

Loder and colleagues in 1996 described SCFEs based on clinical symptoms as *stable* or *unstable*. A hip is considered to have a stable slip if the child is able to bear weight with or without the use of aids such as crutches. It is considered unstable if the child in entirely unable to bear weight. Unstable slips will present clinically in a similar fashion to hip fractures.

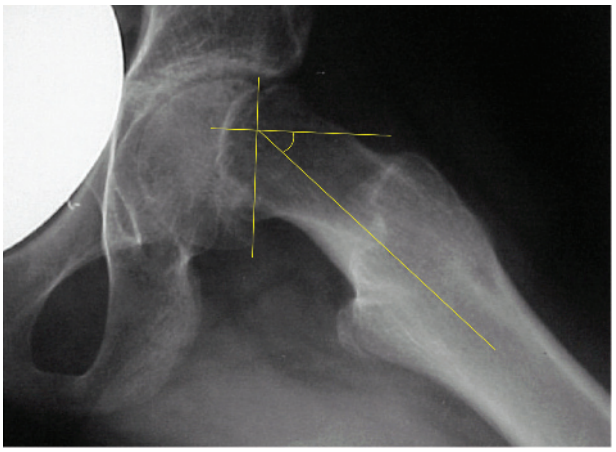


Figure 19.15 SCFE – Southwick angle Angle created by a line perpendicular to the physis and a line parallel to the femoral shaft.

This classification system has been found to be highly prognostic. Unstable slips are at significantly higher risk of avascular necrosis of the femoral head with a reported rate around 47% compared to 10% in stable slips. This classification system has proven to be the most indicative of prognosis.

RADIOGRAPHIC – SOUTHWICK

Radiographic severity can be determined based on the difference in Southwick angle between the pathological and normal hip. A difference of less than 30 degrees is considered a *mild* slip, 30–50 degrees is considered *moderate*, and a difference of greater than 50 degrees in the Southwick angle between both hips is considered *severe*.

RADIOGRAPHIC – LATERAL SLIP GRADE

Slip severity can also be graded based on the percentage epiphyseal displacement in relation to the metaphysis. *Grade I* describes less than 33% slippage, *grade II* describes 33–50% and *grade III* represents a slip of greater than 50% off the metaphysis. This grading can be performed on either an AP or a frog-lateral image.

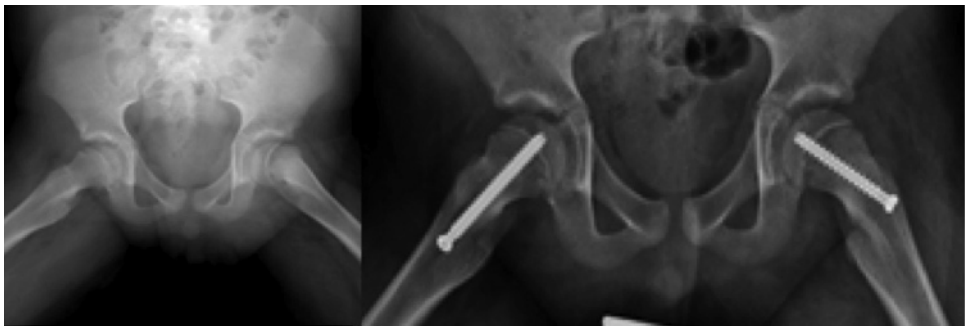
Treatment

IN-SITU PINNING

Treatment of SCFE is focused on stabilization of the epiphysis with induction of physeal fusion and growth arrest. Closed reduction manoeuvres are now considered largely historical and the target of treatment is rather *in-situ fixation of the proximal femoral physis* (Figure 19.16). Aggressive or forced closed reduction manoeuvres are proven to increase the risk of AVN and have therefore been abandoned. However, spontaneous or serendipitous reduction has been described while positioning the child on the surgical bed. This may occur in up to 90% of unstable slips but as few as 8% of stable slips.

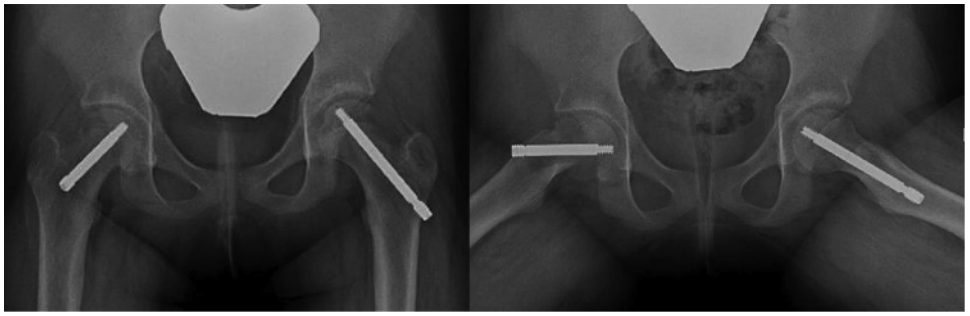
Parsch has advocated gentle open reduction of the acute (unstable) component of acute-on-chronic slips. This is accomplished through a Watson-Jones approach and anterior capsulotomy followed by gentle posterior pressure on the epiphysis by the surgeon's finger. Goals of in-situ fixation include prevention of further displacement and induction of physeal arrest.

The predominant surgical technique uses cannulated screws placed percutaneously into the centre of the epiphysis and traversing perpendicular to the physis. For optimal stability a minimum of five screw threads should be contained within the physis in order to provide adequate stability and prevent slip progression. It is critical that the joint is not penetrated due to the risk of damage to the articular cartilage of the hip. The use of one versus two screws remains controversial.



(a) (b)

Figure 19.16 SCFE – in-situ pinning (a) Preoperative frog-lateral X-ray of child with left-sided SCFE. (b) Frog-lateral X-ray following in-situ pinning of left hip and prophylactic pinning for right hip in the same child.



(a) (b)

Figure 19.17 SCFE – in-situ pinning in young children (a) AP and (b) frog-lateral X-ray of sliding screws used in younger children when significant growth is expected from proximal femoral physis.

Prophylactic fixation of an asymptomatic hip has been advocated in children who have a high risk of contralateral slip and is becoming more widely accepted. These children include those who are young at the time of primary diagnosis (<10 years), who have endocrine disorders or who are obese with delay in presentation. In young children, when there is still a significant contribution to growth expected from the proximal femoral physis, the goal of treatment is to hold the slip without arresting growth. One way to address the concern of growth arrest has been the use of sliding screw devices (Figure 19.17), which achieve fixation in both the epiphysis and diaphysis. This allows growth along the length of the screw, while maintaining the position of the slipped physis.

OPEN REDUCTION

Patients with highly displaced, unstable SCFE are at increased risk of developing femoroacetabular impingement (FAI) as well as AVN. In these cases FAI can result from both cam and pincer impingement. Cam impingement is caused by posterior displacement of the epiphysis leading to exposure of the anterior metaphysis, while the deep acetabulum simultaneously risks pincer impingement.

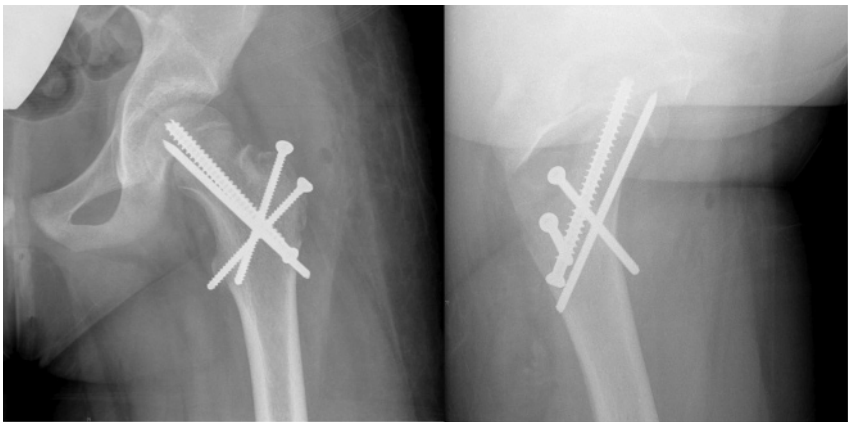
The surgical hip dislocation was first described by Ganz and colleagues in 2001. This technique uses an anterior incision with dissection proximal to piriformis, in the interval between piriformis and gluteus minimus, in order to avoid the blood supply to the femoral head. Once access is gained to the hip joint, a modified Dunn procedure can be used to reduce the epiphysis. This technique allows controlled reduction of the epiphysis while providing an opportunity to assess and confirm blood flow to the femoral head, thereby potentially reducing the risks of both FAI and AVN (Figures 19.18 and 19.19).

PROXIMAL FEMORAL OSTEOTOMY

Traditionally, proximal femoral osteotomy has been performed in cases of severe slip (i.e. those that cannot be pinned *in situ*) without surgical dislocation. The closer to the joint and therefore to the site of deformity that the osteotomy is performed the less correction is required, but at increased risk of AVN. The sites of osteotomy are *subcapital* (*Dunn*) and *basicervical* (*Kramer*), both usually performed through a Watson-Jones approach, or a *Southwick intertrochanteric* osteotomy.

Dunn osteotomy

In some cases, where the slip is severe, in-situ pinning may be technically unfeasible or inadequate for normal or near-normal joint function. In this case a proximal femoral (Dunn) osteotomy may be performed anteriorly or via surgical dislocation of the hip. The osteotomy is performed at the proximal metaphysis to allow reduction of the epiphysis back onto the metaphysis. This osteotomy is performed at the site of maximal deformity and therefore allows for the greatest reduction. This is also called a *capital realignment procedure*. The femoral neck may be shortened slightly to reduce tension of the posterior vessels and thereby reduce



(a) (b)

Figure 19.18 Open reduction (a) AP and (b) lateral X-rays following open reduction and in-situ pinning.

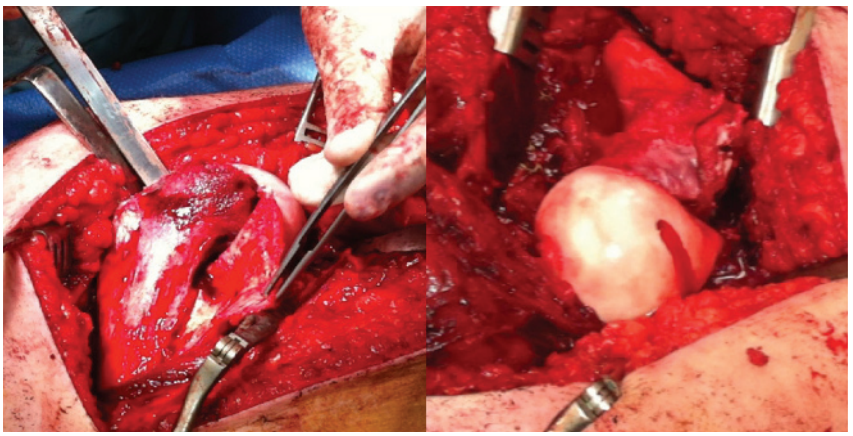
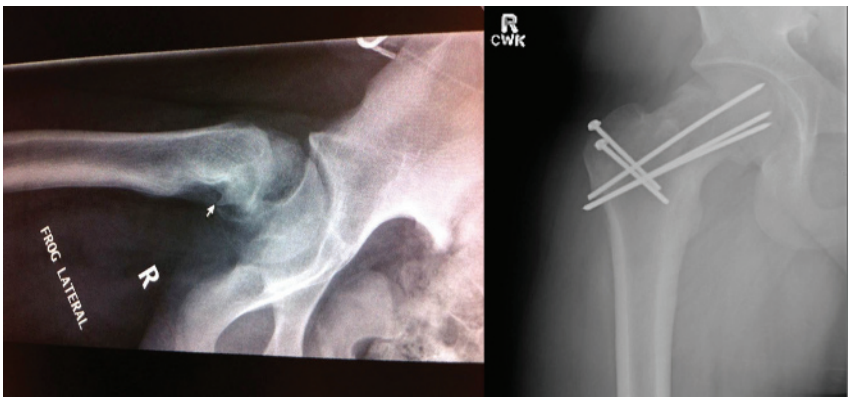


Figure 19.19 Ganz technique Intraoperative images of Ganz surgical hip dislocation allowing direct observation of preservation of the blood supply to the femoral head post reduction and dislocation.



(a) (b)

Figure 19.20 SCFE (a) Lateral X-ray of severe preoperative slip. (b) AP X-ray following Dunn osteotomy and in-situ fixation.

risk of AVN. Reduction is held with smooth pins. The most common complication of this procedure is AVN of the femoral head (Figure 19.20).

Kramer osteotomy In severe chronic slips, deformity of the proximal femur may significantly limit physical function. The Kramer osteotomy is an anterosuperior-based wedge osteotomy at the base of the femoral neck used to correct deformity. Osteotomy at this location has a lower risk of AVN and is beneficial in children with a Trendelenburg gait preoperatively. However, the degree of reduction that can be achieved is limited to 35–55 degrees and there is a higher risk of leg length discrepancy than with the Dunn osteotomy.

Southwick osteotomy If significant deformity remains within the proximal femur following remodelling of SCFE, an intertrochanteric wedge osteotomy can be performed at the level of the lesser trochanter. The goals of this Southwick osteotomy are flexion, valgus and internal rotation in order to allow increased range of motion in the hip and reduce impingement. The osteotomy is extracapsular and therefore carries a low risk of AVN.

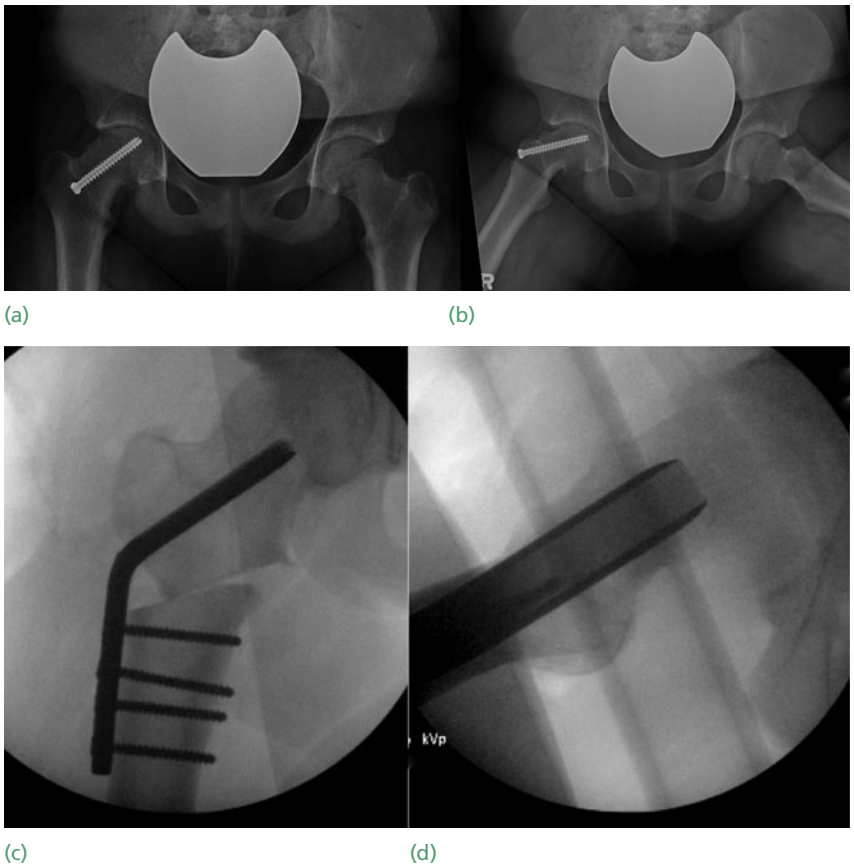


Figure 19.21 SCFE (a) AP and (b) frog-lateral X-rays following healed SCFE with residual deformity. (c) AP and (d) lateral intraoperative images showing removal of screw fixation and subtrochanteric osteotomy to reduce impingement.

Late osteotomy Long-standing SCFE with delayed presentation leads to proximal femoral deformity with extension, external rotation and shortening. Corrective late osteotomy can be performed to address this deformity. Subtrochanteric osteotomy produces flexion, internal rotation and valgus to restore normal anatomy of the proximal femur (Figure 19.21).

Complications

The most significant risk associated with SCFE is avascular necrosis of the femoral head. Surgical complications include chondrolysis and early osteoarthritis secondary to screw penetration, although the risk of these complications is decreasing with increasing use of intraoperative multisequence imaging.

THE ADULT HIP

CLINICAL ASSESSMENT

Symptoms

Hip girdle pain

This can be considered as arising from inside the hip joint itself (intra-articular), arising from structures related to or close-by the hip but outside the joint (extra-articular), or referred to the hip girdle by a pathological process remote to the hip joint.

1 *As a result of intra-articular hip pathology (e.g. osteoarthritis of the hip)*
Pain arising from the hip joint itself is generally felt in the groin. It can radiate down the front of the thigh and can sometimes be felt in the knee. Pain can also radiate around the hip girdle and patients can characteristically localize symptoms using the 'C-sign' (cupping the hand around the anterior, lateral and posterior margins of the hip) when asked to point at the area where pain is experienced.
2 *As a result of extra-articular hip pathology (e.g. abductor tendonitis)*
Pain can also be felt around the hip girdle as a result of conditions local to but outside the hip joint itself, for example trochanteric bursitis +/– abductor tendonitis. Pain arising as a consequence of extra-articular pathology is generally felt on the lateral side of the upper thigh. In the example of abductor tendonitis, the patient may well report localized tenderness and inability to lie on the affected side.
3 *As a result of pathology remote to the hip joint (e.g. lumbar spine facet joint osteoarthritis)*
It is not uncommon for pain to radiate to the hip girdle as a consequence of pathological

BOX 19.1 DIFFERENTIAL DIAGNOSIS OF PAIN AROUND THE HIP GIRDLE OUTSIDE THE HIP JOINT

Within the musculoskeletal system

- Trochanteric bursitis
- Gluteus medius tendinitis
- Stress fracture
- Osteitis pubis
- Iliopsoas tendinitis or bursitis
- Iliopsoas abscess
- Adductor longus strain or tendinitis
- Referred from the spine
- Metastatic disease

Outside the musculosketetal system

- Inguinal hernia
- Inguinal lymphadenopathy
- Gastrointestinal
- Genitourinary
- Gynaecological

processes quite separate from the hip joint itself. These may be musculoskeletal in origin, most commonly from the lumbar-sacral spine. They may also be outside the musculoskeletal system (Box 9.1). Pain radiating to the hip girdle from the lumbar spine generally affects the buttock area and it may or may not be associated with low back pain. One should consider whether the pain radiates more posteriorly in the thigh and is associated with motor weakness and/or sensory changes that could suggest associated lumbar spine nerve root entrapment.

The assessment of pain and its related symptoms forms an important part of a structured medical history. In particular, pain in the hip along with the spine can arise from very diverse pathology and it is important to have a systematic method to consider the pain history itself. A commonly used mnemonic acronym to aid this structured approach is SOCRATES (Box 19.2).

It is important to remember that, when considering symptoms that affect the musculoskeletal system, both the lower back and hip girdle are common sites where patients often present with pain. The specific detail of that pain is important to consider but one should also remember that patients can present with more than one problem, and dual pathology, especially osteoarthritis in more than one site, is not uncommon. The other features of the patient's presenting complaint can help this differentiation and guide subsequent investigations.

BOX 19.2 SOCRATES: A STRUCTURED APPROACH TO PAIN ASSESSMENT OF THE HIP

Site – Where is the pain or the maximal site of the pain?
Onset – When did the pain start, and was it sudden or gradual?
Character – What is the pain like? An ache? Stabbing? Burning?
Radiation – Does the pain radiate anywhere?
Associations – Are any other symptoms associated with the pain?
Time course – Does the pain follow any pattern?
Exacerbating/relieving factors – Does anything change the pain?
Severity – How bad is the pain?

Stiffness

Loss of movement is most commonly associated with intra-articular pathology. Early loss of movement can be well compensated for by pelvic mobility and movement at the lumbar spine. The earliest and most reliable marker of intra-articular pathology is the loss of internal rotation (see 'Clinical examination' below).

As loss of movement progresses, patients with stiffness of the hip commonly report difficulties putting on their shoes and socks. They often have very significant difficulty or indeed find it impossible to cut their toenails. Specific questioning on these issues can sometimes be helpful in differentiating the source of the hip girdle pain. Marked hip joint stiffness can contribute to a patient limping. The difficulties that patients report with the activities of daily living may be secondary to pain but also commonly these are a result of joint stiffness (e.g. when climbing stairs, moving from sit to stand, and washing or drying themselves all over. Stiffness is most marked after inactivity.

Limp (gait disturbance)

The hip joint plays a fundamental role in locomotion, and pathology affecting the hip joint can commonly result in the patient presenting with a significant limp. This limp may simply be a way of coping with pain, or it may be due to a change in limb length, weakness of the hip abductors or joint instability. Most commonly the gait has an antalgic pattern. An antalgic gait is characterized by pain leading to an uneven cadence whereby the patient spends less time with the painful leg touching the ground in the gait cycle. As well as the limp itself, patients may well report a reduction in their walking ability, in terms of both distance and stride length.

Snapping or clicking

Patients may report an audible sound related to their hip. This may be a cluck, click or snap. Rather than actually hearing the sound, they may also report the 'feeling' of a clunk, click or snap that, while it cannot be heard by others, the patient can feel transmitted from their hip joint. These sounds can be associated with pain or can be pain-free. Again, these symptoms can be considered as the result of intra-articular (e.g. labral tear or loose body) or extra-articular pathology (e.g. snapping tendon).

It is important to consider the anatomical site where the patient experiences the snapping as this will most likely help in determining the underlying diagnosis. Lateral-sided clicking or snapping is most commonly associated with a snapping iliotibial band whereas snapping in the groin is more likely originating from the psoas tendon.

Signs with the patient upright

The fundamental principles of *look*, *feel* and *move* are applied to clinical examination of the hip joint. As the hip is a weight-bearing joint, it is critical to assess the patient both standing and lying in a suitable state of undress.

With the patient standing, posture and gait are examined and the Trendelenberg test is performed, as explained below. The patient should also be inspected for scars or sinuses, and comparison made of the two sides for muscle wasting or swelling.

Gait

Limp is a common presenting feature of patients with hip pathology. That limp can be characterized by its effect on the overall pattern of walking and also its effect on the gait cycle (the stance phase, the swing phase or indeed both). The more commonly seen abnormal gait patterns associated with hip pathology are the following.

Antalgic gait The fundamental feature of this abnormal gait pattern is that the patient spends less time on the painful limb so that stance phase is reduced when the painful side is in contact with the floor. Simply put, the patient wants to get off the painful leg as quickly as possible.

Stiff leg gait Movement of the hip joint in the sagittal lane (i.e. flexion and extension) contributes to the swing phase of the gait cycle. When this movement is lost or markedly restricted, the patient will tend to circumduct and swing their leg. This is achieved by the recruitment of more spinal movement.

Short leg gait When the patient is weight-bearing on the shorter leg, he or she dips down. This is most obvious by observing the head of the patient moving up and down depending on whether the shorter or longer limb is in the stance phase in the gait cycle.

Trendelenburg gait This is a dynamic representation of the Trendelenberg test. Put simply, with each step forward that the patient takes with the affected limb they lurch towards the unaffected limb. Patients commonly describe this as 'rolling over' on their hip as they walk. To understand the biomechanical basis of the Trendelenberg gait one first has to understand the basis of the Trendelenberg test which assesses the patient's postural stability when they stand on one leg. In biomechanical terms it is an assessment of the motor, lever arm and fulcrum of the hip joint.

Trendelenburg test In normal two-legged stance, the body's centre of gravity is placed midway between the two feet acting from the centre of the second sacral vertebra. In normal single-legged stance, the centre of gravity has to shift so that it lies over the weight-bearing leg (i.e. the centre of gravity needs to be supported to stop you falling over). To allow this to occur efficiently, the pelvis is effectively pulled up (by the motor) on the unsupported side and the centre of gravity is shifted directly over the standing foot. In clinical examination this is revealed by localizing and placing a finger on each anterior superior iliac spine (ASIS). As the patient stands on one leg the finger on the ASIS of the unsupported leg will rise. This is a normal response and would be recorded as Trendelenburg negative.

If the pelvis drops on the unsupported side, to avoid falling, the person has to throw his body towards the loaded side so that the centre of gravity is again over that foot. In this instance the finger on the ASIS of the unsupported leg will fall. This is recorded as Trendelenberg positive (Box 19.3) and a simple aide-memoire is the 'sound side sags'.

BOX 19.3 POSSIBLE CAUSES OF A POSITIVE TRENDELENBURG SIGN

- **Problem with the motor** – e.g. abductor muscle weakness, superior gluteal nerve palsy.
- **Problem with the lever arm** – shortening of the femoral neck, abductor tendon rupture
- **Problem with the fulcrum** – pain arising from intra-articular pathology, dislocation or subluxation of the hip

Signs with the patient lying

Look

With the patient then lying supine it is important to check that the pelvis is horizontal (both ASIS are at the same level) and the legs placed in a symmetrical position. Limb length can be gauged by looking at the heels, but measurement is more accurate. With the two legs in identical positions, measure the distance from the ASIS to the medial malleolus on each side. An apparent leg length discrepancy (LLD) is the amount by which one leg 'appears' shorter than the other; a true leg length discrepancy is determined by measurement from a fixed midline bony landmark (such as the xiphisternum) to the medial malleoli and is present when there is a genuine difference in leg length. Hence, a patient can have an apparent LLD without a true LLD, the most common causes being a scoliosis, pelvic obliquity or an adductor contracture. In cases where the fixed posture of the limb makes one leg appear shorter, this may indeed also cause a function LLD to the patient although there is no actual measurable LLD. This is an important consideration for patients who progress to total hip arthroplasty, where change in patient leg length is a common patient complaint following surgery.

Feel

The hip joint is seated deep in the groin, which means that the bony margins or joint line cannot be palpated in the 'feel' component of the clinical examination. With regard to palpation around the hip girdle, the examiner is therefore most often considering pathology outside the hip joint as part of the differential diagnosis. For example, tenderness on palpation of the greater trochanter can be easily detected and, if the patient is complaining of lateral-sided hip girdle pain, this points strongly towards an underlying diagnosis of trochanteric bursitis +/– abductor tendonitis.

Move

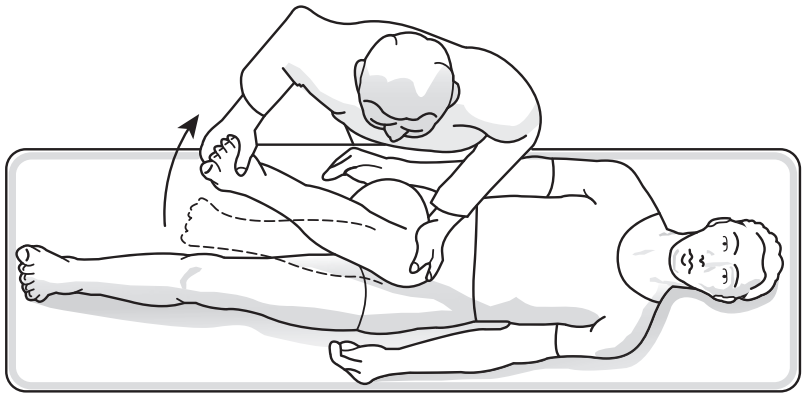
Assessment of a patient's range of motion is an important part of the clinical examination and very helpful in determining whether the underlying problem is intra-articular or not. As for all joints, comparisons can be made to the unaffected side. The loss of internal rotation of the hip joint (Figure 19.22) is the most reliable early clinical sign of intra-articular hip pathology. This may well be revealed on passive hip flexion where there may be a tendency to external rotation of the hip. With progress of joint stiffness, the development of a fixed external rotation contracture is common and thereafter fixed flexion of the hip.

As well as assessing the range of motion it is very important to document any pain that is associated with movement. Hip movement that causes pain which reproduces a patient's symptoms again can be very helpful in discriminating between intra-articular causes and pathology outside or referred to the hip girdle.

Imaging investigations

X-rays

Plain film radiographs remain the first-line and mainstay diagnostic imaging test utilized for hip assessment. Anteroposterior pelvis views together with a lateral projection of the affected side hip joint are the routine requested views. As for all X-ray interpretation, a standard workflow that considers adequacy, alignment and abnormalities (three A's) is a valuable tool to ensure a systematic assessment.



(a)

(b)

Figure 19.22 Clinical examination – internal rotation (a) How to assess internal rotation of the hip with the joint flexed. (b) A patient with normal 40° of internal rotation.

CT and MRI

With rapid technical advances over the last two decades, cross-sectional imaging, most notably CT and MRI, have become integral tools in the diagnosis and treatment of musculoskeletal disease. The pelvis is a complex 3D shape and the principal benefit of CT and MRI is the multiplanar evaluation of the hip joint and surrounding structures. Both CT and MRI have their own strengths and relative weaknesses, and these characteristics typically favour one modality over the other in evaluation of specific pathological conditions.

Modern CT scanning is a very rapid process and multidetector or spiral CT scanners have greatly reduced scanning time down to just several minutes. It is widely available and accessible. High-resolution multiplanar reformatted images can be performed. CT also has the advantage that there are no compatibility issues with metallic prostheses or devices such as pacemakers. CT does, of course, expose the patient to ionizing radiation. Cross-sectional imaging now not only continues to play an important role in diagnosis; there is an expanding role for its use in surgical planning (Figures 19.23–26), helping to provide the surgeon with the 'route map' to surgery.

MRI produces excellent soft-tissue imaging compared with the greyscale images of CT. It is an outdated concept to consider MRI as only a soft-tissue imaging modality as it does allow evaluation of the bony integrity of the hip and any abnormalities, and the physiological state of structures, such as in bone marrow oedema after a traumatic event or vascularity of the femoral head in AVN. One disadvantage of MRI is the length of the examination (typically 30–45 minutes) and that it requires the patient to remain motionless for long periods to obtain the best images. MRI may well be contraindicated in patients with pacemakers and some other implantable devices, and claustrophobia is an issue with many MRI systems.

Historically a limitation of MRI and CT is the artifact generated by orthopaedic hardware. However, modern *metal artifact reduction sequences* (*MARS*) MRI have led to a major improvement in images obtained around implants and as such in the last 5 years the use of MARS MRI in the assessment of the problematic hip replacement has become a very commonplace investigation (Figure 19.27).

Ultrasound scanning

Ultrasound scanning can be very useful in soft-tissue assessment around the hip. It is of particular use in the assessment of the abductor muscle insertion onto the greater trochanter and in identifying soft-tissue inflammation and fluid collections in this area. It has the major advantage that it is dynamic, the patient can interact with the operator to localize sites of tenderness, and these can be imaged directly. In addition, the patient can move the joint such that the dynamic behaviour of structures (e.g. snapping tendon) can be demonstrated. Ultrasound is also a helpful modality to deliver image-guided injections.

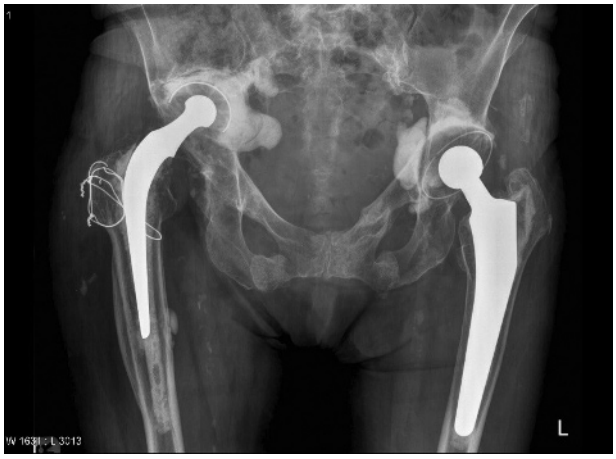


Figure 19.23 Imaging – X-ray Radiograph of a failed right THA with severe migration of the cemented acetabular socket. While it is clear that there is a major problem with the right socket, more detailed assessment is required to investigate the effect on surrounding structures. CT scans are extremely helpful to decide upon reconstruction technique and plan surgery.

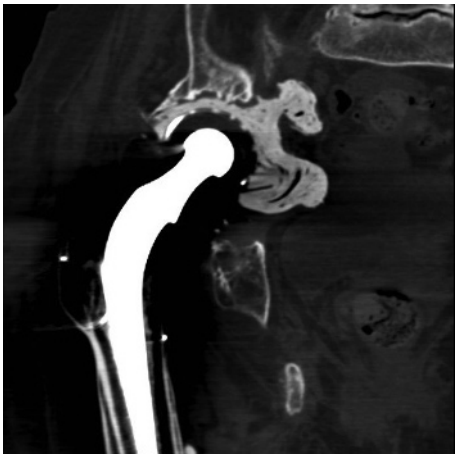


Figure 19.24 Imaging – CT Reformatted image obtained from multiplanar CT scan. This coronal slice demonstrates the enhanced detail available and allows accurate assessment of the amount and site of bone stock loss.

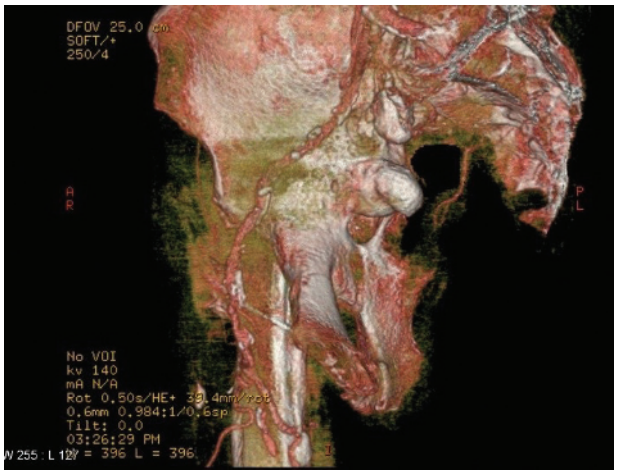


Figure 19.25 Imaging – CT angiogram In the same case as Figure 19.24, due to massive component migration into the pelvis, there was clinical concern about adjacent blood vessels. During the same CT scan the injection of intravenous contrast produces a vascular study – a CT angiogram, so proximity to major blood vessels can be assessed. Clearly, in this case the internal iliac vessels are at risk during surgery. This can have major intraoperative implications and may need the assistance of a vascular surgeon during the procedure.

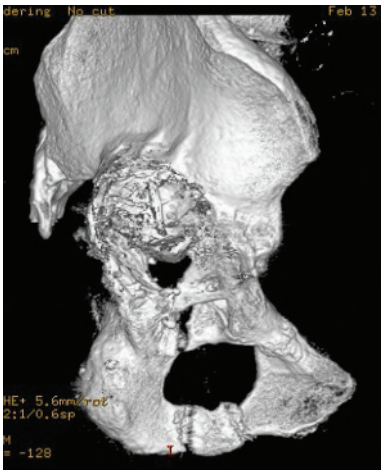
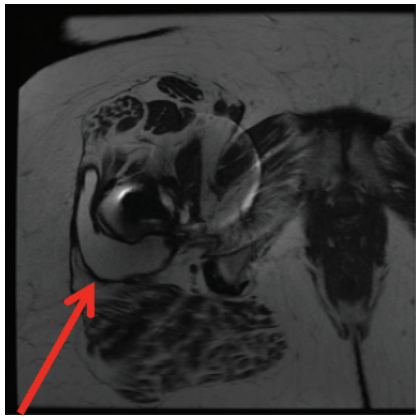


Figure 19.26 Surgical planning Modern CT images allow for significant post-investigation manipulation. In this same case the femur has been digitally subtracted from the image so that the surgeon has a view of the pelvis and acetabulum akin to what they actually see at the time of surgery and can represent a major aid to preoperative planning.



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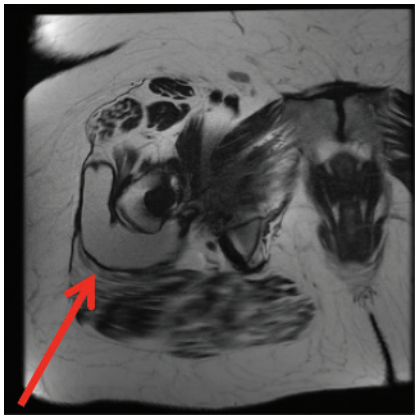


(b)

Figure 19.27 MARS MRI The patient here has a MOM THA and there is clinical concern about a potential adverse soft-tissue reaction. There is a 1-year interval between the left image (a) and the right image (b). As demonstrated, very clear, reproducible images can be obtained that are non-operator dependent. In this case the red arrows show that there has been an increase in the size of the abnormal fluid collection. This allows direct comparison by the surgeon in the clinic setting and also enables the surgeon to show this to the patient to aid explanation.

Arthrogram +/– local anaesthetic

Although modern cross-sectional imaging is very reliable, intra-articular hip lesions can be missed by radiological studies. The addition of contrast agents such as gadolinium in conjunction with CT and MRI has been shown to increase the diagnostic yield principally in the detection of labral lesions. An arthrogram can also represent a useful test when combined with the injection of local anaesthetic +/– steroid. In patients where clinical history and examination fail to confirm the diagnosis, an 'anaesthetic arthrogram' is helpful as a diagnostic test (Figure 19.28).

Following the injection of local anaesthetic, patients are requested to keep a pain diary, and a significant improvement in pain following hip-joint injection is a very helpful pointer that intra-articular pathology is causing the patient's symptoms. This can be particularly helpful when both lumbar spine and hip pathology coexist and the patient is unable to discriminate between the pain arising from the two anatomical locations. In patients with a significant hip joint effusion +/– synovitis, the addition of steroid to the arthrogram can also have a significant therapeutic benefit when the hyaline cartilage is preserved.

Modern surgical planning tools and 3D printing

The most up-to-date method of surgical planning involves transferring the data obtained from CT scan and cross-sectional imaging and the production of

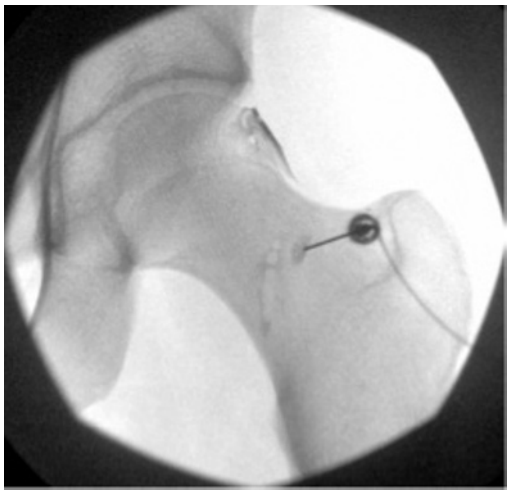


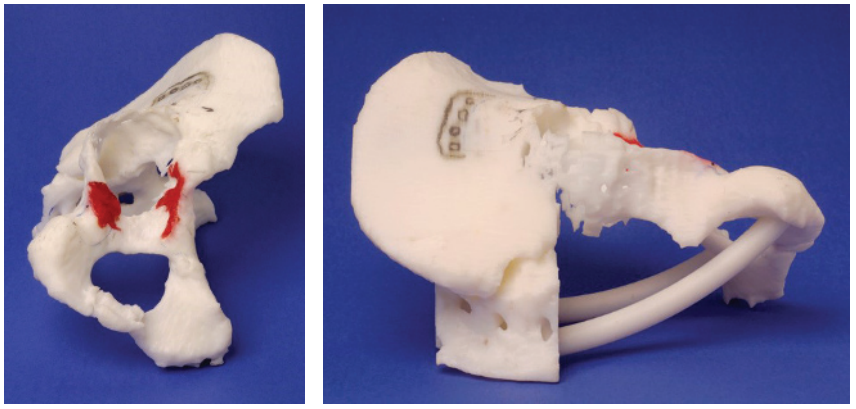
Figure 19.28 Anaesthetic arthrogram Fluoroscopy image obtained during an anaesthetic arthrogram. The needle visible on the image injects contrast to confirm the intra-articular placement prior to injection of local anaesthetic +/– steroid.

3D printed models. This again represents a significant advancement in surgical planning and allows the surgeon to perform a simulated procedure before entering the operating theatre. It is particularly helpful in complex cases in determining the feasibility of revision total hip arthroplasty (THA) preoperatively (Figures 19.29 and 19.30). Some implant companies use 3D models and planning tools to decide on implant type and size and, to replicate preoperative planning, patient-specific pre-jigs are provided. This technology is currently expensive, which prohibits widespread adoption, but with continuing advances and cost reduction it is easy to envisage the future applications.

Arthroscopy of the hip

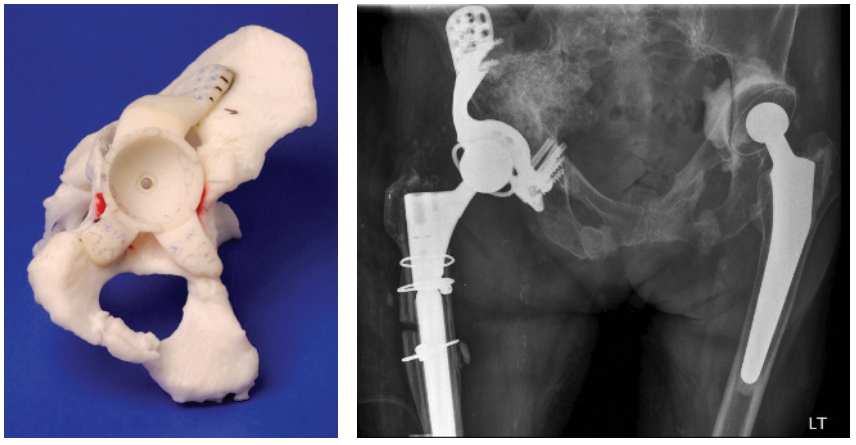
Over the past decade the number of hip arthroscopy procedures performed per annum has increased significantly. The role of hip arthroscopy, as for other joints, can be considered as either diagnostic or therapeutic. With the improvements in cross-sectional imaging, purely diagnostic hip arthroscopy has declined. This has been mirrored by improved instrumentation and surgical technique, such that therapeutic indications have also grown.

Contemporary arthroscopy of the hip is best considered as two phases: initial inspection and direct visualization of the joint that for the most part serves to confirm preoperative diagnosis, followed by therapeutic intervention where indeed both



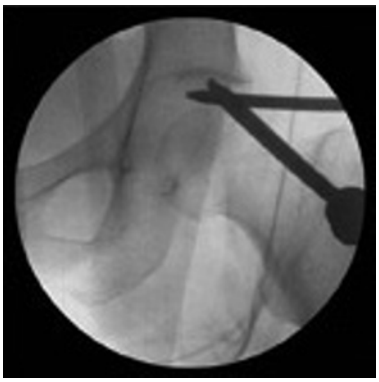
(a) (b)

Figure 19.29 3D modelling This is a 3D model of the case presented in Figures 19.24–26. It enabled the feasibility of revision total hip arthroplasty to be confirmed and significantly aided preoperative planning.



(a) (b)

Figure 19.30 Modelling in preoperative planning In this instance, due to the massive bone loss a custom-made implant was used.



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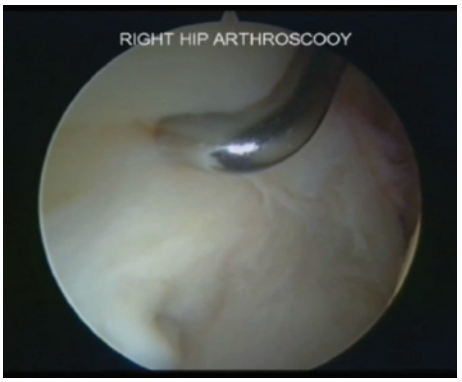


(b)

Figure 19.31 Arthroscopy
(a) Intraoperative image intensifier view of portal establishment set for hip arthroscopy. Note the distraction of the hip joint. (b) View obtained of the hip joint central compartment with probe demonstrating labral tear.

intra- and extra-articular pathology can be addressed and successfully treated.

The appeal of hip arthroscopy, as for other joints, is its ability to facilitate a minimally invasive method of treatment for hip disorders. Arthroscopic surgeons utilize a standard workflow to gain access to the hip joint following anatomical landmarks that allow cannula placement with the aid of an image intensifier (Figure 19.31). Modern hip arthroscopy has evolved to become a much more reproducible procedure. Specific traction tables aid the controlled joint distraction that is required to gain access to the hip joint. Pressure-controlled irrigation systems allow distention of the joint to be maintained, and improvements in optics and scope technology result in significantly improved visualization.

Once visualization has been achieved and working portals have been established, the procedure of hip arthroscopy generally follows a sequence of inspection of the two intra-articular hip joint compartments. The central compartment of the hip comprises the articulating hyaline cartilage surfaces of the acetabulum and femoral head together with the ligamentum teres. The peripheral compartment comprises the intra-capsular recess outside the hip articulation.

Patients who are candidates for hip arthroscopy typically present with mechanical symptoms as well as groin pain. These mechanical symptoms include clicking, catching, locking or buckling; this can also compromise function. Indications for hip arthroscopy and potential complications of the procedure are listed in Boxes 19.4 and 19.5.

BOX 19.4 INDICATIONS FOR HIP ARTHROSCOPY

Intra-articular

- Femoroacetabular impingement (e.g. osteochondroplasty)
- Labral tears
- Loose/foreign bodies
- Cartilage lesions
- Osteochondritis dissecans
- Ligamentum teres injuries
- Total hip replacement assessment

Extra-articular

- Iliopsoas tendinopathy
- Snapping iliotibial band
- Greater trochanteric pain syndrome

BOX 19.5 COMPLICATIONS OF HIP ARTHROSCOPY

Anaesthetic complications – procedure performed under general anaesthetic
Traction-related injury – either from pressure (e.g. pudendal nerve in groin) or, less commonly, directly from over-stretch
Neurovascular injury – most commonly lateral cutaneous nerve of the thigh from portal placement
Extravasation of irrigation fluid
Infection

INTRA-ARTICULAR HIP PATHOLOGY – OSTEOARTHRITIS OF THE HIP JOINT

The hip joint is one of the commonest sites of osteoarthritis (OA). In the past the traditional approach has been to consider hip OA as either primary or secondary. Primary hip osteoarthritis was thought to occur when no underlying cause was apparent, and, when an obvious underlying cause was identified, the term secondary osteoarthritis was applied. This system of classification is outdated, as is the use of the term 'wear-and-tear' arthritis. Over the past two decades there has been major progress in our understanding of the pathoanatomy that leads some patients to develop OA while others develop no problems at all.

BOX 19.6 MECHANICAL AND NON-MECHANICAL CAUSES OF HIP OA

Mechanical

- Developmental dysplasia of the hip (under-coverage)
- Femoroacetabular impingement (over-coverage)
- Perthes disease, slipped capital femoral epiphysis (loss of sphericity)
- Post-traumatic (loss of congruency)

Non-mechanical

- Avascular necrosis of the femoral head
- Ankylosing spondylitis
- Inflammatory arthritis (e.g. rheumatoid arthritis, psoriatic arthritis and systemic lupus erythematosus)
- Primary disorders of cartilage and the synovium (e.g. synovial chondromatosis)

A much more modern approach to the classification of hip OA is to consider the causes as *mechanical* or *non-mechanical* in origin (Box 19.6).

Mechanical causes

The hip is a ball-and-socket joint; as such, when there is 'matched sphericity' between the two articulating surfaces, a mechanical problem with the joint is unlikely. When there is loss of sphericity, then the articular surface is exposed to abnormal loads and contact forces that can initiate and then cause progressive hyaline cartilage damage. The common mechanical causes of hip OA include developmental dysplasia of the hip (DDH) where acetabular under-coverage leads to a loss of sphericity, and femoroacetabular impingement (FAI) where acetabular over-coverage or abnormal femoral shape leads to a loss of sphericity. A similar loss of sphericity is seen as a long-term consequence of Perthes and SCFE/SUFE. In post-traumatic OA there is loss of congruency of the articulating surface as a consequence to intra- or peri-articular fractures.

Non-mechanical causes

These are conditions or processes that can affect the hip joint in isolation or indeed be part of a more widespread musculoskeletal disorder with the common end-stage effect being the destruction of hyaline cartilage. These non-mechanical causes of hip OA include avascular necrosis of the femoral head also termed femoral head osteonecrosis (FHON), ankylosing spondylitis, primary disorders of cartilage and the synovium (e.g. synovial chondromatosis) and inflammatory arthropathy including seronegative inflammatory arthritis.

Clinical presentation

The classic presentation of hip joint OA is groin pain associated with progressive stiffness and limp. Initially this pain may be activity-related but later it is more persistent and can cause disturbed sleep. While an antalgic gait is most common, patients can present with any of the gait abnormalities described and not infrequently these can overlap. Loss of internal rotation of the hip is one of the most consistent clinical findings. As loss of movement progresses, the patient may develop both a fixed external rotation contracture and fixed flexion deformity. At the extreme of passive range of motion it is common for the patient to experience pain that generally reproduces their symptoms. As such, the reproduction pain felt on rotation of the hip joint can be useful in differentiating from intra- or extra-articular sources or indeed referred pain.

Radiological investigations

The *plain film radiograph* overwhelmingly represents the most common radiological investigation that is used to confirm the clinical suspicion of OA. The four important classical signs of OA equally apply to the hip joint (Figure 19.32). The earliest sign is typically decreased joint space that represents loss of hyaline cartilage. Differing initial patterns of cartilage loss can relate to the underlying pathological mechanical process and hip joint anatomy.

This is usually maximal in the superior weight-bearing zone when it is often termed *superior pole hip*

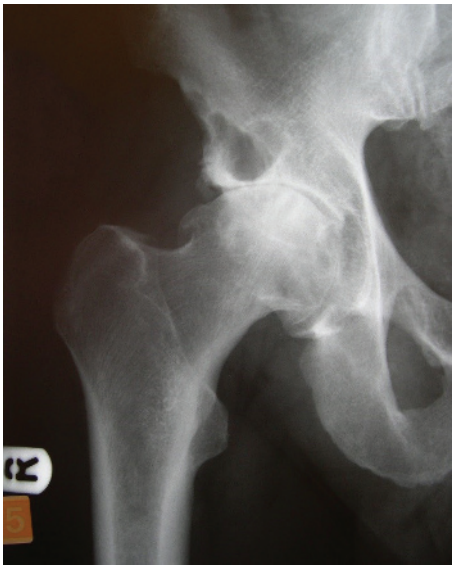
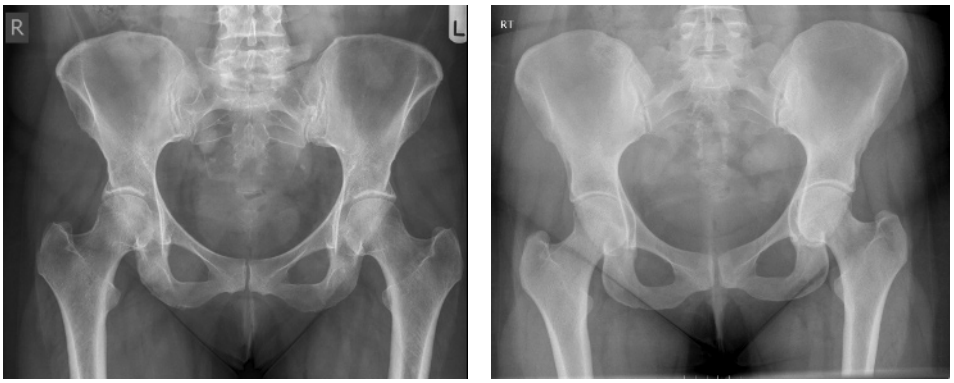


Figure 19.32 Hip osteoarthritis The four cardinal radiographic features of OA are demonstrated. Note the loss of joint space due to hyaline cartilage loss and osteophyte formation, and subchondral sclerosis and subchondral cysts. In this example there is a large acetabular subchondral cyst that is termed a geode.



(a) (b)

Figure 19.33 Femoroacetabular impingement (FAI) (a) Coxa profunda where the acetabular fossa is medial to the ilioischial line. (b) Protrusio where the medial aspect of the femoral head lies medial to the ilioischial line. Both of these anatomical patterns are a cause of global over-coverage of the hip joint and predispose to pincer-type impingement.

OA (e.g. in cam type FAI) or it can also occur more centrally termed *medial pole hip OA* (e.g. in protrusio). This cartilage loss generally progresses to affect the entire joint. The later X-ray signs of OA are subarticular sclerosis, subchondral cysts and osteophyte formation.

Inspection of radiographs often reveals the underlying hip morphological problem that predisposed that patient to development of OA. It is commonplace that the first presentation of a mechanical precipitating cause is end-stage OA. In younger women it is common to find the hallmarks of more subtle dysplasia that did not present earlier in life whereas in men in particular radiological hallmarks of FAI are often present.

MECHANICAL CAUSES OF HIP OA

FEMOROACETABULAR IMPINGEMENT AND OA

The concept of femoroacetabular impingement (FAI) as a cause of hip OA is relatively new and its pathogenesis has been elaborated significantly only in the last 10–15 years. It is considered a mechanical cause of hip OA due to loss of sphericity and essentially over-coverage of the ball-and-socket joint. The work of Ganz and co-workers has provided a major understanding into the pathomechanics of FAI that can be summarized by the '*pathological abutment*' that occurs in two main subtypes of FAI: *pincer* and *cam* types.

In the *pincer type* (Figures 19.33 and 19.34) there is either global over-coverage of the femoral head (circumferentially as in coxa profunda or protrusio) or focal anterior over-coverage (by the anterior part of the acetabular rim or by acetabular retroversion). In the *cam type* there is loss of sphericity at the femoral head neck junction that causes jamming of the femoral neck against the front of the acetabulum. The common pathway of these FAI types is the pathological impact of bone against cartilage that can result in initial damage to the acetabular labrum followed by abrasion or delamination of the hyaline cartilage. It is

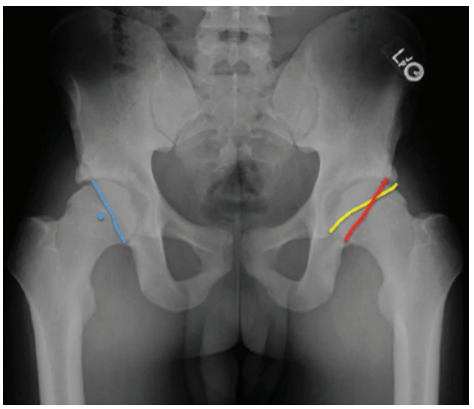


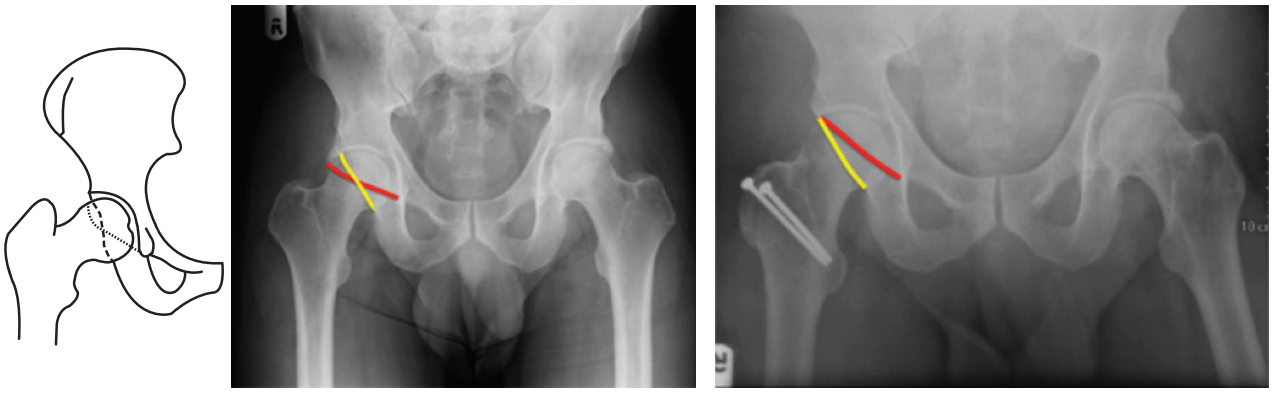
Figure 19.34 Pincer-type FAI due to focal anterior acetabular over-coverage As is common in FAI, the same abnormalities are present bilaterally. In the left hip the radiological crossover sign is demonstrated by tracing the overlap of the anterior (yellow line) and posterior (red line) acetabular walls. On the right hip it is possible to see how the centre of rotation is lateral to the posterior wall (blue line).

common for FAI hips to demonstrate a mixed picture of both pincer and cam impingement.

Clinical features

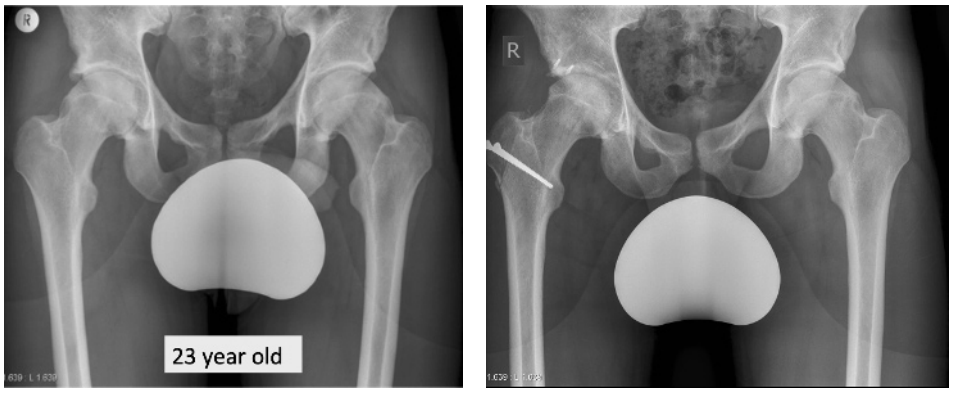
Groin pain and *decreased range of movement* are the usual presenting symptoms. During the initial stages of the disease, groin pain is predominantly activity-related especially with impact loading. It is therefore not uncommon for this condition to present at an earlier stage in its natural history in elite athletes who place increased demands on their hip joints. As the pathological abutment progresses and cartilage damage occurs symptoms become more persistent until eventually the patient presents with the classical symptoms of established hip OA.

The typical patient with pincer-type FAI is female and over 40 years of age while the typical patient with cam-type FAI is a male and about 10–15 years younger. The typical clinical feature is a restriction of internal rotation in flexion that may well affect



(a) (b)

Figure 19.35 Pincer-type FAI secondary to focal anterior over-coverage (a) As in Figure 19.34, a positive crossover sign is demonstrated. (b) Following open surgical dislocation of the left hip via a Ganz osteotomy acetabular rim resection has been performed and labral refixation. Note correction of the crossover sign and screw fixation of osteotomy.



(a) (b)

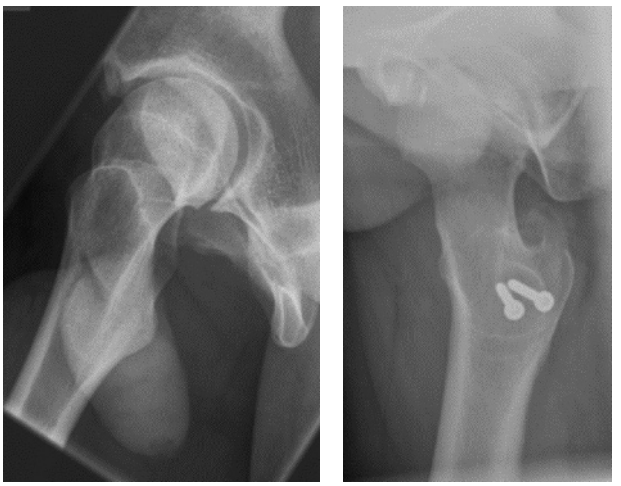
Figure 19.36 Predominantly cam-type FAI (a) Note the large bump at the superior head-neck junction. (b) Following open surgery, at which time femoral osteochondroplasty 'reshaped' the head-neck junction.

both hips. This test (often referred to as an impingement test) can commonly recreate groin pain, especially when there is chondral or labral damage present.

Investigations

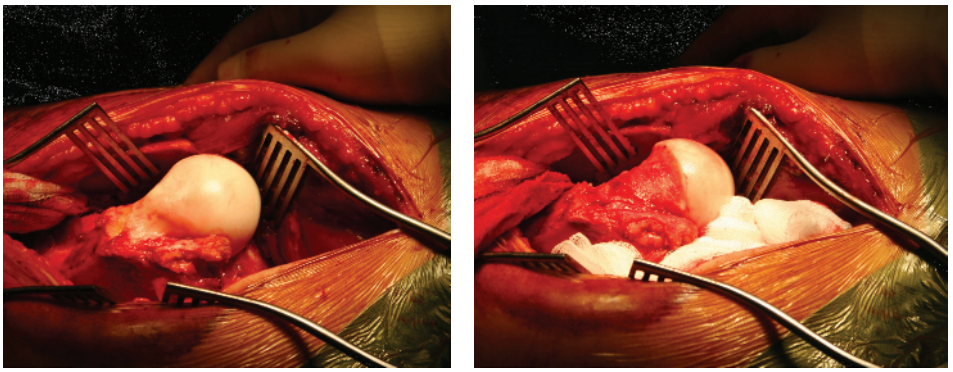
Identifying the subtle bony abnormalities in patients presenting with hip pain is becoming increasingly important for hip surgeons. It allows earlier intervention to relieve symptoms and may enable in the future the prevention of the long-term sequelae of end-stage hip arthritis.

A well-centred anteroposterior *radiograph* of the pelvis and a cross-table lateral radiograph of the hip are required. The pelvis should be pictured with the coccyx pointing towards the symphysis and a distance of 1–2 cm between them. A good quality X-ray projection is essential for assessing acetabular version as it needs to allow visualization of the anterior and posterior rims of the acetabulum. Abnormal signs related to anterior acetabular over-coverage include a cross-over sign and ischial spine sign (Figures 19.34 and 19.35). In cam-type FAI there is a characteristic 'bump' at the superior head-neck junction that may be accompanied by a 'pistol-grip' deformity of the proximal femur (Figures 19.36 and 19.37). Global acetabular



(a) (b)

Figure 19.37 FAI (a) Preoperative and (b) postoperative lateral view of the same case shown in Figure 19.36. Note again on the left image the anterior 'bump' at the head-neck junction and also os acetabulare. The right postoperative image clearly demonstrates how removal of the bump has recreated the anterior offset of the femoral head. The os acetabulare has been excised and a labral repair performed with suture anchors.



(a) (b)

Figure 19.38 FAI - surgery Open surgical dislocation of the hip performed via a Ganz osteotomy. (a) Loss of the head-neck junction. (b) Recreation of the anterior offset by bone resection (osteochondroplasty).

over-coverage is associated with coxa profunda or protrusio.

MRI scans allow a more detailed assessment of hip morphology, and in particular radial slices are used to visualize the labrum and cartilage as well as the femoral head/neck morphology. MRI scans are also helpful to assess the extent of cartilage damage within the joint.

The intra-articular injection of contrast into the hip prior to MRI (*MRI arthrogram*) has been demonstrated to be both more sensitive and more specific for detecting labral and some chondral lesions; however, there are limitations in detecting undisplaced delamination of the acetabular cartilage. New advanced imaging techniques such as the 3-Tesla system and delayed gadolinium-enhanced MRI of cartilage (dGEMRIC) may in the future be able to detect early cartilage lesions.

Treatment

NON-OPERATIVE TREATMENT

The non-operative management of FAI revolves around symptom management and modification of activity. The extent of hyaline cartilage damage within the hip joint has a profound effect on both the outcome and the treatment options for FAI. In cases where hyaline cartilage is preserved joint preservation surgery can be considered.

OPERATIVE TREATMENT

Joint preservation of the hip can be performed either by open or by arthroscopic surgery. The fundamental principal is the resection of the 'impingement lesion' called *osteochondroplasty*. In cam-type FAI this involves recontouring of the head-neck junction to re-establish the sphericity of this area and allow adequate clearance during hip flexion. In pincer-type FAI for focal anterior over-coverage this involves rim resection. Associated labral tears are repaired if possible in an attempt to restore the suction-seal of the hip joint. For more global over-coverage, realignment osteotomies of the acetabulum may be considered. As advances have been made in both instrumentation and technique, arthroscopic methods have become more commonplace than open procedures. Open surgical preservation operations generally involve the surgical dislocation of the hip joint with careful attention to the preservation of the blood supply to the femoral head (Figure 19.38).

Hip dysplasia and OA

Developmental dysplasia of the hip (DDH) in the adult can present a spectrum of challenges to the hip surgeon. Abnormal anatomy is present that is often associated with a hypoplastic pelvis and/or femur together with frequent issues as a result of leg-length discrepancy. It is common for patients with DDH to have had prior surgical interventions, such as osteotomies, which can further increase the complexity of surgery due to retained metalwork. The key principle of joint replacement in the setting of DDH is restoration of the hip centre of rotation. There may be significant challenges to joint replacement in the setting of DDH that include bone stock deficiency, small socket and femur size, excessive femoral anteversion and residual deformity (Figures 19.39 and 19.40).

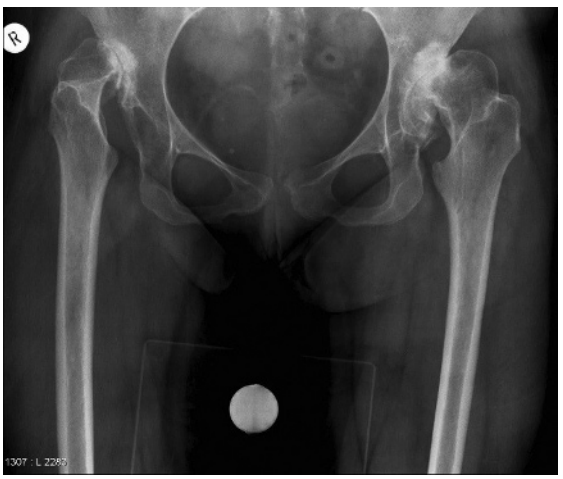


Figure 19.39 Hip dysplasia and OA Bilateral DDH with resulting mechanical OA secondary to under-coverage.

BOX 19.7 THE CROWE CLASSIFICATION

Measurement of proximal migration from the inter-tear drop line

Crowe I	Proximal subluxation of <50% of the height of the femoral head
Crowe II	50–75% subluxation
Crowe III	75–100% subluxation
Crowe IV	>100% subluxation

BOX 19.8 THE HARTOFILAKIDIS CLASSIFICATION

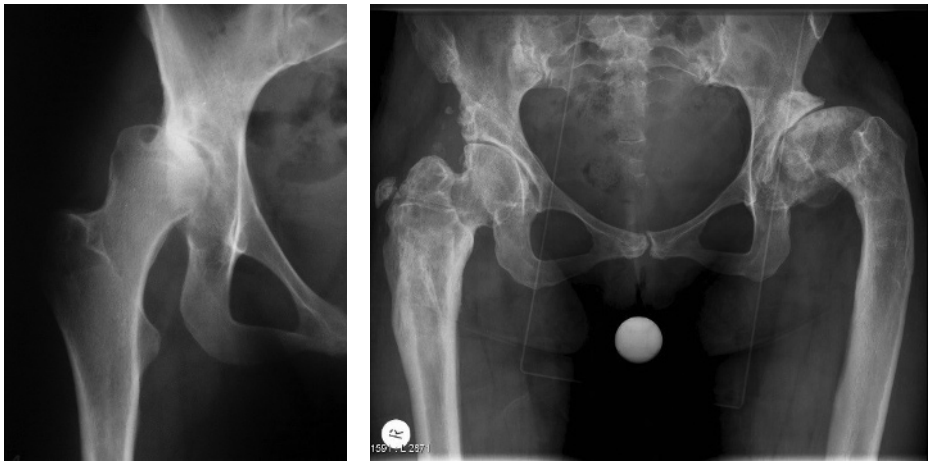
Dysplasia	Femoral head subluxated but still contained in the original acetabulum
Low	Femoral head articulates with the false acetabulum that partially overlaps the true acetabulum
High	Femoral head articulates with a hollow in the acetabular wing

The level of the 'false acetabulum', which degenerates into end-stage osteoarthritis, generally determines to a large part the complexity of the surgery. DDH in the adult can be classified according to the amount of hip subluxation. The classification systems include those of Crowe and colleagues and Hartofilakidis and his co-workers (Boxes 19.7 and 19.8). Outcome after THA and implant survivorship are influenced by the extent of the dysplasia. Classification can help guide treatment and predict outcome. In severe cases of DDH a subtrochanteric shortening osteotomy may be required, as a general rule when more than 4 cm of leg-length discrepancy is present. Major changes in leg length increase risk of nerve injury. Implant selection often necessitates the use of small component sizes with modularity that can allow correction of established deformity (Figures 19.40–43).

Post-traumatic hip OA

Trauma to the hip joint can result in disruption of the joint surface congruity that leads to the progressive loss of hyaline cartilage and mechanical OA. Trauma may cause direct cartilage damage at the time of injury, such as an intra-articular fracture of either acetabulum (Figure 19.44) or femoral head fracture, or indeed native hip dislocation. Cartilage damage may then be progressive when joint surface irregularities persist. Extra-articular trauma can result in proximal femoral deformity that can alter the biomechanics of the hip and joint loading, resulting in progressive cartilage loss. Avascular necrosis (AVN) is also a potential sequela of significant hip joint trauma, especially fracture-dislocations of the hip joint, and secondary collapse as a result of femoral head AVN that can exacerbate loss of joint congruity.

Total hip arthroplasty (THA) is the most frequent treatment for end-stage post-traumatic OA. In such cases of THA the complexity of the surgery can be increased significantly by post-traumatic deformity, retained metalwork from previous internal fixation, soft-tissue scarring and heterotopic ossification.



(a) (b)

Figure 19.40 The spectrum of complexity associated with OA in DDH
(a) A patient with no prior intervention and whose first presentation is with mechanical OA in mid adulthood. (b) A younger patient who has had more than ten previous surgical procedures and has significant femoral deformity on the left hip and obliteration of the femoral canal on the right hip.

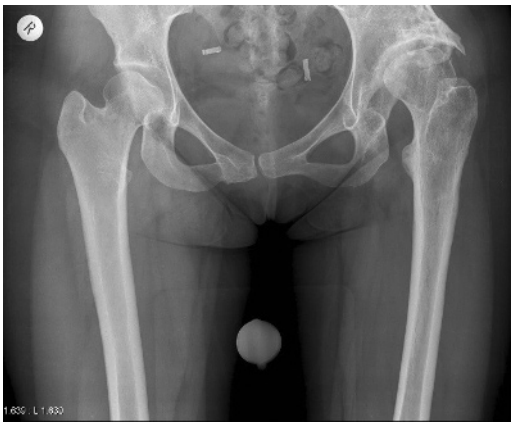


Figure 19.41 Mechanical OA as a result of DDH Preoperative X-ray of a 32-year-old female patient. She has had a previous shelf procedure. It would be classified as a Crowe II, or a low dislocation according to Hartofilakidis.

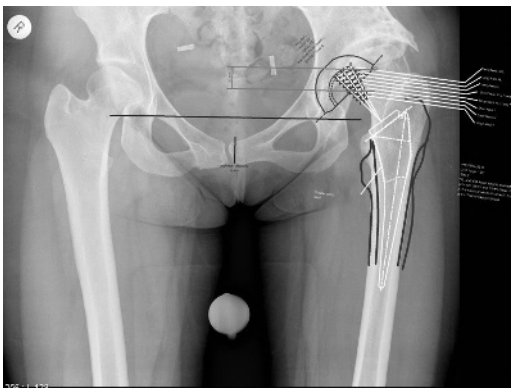


Figure 19.42 DDH and OA Same patient as in Figure 19.41. Preoperative digital templating highlights the uncovering of the acetabular socket.

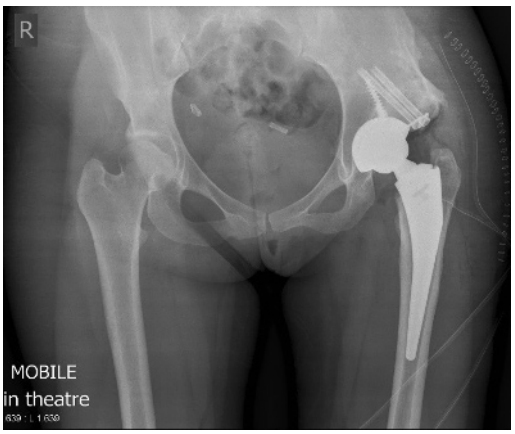


Figure 19.43 DDH and OA Same patient as in Figure 19.41. X-rays in theatre following completion of the complex primary THA. The resected femoral head was used as a superior bulk structural allograft which, as such, aids the stability of the socket and provides greater bone stock for the future. A modular femoral component was used to correct the excess femoral anteversion.

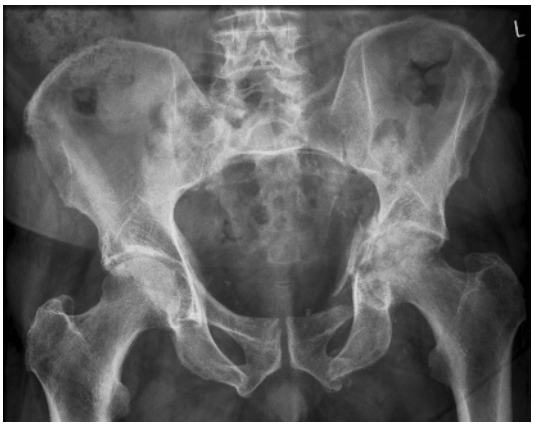


Figure 19.44 Post-traumatic hip OA as a mechanical cause of OA A previous acetabular fracture has caused significant articular surface disruption with loss of joint congruity.

NON-MECHANICAL CAUSES OF HIP OA

AVASCULAR NECROSIS OF THE FEMORAL HEAD

The femoral head is the commonest site of avascular necrosis (AVN) as its blood supply renders it vulnerable to ischaemia. Multiple hypotheses remain to explain the development of AVN, which include ischaemia, direct cellular toxicity and altered differentiation of mesenchymal stem cells. Most recently idiopathic AVN has been linked to anatomical abnormalities including decreased femoral neck angle, increased femoral anteversion, reduced centre edge angle and reduced acetabular anteversion.

The causes of hip AVN (Box 19.9) include excess alcohol consumption and the use of corticosteroids; trauma, in particular displaced intracapsular hip

BOX 19.9 CAUSES OF FEMORAL HEAD AVASCULAR NECROSIS

Idiopathic
Excess alcohol consumption
Use of corticosteroids
Trauma, in particular displaced intracapsular hip fractures and traumatic hip dislocations
Sickle-cell anaemia
Gaucher's disease
Thrombophilia
Primary Cushing disease
Chemo- and radiotherapy
Decompression sickness

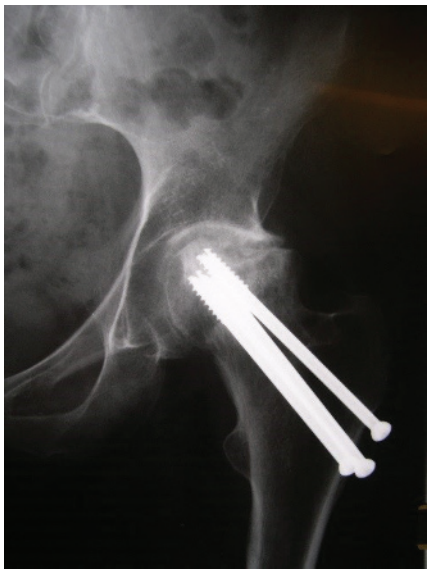


Figure 19.45 Avascular necrosis (AVN) Post-traumatic AVN following intracapsular hip fracture.

fractures (Figure 19.45) and traumatic hip dislocations or AVN may well be idiopathic. Other less common causes of AVN include sickle-cell anaemia (Figure 19.46), Gaucher's disease, thrombophilia, primary Cushing disease and decompression sickness. The true incidence of hip AVN is difficult to determine as many early cases progress undiagnosed and only later present as end-stage osteoarthritis. AVN eventually results in the collapse of subchondral bone that causes joint incongruity.

Diagnosis

Plain X-rays are often normal in the early stages of AVN and the first signs usually occur 6 months after the occurrence of bone death. When present, these changes reflect increased density in the adjacent living bone and are most commonly seen as increased density/sclerosis. As time progresses the classic appearance is of a thin subchondral fracture line (the '*crescent sign*'), which is an important differentiating factor in the management of AVN. Prior to any disruption of the articular surface, the AVN is termed *pre-collapse*. Following the appearance of a crescent sign, flattening of the affected weight-bearing zone occurs, and at this stage the AVN is termed *post-collapse*. With the continued collapse of subchondral bone there is loss of support for the articular surface, such that progressive hyaline cartilage loss occurs.

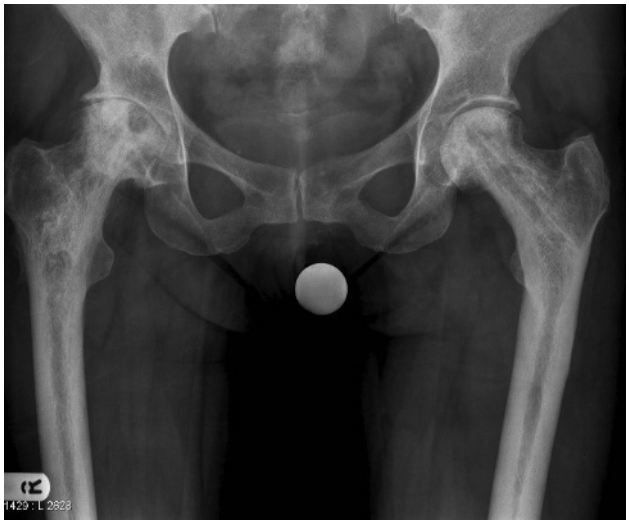
MRI scans facilitate earlier diagnosis and show characteristic changes in the marrow long before any abnormality is visible on plain radiographs. The diagnostic feature on MRI is a band of altered signal intensity running through the femoral head that represents the reactive zone between living and dead bone. This demarcates the ischaemic area, the extent and location of which are important factors in several of the classifications and clinical staging of AVN.

There are many proposed classifications. A key watershed that unifies all the proposed systems is the differentiation of the AVN into pre-collapse and post-collapse as this has very significant implications for successful treatment.

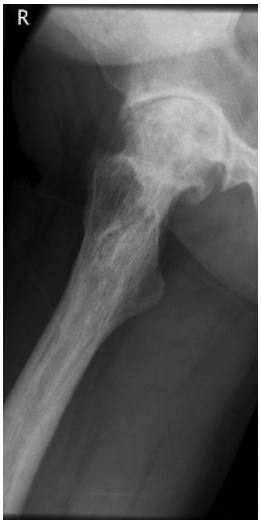
Treatment

PRE-COLLAPSE AVN

Pre-collapse AVN offers the opportunity for joint preservation and the *non-surgical management* of AVN for symptomatic or non-symptomatic patients. Protected weight-bearing, bisphosphonates and anticoagulation for AVN associated with hypercoagulability have been reported with variable success. Although investigation into non-operative management continues, a general conclusion from available data would suggest that larger areas of AVN (greater than 25% of



(a)



(b)

Figure 19.46 AVN secondary to sickle-cell anaemia Note on the lateral view the classic bone within a bone appearance. The right hip demonstrates established non-mechanical OA and in the left hip the tract from previous core decompression is visible in the femoral neck.

the femoral head) located in the weight-bearing zone will tend to progress if not treated surgically.

In the *surgical management* of pre-collapse AVN numerous procedures have been employed. The majority are variations on the theme of percutaneous decompression of the AVN lesion accompanied by bone grafting. Contemporary practice also often includes the adjuvant use of potential biological agents such as concentrated stem cells, bone morphogenetic protein (BMP) and platelet-rich plasma (PRP). There are limited available data on the use of these biological agents at this stage and they have not been assessed in a randomized control trial. Vascularized bone grafts have been used previously but are now employed less commonly. The same is true for femoral osteotomies.

POST-COLLAPSE AVN

There is a general consensus that hip preservation procedures are not recommended in post-collapse AVN. In these cases it is more judicious to consider *total hip arthroplasty* because the results are more reliable. However, it should be appreciated that higher failure rates, accounted for by several factors, are often encountered in joint replacement for severe AVN of the femoral head compared with THA for other causes of osteoarthritis. The factors that often influence outcome are of those that were the underlying cause for AVN (e.g. previous radiotherapy to hip/pelvis) and young patient age.

Ankylosing spondylitis and hip OA

While the primary effect of ankylosing spondylitis (AS) is in the spine, the hip joint is the most common site of peripheral joint involvement. The hip joint can be involved in 30–50% of patients with AS. In AS patients the movement that occurs at the hip joint provides an important compensation for the progressive spinal stiffness that patients experience. With progressive hip involvement this compensation is lost, and deteriorating hip function can have a profound effect on a patient's balance, particularly in the sagittal plane. Patients with extensive hip involvement as a result of AS often have severe soft-tissue contractures, especially fixed flexion contracture.

The indications for THA in non-mechanical hip OA due to AS are as for other causes of OA based on an individualized patient assessment. Some complications, such as the formation of heterotopic ossification, are more frequent after total hip replacement for AS.

Inflammatory arthritis of the hip

Inflammatory arthritis results from progressive hyaline cartilage destruction of the hip joint via the synovium. Examples include rheumatoid arthritis (RA) and systemic lupus erythematosus (SLE). RA has an incidence of around 1–3% in the population, affects the hip joints in 15–20% of patients, and often affects both hip joints.

Huge progress has been made in the non-surgical management of RA over the past decade with established drug regimes that often include disease-modifying drugs. Disease-modifying antirheumatic drugs (DMARDs) are a group of medications commonly used in patients with rheumatoid arthritis. They work to decrease pain and inflammation, to reduce or prevent joint damage, and to preserve the structure and function of the joints. Some of these drugs are also used in treating other conditions such as AS, psoriatic arthritis and SLE.

Initial findings on X-ray (Figure 19.47) may be minimal because synovitis typically causes the initial pain at presentation. Osteophyte formation typically is not observed, but progressive hyaline cartilage destruction leads to joint space narrowing. Other common radiographic findings include cyst formation and acetabular protrusio-type deformity.

Failure of non-surgical management is an indication for THA. As a consequence of the underlying disease process, coupled with side effects from often long-standing medical management, there are particular challenges that THA in patients with inflammatory arthritis presents to the hip surgeon. Poor bone stock and osteopenia as a result of disuse and osteoporosis from long-term steroid use is common. This can significantly affect the risk of fracture related to THA. These patients exhibit a degree of immunosuppression which increases their risk of periprosthetic joint infection. This patient group is best managed as part of a multidisciplinary team with the key involvement of a rheumatologist alongside the orthopaedic surgeon in order to best optimize their perioperative

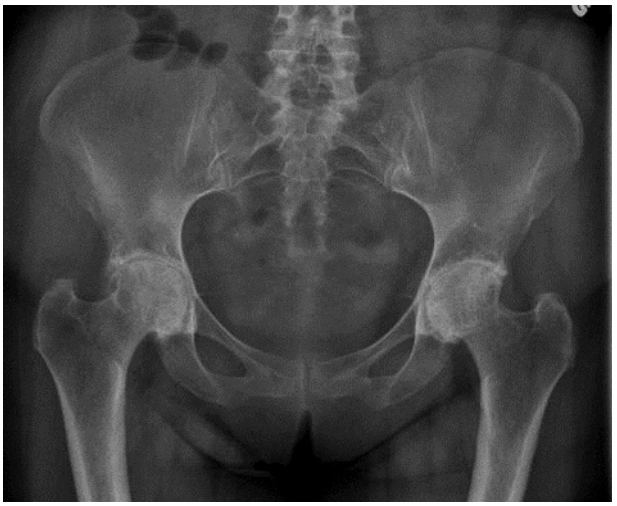


Figure 19.47 Inflammatory arthritis Note severe loss of joint space and periarticular cysts.

management. Anti-TNF agents should be stopped in the perioperative period as they inhibit wound healing, but cessation of other disease-modifying drugs such as Methotrexate is controversial, as this may result in a generalized flare-up of symptoms. Often other joint involvement can affect postoperative rehabilitation (e.g. shoulder inflammatory disease may restrict use of crutches) such that structured occupational and physiotherapy input is required.

NON-OPERATIVE TREATMENT OF HIP OA

As the main presenting symptom is most often pain, the cornerstone of non-operative management of hip OA focuses on effective *pain management*. Simple oral pharmacological agents such as non-steroidal anti-inflammatory drugs (NSAIDs) are often among the most effective and avoid the undesirable side effects from opioid-based drugs. Paracetamol has been shown to be no more effective than placebo, but the placebo effect of drugs should not be underestimated.

Life-style modification to reduce impact loading and also weight reduction for overweight patients is also important. The absolute weight is probably more important than the body mass index, as the patient's weight is directly proportional to the forces transmitted through the hip joint. The use of a walking stick or walking aids assist in maintaining mobility.

The more common natural history of hip OA is that of steady decline with increasing pain and decreasing function, although a small group of patients can fall into a subgroup of rapidly progressive symptoms determined by the underlying pathological cause for the OA. Some patients experience flares of pain while some have a remitting course where symptoms can improve for protracted periods.

TOTAL HIP ARTHROPLASTY

Total hip arthroplasty (THA) has established itself as one of the most successful operations performed across all surgical specialties and is the definitive treatment of end-stage OA of the hip. Its success is due in no small part to its reproducibility where excellent results can be obtained using differing surgical approaches and various designs of implant. The options of osteotomy and arthrodesis to treat hip OA are now performed very rarely indeed, and excision arthroplasty of the hip is considered a salvage procedure only in the setting of persistent hip infection. These now almost historical techniques for hip OA played a significant role in the past by virtue of the fact that they presented a reasonable option for pain relief. They did not, however, restore the biomechanics of the hip – in fact, they most often restricted joint movement – and as a result the trade-off for pain relief was poor function. The success of THA is in major part due to there being no such trade-off in function. The fact that THA addresses pain, stiffness and loss of function has led to its being labelled the 'operation of the century' by the *Lancet*. In joint registries all around the world without exception OA is the leading indication for THA. In European countries it accounts for over 90% of hip replacements.

Indications

THA for OA is best thought of as a lifestyle intervention. In general, when non-operative management has failed and symptoms have reached a stage where they are producing an unacceptable impingement on an individual's lifestyle and participation in society, THA should be considered. Although THA is generally highly successful, its risks must be weighed against the indications for the procedure and functional limitations facing the patient. So, the decision to proceed to THA is based on an individualized patient assessment, including pain, level of disability and loss of function coupled, importantly, with their own expectations of potential outcome following surgery. The most common indications for THA include the following: severe pain on movement, disturbed sleep due to pain, pain at rest, functional limitations during activities of daily living and reduced walking distance. Preoperative assessment needs also to consider an individual patient's medical comorbidities, which may preclude THA or have a significant effect on perioperative risk of both mortality and morbidity.

Age is an important factor to consider when considering a patient for THA. The average age of a patient undergoing THA in Europe is 68 years. Although younger patients, of course, may equally benefit from the pain relief and functional improvement from a THA, they need to be informed and understand the significant risk of single or multiple revisions that may be required during their lifetime.

In a 40-year observational study researchers at the Mayo Clinic, in the United States of America, have provided some useful benchmark data for the lifetime risk for revision THA. With almost all the patients followed up till either death or revision, a 'rule of thumb' for the lifetime likelihood of revision is reported based on the age at implantation of the index THA. This is 1 in 3 for patients aged <50 years, 1 in 5 for patients 50–59 years, 1 in 10 for patients 60–69 years, and 1 in 20 for patients ≥70 years.

Surgical approach

A summary of the commonly used surgical approaches to perform THA is provided in Table 19.3. The surgeon most commonly chooses the choice of approach based on their training and competence. Other factors may include previous incisions, obesity, risk of dislocation and deformity. Each surgical approach has its perceived risks and benefits. There has been a trend in the past decade to reduced incision size and more minimally invasive techniques have become popular.

Implant types

A total hip replacement comprises an acetabular cup and a femoral stem that articulate with each other at the bearing surface. These are broadly classified according to the type of fixation that stabilizes the implant within the bone, either cemented or uncemented. In this traditional subdivision *cemented fixation* provides mechanical fixation by interlocking with bone and *uncemented fixation* relies on osseointegration, a biological bonding of the component to host bone. THA can also be performed using a combination of fixation methods for different components that constitute the construct; a *hybrid THA* utilizes an uncemented acetabular component and a cemented femoral stem, while in a *reverse hybrid THA* a cemented socket and uncemented stem are used.

The THA articulation or bearing can also be considered as two broad categories according to the materials used, i.e. hard-on-soft bearings (metal-on-polyethylene or ceramic-on-polyethylene) or hard-on-hard bearings (ceramic-on-ceramic or metal-on-metal). Each of these bearing choices has its own advantages and disadvantages (Table 19.4). Metal-on-polyethylene remains the most commonly used bearing surface in THA. Ceramic-on-ceramic bearings produce the lowest wear rates, but overall ceramic-on-polyethylene bearings are associated with the lowest risk of revision.

CEMENTED THA

In general terms the progress made in cemented THA over the past four decades has been in the evolution of surgical technique whereas in uncemented THA there has been much more significant progress in implant design. In cemented THA, fixation is achieved by embedding the implant in polymethylmethacrylate (PMMA) cement. PMMA acts not as a glue but as a grouting material filling the interstices and the technique of 'cementing' an implant has evolved through several generations to its current form.

There are several essential steps in a modern *cementing technique*: the cement is vacuum-mixed in

Table 19.3 Surgical approaches to the hip used to perform THA

Approach	Eponymous name	Intermuscular plane	Internervous plane
Direct anterior	Smith–Petersen	Sartorius and tensor fascia lata	Femoral nerve and superior gluteal nerve
Anterolateral	Watson–Jones	Tensor fascia lata and gluteus medius	Both supplied by superior gluteal nerve
Direct lateral / Transgluteal	Hardinge / Omega	Muscle-splitting approach: gluteus medius	Superior gluteal nerve
Posterolateral	Moore / Southern	Reflection of short external rotators	*No inter-nervous plane*

Table 19.4 Bearing choices in THA

Bearing	Advantages	Disadvantages
Metal on polyethylene (MOP)	Longest track record of bearing surfaces Lowest cost Most modularity	Higher wear and osteolysis rates compared to ceramic Smaller head sizes used reduce wear but increase risk of dislocation Potential for corrosion at modular junctions
Metal on metal (MOM)	Lower wear properties than MOP Larger head sizes allow for increased ROM and decrease dislocation	More expensive than metal-on-polyethylene bearings Adverse reaction to metallic debris (ARMD) Particular concern in pregnant women / renal disease /metal hypersensitivity
Ceramic on ceramic (COC)	Best wear properties of all bearing surfaces Inert particles	Expensive Fractures of liner and/or head due to brittle nature of material Squeaking Less modularity with fewer neck length options
Ceramic on polyethylene (COP)	Potentially better wear profile than MOP Avoids risk of ceramic liner fracture Avoids risk of squeak Minimizes the risk of corrosion at modular junction	Increased cost compared to metal head Risk of head fracture

a sealed system; lavage and brushing of the endosteal surface are carried out to remove any loose cancellous bone; in the femur a distal plug and retrograde insertion of cement is utilized; pressurization of the PMMA in both socket and femur is important to drive cement into the trabecular bone network and achieve the best interdigitation at the bone-cement interface. Hypotensive anaesthesia is of benefit to aid this process to avoid blood compromising the interface, and care should be taken not to move the prosthesis until the cement has hardened.

Cemented cup design has changed very little and essentially involves the insertion of a single-piece ultra-high molecular weight polyethylene (UHMWPE) implant. *Cemented stems* are generally made of either stainless steel or cobalt-chrome. The majority of modern designs are modular, allowing exchange of the head and different materials to be used.

There are two schools of thought regarding the design of cemented stems (Figure 19.48). The 'sit-up and stay' theory applies to the shape-closed implants popularized by *Charnley* that have historically been used in greater numbers. The 'slip and slide' force-closed or taper-slip designs are now the more commonly utilized cemented stem design, proposed initially by Robin Ling and Clive Lee in Exeter. The *Exeter* philosophy, as it is often termed, has demonstrated that a smooth, collarless, highly polished tapered femoral stem will continue settling within the cement mantle even after polymerization, thereby maintaining expansile pressure between cement and bone and thus maintaining fixation. Proponents of cemented stems in particular feel that it confers significant advantages over cementless designs because the fixation achieved is independent of stem size that may allow more accurate recreation of the hip biomechanics. Both designs of cemented stem have demonstrated excellent longevity.

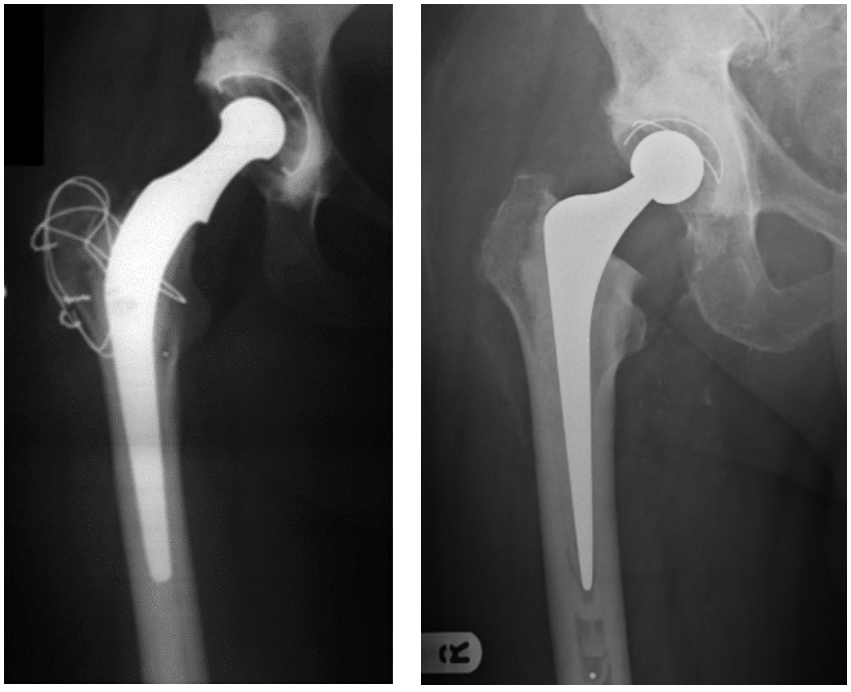
Cement has the potential advantage that antibiotics can be added; indeed, antibiotic-loaded PMMA is used commonly in both the prevention and the treatment of periprosthetic joint infection (see below). In the revision setting, if only minimal damage has occurred to the cement mantle, then another potential advantage is that this can be reutilized in what is termed a 'cement-on-cement' revision, thus reducing the magnitude of the revision procedure.

UNCEMENTED THA

The success of modern uncemented THA owes much to the evolution of successful implant designs. While previously cobalt-chrome was widely used, now the overwhelming majority of both acetabular sockets and femoral stems are made of titanium alloy due to its more favourable biomechanical and biological properties. Both uncemented sockets and stems can be subdivided based on their surface characteristics that permit osseointegration. *Ingrowth uncemented implants* have a porous surface that allows for the ingress of new bone formation; *ongrowth uncemented implants* have a roughened surface that allows bone to grow onto but not into the implant. In both types, these surfaces can be coated, for example with calcium hydroxyapatite to potentially aid osseointegration.

Uncemented acetabular implants

Uncemented acetabular implants generally comprise a modular titanium alloy shell into which a liner is inserted. The



(a) (b)

Figure 19.48 Cemented THA
(a) Charnley cemented THA 'sit-up and stay' philosophy stem design; (b) Exeter cemented THA 'slip and slide' philosophy stem design.

design of the shell can differ in terms of its shape and geometry, which in turn affect the bone preparation of the acetabulum prior to insertion.

Socket preparation is with hemispherical reamers. Acetabular uncemented sockets that are hemispherical in design are inserted with a 1–2 mm press-fit between the size of the acetabular reamer and implant size selected. It is the strain energy created on impaction of the socket that maintains initial stability. Peripherally expanded socket designs are implanted line-to-line and it is the rim fit that provides maximal early stability. In both designs supplemental screw fixation can aid primary stability. Thereafter, secondary stability is achieved once either bone ingrowth or ongrowth has occurred, which is generally achieved within 3 months. The long-term fixation of uncemented implants is dependent upon achieving secondary stability via osseointegration. If initial implant stability is inadequate and there is significant micro-motion at the implant interface, osseointegration will not occur. Both designs of uncemented socket have demonstrated excellent longevity.

Uncemented femoral stems These are most commonly subdivided according to the implant shape and extent of surface of the available for biological fixation. In terms of shape, the now most commonly used designs are tapered (Figure 19.49), other stem shapes include anatomical and cylindrical designs. The stem may be designed for proximal fixation only with surface transformation of the upper part of the prosthesis or the stem extensively coated throughout its length. More modern designs of titanium alloy femoral stems are associated with a reduction in thigh pain and stress-shielding, two complications that were commonly associated with historical cobalt-chrome femoral stems.

HIP RESURFACING

Hip resurfacing (HR) involves the preservation of the femoral head and therefore has the attractive proposition of avoiding a stemmed femoral component. Historically, HR failed due to poor implant materials that utilized a large femoral head on polyethylene which produced high volumetric wear and osteolysis. Modern resurfacing devices have a metal-on-metal articulation (Figure 19.50). Therefore, for younger patients, HR potentially presents a more conservative implant where femoral bone stock is preserved for the future. Other potential advantages include improved restoration of hip biomechanics with lower risk of limb-length discrepancy and ability to engage in high-demand activities.

The past decade, however, has seen the rise and fall of both HR and the use of metal-on-metal THA. The 2008 report of the National Joint Registry of England and Wales recorded that 46% of patients under the age of 55 years undergoing hip arthroplasty received a MOM implant, compared to the 2016 report where less than 1% of all THAs performed received MOM. This dramatic change in practice following early enthusiasm with modern HR devices occurred because of the widespread problems with adverse reactions to metal debris (ARMDs) associated with MOM bearings. Specific MOM implants were either recalled or withdrawn from the marketplace from 2010 onwards because of higher than anticipated failure rates. When ARMD is present due to the local toxicity of cobalt and chromium, it can present in a spectrum of soft-tissue damage ranging from fluid collections to severe muscle and soft-tissue necrosis. The current indications for HR are restricted to male patients under the age of 55 years with favourable anatomy and femoral heads large enough to allow a component size over 50 mm.

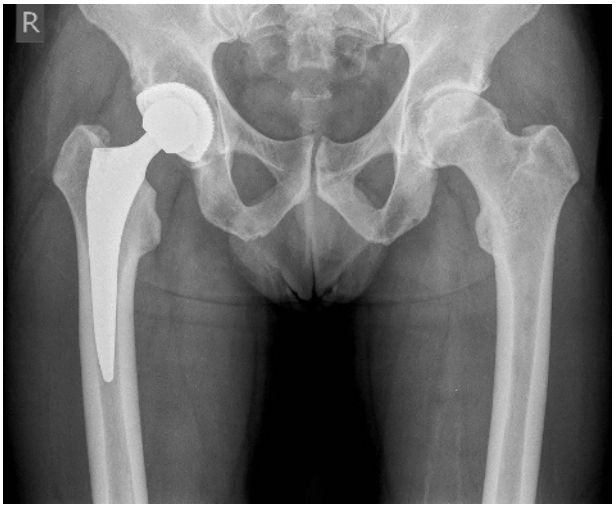


Figure 19.49 Uncemented THA Note the proximally coated tapered stem using a ceramic-on-polyethylene bearing.

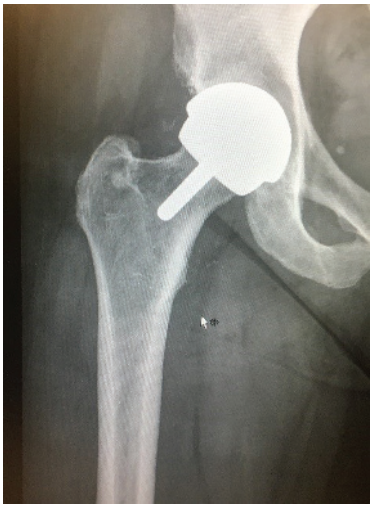


Figure 19.50 Hip resurfacing arthroplasty This utilizes an uncemented acetabular socket and a large diameter metal-on-metal bearing.

Rehabilitation following THA

In recent years the inpatient stay following THA has reduced dramatically. Enhanced recovery protocols are commonplace and in both North America and mainland Europe the concept of day-case THA has become a reality in certain centres. The mean length of inpatient stay has been reduced to less than 3 days in most hospitals. Early mobilization not only aids discharge but reduces the risk of venous thromboembolism. Patients mobilize on crutches at time of discharge and will have negotiated stairs independently. In general, progress to full weight-bearing without support will usually take 4–6 weeks at the patient's own pace.

Complications

As for any surgical procedure, complications can be considered as immediate, early or late, or those specific to the procedure itself and those that can occur following surgery in general (Box 19.10). THA is often performed on older patients with significant comorbidities. These may, of course, have a significant effect on perioperative risk, for example diabetes, where optimal blood sugar control is imperative to reduce perioperative morbidity and reduce the risk of early and late periprosthetic joint infection (PJI). Examples of procedure-specific factors that increase the risk of complications include previous hip operations, severe deformity, osteoporosis and lack of preoperative planning.

BOX 19.10 POTENTIAL COMPLICATIONS FOLLOWING THA

Immediate

- Anaesthetic-related (e.g. airway problems)
- Anaphylaxis (e.g. to antibiotic prophylaxis or anaesthetic agents/drugs)
- Blood loss and haemorrhage
- Fracture and/or perforation
- Nerve injury
- Leg-length discrepancy

Early

- Venous thromboembolism (DVT and PE)
- Urinary retention
- Cardio / respiratory issues
- Poor wound healing
- Early periprosthetic joint infection (PJI)
- Dislocation
- Fracture

Late

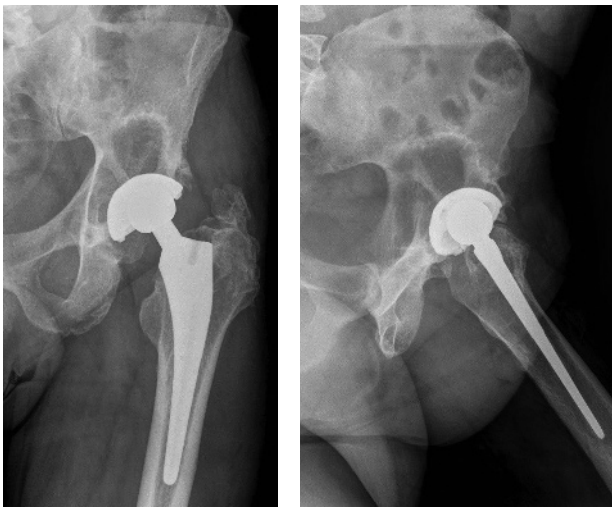
- Wear and osteolysis
- Aseptic implant loosening
- Dislocation and recurrent instability
- Late periprosthetic infection
- Periprosthetic fracture
- Heterotopic bone formation

Immediate *During surgery itself* complications can occur that may be related to the anaesthetic or the surgical procedure itself. Neurovascular injury is an inherent risk in the majority of musculoskeletal surgery and equally applies to THA where the sciatic nerve is the most commonly affected. Intraoperative technical issues or misadventure can be minimized by appropriate preoperative planning and implant selection.

Early *Within the first 6 weeks of surgery* venous thromboembolism is a major early concern following THA. All patients should undergo an individual risk assessment, and the cornerstones of prevention are early mobilization, together with the use of mechanical and chemical prophylaxis. If early THA dislocation occurs, the surgeon should be alerted with regard to component malposition. Early fracture is more common following the use of an uncemented femoral stem and in conditions that affect bone density such as osteoporosis. Infection is the most serious postoperative complication and can occur either early or late and is considered in more detail below.

Late *Beyond 6 weeks for the lifetime of the implant.* Heterotopic bone formation following THA is relatively common and is seen in about 20% of patients 5 years after joint replacement. For the large majority it is of little concern and represents just a small island of bone picked up as an incidental finding on a follow-up radiograph. Infrequently and in severe cases heterotopic ossification (HO) can present with pain and stiffness. While the underlying cause is unknown, patients with diffuse idiopathic skeletal hyperostosis (DISH) and ankylosing spondylitis are particularly at risk. Prophylaxis to prevent HO should be considered in high-risk patients by giving either a course of non-steroidal anti-inflammatory drugs or a single dose of irradiation to the hip.

In the longer term aseptic loosening either of the acetabular socket or the femoral stem is the commonest cause of failure and is the most common reason for revision THA. With modern implant fixation aseptic loosening rates are generally less than 10% at 15 years after operation. Patients with aseptic loosening may present with 'start-up pain', a term used to describe pain in the groin or upper thigh that starts when first weight-bearing on the leg after sitting or lying. The diagnosis is based on the X-ray signs of a progressive radiolucency around the implant, fracturing of cement or change in position of the implant. Radionuclide



(a) (b)

Figure 19.51 Osteolysis Severe periacetabular osteolysis and symmetrical wear are visible where the femoral head does not sit centrally in the socket.

scanning can also show increased activity around components where fixation is compromised.

Osteolysis is the biological consequence of wear on the bearing surface. The wear particles that are produced interact with active macrophages, which results in cytokine release and the subsequent resorption of periprosthetic bone in the effective joint space. It is associated with granuloma formation at the interface between implant and bone. Importantly, osteolysis can be asymptomatic and associated with or without aseptic loosening. Osteolysis is diagnosed on X-rays as lucent areas adjacent to implants (Figure 19.51). Modern bearing surfaces, in particular the widely used highly cross-linked ultra-high molecular weight polyethylene (UHMWPE), have reduced wear rates and osteolysis.

Dislocation and subluxation are related to implant design, patient factors such as cognitive impairment and neurological disease, and surgeon factors including surgical approach and component orientation. The integrity of the soft tissue, especially abductor muscle deficiency, can risk recurrent instability.

Periprosthetic fracture can occur immediately, early and later following THA. Treatment is dependent on the location of the fracture and its effect on implant fixation. When implant fixation is unaffected, internal fixation is most commonly performed, but when the fracture compromises the implant fixation then revision THA is performed (Figure 19.52).

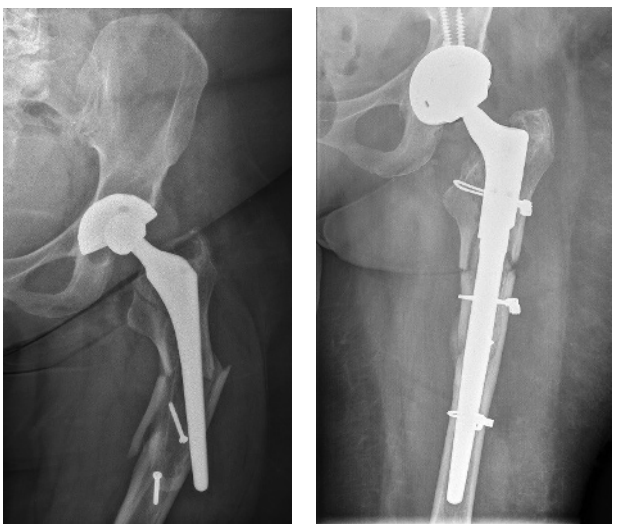
PERIPROSTHETIC JOINT INFECTION (PJI)

PJI represents the most serious complication of THA, other than mortality. Infection results in pain and poor quality of life and, if untreated, it can progress to cause major disability, amputation and death. The incidence of deep infection necessitating revision surgery is currently 1–2%. PJI following THA can occur early or late, with late infection also associated with haematogenous spread to THA from a distant anatomical site.

Prevention of PJI is integrated into the care pathway of patients undergoing THA. *Preoperatively* this starts at the time of pre-assessment where modifiable risk factors are improved. For example, good dental hygiene, no areas of chronic broken skin or ulceration and optimization of medical comorbidities (e.g. ensure in diabetic patients HbA1c is less than 7.5%) should be ensured. Preoperative screening of patients for MRSA is commonplace as well as a growing role for methicillin-sensitive *Staphylococcus aureus* (MSSA) screening in high-risk patients.

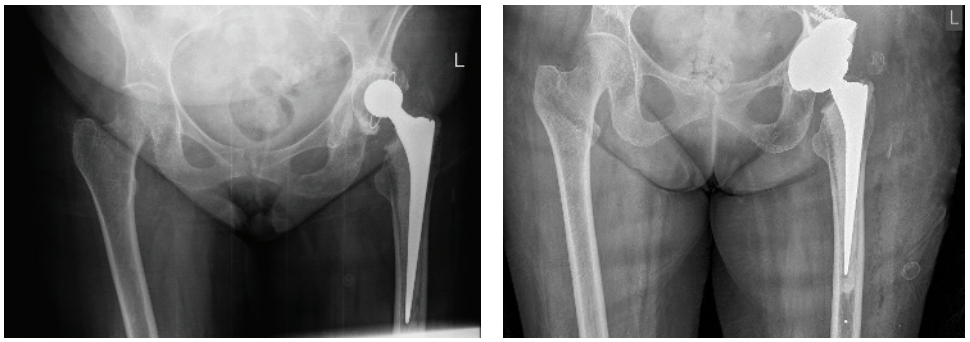
Intraoperative prevention of PJI is focused on trying to prevent bacterial contamination that occurs when colony-forming units commonly found in circulating shed skin cells land in the surgical wound. Implant surgery is best performed in specific operating theatres with ultra-clean laminar airflow, strict aseptic technique and reduced operating room personnel and theatre traffic, along with the use of specific surgical hoods and gowns. The anaesthetist plays an important role in the prevention of PJI. Prophylactic antibiotics should be given prior to the induction of anaesthetic and intraoperatively fluid balance should be optimized and patient-warming methods employed. Good surgical technique is also important, with blood management regimes that often include the use of tranexamic acid significantly reducing the risk of blood transfusion.

Postoperatively good wound care, pain management and nutrition together with standardized protocols to minimize postoperative complications such as chest infections can also help prevent PJI.



(a) (b)

Figure 19.52 Revision THA Periprosthetic fracture with stem loosening (a) treated by revision THA using long uncemented tapered stem that bypasses the zone of injury (b).



(a) (b)

Figure 19.53 Revision THA (a) Aseptic loosening of a cemented acetabular socket with proximal migration and loss of superior bone stock. (b) Following revision THA using high porous metal augment and uncemented hemispherical acetabular socket. Note the restoration of the hip centre of rotation.

The *surgical treatment* of hip PJI involves debridement and implant exchange that can be accomplished in a single operation or more commonly as a two-stage procedure. These procedures are associated with significant patient morbidity and indeed mortality. Failure to eradicate infection is a contraindication to reimplantation of a hip prosthesis and in such circumstances an excision arthroplasty is a salvage procedure.

Contraindications

Active infection is a key important contraindication to THA since it significantly increases the risk of PJI with the potential dire consequences described above. Patients who are non-ambulant or have severe neurological problems such as post-neurological injury may not be good candidates for THA. Similarly, patients with severe cognitive impairment or dementia need very careful consideration prior to THA as very marginal gains may well be outweighed by major surgical risks.

Revision THA

The 13th National Joint Registry reported that there were over 8300 revision THA procedures performed in England and Wales in 2015. Revision THA has changed dramatically over the past two decades and has evolved from a procedure that had a very limited armamentarium to a much more reproducible procedure with a wide range of strategies to deal with various degrees of surgical complexity. Nevertheless, an overriding rule of thumb is that, with each revision THA procedure that a patient undergoes, the functional outcome diminishes and the risk of complications increases.

The main reasons for revision surgery are loosening, infection and dislocation. Revision implants more commonly employ cementless fixation and specific techniques are employed to restore missing bone stock. This can involve the use of bone graft, which can be in the form of bone chips and impacted (impaction bone grafting) or structural bone grafts. Contemporary revision THA practice more commonly uses highly porous metal augments (Figure 19.53) in cases of severe bone loss, and promising medium-term results over the past decade have been reported.

NOTE: Probably the most important factor in determining the successful outcome after hip replacement is good and informed decision making.

The knee

20

Andrew Price, Nick Bottomley & William Jackson

CLINICAL ASSESSMENT

Symptoms

Pain, with acute or gradual onset, is the most common knee symptom. With inflammatory or degenerative disorders it is usually diffuse, but with mechanical disorders (and especially after injury) it is often localized and the patient is more likely to point to a specific region of the knee.

If the patient can describe the mechanism of the injury, this is extremely useful: a direct blow to the front of the knee may damage the patellofemoral joint or the posterior cruciate ligament; a blow to the side may rupture the collateral ligament; twisting injuries are more likely to cause a torn meniscus or a cruciate ligament rupture.

Swelling may be diffuse or localized. If there was an injury, it is important to ask whether the swelling appeared immediately (suggesting a haemarthrosis) or only after some hours (typical of a torn meniscus or articular cartilage injury). A complaint of recurrent swelling, with more or less normal periods in between, suggests a long-standing internal derangement – possibly an old meniscal tear, degeneration of the meniscus, a small osteoarticular fracture, loose bodies in the joint or early osteoarthritic change. Chronic swelling is typical of synovitis or more established arthritis.

A small, localized swelling on the anteromedial or anterolateral side of the joint makes one think of a cyst of the meniscus or possibly a loose body. Swelling over the front of the knee could be due to a pre-patellar bursitis; a localized bulge in the popliteal fossa can also be caused by a bursal swelling, but is more often due to ballooning of the synovial membrane and capsule at the back of the joint. However, tumour (benign or malignant) can occur around the knee and is a differential diagnosis until proven otherwise.

Stiffness is a common complaint, but it must be distinguished from inhibition of movement due to pain, or simple weakness of the extensor apparatus. Particularly characteristic is stiffness that appears regularly after periods of rest – so-called 'post-inactivity stiffness' – sometimes called gelling, which suggests some type of chronic arthritis, commonly osteoarthritis.

Locking is different from stiffness. The knee, quite suddenly, cannot be straightened fully, although flexion is still possible. This happens when a torn meniscus or loose body is caught between the articular surfaces. By gently moving the knee around, the patient may be able to 'unlock' it; sudden *unlocking* is the most reliable evidence that something mobile had previously obstructed full extension. Do not be misled by 'pseudo-locking', when movement, usually flexion, is suddenly stopped by pain or the fear of impending pain.

Deformity is seldom a leading symptom; patients are not keen to admit to having 'knock knees' or 'bandy legs'. However, a unilateral deformity, especially if it is progressive, will be more worrying. Some patients may not have noticed slowly developing deformity (such as genu varus) but it is reported by partners or other close relatives.

Giving way, a feeling of instability, or a lack of trust in the knee are a set of symptoms that suggest a mechanical disorder caused by ligamentous, meniscal or capsular injury, or simple muscle weakness. However, giving way may also be due to painful arthrosis in the knee. Excessive use of an unstable knee produces post-exercise swelling (effusion or haemarthrosis) and diffuse pain within the joint.

Limp may be due to pain, instability or deformity. An antalgic gait may also be caused by pain referred from the hip.

Loss of function manifests as a progressively diminishing walking distance, inability to run and difficulty going up and down steps. Squatting or kneeling may be painful, either because of pressure on the patellofemoral joint or because the knee cannot flex fully. More generally the patient may report

a reduction in participation in activities that are important to them and a reduction in their overall quality of life.

Signs with the patient upright

For the examination, both lower limbs must be exposed from groin to toe; a mere hitching up of the skirt or rolling back of a trouser leg is unlikely to expose the limb adequately.

Deformity (valgus or varus or fixed-flexion or hyperextension) can often be best seen with the patient standing and attempting to bear weight symmetrically with their legs together (if possible!). Normally the knees and ankles can touch in the midline; this means that the knees must be in slight valgus (typically about 7 degrees in women and 5 degrees in men), because the hips are wider than the knees. *Genu valgum* and *genu varum* are determined in relation to this normal anatomical alignment. But look carefully to see whether the deformity is really in the knee (often a sign of arthritis) and not in the lower end of the femur (a previous fracture or a bone tumour) or the upper end of the tibia (such as a malunited fracture or Paget's disease – see Figure 20.1e).

Alignment of the extensor mechanism (quadriceps, patella and patellar ligament) can also be measured with the patient standing but is probably more conveniently done with the patient seated where the relative orientation of the patella and quadriceps tendons can be established (see below).

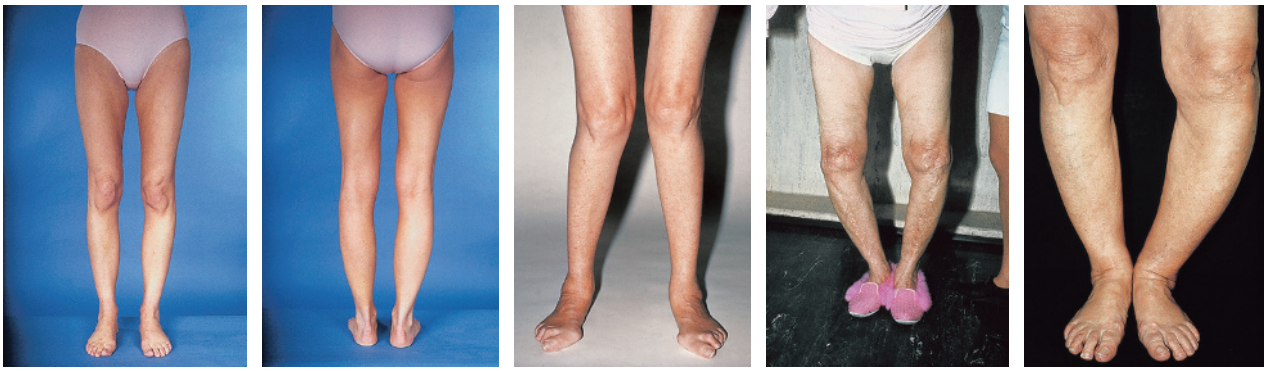
Gait is important; the patient should also be observed walking with and without any support such as a stick or crutch. In the *stance phase* note whether the knee extends fully (is there a fixed flexion deformity or a hyperextension deformity?) and see if there is any lateral or medial thrust signifying instability. In the *swing phase* note whether the knee moves freely or is held in one position – usually because the joint is painful but perhaps because it really is ankylosed! When the patient walks, is there any sign of a limp? And if so, does it stem from the knee? Or perhaps the hip, or the foot? Foot drop caused by common peroneal injury around the knee can be identified during the gait cycle.

Signs with the patient sitting

With the patient sitting upright on the examination couch with their legs dangling over the edge, the outlines of the patellae and patellar ligaments, as well as the general shape and symmetry of the two knees and the tibial tubercles, can be made out quite easily. With the knees at 90 degrees of flexion, the patellae should be facing straight forwards; note if they appear to be seated higher than usual (*patella alta*) or lower than usual (*patella baja*). Patella alta is believed to be associated with a higher than normal incidence of chondromalacia patellae.

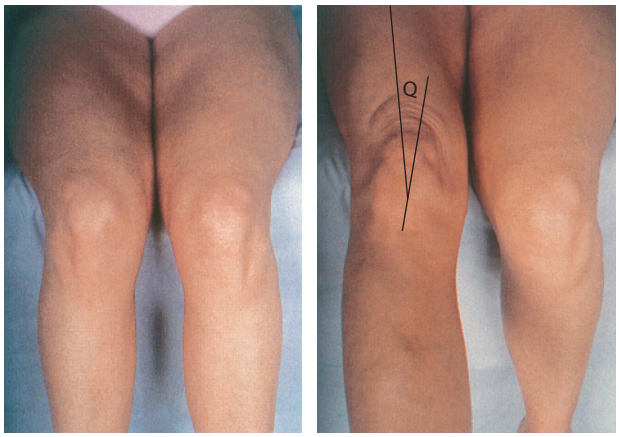
Next, ask the patient to straighten each knee in turn and observe how the patella moves upwards. Does it remain centred over the femoral condyles or does it veer off towards one side in the early phase of knee extension and then slide back to the centre with full extension – suggesting a tendency to subluxation?

Patellar alignment can also be assessed by measuring the *Q-angle* (quadriceps angle). This is the angle subtended by a line drawn from the anterior superior iliac spine to the centre of the patella and another from the centre of the patella to the tibial tubercle (Figure 20.2b); it normally averages about 14 degrees in men and 17 degrees in women. An increased Q-angle is regarded as a predisposing factor in the development of chondromalacia; however, small variations from the norm are not a reliable indicator of future pathology.



(a) (b) (c) (d) (e)

Figure 20.1 Examination standing (a,b) Look at the general shape and posture, first from in front and then from behind. Normally the knees are in slight valgus. Look for swelling of the joint or wasting of the thigh muscles; quadriceps wasting occurs very quickly. (c) This patient has rheumatoid arthritis and bilateral valgus deformities; in contrast, osteoarthritis is likely to lead to varus deformities (d). Unilateral deformity is easier to notice and almost always pathological – this man has Paget's disease of the tibia (e).



(a) (b)

Figure 20.2 Examination with the patient sitting
The two knees are compared for shape and symmetry. Note the position of the patellae (a) in relaxation; (b) in full extension and by measuring the Q-angle.

Signs with the patient lying supine

The knees are the most visible and accessible of all the large joints; with the legs lying side by side, features on one side can be constantly compared with those on the other.

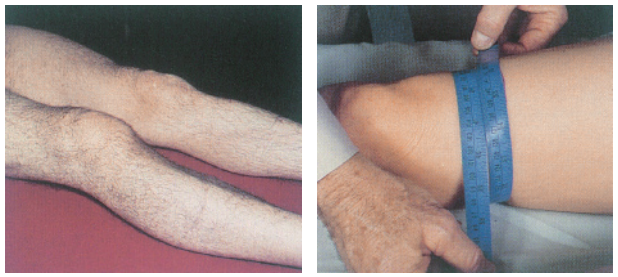
Look

The first things that strike one are the *position of the knee*. Is it symmetrical with the normal side? Is it held in valgus or varus, incompletely extended, or hyperextended? Note also the presence of *swelling*, either of the joint as a whole or as *lumps or bumps* in localized areas.

Wasting of the quadriceps is a sure sign of joint disorder (Figure 20.3). The visual impression obtained as a patient attempts to contract the muscle group by pressing the knee into the couch can be checked by objectively *measuring* the girth of the thigh at the same level (e.g. a fixed distance above the joint line or a hand's breadth above the patella) in each limb.

Look more closely for signs of *bruising*, and for *old scars or sinuses*, signifying previous infection or operations.

Take note of the *shape and position of the patella*, both with the knee at rest and during movement. Always compare the symptomatic with the normal side.



(a) (b)

Figure 20.3 Examination with the patient supine (a) Wasting of the quadriceps occurs rapidly after any internal derangement of the knee. (b) The girth is measured at the same level in both limbs, about a hand's breadth above the patella.

Feel

As with all joints, palpation of the knee – if it is to be rewarding – demands a sound knowledge of the local anatomy.

Start by running your hand down the length of the limb, feeling for changes in *skin temperature* and comparing the symptomatic with the normal side. There is normally a gradual decrease in skin temperature from proximal to distal. Increased warmth over the knee signifies increased vascularity, usually due to inflammation.

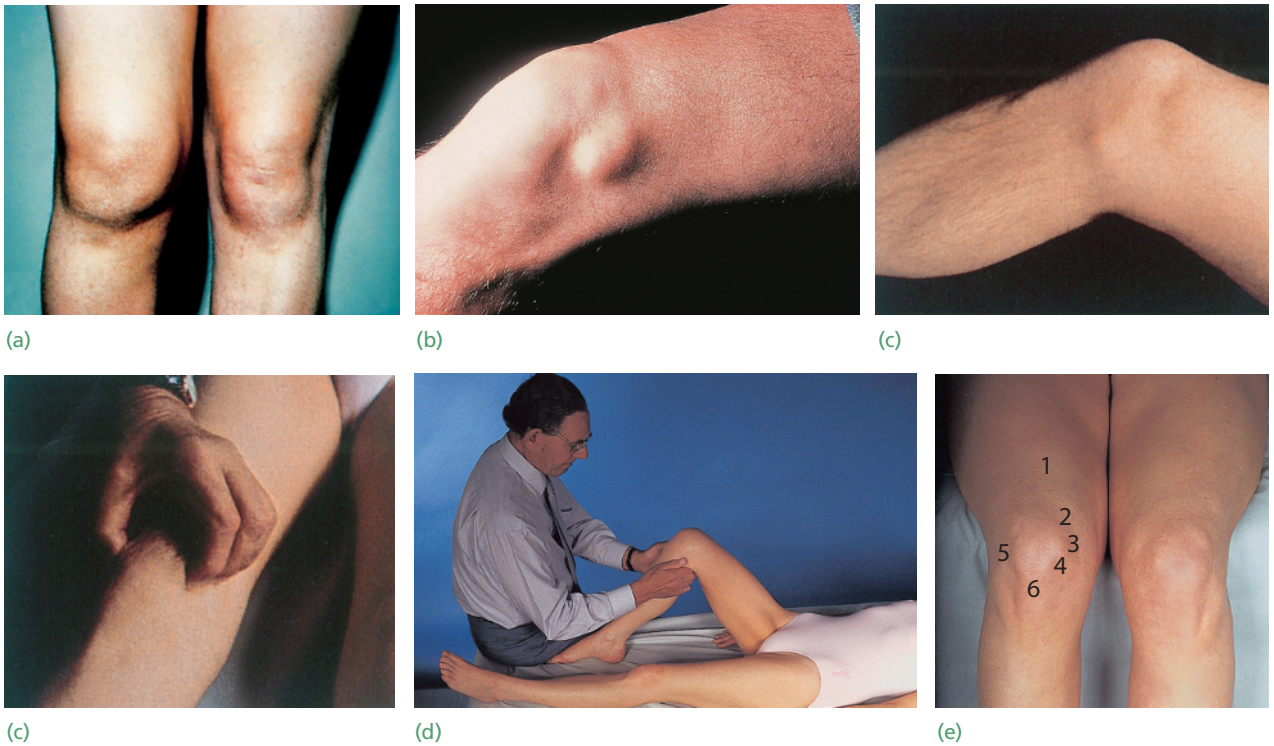
The *soft tissues and bony outlines* are then palpated systematically, feeling for abnormal outlines and localized tenderness. This is easier if the joint is flexed and the examiner sits on the edge of the couch facing the knee (Figure 20.4). By placing both hands over the front of the knee, the outlines of the joint margins, the patellar ligament, the collateral ligaments, the iliotibial band and the pes anserinus are then easily traced with the fingers. The point of maximum tenderness will suggest at least the anatomical site of pathology if not the precise diagnosis.

Synovial thickening is best appreciated as follows: placing the knee in extension, the examiner grasps the edges of the patella in a pincer made of the thumb and middle finger, and tries to lift the patella forwards; normally the bone can be grasped quite firmly, but if the synovium is thickened, the fingers simply slip off the edges of the patella. In a monoarthropathy the suprapatellar pouch can demonstrate a boggy swelling of the synovium.

Move

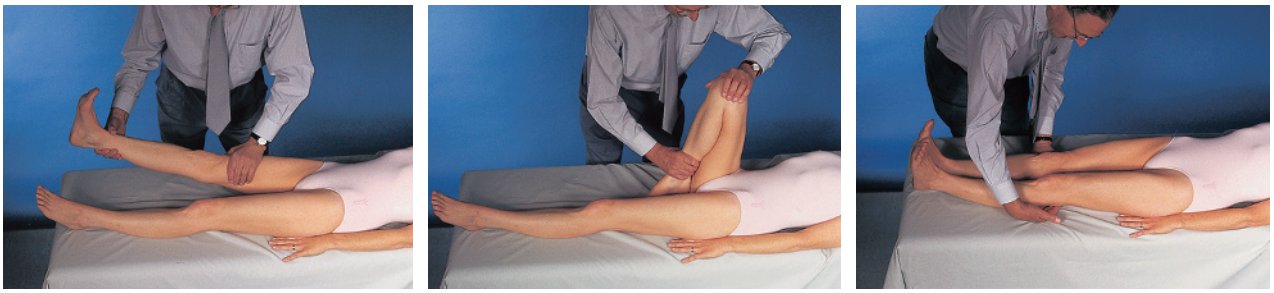
Passive extension can be tested by the examiner simply holding both legs by the ankles and lifting them off the couch; the knees should straighten fully (or even into a few degrees of hyperextension) and symmetrically.

Active extension can be roughly tested by the examiner slipping a hand under each knee and then asking the patient to force the knees into the surface of the couch; it is usually easy to feel whether the hands are trapped equally strongly on the two sides (Figure 20.5c). Another way is to have the patient sitting on the edge of the couch with his or



(a) (b) (c) (c) (d) (e)

Figure 20.4 Examination with the patient supine Swelling may involve either the whole joint, as in patient (a) with acute synovitis, or may be due to some localized lesion as in patient (b) with a large loose body in the joint and patient (c) with a small meniscal cyst protruding at the medial joint line. (d) Feeling for synovial swelling: try to 'lift' the patella – if the synovium is thickened your fingers will slip off the edges. (e) Feeling for tenderness: sit facing the patient's knee and try to identify the exact site of pain/tenderness. A superficial 'map' is shown in (f): 1 quadriceps tendon; 2 edge of patella; 3 medial collateral ligament; 4 joint line; 5 lateral collateral ligament; 6 patellar ligament.



(a) (b) (c)

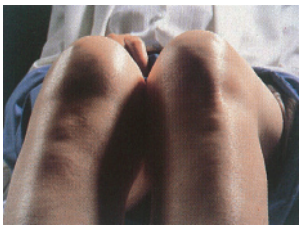
Figure 20.5 Movement The knee should move from full extension (a) through a range of 150° to full flexion (b). Small degrees of flexion deformity (loss of full extension) can be detected by placing the hands under the knees while the patient forces the legs down on the couch (c); if your hand can be extracted more easily on one side than the other, this indicates loss of the final few degrees of complete extension.

her legs hanging over the side and then asking them to extend each knee as far as possible; the test can be repeated with the patient extending the knees against resistance.

Passive and *active flexion* are tested with the patient lying supine. Normally the heel can be pulled up close to the buttock, with the knee moving through a range of 0–150 degrees. The 'heel-to-buttock' distance is compared on the two sides.

Internal and *external rotation*, though normally no more than about 10 degrees, should also be assessed. The patient's hip and knee are flexed to 90 degrees; one hand steadies and feels the knee, the other rotates the foot.

Crepitus during movement may be felt with a hand placed on the front of the knee. It usually signifies patellofemoral roughness.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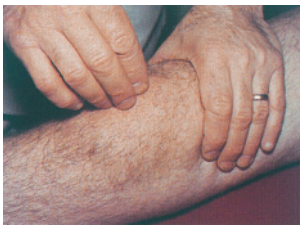
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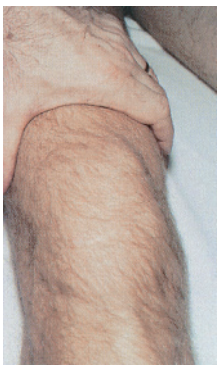
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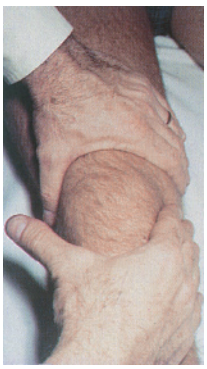
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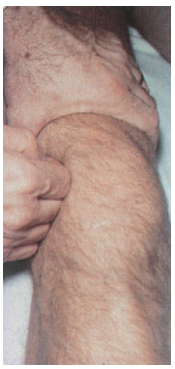

(e)

Figure 20.6 Testing for intra-articular fluid (a) The juxtapatellar hollow, which disappears in flexion if there is fluid in the knee. (b) Patellar tap test. (c,d,e) Doing the bulge test: compress the suprapatellar pouch (c), empty the medial compartment (d), push fluid back from the lateral compartment and watch for the bulge on the medial side (e).

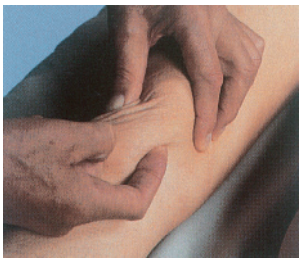
Tests for intra-articular fluid (Figure 20.6)

Cross fluctuation This test is applicable only if there is a large effusion. The left hand compresses and empties the suprapatellar pouch while the right hand straddles the front of the joint below the patella; by squeezing with each hand alternately, a fluid impulse as synovial fluid moves between compartments is transmitted across the joint.

The patellar tap test In a patient with a large effusion the suprapatellar pouch is compressed with the left hand to squeeze any fluid from the pouch into the joint. With the other hand the patella is then tapped sharply backwards onto the femoral condyles. In a positive test the patella can be felt striking the femur and bouncing off again (a type of ballottement).

The bulge test This is a useful method of testing when there is very little fluid in the joint, though it takes some practice to get it right! After squeezing any fluid out of the suprapatellar pouch, the medial compartment is emptied by pressing on the inner aspect of the joint; that hand is then lifted away and the lateral side is sharply compressed – a distinct ripple is seen on the flattened medial surface as fluid is shunted across.

The juxtapatellar hollow test Normally, when the knee is flexed, a hollow appears lateral to the patellar ligament and disappears with further flexion; if there is excess fluid, the hollow fills and disappears at a lesser angle of flexion. Comparison between knees is advised at all times when examining for an effusion.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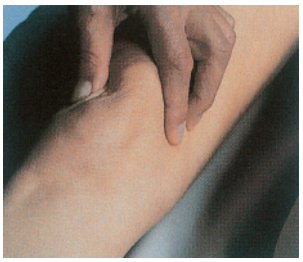
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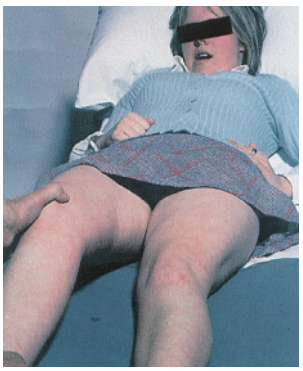

(c)

Figure 20.7 Patellofemoral joint (a) Feeling under the edge of the patella. (b) Testing for patellofemoral tenderness. (c) The patellar apprehension test.

The patellofemoral joint

The *size, shape and position* of the patella are noted. The bone is felt, first on its anterior surface and then along its edges and at the attachments of the quadriceps tendon and the patellar ligament. Much of the posterior surface, too, is accessible to palpation if the patella is pushed first to one side and then to the other; tenderness suggests synovial irritation or articular cartilage softening (Figure 20.7).

Moving the patella up and down while pressing it lightly against the femur (*the 'friction test'*) causes painful grating if the central portion of the articular cartilage is damaged.

The '*patellar apprehension test*', performed by pressing the patella laterally with the thumb while flexing the knee slowly, may induce anxiety and sharp resistance to further movement; it is diagnostic of recurrent patellar subluxation or dislocation.

Tests for stability

Collateral ligaments The medial and lateral ligaments are tested by stressing the knee into valgus and varus: this is best done by tucking the patient's foot under your arm and holding the extended knee firmly with one hand on each side of the joint; the leg is then angulated alternately towards abduction and adduction (Figure 20.8).

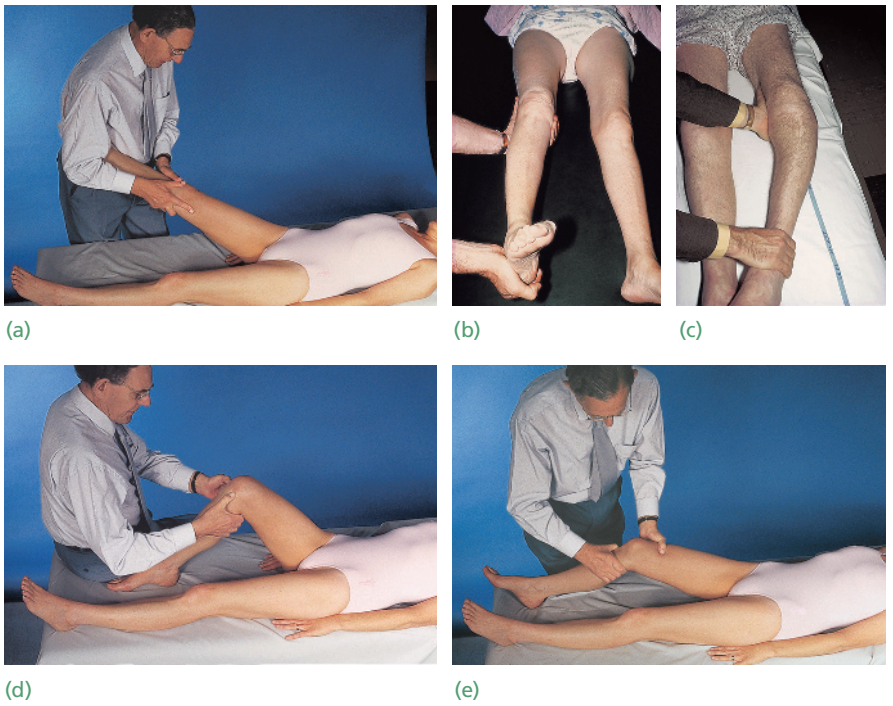


Figure 20.8 Testing for instability There are two ways of testing the collateral ligaments (side-to-side stability): (a) by gripping the foot close to your body and guiding the knee alternately towards valgus and varus; (b) by gripping the femoral condyles (provided your hand is big enough) and then forcing the leg alternately into valgus and varus. (c) In this case there was gross instability on the lateral side, allowing the knee to be pulled into marked varus. Cruciate ligament instability can be assessed by either the drawer test (d) or the Lachman test (e), as described in the text.

The test is performed at full extension and again at 30 degrees of flexion. There is normally some medial–lateral movement at 30 degrees, but if this is excessive (compared to the normal side) it suggests a damaged collateral ligament. Sideways movement in full extension is always abnormal; it may be due to either torn or stretched ligaments and capsule or loss of articular cartilage or bone, which allows the affected compartment to collapse.

Cruciate ligaments Routine tests for cruciate ligament stability are based on examining for abnormal gliding movements in the sagittal plane. With both knees flexed 90 degrees and the feet resting on the couch, the upper tibia is inspected from the side; if its upper end has dropped back, or can be gently pushed back, this indicates a tear of the posterior cruciate ligament (the 'sag sign'). With the knee in the same position, the foot is anchored by the examiner sitting on it (provided this is not painful); then, using both hands, the upper end of the tibia is grasped firmly and rocked backwards and forwards to see if there is any anteroposterior glide (the '*drawer test*'). Excessive anterior movement (a positive anterior drawer sign) denotes anterior cruciate laxity; excessive posterior movement (a positive posterior drawer sign) signifies posterior cruciate laxity.

More sensitive is the *Lachman test*, where the patient's knee is flexed 20 degrees; with one hand grasping the lower thigh and the other the upper part of the leg, the joint surfaces are shifted backwards and forwards upon each other. Abnormal movement suggests an anterior cruciate ligament (ACL) injury. If the knee is stable, there should be no gliding. In both the drawer test and the Lachman test, note whether the endpoint of abnormal movement is 'soft' or 'hard'.

Complex ligament injuries When only a collateral or cruciate ligament is damaged, the diagnosis is relatively easy: the direction of unstable movement is either sideways or front-to-back. With combined injuries the direction of instability may be oblique or rotational. Special clinical tests have been developed to detect these abnormalities (see Chapter 31); the best known is the *pivot shift test*. The patient lies supine with the lower limb completely relaxed. The examiner lifts the leg with the knee held in full extension and the tibia internally rotated (the position of slight rotational subluxation). A valgus force is then applied to the lateral side of the joint as the knee is flexed; a sudden posterior movement of the tibia is seen and felt as the joint is fully relocated. The test is sometimes quite painful and is generally best performed with the patient under general anaesthesia.

Tests for meniscal injuries

McMurray's test This classic test for a torn meniscus is seldom used now that the diagnosis can easily be made by MRI. However, advanced imaging is not always available and the clinical test has not been altogether discarded.

The test is based on the fact that the loose meniscal tag can sometimes be trapped between the articular surfaces and then induced to snap free with a palpable and audible click. The knee is flexed as far as possible; one hand steadies the joint and the other rotates the leg medially and laterally while the knee

Figure 20.9 Meniscal injury – Thessaly test Picture showing how the patient is positioned during the Thessaly test.

is slowly extended. The test is repeated several times, with the knee stressed in valgus or varus, feeling and listening for the click.

A positive test is helpful but not pathognomonic; a negative test does not exclude a tear. It has been shown to not be a specific or sensitive test.

Thessaly test This test is based on a dynamic reproduction of load transmission in the knee joint under normal or trauma conditions. With the affected knee flexed to 20 degrees and the foot placed flat on the ground, the patient takes his or her full weight on that leg while being supported (for balance) by the examiner (Figure 20.9). The patient is then instructed to twist his or her body to one side and then to the other three times (thus, with each turn, exerting a rotational force in the knee) while keeping the knee flexed at 20 degrees. Patients with meniscal tears experience medial or lateral joint line pain and may have a sense of locking. The test has shown a high diagnostic accuracy rate at the level of 95% in detecting meniscal tears, with a low number of false positive and negative recordings. A simple version of this test in the 'squat test' where a patient performs a deep squat and this reproduces a mechanical pain in the affected knee.

Signs with the patient lying prone

Scars or lumps in the popliteal fossa are noted. If there is a swelling, is it in the midline (most likely a bulging capsule) or to one side (possibly a bursa)? A semimembranous bursa is usually just above the joint line, a Baker's cyst below it.

The popliteal fossa is carefully palpated. If there is a lump, where does it originate? Does it pulsate? Can it be emptied into the joint?

Apley's test With the patient prone, the knee is flexed to 90 degrees and rotated while a compression force is applied; this, the *grinding test*, reproduces symptoms if a meniscus is torn. Rotation is then repeated while the leg is pulled upwards with the surgeon's knee holding the thigh down; this, the *distraction test*, produces increased pain only if there is ligament damage.

Lachman test The Lachman test can be readily performed with the patient prone.

Imaging

X-rays

Anteroposterior and lateral views are routine; it is often useful also to obtain tangential ('skyline') patellofemoral views and intercondylar (or tunnel) views. *The anteroposterior view should always be taken with the patient standing* (Figure 20.10); unless the femorotibial compartment is loaded, narrowing of the articular space may be missed. Both knees should be X-rayed, so as to compare the abnormal with the normal side. A Rosenberg view is used to specifically assess the lateral compartment.

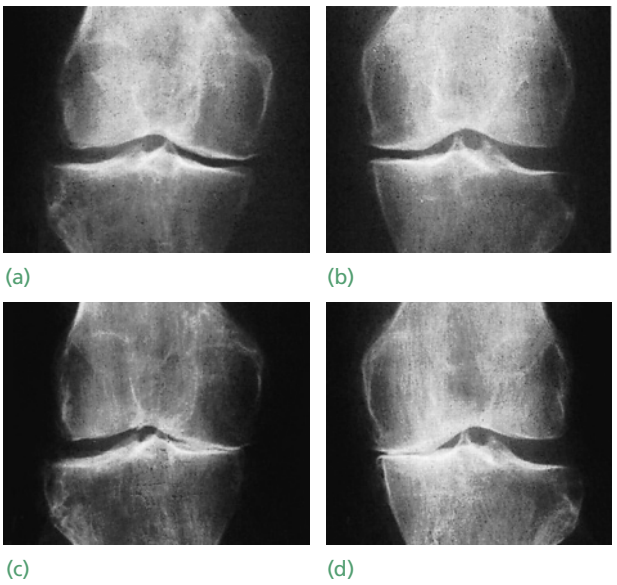


Figure 20.10 X-rays Anteroposterior views should always be taken with the patient standing. (a,b) X-rays with the patient lying down show only slight narrowing of the medial joint space on each side; but with weight-bearing (c,d) it is clear that these changes are much more marked.

Tibiofemoral alignment can be measured on full-length standing views. Normal indices have also been established for patellar height and patellofemoral congruence. These features are discussed in the relevant sections of the chapter. If available, previous X-ray imaging should always be used for comparison to determine progression of changes (typically osteoarthritic) in the knee.

Other forms of imaging

Ultrasound imaging has become an important mode of investigating a wide range of musculoskeletal conditions (e.g. guiding knee joint aspiration or synovial biopsy, and identifying tendon pathology in the knee).

Radioscintigraphy is also helpful in showing 'hot spots' due to the spread of malignancy or loosening of components after joint replacement.

CT is useful for showing patellofemoral congruence at various angles of flexion and for detailed understanding of 3D bone structure.

MRI provides a reliable means of diagnosing injury or damage to the soft tissues of the knee, such as meniscal tears or cruciate ligament injuries in the knee (Figure 20.11). It is also helpful in identifying the early stages of osteoarthritis within the knee and osteonecrosis of the femoral or tibial condyles. In addition, MRI scans are an essential part of the investigation of musculoskeletal tumours.

PET scans are a useful way of imaging musculoskeletal tumours.

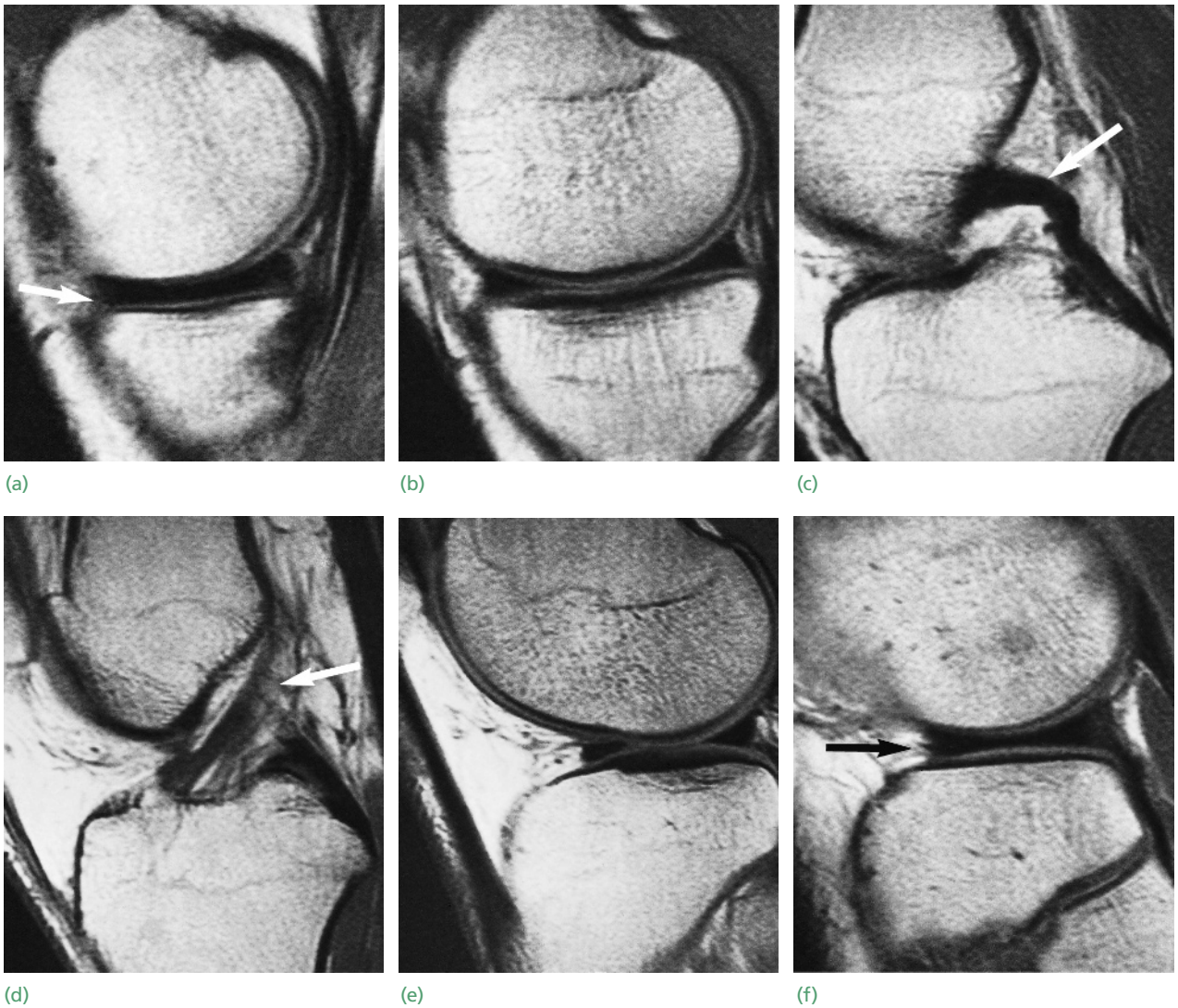


Figure 20.11 MRI A series of sagittal T1 weighted images proceeding from medial to lateral show the normal appearances of (a,b) the medial meniscus; (c) the posterior cruciate ligament; (d) the somewhat fan-shaped anterior cruciate ligament and (e,f) the lateral meniscus.

ARTHROSCOPY

With the development of MRI technology, arthroscopy is much less often used as a diagnostic tool (Figure 20.12). However, in certain situations arthroscopy can be used effectively in this way:

1 to establish or refine the accuracy of diagnosis (e.g. to locate and perform excision biopsy of a pigmented villonodular synovitis (PVNS) lesion in the knee)
2 to help in deciding whether to operate, or to plan the operative approach with more precision (e.g. to determine the degree of articular cartilage damage in planning osteotomy around the knee)
3 to confirm diagnosis and treat certain joint conditions with specific operative procedures (e.g. repair or excision of a bucket-handle tear within a locked knee joint or assessment and repair of a rotator cuff tear).

There is increasing evidence to suggest that in certain conditions arthroscopic intervention is less effective than previously thought. For example, washout of the osteoarthritic knee and meniscectomy in the degenerate knee, without the presence of mechanical symptoms, are both procedures where surgery may not be effective.

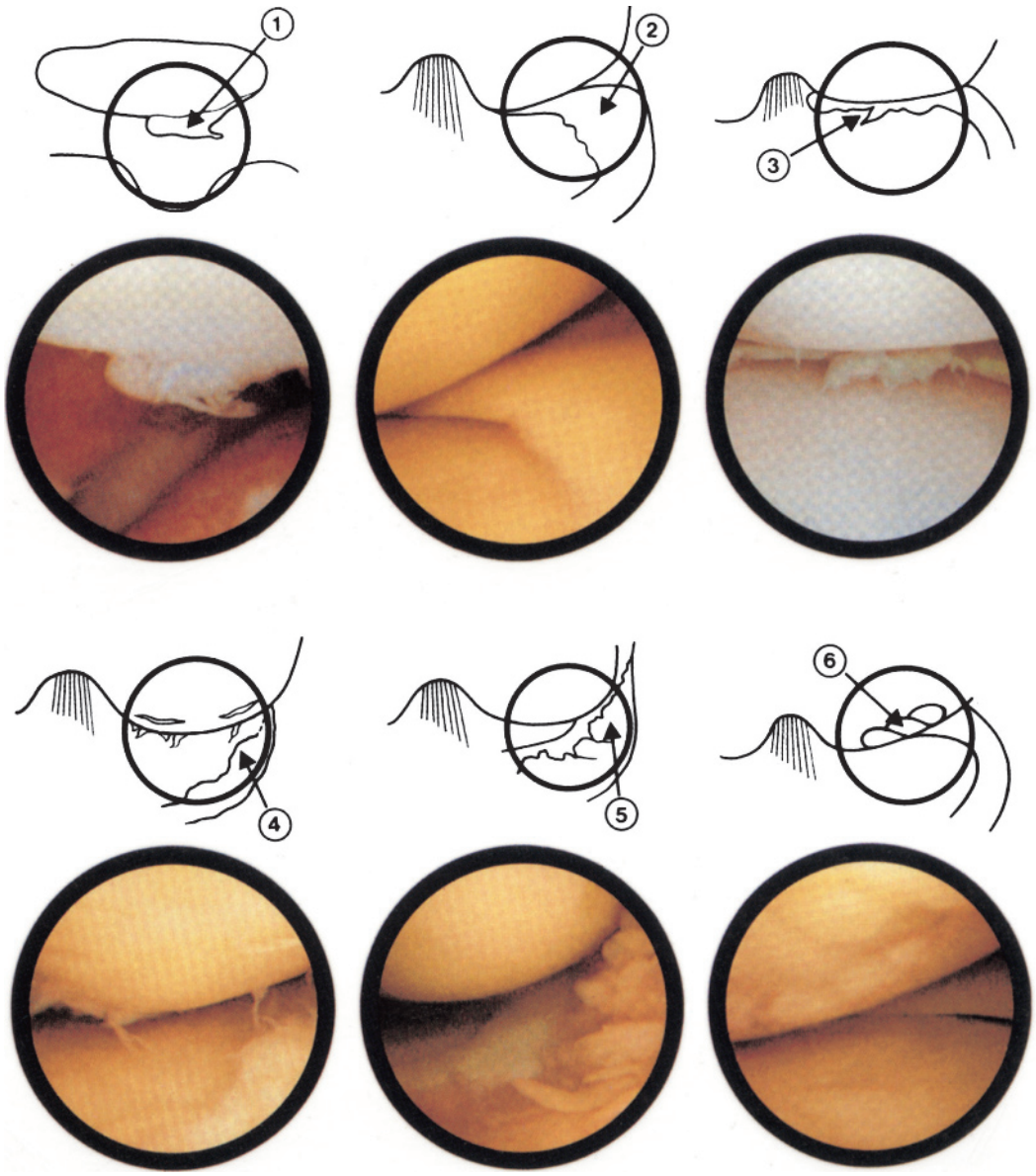


Figure 20.12 Arthroscopy Arthroscopic images of the interior of the right knee from the lateral side, showing (1) chrondomalacia patellae; (2) normal medial meniscus; (3) torn medial meniscus; (4) degenerate medial meniscus and osteoarthritic femoral condyle; (5) rheumatoid synovium; (6) osteochondritis dissecans of medial femoral condyle.

THE DIAGNOSTIC CALENDAR

While most disorders of the knee can occur at any age, certain conditions are more commonly encountered during specific periods of life.

Congenital knee disorders may be present at birth or may become apparent only during the first or second decade of life.

Adolescents with anterior knee pain are usually found to have chondromalacia patellae, patellar instability, osteochondritis or a plica syndrome. But remember – knee pain may be referred from the hip.

Young adults engaged in sports are the most frequent victims of meniscal tears and ligament injuries. Examination should include a variety of tests for ligamentous instability that would be quite inappropriate in elderly patients.

Patients above middle age with chronic pain and stiffness probably have osteoarthritis. With primary osteoarthritis of the knees, other joints also are often affected; polyarthritis does not necessarily (nor even most commonly) mean rheumatoid arthritis.

DEFORMITIES OF THE KNEE

By the end of growth the knees are normally in 5–7 degrees of valgus, although there is significant variation in the range of coronal alignment seen in normal subjects without knee pathology. However, in some circumstances excessive deformity is seen in one of three patterns: *bow leg* (genu varum), knock knee (genu valgum) and *hyperextension* (genu recurvatum).

BOW LEGS AND KNOCK KNEES IN CHILDREN

Deformity is usually gauged from simple observation. Bilateral bow leg can be recorded by measuring the distance between the knees with the child standing and the heels touching; it should be less than 6 cm. Similarly, knock knee can be estimated by measuring the distance between the medial malleoli when the knees are touching with the patellae facing forwards; it is usually less than 8 cm.

Physiological bow legs and knock knees

Bow legs in babies and knock knees in 4-year-olds are so common that they are considered to be *normal stages of development* (Figure 20.13). Other postural abnormalities such as 'pigeon toes' and flat feet may coexist but these children are normal in all other respects; the parents should be reassured and the child should be seen at intervals of 6 months to record progress.

In the occasional case where, by the age of 10, the deformity is still marked (i.e. the intercondylar distance is more than 6 cm or the intermalleolar distance more than 8 cm), operative correction should be advised.

Stapling of the physes on one or other side of the knee can be done to restrict growth on that side and allow correction of the deformity (the staples are removed once the knee has over-corrected slightly); there is a risk, however, that normal growth will not resume when the staples are removed.

Hemi-epiphysodesis (fusion of one-half of the growth plate) on the 'convex' side of the deformity will achieve similar correction; this requires careful timing, guided by charting the child's bone age and estimating the corrective effect of arresting further growth on one side of the bone.

Corrective osteotomy (supracondylar osteotomy for valgus knees and high tibial osteotomy for varus knees) may sound sensible; however, the child (and the parents) will have to put up with the 'deformity' until growth is complete before undergoing the operation, otherwise there is a risk of the deformity recurring while the child is still growing.

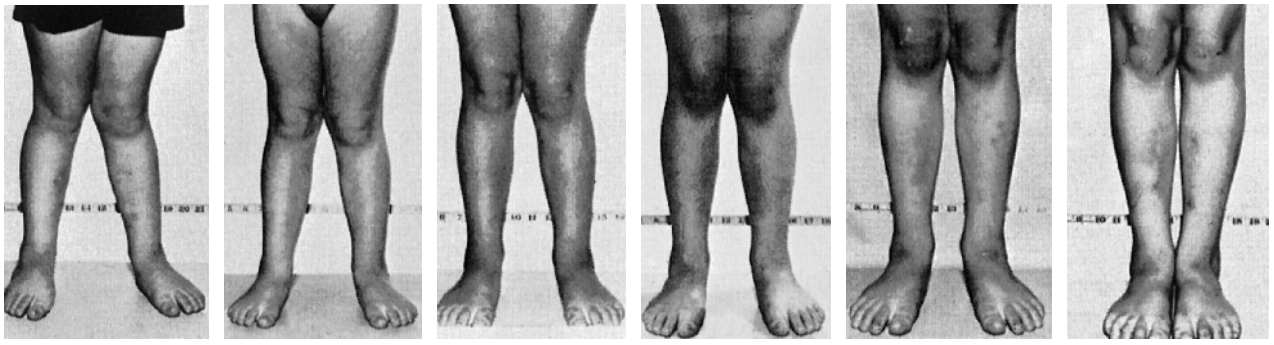


Figure 20.13 Physiological genu valgum 'Knock-knees' in young children usually correct spontaneously. These pictures of the same child were obtained at various ages between 3 and 7 years.

Compensatory deformities

Varus, valgus and rotational deformities of the proximal femur may give rise to complex compensatory deformities of the knees and legs once the child starts to walk. Thus, persistent anteversion of the femoral neck may come to be associated with 'squinting knees' (the patellae face inwards when the hips are fully located), genu valgum, tibial torsion and valgus heels. It is essential to analyse all components of these deformities before focusing on the knees. Often they correct spontaneously by the end of growth, or if some elements persist, they cause little or no problem; only in severe cases – and after the most meticulous preoperative planning – are osteotomies undertaken (Figure 20.14).

Pathological bow legs and knock knees

Disorders which cause distorted epiphyseal and/or physeal growth may give rise to bow leg or knock knee; these include some of the skeletal dysplasias and the various types of rickets, as well as injuries of the epiphyseal and physeal growth cartilage (Figure 20.15). A unilateral deformity is likely to be pathological, but it is essential in all cases to look for signs of injury or generalized skeletal disorder. If angulation is severe, operative correction will be necessary, but it should be deferred until near the end of growth lest the deformity recur with further growth.

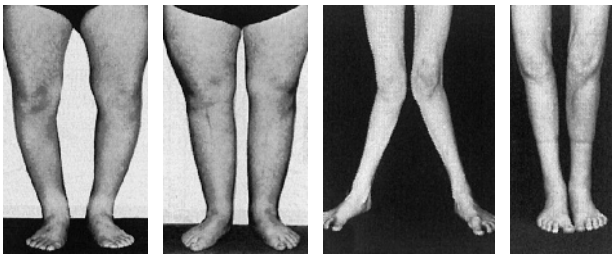
Blount's disease

This is a progressive bow-leg deformity associated with abnormal growth of the posteromedial part of the proximal tibia. The children are usually overweight and start walking early; the condition is bilateral in 80% of cases. Children of Afro-Caribbean descent appear to be affected more frequently than others. Deformity is noticeably worse than in physiological bow legs and may include internal rotation of the tibia. The child walks with an outward thrust of the knee; in the worst cases there may be lateral subluxation of the tibia.

X-rays The proximal tibial epiphysis is flattened medially and the adjacent metaphysis is beak-shaped. The medial cortex of the proximal tibia appears thickened; this is an illusory effect produced by internal rotation of the tibia. The tibial epiphysis sometimes looks 'fragmented'; occasionally the femoral epiphysis is also affected. In the late stages a bony bar forms across the medial half of the tibial physis, preventing further growth on that side. The degree of proximal tibia vara can be quantified by measuring the metaphyseal–diaphyseal angle (see Figure 20.16).

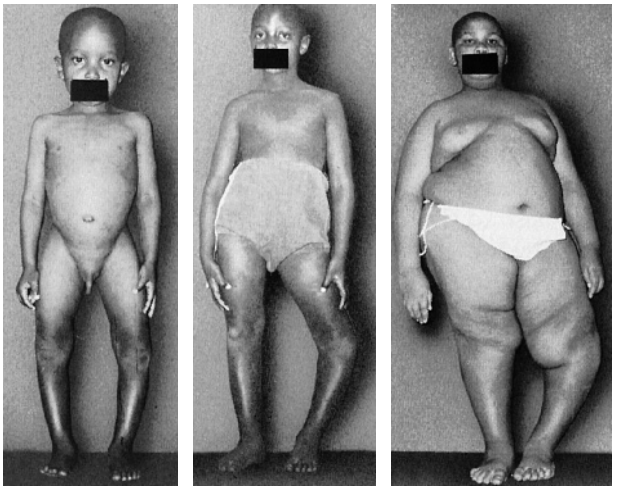
In contrast to physiological bowing, abnormal alignment occurs in the proximal tibia and not in the joint.

Treatment Spontaneous resolution is rare and, once it is clear that the deformity is progressing, a corrective osteotomy should be performed, addressing both the varus and the rotational components. A preoperative (or perioperative) arthrogram, to outline the misshapen epiphysis, will help in planning the operation. Slight over-correction should be aimed for as some recurrence is inevitable. In severe cases it may be necessary also to elevate the depressed medial tibial plateau using a wedge of bone taken from the femur. If a bony bar has formed, it can be excised and replaced by a free fat graft. In older children it may be easier to perform a surgical correction and then (if necessary) lengthen the tibia by the Ilizarov method. All these procedures should be accompanied by fasciotomy to reduce the risk of a postoperative compartment syndrome.



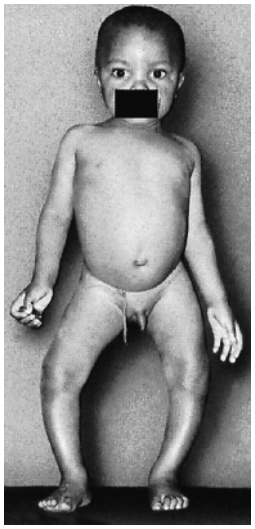
(a) (b) (c) (d)

Figure 20.14 Persistent deformities (a,b) Persistent genu varum before and after corrective osteotomy. (c,d) Before and after osteotomy for severe genu valgum.



(a) (b) (c)

Figure 20.15 Pathological bow legs (a) Child with healed rickets. (b) Growth deformity following a fracture involving the proximal tibial physis. (c) The deformity here was due to a 'slipped' proximal tibial epiphysis in a child with an endocrine disorder.



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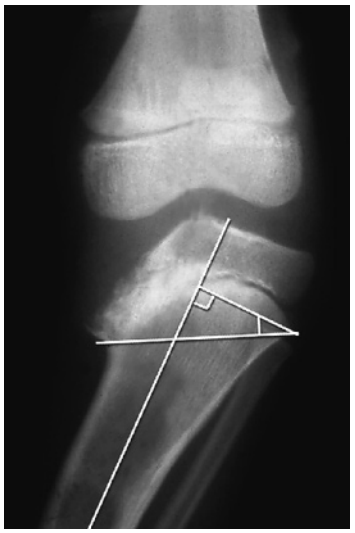


(b)

Figure 20.16 Blount's disease In contrast to the children in Figure 20.15, this young boy developed progressive bow-legged deformities from the time he started walking. X-rays showed the typical features of Blount's disease: marked distortion of the tibial epiphysis, as if one half of the growth plate (physis) had fused and stopped growing. Changes can be accurately assessed by measuring the metaphyseo-diaphyseal angle: a line is drawn perpendicular to the long axis of the tibia and another across the metaphyseal flare as shown on the X-ray; the acute angle formed by these two lines should normally not exceed 11°.

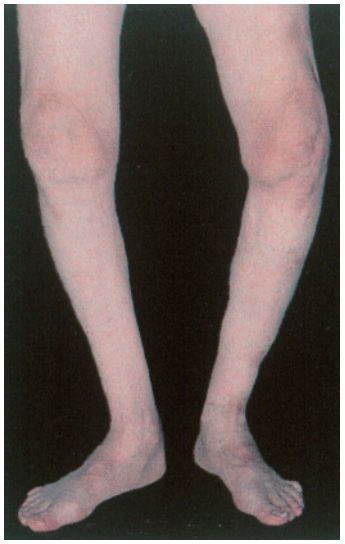
DEFORMITIES OF THE KNEE IN ADULTS

Genu varum and genu valgum

Angular deformities are common in adults (usually bow legs in men and knock knees in women) (Figure 20.17). They may be the *sequel to childhood deformity* and, if so, usually cause no problems. However, if the deformity is associated with early osteoarthritis (medial compartment in genu varum knees and the lateral compartment in genu valgum knees), patients may present with significant symptoms often as pronounced as those with more advanced joint damage. Even in the absence of overt osteoarthritis, if the patient complains of persistent severe pain and there are radiological signs of early joint damage (usually seen on MRI), an osteotomy can be performed – above the knee for valgus deformity and below the knee for varus. Preoperative planning should include radiographic measurements to determine the mechanical and anatomical axes of both bones and the lower limb, as well as estimation of the centre of rotation of angulation.

Deformity may be entirely *secondary to arthritis* – usually varus in osteoarthritis and valgus in rheumatoid arthritis. Stress X-rays or MRI can be useful in the assessment of these cases. Depending on the degree of joint damage, osteotomy or joint replacement may be considered in these patients.

Other causes of varus or valgus deformity are *ligament injuries, malunited fractures* and *Paget's disease*. Where possible, the underlying disorder should be dealt with; provided the joint is stable, corrective osteotomy may be all that is necessary.



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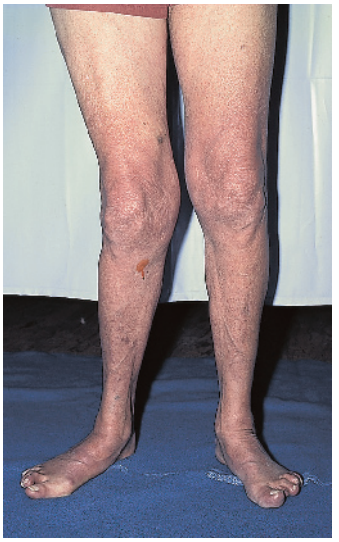

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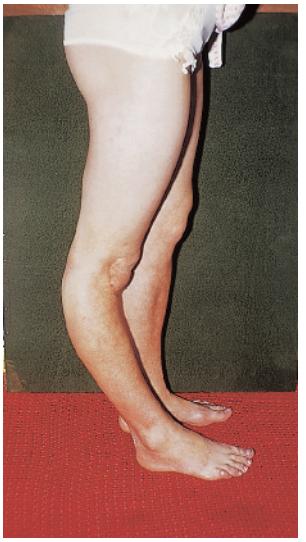


(c)

Figure 20.17 Knee deformities in adults Genu varum is usually associated with osteoarthritis (a); genu valgum with rheumatoid arthritis (b); and genu recurvatum (c) with severe destructive arthritis (e.g. Charcot's disease) or a flail joint (e.g. postpoliomyelitis).

Genu recurvatum (hyperextension of the knee)

Congenital recurvatum This may be due to abnormal intra-uterine posture; it usually recovers spontaneously. Rarely, gross hyperextension is the precursor of true congenital dislocation of the knee.

Lax ligaments Normal people with generalized joint laxity tend to stand with their knees back-set. Prolonged traction, especially on a frame, or holding the knee hyperextended in plaster, may overstretch ligaments, leading to permanent hyperextension deformity. Ligaments may also become overstretched following chronic or recurrent synovitis (especially in rheumatoid arthritis), the hypotonia of rickets, the flailness of poliomyelitis or the insensitivity of Charcot's disease.

In paralytic conditions such as poliomyelitis, recurvatum is often seen in association with fixed equinus of the ankle: in order to set the foot flat on the ground, the knee is forced into hyperextension. In moderate degrees, this may actually be helpful (e.g. in stabilizing a knee with weak extensors). However, if excessive and prolonged, it may give rise to a permanent deformity. If bony correction is undertaken, the knee should be left with some hyperextension to preserve the stabilizing mechanism. If quadriceps power is poor, the patient may need a caliper. Severe paralytic hyperextension can be treated by fixing the patella into the tibial plateau, where it acts as a bone block.

Miscellaneous Other causes of recurvatum are *growth plate injuries* and *malunited fractures*. These can be safely corrected by osteotomy.

Lesions of the menisci

The menisci have an important role in (1) improving articular congruency and increasing the stability of the knee; (2) controlling the complex rolling and gliding actions of the joint; and (3) distributing load during movement. During weight-bearing, at least 50% of the contact stresses are taken by the menisci when the knee is loaded in extension, rising to almost 90% with the knee in flexion. If the menisci are removed, articular stresses are markedly increased; even a partial meniscectomy of one-third of the width of the meniscus will produce a threefold increase in contact stress in that area.

The medial meniscus is much less mobile than the lateral, and it cannot as easily accommodate to abnormal stresses. This may be why meniscal lesions are more common on the medial side than on the lateral.

Even in the absence of injury, there is gradual degeneration and change in the material properties of the menisci with age, so splits and tears are more likely in later life. In many cases these are associated with osteoarthritic articular cartilage damage or chondrocalcinosis. In younger people, meniscal tears are usually the result of trauma, with a specific injury identified in the history.

Tears of the meniscus

The meniscus consists mainly of circumferential collagen fibres held by a few radial strands. It is therefore more likely to tear along its length than across its width. The split is usually initiated by a rotational and shearing force, which occurs (for example) when the knee is flexed and twisted while taking weight; hence the frequency in footballers. In middle life, when fibrotic change has restricted mobility of the meniscus, tears can occur with relatively little force.

Pathology

The medial meniscus is affected far more frequently than the lateral, partly because its attachments to the capsule make it less mobile. Tears of both menisci may occur with severe ligament injuries.

In some cases the split is *vertical* in the length of the meniscus. If the separated fragment remains attached front and back, the lesion is called a *bucket-handle tear*. The torn portion can sometimes displace towards the centre of the joint and becomes jammed between femur and tibia. This causes a block to movement with the patient describing a 'locked knee'. Other patterns of tear can be identified: posterior or anterior horn tears, a geographical description of the tear where the very back or front of the meniscus is damaged, and parrot beak tears where an oblique tear pattern creates a flap of meniscus that may be stable (unlikely to displace) or unstable (displaced or likely to displace).

Horizontal tears are usually 'degenerative' or due to repetitive minor trauma and on the whole are stable. Some are associated with meniscal cysts (see below). Often the degenerate meniscus tears are described as complex, with the tear pattern lying in many planes. These tears may be displaced or likely to displace. If the loose piece of meniscus can be displaced, it acts as a mechanical irritant, giving rise to recurrent synovial effusion and mechanical symptoms (e.g. a sudden catch or sense of giving way in the knee). All except the most peripheral part of the meniscus is avascular and spontaneous repair does not occur unless the tear is in the outer third, which is vascularized from the attached synovium and capsule.

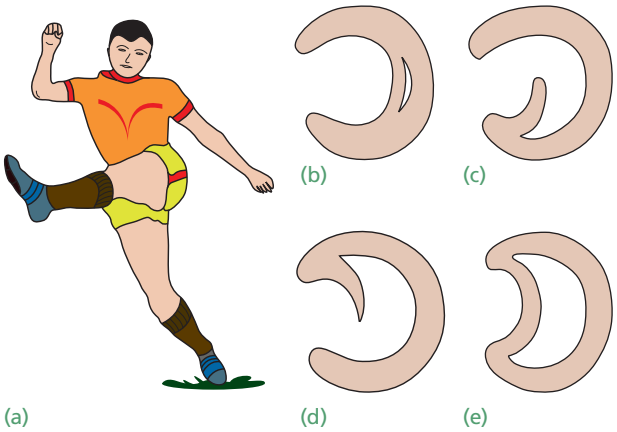


Figure 20.18 Torn medial meniscus (a) The meniscus is usually torn by a twisting force with the knee bent and taking weight; the initial split (b) may extend anteriorly (c); posteriorly (d) or both ways to create a 'bucket-handle' tear (e).

Clinical features

The patient is usually a young person who sustains a twisting injury to the knee on the sports field (Figure 20.18). Pain is often severe and further activity is avoided; occasionally the knee is 'locked' in partial flexion. Almost invariably, swelling appears some hours later, or perhaps the following day.

With rest, the initial symptoms subside, only to recur periodically after trivial twists or strains. Sometimes the knee gives way spontaneously and this is again followed by pain and swelling.

It is important to remember that in patients aged over 40 the initial injury may be unremarkable and the main complaint is of recurrent 'giving way' or 'a catch in the knee'. 'Locking' – i.e. the sudden inability to extend the knee fully – suggests a bucket-handle tear. The patient sometimes learns to 'unlock' the knee by bending it fully or by twisting it from side to side.

On examination the joint may be held slightly flexed and there is often an effusion. In long-standing cases the quadriceps will be wasted. Tenderness is localized to the joint line, in the vast majority of cases on the medial side. Flexion is usually full but extension is often slightly limited.

Between attacks of pain and effusion there is a disconcerting paucity of signs. The history is helpful, and McMurray's test, Apley's grinding test or the Thessaly test may be positive (Figure 20.19).

Investigations

Plain X-rays are usually normal, but *MRI* is a reliable method of confirming the clinical diagnosis, and it may even reveal tears that are missed by arthroscopy.

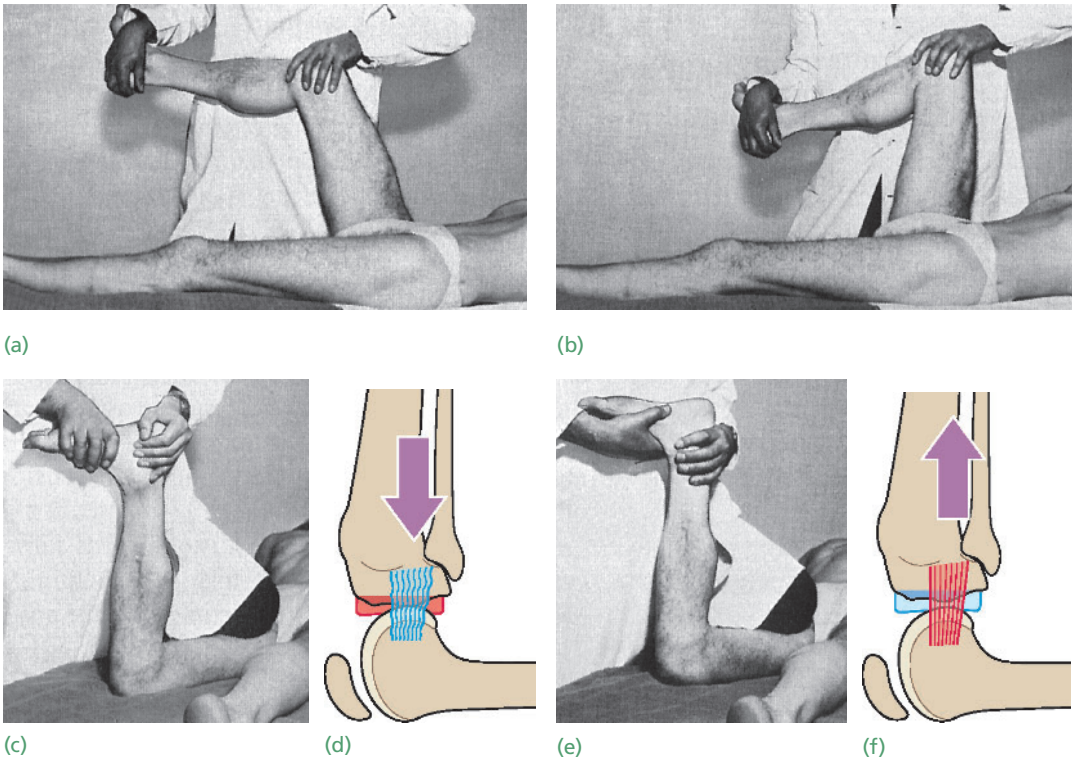


Figure 20.19 Torn medial meniscus – tests (a,b) McMurray's test is performed at varying angles of flexion. (c,d) The grinding test relaxes the ligaments but compresses the meniscus – it causes pain with meniscus lesions. (e,f) The distraction test releases the meniscus but stretches the ligaments and causes pain if these are injured.

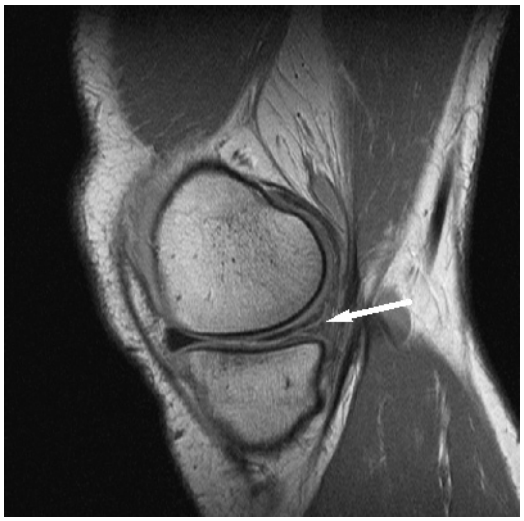


Figure 20.20 Torn meniscus – MRI Sagittal MRI showing a tear in the medial meniscus.

The accessibility of MRI scanning means that the vast majority of patients with suspected meniscal pathology should be investigated with an MRI scan (Figure 20.20). *Diagnostic knee arthroscopy is now infrequently performed due to the increased use of MRI scanning.*

Differential diagnosis

Loose bodies The presence of loose bodies in the joint may cause true locking. The history is much more insidious than with meniscal tears and the attacks are variable in character and intensity. A loose body may be palpable and is often visible on X-ray.

Recurrent dislocation of the patella This causes the knee to give way; typically the patient is caught unawares and collapses to the ground. In many cases the patella spontaneously reduces, although in some patients, reduction of the patella by extension of the knee is required. The traumatic nature of the dislocation can produce an osteochondral fracture of the patella or femur with potential loose body formation. Following relocation the knee demonstrates tenderness localized to the medial edge of the patella and the apprehension test is positive.

Fracture of the tibial spine A fracture can follow an acute injury and may cause a block to full extension. However, swelling is immediate and the knee becomes tense as a haemarthrosis forms. X-ray may show the fracture.

Partial tear of the medial collateral ligament A partial tear may heal with adhesions where it is attached to the medial meniscus, so that the meniscus loses mobility. The patient complains of recurrent attacks of pain and giving way, followed by tenderness on the medial side, which can persist for many months despite the knee regaining full stability. Sleep may be disturbed if the medial side rests upon the other knee or the bed. As with a meniscus injury, rotation is painful; but unlike a meniscus lesion, the grinding test gives less pain and the distraction test more pain.

Torn anterior cruciate ligament This can produce instability, and a sense of the knee 'giving way' or buckling when the patient turns sharply towards the side of the affected knee. Patients often report feeling or hearing a 'pop' from the knee at the time of the injury. Careful examination should reveal signs of rotational instability, a positive Lachman test or a positive anterior drawer sign. MRI or arthroscopy will settle any doubts.

Treatment

DEALING WITH THE LOCKED KNEE

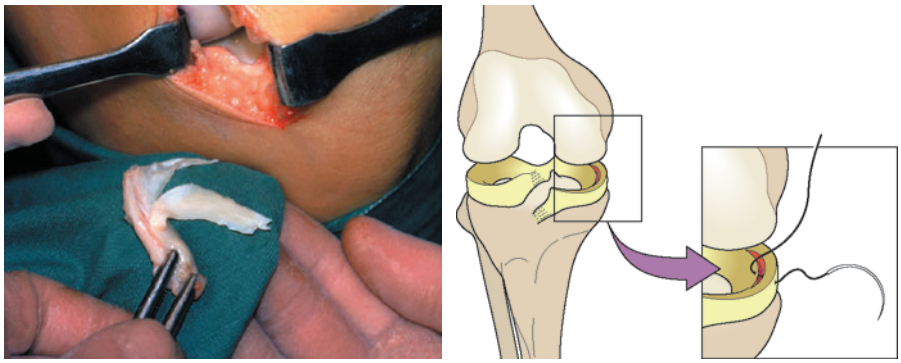
Usually the knee 'unlocks' spontaneously; if not, gentle passive flexion and rotation may do the trick. Forceful manipulation is unwise (it may do more damage) and is usually unnecessary; after a few days' rest the knee may well unlock itself. However, if the knee does not unlock, or if attempts to unlock it cause severe pain, arthroscopy is indicated. If symptoms are not marked, it may be better to wait a week or two and let the synovitis settle down, thus making the operation easier; if the tear is confirmed, the offending fragment is removed or repaired if possible.

CONSERVATIVE TREATMENT

If the joint is not locked, MRI scanning now plays a critical role in planning further treatment. If a peripheral tear has been identified and the lesion may be repairable, then arthroscopy and suture repair of the meniscus can be employed, particularly in the younger patient. However, other unstable or potentially unstable tears, identified on MRI, often present with infrequent symptoms that are not disabling. Non-operative care should be instigated as many patients will settle, with resolution of the acute flare of pain and swelling, despite the continued presence of the meniscal tear.

OPERATIVE TREATMENT

Surgery is indicated (1) if the joint cannot be unlocked and (2) if mechanical symptoms (locking or catching) are recurrent and non-operative treatment has failed. In most cases, if available, an MRI scan should be obtained to determine the pattern of tear and to plan treatment with the patient. As a result 'diagnostic' arthroscopy is now infrequently performed. Tears close to the periphery, which have the capacity to heal, can be sutured; at least one edge of the tear should be red (i.e. vascularized). In appropriate cases



(a) (b)

Figure 20.21 Torn meniscus – operation (a) Removal of a torn medial meniscus. (b) Repair is appropriate if at least one edge of the tear is vascularized. This can be done arthroscopically.

the success rate for both open and arthroscopic repair can be high.

Tears other than those in the peripheral third are dealt with by excising the torn portion (or the bucket handle) (Figure 20.21). Total meniscectomy is thought to cause more instability and so predispose to late secondary osteoarthritis; certainly in the short term it causes greater morbidity than partial meniscectomy and has no obvious advantages.

Arthroscopic meniscectomy has distinct advantages over open meniscectomy: shorter hospital stay, lower costs and more rapid return to function. However, it is by no means free of complications.

Postoperative pain and stiffness are reduced by prophylactic non-steroidal anti-inflammatory drugs. In some patients a flare of pain can occur, which can persist for a number of months. In some cases there is a rapid progression of articular cartilage damage and the development of arthritis.

Outcome

Neither a meniscal tear by itself nor removal of the meniscus necessarily leads to secondary osteoarthritis. However, the likelihood is increased if the patient has (1) a pre-existing varus deformity of the knee, (2) signs of cruciate ligament insufficiency or (3) features elsewhere of a generalized osteoarthritis.

Meniscal degeneration

Patients over 45 years old may present with symptoms and signs of a meniscal tear. Often, though, they can recall no preceding injury. MRI scanning reveals a horizontal cleavage in the medial meniscus – the characteristic 'degenerative' lesion – or detachment of the anterior or posterior horn without an obvious tear. Associated osteoarthritis or chrondrocalcinosis is common. Meniscectomy is often not helpful in these cases and is only indicated if mechanical symptoms are marked, such as a mechanical block to movement or recurrent sharp pains in the knee.

Discoid lateral meniscus

In the foetus the meniscus is not semilunar but disc-like. If this disc-like shape persists postnatally symptoms can occur if the whole meniscus is unstable or more typically where a tear occurs. Usually a young patient complains that, without any history of injury, the knee gives way and 'thuds' loudly. A characteristic clunk may be felt at 110 degrees as the knee is bent and at 10 degrees as it is being straightened. The diagnosis is easily confirmed by MRI.

If there is only a clunk, treatment is not essential. If pain is intrusive, the meniscus may be partially excised, leaving a normally shaped meniscus.

Meniscal cysts

Cysts of the menisci most often arise from horizontal cleavage tears. It is also suggested that synovial cells infiltrate into the vascular area between meniscus and capsule and multiply there. The multilocular cyst contains gelatinous fluid and is surrounded by thick fibrous tissue.

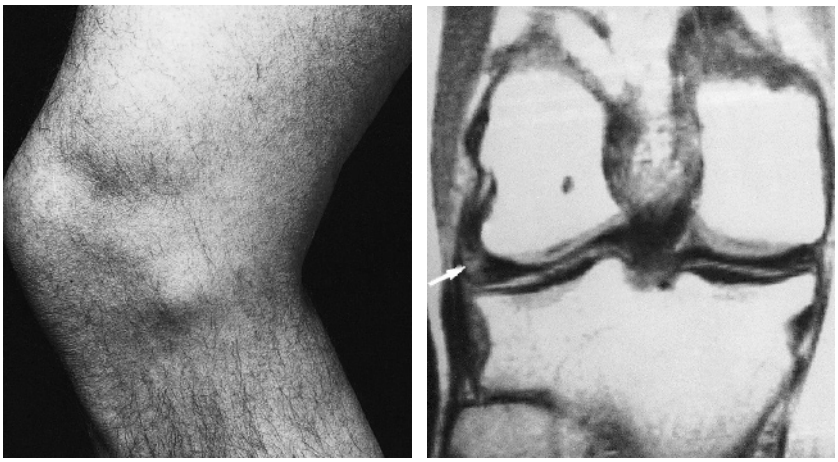
Clinical features

The lateral meniscus is affected much more frequently than the medial. The patient complains of an ache or a small lump at the side of the joint. Symptoms may be intermittent, or worse after activity.

On examination the lump is situated at or slightly below the joint line, usually anterior to the collateral ligament. It is seen most easily with the knee slightly flexed (Figure 20.22); in some positions it may disappear altogether. Lateral cysts are often so firm that they are mistaken for a solid swelling. Medial cysts are usually larger and softer.

Differential diagnosis

Apart from cysts, various conditions may present with a small lump along the joint line.



(a) (b)

Figure 20.22 Meniscal cyst
(a) Typical appearance of a small, firm swelling at or just below the joint line. (b) MRI showing the cyst arising from the edge of the meniscus (arrow).

Ganglion A ganglion lump is quite superficial, usually not as 'hard' as a cyst, and unconnected with the joint.

Calcific deposits in the collateral ligament These usually appear on the medial side, are intensely painful and tender, and often show on the X-ray.

Prolapsed, torn meniscus This occasionally presents as a rubbery, irregular lump at the joint line. In some cases the distinction from a 'cyst' is largely academic.

Tumour Various tumours, both of soft tissue (lipoma, fibroma) and of bone (osteochondroma), may produce a medial or lateral joint lump. Careful examination will show that the lump does not arise from the joint itself.

Treatment

In many cases, if the cyst is asymptomatic, no treatment is required. However, a tense cyst can be symptomatic and can be addressed surgically. The role of surgery is to decompress the cyst by excising the cleavage tear. In the past this was usually total meniscectomy, in order to prevent an inevitable recurrence of the cyst. A more contemporary approach is to resect the lower half of the cleavage tear, decompressing the cyst. The outcome is often successful but the cyst may recur with recurrence of mechanical symptoms.

CHRONIC LIGAMENTOUS INSTABILITY

The knee is a complex hinge which depends heavily on its ligaments for mediolateral, anteroposterior and rotational stability. Ligament injuries, from minor strains through partial ruptures to complete tears, are common in sportsmen, athletes and dancers. Whatever the nature of the acute injury, the victim may be left with chronic instability of the knee – a sense of the joint wanting to give way, or actually giving way, during unguarded activity. Sometimes this is accompanied by pain and recurrent episodes of swelling. There may be a meniscal tear, but meniscectomy is likely to make matters worse; sometimes patients present with meniscectomy scars on both sides of the knee.

Examination should include special tests for ligamentous instability as well as radiological investigation and arthroscopy. It is important not only to establish the nature of the lesion but also to measure the level of functional impairment against the needs and demands of the individual patient before advocating treatment.

The subject is dealt with in detail in Chapter 31.

RECURRENT DISLOCATION OF THE PATELLA

Acute dislocation of the patella is dealt with in Chapter 31. In 15–20% of cases (mostly children) the first episode is followed by recurrent dislocation or subluxation after minimal stress. This is due, in some measure, to disruption or stretching of the medially based ligamentous structures (e.g. the medial patellofemoral ligament – MPFL) which normally stabilize the extensor mechanism. However, in a significant proportion of cases there is no history of an acute strain and the initial episode is thought to have occurred 'spontaneously'. It is now recognized that in all cases of recurrent dislocation, but particularly in the latter group, one or more *predisposing factors* are often present: (1) generalized ligamentous laxity; (2) underdevelopment of the lateral femoral condyle and flattening of the intercondylar groove; (3) maldevelopment of the patella, which may be too high or too small; (4) valgus deformity of the knee; (5) external tibial torsion or (6) a primary muscle defect.

Repeated dislocation damages the contiguous articular surfaces of the patella and femoral condyle; this may result in further flattening of the condyle, so facilitating further dislocations.

Dislocation is almost always towards the lateral side; medial dislocation is seen only in rare iatrogenic cases following overzealous lateral release or medial transposition of the patellar tendon.

Clinical features

Girls are affected more commonly than boys and the condition may be bilateral. Dislocation occurs unexpectedly when the quadriceps muscle is contracted with the knee in flexion. There is acute pain, the knee is stuck in flexion and the patient may fall to the ground.

Although the patella always dislocates laterally, the patient may think it has displaced medially because the uncovered medial femoral condyle stands out prominently. If the knee is seen while the patella is dislocated, the diagnosis is obvious. There is a lump on the lateral side, while the front of the knee (where the patella ought to be) is flat (Figure 20.23). The tissues on the medial side are tender, the joint may be swollen and aspiration may reveal a blood-stained effusion.

More often the patella has reduced by the time the patient is seen. Tenderness and swelling may still be present and *the apprehension test* is positive: if the patella is pushed laterally with the knee slightly flexed, the patient resists and becomes anxious, fearing another dislocation. The patient will normally volunteer a history of previous dislocation.

Between attacks the patient should be carefully examined for features that are known to predispose to patellar instability (see above).

Imaging

X-rays may reveal loose bodies in the knee, derived from old osteochondral fragments. A lateral view with the knee in slight flexion may show a high-riding patella and tangential views can be used to measure the sulcus angle and the congruence angle.

MRI is helpful and may show signs of the previous patellofemoral soft-tissue disruption on the medial side of the knee or trochlear dysplasia.

Treatment

NON-OPERATIVE TREATMENT

If the patella is still dislocated, it is pushed back into place while the knee is gently extended. The only indications for immediate surgery are (1) inability to reduce the patella (e.g. with a rare 'intra-articular' dislocation); and (2) the presence of a large, displaced osteochondral fragment.

Usually a *knee brace* is applied and retained for 2–3 weeks to allow the soft tissues to heal; isometric quadriceps-strengthening exercises are encouraged and the patient is allowed to walk with the aid of crutches.

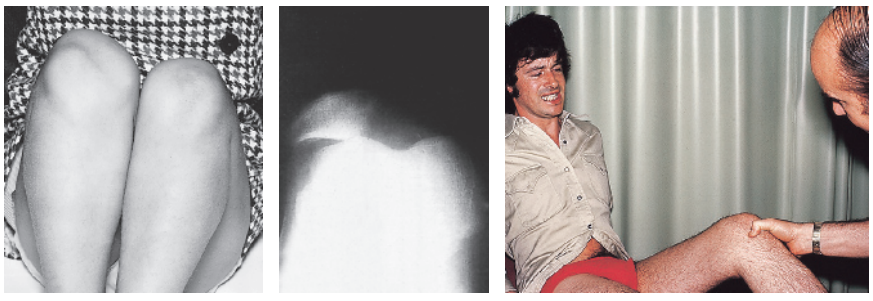
Exercises should be continued for at least 3 months, concentrating on strengthening the vastus medialis muscle. If recurrences are few and far between, conservative treatment may suffice; as the child grows older the patellar mechanism tends to stabilize. However, about 15% of children with patellar instability suffer repeated and distressing episodes of dislocation, and for these patients surgical reconstruction is indicated.

OPERATIVE TREATMENT

The principles of operative treatment are (1) to repair or strengthen the medial patellofemoral ligaments; (2) to realign the extensor mechanism so as to produce a mechanically more favourable angle of pull; and (3) to treat trochlear dysplasia if present and predisposing to instability. This can be achieved in several ways (see Figure 20.24).

Direct medial patellofemoral ligament repair
Occasionally it is possible to perform a direct repair or reconstruction of an attenuated medial patellofemoral ligament.

Suprapatellar realignment (Insall) The lateral retinaculum and capsule are divided. The quadriceps tendon adjacent to the vastus medialis is split longitudinally to the level of the tibial tubercle; the free edge is then sutured over the middle of the patella,



(a) (b) (c)

Figure 20.23 Patellofemoral instability (a,b) This young girl presented with recurrent subluxation of the right patella. The knee looks abnormal and the X-ray shows the patella riding on top of the lateral femoral condyle. (c) The apprehension test: watch the patient's face!

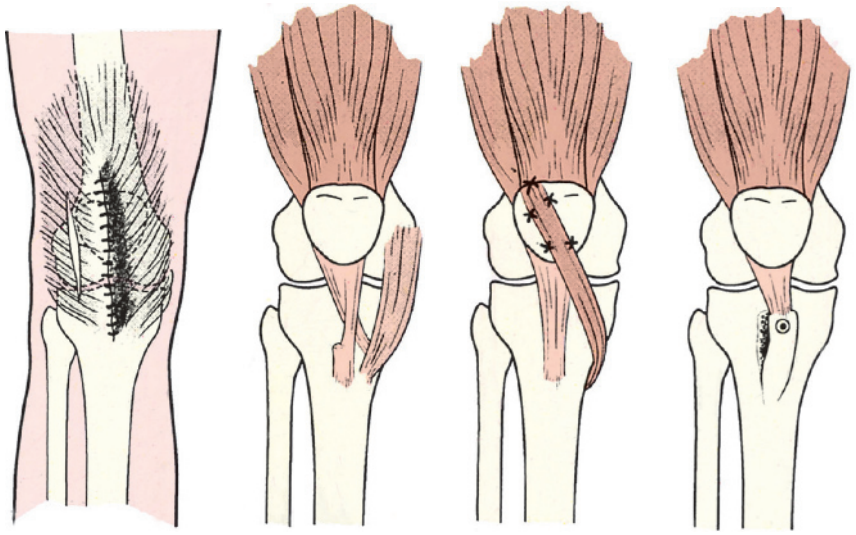


Figure 20.24 Realignment for recurrent patellar dislocation There are several methods popularly used. Most involve a lateral release of the capsule and some form of 'tether' medially. This check-rein may be created from (a) vastus medialis (Insall), (b) by transposing the lateral half of the patellar ligament medially (Roux–Goldthwait) or by (c) the semitendinosus tendon (Galleazzi). (d) In adults, bony operations which shift the position of the patellar tubercle may be tried (Elmslie–Trillat).

thus bringing vastus medialis distally and closer to the midline.

Infrapatellar soft-tissue realignment (Roux-Goldthwait) The lateral half of the patellar ligament is detached, threaded under the medial half and reattached more medially and distally. This operation is seldom used by itself but may be combined with suprapatellar realignment.

Infrapatellar bony realignment (Elmslie–Trillat) The tibial tubercle is osteotomized and moved medially, thus improving the angle of pull on the patella. This procedure is only appropriate after closure of the proximal tibial physis; if growth is incomplete, damage to the physis may result in a progressive recurvatum deformity. The presence of trochlear dysplasia may require a trochleoplasty, where the shallow groove within the knee is deepened to allow the patellar to track properly.

NOTE: All these procedures can be combined with repair or tightening of the medial patellofemoral ligament. At the end of the operation it is essential to check that the patella moves smoothly to at least 60 degrees of knee flexion; excessive tightening or uneven tension may cause maltracking (and, occasionally, even medial subluxation!) of the patella.

Patellectomy Occasionally the patellofemoral cartilage is so damaged that patellectomy is indicated, but this operation should be avoided if possible. There is a small risk that after patellectomy the patellar tendon may continue to dislocate and require realignment by the tibial tubercle transfer.

Recurrent subluxation

Patellar dislocation is sometimes followed by recurrent subluxation rather than further episodes of complete displacement. This is the borderline between frank instability and maltracking of the patella (see below).

Other types of non-traumatic dislocation

Congenital dislocation, in which the patella is permanently displaced, is fortunately very rare. Reconstructive procedures, such as semitendinosus tenodesis, have been tried but the results are unpredictable.

Habitual dislocation differs from recurrent dislocation in that the patella dislocates every time the knee is bent and reduces each time it is straightened. In long-standing cases the patella may be permanently dislocated. The probable cause is *contracture of the quadriceps,* which may be congenital or may result from repeated injections (usually antibiotics) into the muscle.

Treatment requires lengthening of the quadriceps. Additionally a lateral capsular release and medial plication may be needed to hold the patella in the intercondylar groove.

Patellofemoral pain syndrome (chondromalacia of the patella; patellofemoral overload syndrome)

There is no clear consensus concerning the terminology, aetiology and treatment of pain and tenderness in the anterior part of the knee. This syndrome is

common among active adolescents and young adults. It is often (but not invariably) associated with softening and fibrillation of the articular surface of the patella – *chondromalacia patellae*. Having no other pathological label, orthopaedic surgeons have tended to regard chondromalacia as the cause (rather than one of the effects) of the disorder. Against this are the facts that (1) chondromalacia is commonly found at arthroscopy in young adults who have no anterior knee pain; and (2) some patients with the typical clinical syndrome have no cartilage softening.

Pathogenesis and pathology

Pain over the anterior aspect of the knee occurs as one of the symptoms in a number of well-recognized disorders, the commonest of which are bursitis, Osgood–Schlatter disease, a neuroma, plica syndromes, patellofemoral arthritis and tendinitis affecting either the insertion of the quadriceps tendon or the patellar ligament – Sinding-Larsen disease. When these are excluded and no other cause can be found, one is left with a clinically recognizable syndrome that has earned the unsatisfactory label of 'anterior knee pain' or 'patellofemoral pain syndrome'.

It is believed that the basic disorder is probably mechanical overload of the patellofemoral joint.

Rarely, a single injury (sudden impact on the front of the knee) may damage the articular surfaces. Much more common is repetitive overload which may be due to:

1 *malcongruence of the patellofemoral surfaces* because of some abnormal shape of the patella or intercondylar groove
2 *malalignment of the lower extremity and/or the patella*
3 *muscular imbalance of the lower extremity* with decreased strength due to atrophy or inhibition, or relative weakness of the vastus medialis, which causes the patella to tilt, or subluxate, or bear more heavily on one facet than the other during flexion extension
4 *overactivity.*

'Overload', as used here, means either direct stress on a load-bearing facet or sheer stresses in the depths of the articular cartilage at the boundary between high-contact and low-contact areas. Personality and chronic pain response issues must also be considered.

Patellofemoral overload leads to changes in both the articular cartilage and the subchondral bone, not necessarily of parallel degree. Thus, the cartilage may look normal and show only biochemical changes such as overhydration or loss of proteoglycans, while the underlying bone shows reactive vascular congestion (a potent cause of pain). Or there may be obvious cartilage softening and fibrillation, with or without subarticular intraosseous hypertension. This would account for the variable relationship between (1) malalignment syndrome; (2) cartilage softening; (3) subchondral vascular congestion; and (4) anterior knee pain.

Cartilage fibrillation usually occurs on the medial patellar facet or the median ridge, remains confined to the superficial zones and generally heals spontaneously. It is not a precursor of progressive osteoarthritis in later life. Occasionally the lateral facet is involved – Ficat's 'hyperpression zone' syndrome – and this may well be progressive.

Clinical features

The patient, often a teenage girl or an athletic young adult, complains of intermittent or persistent pain over the front of the knee or 'underneath the knee-cap'. Symptoms are aggravated by activity or climbing stairs, or when standing up after prolonged sitting. The knee may give way and occasionally swells. It sometimes 'catches' but this is not true locking. Often both knees are affected. Occasionally there is a history of previous injury or recurrent instability.

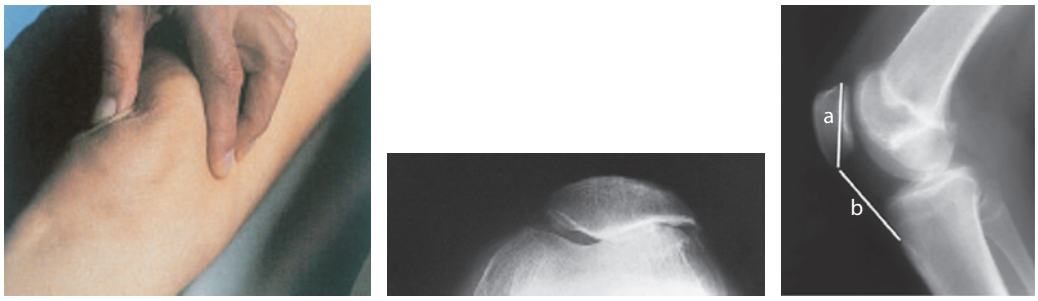
At first sight the knee looks normal but careful examination may reveal malalignment or tilting of the patellae. Other subtle signs include quadriceps wasting, fluid in the knee, tenderness under the edge of the patella and crepitus on moving the knee.

Patellofemoral pain is elicited by pressing the patella against the femur and asking the patient to contract the quadriceps – first with central pressure, then compressing the medial facet and then the lateral. If, in addition, the apprehension test is positive, this suggests previous subluxation or dislocation.

Patellar tracking can be observed with the patient seated on the edge of the couch, flexing and extending the knee against resistance; in some cases subluxation is obvious.

With the patient sitting or lying supine, patellar alignment can be gauged by measuring the quadriceps angle, or Q-angle – the angle subtended by the line of quadriceps pull (a line running from the anterior superior iliac spine to the middle of the patella) and the line of the patellar ligament. It normally averages 14–17 degrees and an angle of more than 20 degrees is regarded as a predisposing factor in the development of anterior knee pain. Another predisposing factor is a high-riding patella (*patella alta*); compressive force on the patellar articular surface during flexion and extension is likely to be greater than normal. Patella alta is best measured on the lateral X-ray.

Lastly, the structures around the knee should be carefully examined for other sources of pain, and the hip is examined to exclude referred pain.



(a) (b) (c)

Figure 20.25 Chondromalacia of the patella There is no pathognomonic feature on which to base the diagnosis of chondromalacia, but several signs are suggestive. (a) Hold the patella against the femoral condyles and ask the patient to tighten the thigh muscles; even in normal people this may be uncomfortable, but patients with chondromalacia experience sudden acute pain in the patellofemoral joint. (b) A 'skyline' X-ray with the knee in partial flexion may show obvious tilting of the patella. (c) In the lateral X-ray, with the knee flexed to 45°, the lengths of the patella and the patellar ligament are normally about equal (a ratio of 1:1); in patella alta the ratio is less than 1:1.

Imaging

X-ray examination should include 'skyline' views of the patella, which may show abnormal tilting or subluxation, and a lateral view with the knee half-flexed to see if the patella is high or small (Figure 20.25). The presence of trochlear dysplasia can be identified on plain X-rays.

The most accurate way of showing and measuring patellofemoral malposition is by *CT or MRI* with the knees in full extension and varying degrees of flexion.

Arthroscopy

Arthroscopy can be used in the process of excluding other causes of anterior knee pain serving to gauge patellofemoral congruence, alignment and tracking. However, often no therapeutic response to this procedure occurs.

Differential diagnosis

Other causes of anterior knee pain must be excluded before finally accepting the diagnosis of patellofemoral pain syndrome (see Box 20.1). Even then, the exact cause of the syndrome must be established before treatment: for example, is it abnormal posture, overuse, patellar malalignment, subluxation or some abnormality in the shape of the bones?

Treatment

CONSERVATIVE MANAGEMENT

In the vast majority of cases the patient will be helped by adjustment of stressful activities, deploying focused physiotherapy, combined with reassurance that most patients eventually recover without surgery. Exercises are directed specifically at strengthening the medial quadriceps so as to counterbalance the tendency to lateral tilting or subluxation of the patella. Some patients respond to simple measures such as providing support for a valgus foot.

BOX 20.1 CAUSES OF ANTERIOR KNEE PAIN

1. **Referred from hip**
2. **Patellofemoral disorders**
   - Patellar instability
   - Patellofemoral overload
   - Osteochondral injury
   - Patellofemoral osteoarthritis
3. **Knee joint disorders**
   - Osteochondritis dissecans
   - Loose body in the joint
   - Synovial chondromatosis
   - Plica syndrome
4. **Periarticular disorders**
   - Patellar tendinitis
   - Patellar ligament strain
   - Bursitis
   - Osgood–Schlatter disease

OPERATIVE TREATMENT

Surgery should be considered only if conservative treatment has lasted at least 6 months and there is a demonstrable abnormality that is correctable by operation (see Figure 20.24).

Extensor mechanism malalignment is treated with a realignment procedure, either *proximal realignment* – a combined open release of the lateral retinaculum and reefing of the oblique part of the vastus medialis – or *distal realignment*, as described earlier. They will improve the tracking angle but run the risk of increasing patellofemoral contact pressures and thus aggravating the patient's symptoms.

Where articular cartilage damage is identified *chondroplasty* can be employed, where shaving of the patellar articular surface is performed arthroscopically using a power tool. Soft and fibrillated cartilage is removed, in severe cases down to the level of subchondral bone; the hope is that it will be replaced by fibrocartilage.

Lateral facet pressure syndrome remains one of the few indications for performing *lateral release*, where the lateral knee capsule and extensor retinaculum are divided longitudinally, either open or arthroscopically. This sometimes succeeds on its own (particularly if significant patellar tilting can be demonstrated on X-ray or MRI), but more often patellofemoral realignment will be needed as well.

Distal elevation of the patellar ligament In Maquet's tibial tubercle advancement operation the tubercle, with the attached patellar ligament, is hinged forwards and held there with a bone-block. This has the effect of reducing patellofemoral contact pressures. Some patients resent the bump on the front part of the tibia and the operation may substitute a new set of complaints for the old. Alternatively, the Fulkerson anteromedial tibial tubercle transfer and elevation can be used with satisfactory mid-term results. These operations are now infrequently performed.

Patellectomy This is a last resort, but patients with severe discomfort are grateful for the relief it brings after other operations have failed.

OSTEOCHONDRITIS DISSECANS

The prevalence of osteochondritis dissecans (OCD) is between 15 and 30 per 100 000 with a male to female ratio of 5:3. An increase in its incidence has been observed in recent years, probably due to the growing participation of young children of both genders in competitive sports.

A small, well-demarcated, avascular fragment of bone and overlying cartilage sometimes separates from one of the femoral condyles and appears as a loose body in the joint. The most likely cause is trauma, either a single impact with the edge of the patella or repeated microtrauma from contact with an adjacent tibial ridge. The fact that over 80% of lesions occur on the lateral part of the medial femoral condyle, exactly where the patella makes contact in full flexion, supports the first of these. There may also be some general predisposing factor, because several joints can be affected, or several members of one family. Lesions are bilateral in 25% of cases.

Pathology

The lower, lateral surface of the medial femoral condyle is usually affected, rarely the lateral condyle, and still more rarely the patella. An area of subchondral bone becomes avascular and within this area an ovoid osteocartilaginous segment is demarcated from the surrounding bone. At first the overlying cartilage is intact and the fragment is stable; over a period of months the fragment separates but remains in position ('OCD lesion *in situ*'); finally the fragment breaks free to become a loose body in the joint. The small crater is slowly filled with fibrocartilage, leaving a depression on the articular surface.

Classification

Osteochondritis dissecans of the knee is classified according to anatomical location, arthroscopic appearance, scintigraphic or MRI findings and chronological age. For prognostic and treatment purposes it is divided into juvenile and adult forms, either stable or unstable.

Clinical features

The patient, usually between 15 and 20 years of age, presents with intermittent ache or swelling. Later, there are attacks of giving way such that the knee feels unreliable; 'locking' sometimes occurs.

The quadriceps muscle is wasted and there may be a small effusion. Soon after an attack there are two signs that are almost diagnostic: (1) tenderness localized to one femoral condyle; and (2) Wilson's sign: if the knee is flexed to 90 degrees, rotated medially and then gradually straightened, pain is felt; repeating the test with the knee rotated laterally is painless.

Imaging

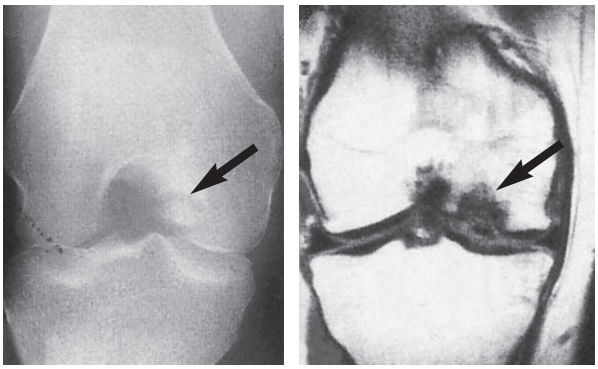
Plain X-rays may show a line of demarcation around a lesion *in situ*, usually in the lateral part of the medial femoral condyle. This site is best displayed in special intercondylar (tunnel) views, but even then a small lesion or one situated far back may be missed (Figure 20.26). Once the fragment has become detached, the empty hollow may be seen – and possibly a loose body elsewhere in the joint.

MRI is the most effective imaging technique to define the site, size and activity of an OCD lesion. An OCD lesion will typically demonstrate an area of low signal intensity in the T1-weighted images, with adjacent bone also appearing abnormal, probably due to oedema.

These investigations usually indicate whether the fragment is 'stable' or 'loose', based on the presence of fluid under the lesion.

Arthroscopy

Arthroscopy is not used for diagnostic purposes if MRI scanning is available, but it can be deployed to determine if an OCD lesion is stable or unstable, prior to fixation or removal.



(a) (b)

Figure 20.26 Osteochondritis dissecans – imaging
The lesion is often missed in the standard anteroposterior X-ray and is better seen in the 'tunnel view', usually along the lateral side of the medial femoral condyle (a). Here the osteochondral fragment has remained in place but sometimes it appears as a separate body elsewhere in the joint. (b) MRI provides confirmatory evidence and shows a much wider area of involvement than is apparent in the plain X-ray.

Differential diagnosis

Avascular necrosis of the femoral condyle – usually associated with corticosteroid therapy or alcohol abuse – may result in separation of a localized osteocartilaginous fragment. However, it is seen in an older age group and on X-ray the lesion is always on the dome of the femoral condyle, and this distinguishes it from osteochondritis dissecans.

Treatment

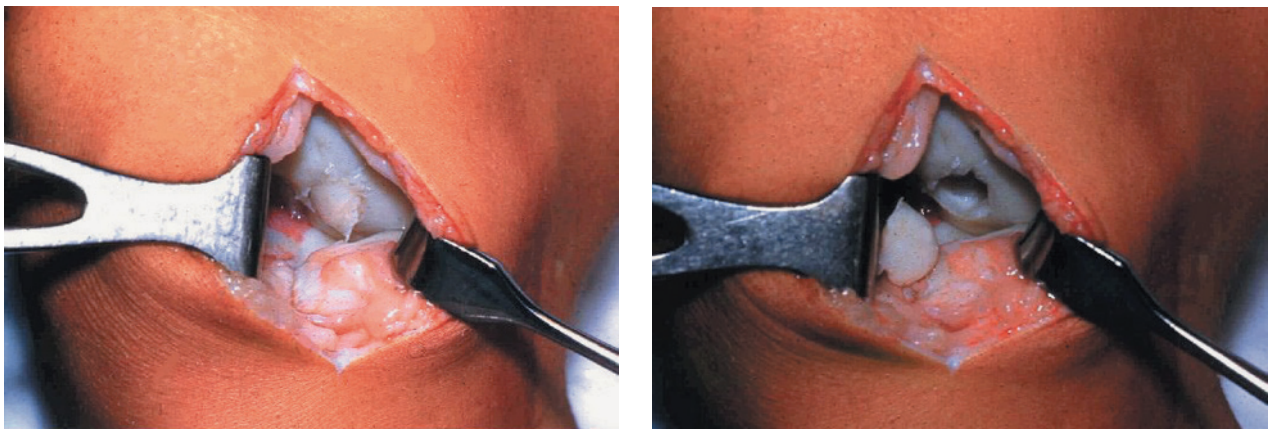
For the purposes of management, it is useful to 'stage' the lesion; hence the importance of MRI and possibly arthroscopy. Lesions in adults have a greater propensity to instability whereas juvenile osteochondritis is typically stable. Those lesions with an intact articular surface have the greatest potential to heal with non-operative treatment if repetitive impact loading is avoided.

In the earliest stage, when the cartilage is intact and the lesion is 'stable', no treatment is needed but activities are curtailed for 6–12 months. Small lesions often heal spontaneously.

If the fragment is 'unstable' (i.e. surrounded by a clear boundary with radiographic 'sclerosis' of the underlying bone), or showing MRI features of separation, treatment will depend on the size of the lesion and the age of the patient. A small fragment should be removed by arthroscopy and the base drilled (Figure 20.27); the bed will eventually be covered by fibrocartilage, leaving only a small defect. A large fragment (say more than 1 cm in diameter) should be fixed *in situ* with pins or Herbert screws, or in the younger patient with open growth plates. In older patients removal of the unstable fragment and cartilage repair techniques (e.g. microfracture or autologous cartilage implantation (ACI) are carried out.

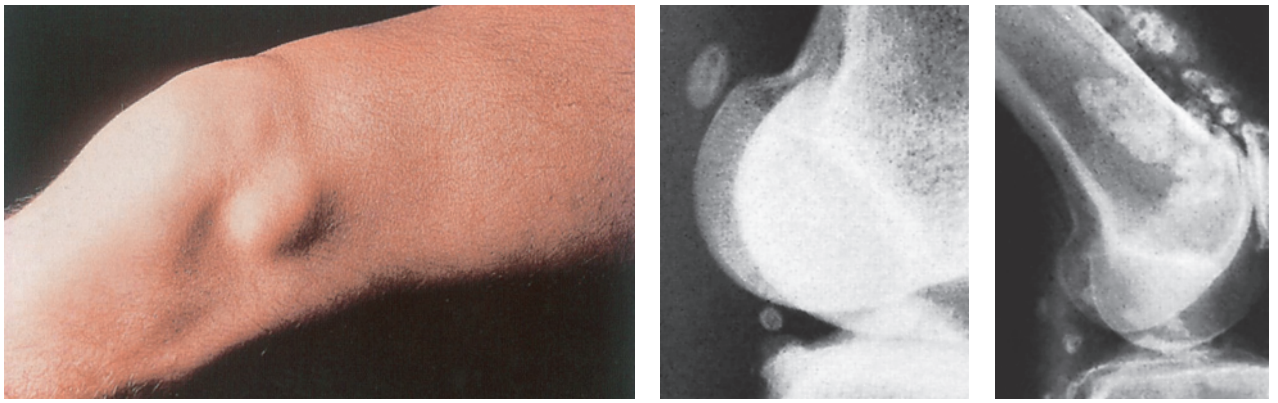
LOOSE BODIES

Loose bodies can be found in the knee produced by: (1) injury (a chip of bone or cartilage); (2) osteochondritis dissecans (which may produce one or two fragments); (3) osteoarthritis (pieces of cartilage or osteophyte); (4) Charcot's disease (large osteocartilaginous bodies); and (5) synovial chondromatosis (cartilage metaplasia in the synovium, sometimes producing hundreds of loose bodies) (Figure 20.28).



(a) (b)

Figure 20.27 Osteochondritis dissecans Intraoperative pictures showing the articular lesion (a) and the defect left after removal of the osteochondral fragment (b).



(a) (b) (c)

Figure 20.28 Loose bodies (a) This loose body slipped away from the fingers when touched; the term 'joint mouse' seems appropriate. (b) Which is the loose body here? Not the large one (which is a normal fabella), but the small lower one opposite the joint line. (c) Multiple loose bodies are seen in synovial chondromatosis, a rare disorder of cartilage metaplasia in the synovium.

Clinical features

Loose bodies may be symptomless, but the commonest complaint is attacks of sudden locking without injury. The joint gets stuck in a position which varies from one attack to another. Sometimes the locking is only momentary and usually the patient can wriggle the knee until it suddenly unlocks. The patient may be aware of something 'popping in and out of the joint'.

In adolescents, a loose body is usually due to osteochondritis dissecans while in adults osteoarthritis is a more frequent cause.

Only rarely is the patient seen with the knee still locked. Sometimes, especially after the first attack, there is synovitis or there may be evidence of the underlying cause. A pedunculated loose body may be felt; one that is truly loose tends to slip away during palpation (the well-named 'joint mouse').

Imaging

Most loose bodies are radio-opaque on *X-ray*. The films may also show an underlying joint abnormality. An *MRI scan* is required to assess the activity of the site of the OCD lesion.

Treatment

A loose body causing symptoms should be removed unless the joint is severely osteoarthritic. This can usually be done through the arthroscope, but finding the loose body may be difficult; it may be concealed in a synovial pouch or sulcus, and a small body may even slip under the edge of one of the menisci.

SYNOVIAL CHONDROMATOSIS

This is a rare disorder in which the joint comes to contain multiple loose bodies, often in pearly clumps resembling sago ('snowstorm knee'). The usual explanation is that myriad tiny fronds undergo cartilage metaplasia at their tips; these tips break free and may ossify. It has, however, been suggested that chondrocytes may be cultured in the synovial fluid and that some of the products are then deposited onto previously normal synovium, so producing the familiar appearance. X-rays reveal multiple loose bodies; on arthrography they show as negative defects.

The loose bodies should be removed arthroscopically and a synovectomy performed.

PLICA SYNDROME

A plica is the remnant of an embryonic synovial partition which persists into adult life. During development of the embryo, the knee is divided into three cavities – a large suprapatellar pouch and beneath this the medial and lateral compartments – separated from each other by membranous septa. Later these partitions disappear, leaving a single cavity, but part of a septum may persist as a synovial pleat or plica (from the Latin *plicare* = to fold). This is found in over 20% of people, usually as a *median infrapatellar fold* (the ligamentum mucosum), less often as a *suprapatellar curtain* draped across the opening of the suprapatellar pouch or a *mediopatellar plica* sweeping down the medial wall of the joint.

Pathology

The plica itself is not pathological. But if acute trauma, repetitive strain or some underlying disorder (e.g. a meniscal tear) causes inflammation, the plica may become oedematous, thickened and eventually fibrosed; it then acts as a tight bowstring, impinging on other structures in the joint and causing further synovial irritation.

Clinical features

An adolescent or young adult complains of an ache in the front of the knee (occasionally both knees), with intermittent episodes of clicking or 'giving way'. There may be a history of trauma or markedly increased activity. Symptoms are aggravated by exercise or climbing stairs, especially if this follows a long period of sitting.

On examination there may be muscle wasting and a small effusion. The most characteristic feature is tenderness near the upper pole of the patella and over the femoral condyle. Occasionally the thickened band can be felt. Movement of the knee may cause catching or snapping.

Diagnosis

There is still controversy as to whether 'plica syndrome' constitutes a real and distinct clinical entity. Some clinicians believe it is a significant cause of anterior knee pain. However, it may closely resemble other conditions such as patellar overload or subluxation; indeed, the plica may become troublesome only when those other conditions are present. The diagnosis is often not made until arthroscopy is undertaken. The presence of a chondral lesion on the femoral condyle secondary to plica impingement confirms the diagnosis.

Treatment

The first line of treatment is rest, anti-inflammatory drugs and adjustment of activities. If symptoms persist, the plica can be divided or excised by arthroscopy.

TUBERCULOSIS

Tuberculosis of the knee may appear at any age, but it is more common in children than in adults.

Clinical features

Early presentation Pain and limp are early features; or the child may present with a swollen joint and a low-grade fever. The thigh muscles are wasted, thus accentuating the joint swelling. The knee feels warm and there is synovial thickening. Movements are restricted and often painful. The Mantoux test is positive and the erythrocyte sedimentation rate (ESR) may be increased.

X-rays show marked osteoporosis and, in children, enlargement of the bony epiphyses (Figure 20.29). Unlike pyogenic arthritis, joint space narrowing is a late sign; this is because cartilage lysis is prevented by the presence of a plasmin inhibitor in the synovial exudate.

Late features If the disease is allowed to persist, the joint surfaces will gradually be eroded and the knee joint will become deformed. The classical picture in neglected cases is a composite deformity: posterior and lateral subluxation or dislocation, external rotation and fixed flexion.

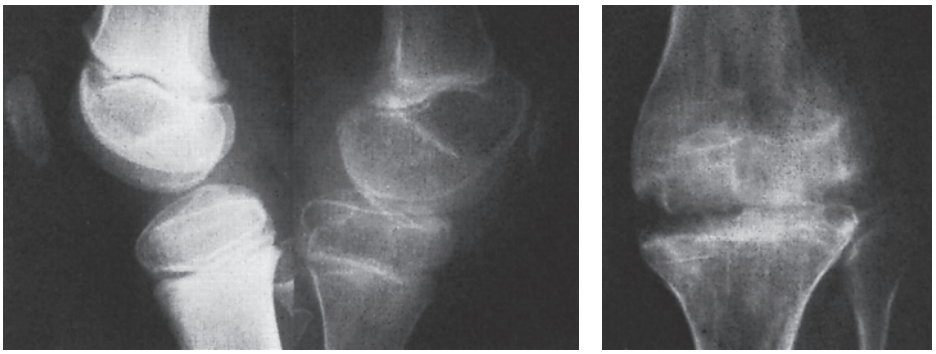
Diagnosis

Mono-articular rheumatoid synovitis, or juvenile chronic arthritis, may closely resemble tuberculosis. A synovial biopsy may be necessary to establish the diagnosis.

Treatment

General antituberculous chemotherapy should be given for 12–18 months (see Chapter 2).

In the active stage the knee is rested in a bed splint. The synovitis usually subsides but, if it does not do so after a few weeks' treatment, surgical debridement



(a) (b)

Figure 20.29 Tuberculosis (a) Lateral views of the two knees. On one side the bones are porotic and the epiphyses enlarged, features suggestive of a severe inflammatory synovitis. (b) Later the articular surfaces are eroded.

will be needed. All obviously diseased and necrotic tissue is removed and bone abscesses are evacuated.

In the healing stage the patient is allowed up wearing a weight-relieving caliper. Gradually this is left off, but the patient is kept under observation for any sign of recurrent inflammation. If the articular cartilage has been spared, movement can be encouraged and weight-bearing is slowly resumed. However, if the articular surface is destroyed, immobilization is continued until the joint stiffens.

In the aftermath the joint may be painful; it is then best arthrodesed, but in children this is usually postponed until growth is almost completed. The ideal position for fusion is 10–15 degrees of flexion, 7 degrees of valgus and 5 degrees of external rotation.

In some cases, once it is certain that the disease is quiescent, joint replacement may be feasible.

RHEUMATOID ARTHRITIS

Clinical features

Occasionally, rheumatoid arthritis starts in the knee as a chronic monarticular synovitis. Sooner or later, however, other joints become involved.

The general features of rheumatoid disease are described in Chapter 3.

The early stage is characterized by synovitis; rheumatoid disease occasionally starts with involvement of a single joint. The patient complains of pain and chronic swelling of the knee; there is usually an effusion and the thigh muscles may be wasted. The thickened synovium is often palpable.

Unless there are obvious signs of an inflammatory polyarthritis, the condition has to be distinguished from other types of inflammatory monarthritis, such as gout, Reiter's disease and tuberculosis; biopsy and microbiological investigations may be needed.

During this early stage, while the joint is still stable and the muscles are reasonably strong, there is a danger of rupturing the posterior capsule; the joint contents are extruded into a large posterior bursa or between the muscle planes of the calf, causing sudden pain and swelling which closely mimic the features of calf vein thrombosis (Figure 20.30).

As the disease progresses the knee becomes increasingly unstable, muscle wasting is marked and there is some loss of flexion and extension.

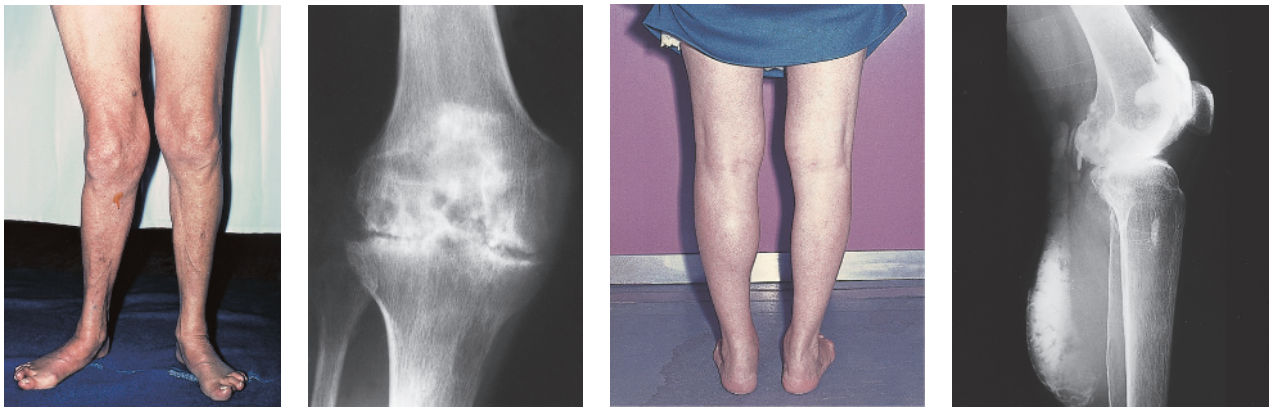
X-rays may show diminution of the joint space, osteopenia and marginal erosions. The picture is easily distinguishable from that of osteoarthritis by the complete absence of osteophytes.

In the late stage pain and disability are usually severe. In some patients stiffness is so marked that the patient has to be helped to stand and the joint has only a jog of painful movement. In others, cartilage and bone destruction predominate and the joint becomes increasingly unstable and deformed, usually in fixed flexion and valgus. X-rays reveal the bone destruction characteristic of advanced disease.

Treatment

CONSERVATIVE MANAGEMENT

The majority of patients can be managed by conservative measures. In addition to general treatment with anti-inflammatory and disease-modifying drugs, local splintage and injection of corticosteroid usually help to reduce the synovitis. A more prolonged effect may be obtained by injecting radiocolloids such as yttrium-90 (90Y). A significant advance in treatment has been the introduction of anti-TNF medication, which can stop the



(a) (b) (c) (d)

Figure 20.30 Rheumatoid arthritis (a) Patient with rheumatoid arthritis showing the typical valgus deformity of the right knee; the feet and toes also are affected. (b) X-ray showing marked erosive arthritis resulting in joint deformity. (c) This patient presented with a painful swelling of the left calf. She was thought at first to have developed deep vein thrombosis – until we examined her knee and recognized this as a posterior synovial rupture, later confirmed by arthrography (d).

inflammatory process within the joint and prevent longer-term joint destruction.

OPERATIVE TREATMENT

Synovectomy and debridement Only if other measures fail to control the synovitis (which nowadays is rare) is arthroscopic synovectomy indicated, at which time articular pannus and cartilage tags are removed. Postoperatively, any haematoma must be drained and movements are commenced as soon as pain has subsided.

Arthroplasty Total joint replacement is useful when joint destruction is advanced. However, it is less successful if the knee has been allowed to become very unstable or very stiff; timing of the operation is important. Care must be taken to preserve the collateral ligaments in this type of reconstruction.

OSTEOARTHRITIS

The knee is the commonest of the large joints to be affected by osteoarthritis, with many members of the population having symptoms and corresponding changes in their knee joints (see Chapter 5). Often there is a predisposing factor: injury to the articular surface, a torn meniscus, ligamentous instability or pre-existing deformity of the hip or knee, to mention a few. However, in many cases no obvious cause can be found. Underlying all of these factors is a significant genetic predisposition to the condition. Curiously, while the male:female distribution is more or less equal in white (Caucasian) peoples, black African women are affected far more frequently than their male counterparts.

Osteoarthritis is often bilateral and in these cases there is a strong association with Heberden's nodes and generalized osteoarthritis.

Pathology

Osteoarthritis can affect all of the soft tissues around the knee but articular cartilage breakdown is a consistent feature which usually starts in an area of excessive loading. These produce a number of typical patterns of articular cartilage damage. Disease localized to the medial compartment is the commonest pattern occurring, producing varus deformity to normal limb alignment. Disease can less commonly be isolated to the lateral or patellofemoral compartments. In more advanced disease the damage extends to more than one compartment. Other intra-articular characteristic features found include peripheral osteophyte formation, bone loss, degenerative change to the menisci and anterior cruciate ligament (ACL) destruction.

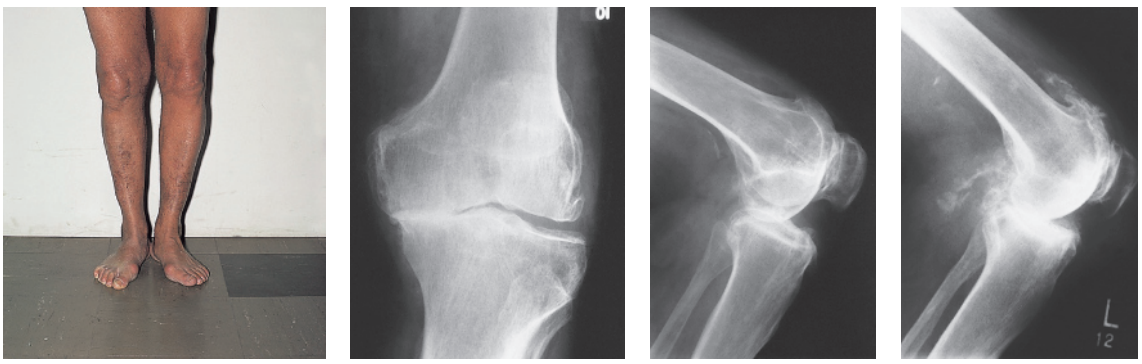
Concurrent chondrocalcinosis is relatively common, but whether this is cause or effect – or quite unrelated – remains unknown.

Clinical features

Patients are usually over 50 years old and are often overweight.

Pain is the leading symptom, worse after use, or (if the patellofemoral joint is affected) on stairs. After rest, the joint feels stiff and it hurts to 'get going' after sitting for any length of time. Swelling is common, and giving way or locking may occur.

On examination there may be an obvious deformity (usually varus as the medial compartment is often more severely affected) or the scar of a previous operation (Figure 20.31). The quadriceps muscle is usually wasted.



(a) (b) (c) (d)

Figure 20.31 Osteoarthritis (a,b) Varus deformity of the left knee suggesting loss of cartilage thickness in the medial compartment. X-ray shows diminished joint space and peripheral osteophytes on the medial side of the knee. (c) Sometimes it is the patellofemoral joint that is mainly affected. (d) Patellofemoral osteoarthritis with long trailing osteophytes is typical of calcium pyrophosphate arthropathy.

During an exacerbation the knee can develop an effusion and an associated synovitis. Movement becomes increasingly limited and is often accompanied by crepitus. One of the earliest signs of osteoarthritis is loss of the last few degrees of terminal extension as a fixed flexion deformity develops due to shortening of the posterior capsule.

In the development of the disease deformity is correctable, but deformities that become more severe can become 'fixed' where correction cannot be achieved passively during examination.

The natural history of osteoarthritis is one of alternating 'bad spells' and 'good spells'. Patients may experience long periods of lesser discomfort and only moderate loss of function, followed by exacerbations of pain and stiffness (perhaps after unaccustomed activity). In general, worsening symptoms tend to produce a significant reduction in a patient's quality of life.

Imaging

Standard X-rays of the knee to investigate OA include an anteroposterior view that is best obtained with the patient standing and bearing weight (only in this way can small degrees of articular cartilage thinning be revealed) (Figure 20.32), and a lateral view. Typically, the tibiofemoral joint space is diminished (often only in one compartment) and there is subchondral sclerosis. Osteophytes and subchondral cysts are usually present and sometimes there is soft-tissue calcification in the suprapatellar region or in the joint itself (chondrocalcinosis).

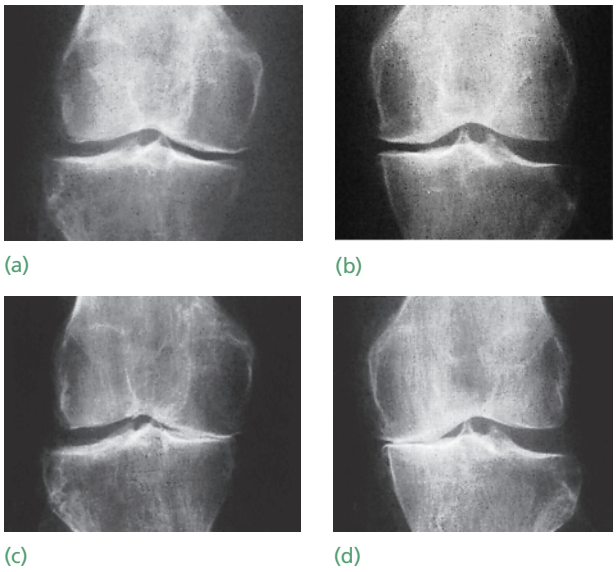


Figure 20.32 Osteoarthritis – X-rays Always obtain weight-bearing views of the knees. X-rays taken with the patient lying down (a,b) suggest only minor cartilage loss on the medial side of each knee. (c,d) Weight-bearing views show the true position: there is severe loss of articular cartilage.

An *MRI scan* can produce additional useful information concerning the soft tissues of the knee (e.g. state of the ACL) and the degree of articular cartilage damage across the whole knee.

Treatment

NON-OPERATIVE TREATMENT

The initial treatment for nearly all patients with OA is non-operative, including good information about the disease, exercise and weight-loss if appropriate. Physiotherapy is important to help produce gradual strengthening of the quadriceps muscles and increase the level of exercise being undertaken by the patient. In addition, analgesics should be prescribed for pain.

Joint loading is lessened by using a walking stick. In addition, a knee off-loader brace may reduce deformity and reduce symptoms around the knee.

Intra-articular corticosteroid injections will often relieve pain and can be used repeatedly.

New forms of medication have been introduced in recent years and intra-articular injection of hyalourans is used as a treatment option. Unfortunately, there is little data to suggest the long-term efficacy of these treatments.

OPERATIVE TREATMENT

Persistent pain unresponsive to conservative treatment, progressive deformity and instability are the usual indications for operative treatment.

Arthroscopic washout This has been shown to be ineffective in treating knee OA and should not be offered to patients.

Patellectomy Once commonly performed, patellectomy is no longer used routinely in current practice.

Realignment osteotomy Realignment osteotomy around the knee is often successful in relieving symptoms and staving off the need for 'end-stage' surgery. The ideal indication is a 'young' patient (under 55 years) with a varus knee and osteoarthritis confined to the medial compartment. In this case a high (opening wedge) tibial valgus osteotomy will redistribute weight to the lateral side of the joint. The degree and accuracy of angular correction are the most important determinants of mid- and long-term clinical outcome.

Replacement arthroplasty Replacement is indicated in patients with bone-on-bone OA changes within the knee who describe moderate to severe symptoms that have not been responsive to at least 3 months of non-operative measures. Total knee replacement (the most commonly used) is effectively a 'resurfacing' procedure, using a metal femoral condylar component and a tibial component that is most typically a metal

and polyethylene modular component but monobloc tibial components are sometimes used. If the disease is largely confined to one compartment, a unicompartmental replacement can be performed as an alternative to osteotomy or total joint replacement. The ACL must be intact for this approach. With modern techniques, and meticulous attention to anatomical alignment of the knee, the results of replacement arthroplasty are excellent, with approximately 85% of patients reporting satisfaction and a large improvement in quality of life after the operation. However, up to 15% of patients do not do well and a proportion of these can be dissatisfied with the outcome of their surgery.

Arthrodesis This is indicated only if there is a strong contraindication to arthroplasty (e.g. previous infection) or to salvage a failed arthroplasty.

OSTEONECROSIS

Osteonecrosis of the knee, though not as common as femoral head necrosis, has the same aetiological and pathogenetic background (see Chapter 6). The usual site is the dome of one of the femoral condyles, but occasionally the medial tibial condyle is affected. Two main categories are identified: (1) *osteonecrosis associated with a definite background disorder* (e.g. corticosteroid therapy, alcohol abuse, sickle-cell disease, hyperbaric decompression sickness, systemic lupus erythematosus (SLE) or Gaucher's disease); and (2) *'spontaneous' osteonecrosis* of the knee, popularly known by the acronym SONK, which is due to a small insufficiency fracture of a prominent part of the osteoarticular surface in osteoporotic bone; the vascular supply to the free fragment is compromised.

A third type, *postmeniscectomy osteonecrosis,* has been reported; its prevalence and pathophysiology are still unclear.

Clinical features

Patients are usually over 60 years old and women are affected three times more often than men. Typically they give a history of sudden, acute pain on the medial side of the joint. Pain at rest also is common.

On examination there is usually a small effusion, but the classic feature is tenderness on pressure upon the medial femoral or tibial condyle rather than along the joint line proper.

The patient may offer a history of similar symptoms in the hip or the shoulder. Whether or not this is the case, those joints should be examined as well.

Imaging

The *X-ray* appearances are often unimpressive at the beginning, but a radionuclide scan may show increased activity on the medial side of the joint. Later the classic radiographic features of osteonecrosis appear (see Chapter 6). On the femoral side, it is always the *dome* of the condyle that is affected (Figure 20.33), unlike the picture in osteochondritis dissecans.

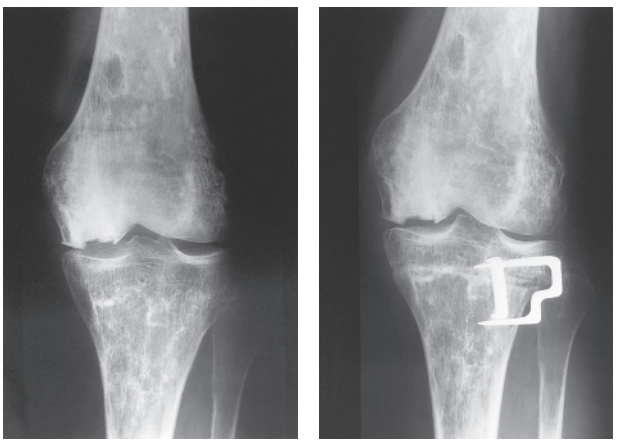
MRI enhances the ability to visualize bone marrow and to separate necrotic from viable areas with a high level of specificity. It shows the area of reactive bone surrounding the osteonecrotic lesion and can demonstrate the integrity of the overlying cortical shell of bone and articular cartilage. Sequential MRI scans are helpful in determining the clinical course of the condition (resolution or collapse).

Special investigations

Once the diagnosis is confirmed, usually after MRI, investigations should be carried out to exclude generalized disorders known to be associated with osteonecrosis (see Chapter 6).

Differential diagnosis

Osteonecrosis of the knee should be distinguished from osteochondritis dissecans, although in truth the two conditions are closely related; however, the age group, aetiology, site of the lesion and prognosis are different, and these factors may influence treatment. Other conditions that can have a sudden, painful onset and tenderness at the joint line are a flare of osteoarthritic pain, disruption of a degenerative



(a) (b)

Figure 20.33 Osteonecrosis (a) X-ray showing the typical features of subarticular bone fragmentation and surrounding sclerosis situated in the highest part (the dome) of the medial femoral condyle. (In osteochondritis dissecans, the necrotic segment is almost always on the lateral surface of the medial femoral condyle.) (b) In this case the medial compartment was 'unloaded' by performing a high tibial valgus osteotomy. The patient remained pain-free for 6 years before dying of leukaemia.

meniscus, a stress fracture, pes anserinus bursitis and a local tendonitis.

Prognosis

Symptoms and signs may stabilize and the patient may be left with no more than slight distortion of the articular surface; or one of the condyles may collapse, leading to osteoarthritis of the affected compartment. The clinical progress depends on the radiographic size of the lesion, the ratio of size of the lesion to the size of the condyle (>40% carries a worse prognosis) and the stage of the lesion.

Treatment

Treatment is conservative in the first instance and consists of measures to reduce loading of the joint and analgesics for pain. If symptoms or signs increase, operative treatment may be considered.

Bisphosphonate medications may have a positive role in encouraging healing but the evidence for their efficacy is only beginning to emerge. Surgical options include drilling, bone grafting, core decompression of the femoral condyle at a distance from the lesion, osteotomy for patients with persistent symptoms and well-marked articular surface damage or unicompartmental arthroplasty if the femoral condyle collapses.

CHARCOT'S DISEASE

Charcot's disease (neuropathic arthritis) is a rare cause of joint destruction. Because of loss of pain sensibility and proprioception, the articular surface breaks down and the underlying bone crumbles. Fragments of bone and cartilage are deposited in the hypertrophic synovium and may grow into large masses. The capsule is stretched and lax, and the joint becomes progressively unstable.

Clinical features

The patient complains chiefly of instability; pain (other than tabetic lightning pains) is unusual. The joint is swollen and often grossly deformed. It feels like a bag of bones and fluid but is neither warm nor tender. Movements beyond the normal limits, without pain, are a notable feature. Radiologically the joint is subluxated, bone destruction is obvious and irregular calcified masses can be seen.

Treatment

Patients often seem to manage quite well despite the bizarre appearances. However, marked instability may demand treatment. In this case a moulded splint or caliper may control the joint, but if pain becomes intolerable, arthrodesis is feasible. In these cases fixation can be difficult and fusion is usually very slow. Replacement arthroplasty is not indicated.

HAEMOPHILIC ARTHRITIS

The knee is the joint most commonly involved in bleeding disorders. Repeated haemorrhage leads to chronic synovitis and articular cartilage erosion. Movement is progressively restricted and the joint may end up deformed and stiff.

Clinical features

Fresh bleeds cause pain and swelling of the knee, with the typical clinical signs of a haemarthrosis (see Chapter 5). Between episodes of bleeding the knee often continues to be painful and somewhat swollen, with restricted mobility. There is a tendency to hold the knee in flexion, and this may become a fixed deformity.

Imaging

Radiographic examination may show little abnormality on X-rays, apart from local osteoporosis. In more advanced cases the joint space is narrowed and large 'cysts' or erosions may appear in the subchondral bone. In the severe late stage marked deformity is often displayed on the imaging.

Treatment

Modern medical management of this condition, coordinated by haematologists, has been revolutionized by access to clotting factors that can prevent recurrent haemarthrosis and the ensuing joint damage.

Both the haematologist and the orthopaedic surgeon should participate in treatment, which may involve surgical intervention. The acute bleed may need aspiration, but only if this can be 'covered' by giving the appropriate clotting factor; otherwise it is better treated by splintage until the acute symptoms settle down.

Flexion deformity must be prevented by gentle physiotherapy and intermittent splintage. If the joint is painful and eroded, operative treatment may be considered. However, although replacement arthroplasty is feasible, this should be done only after the most searching discussion with the patient, where all the risks are considered, and only if a full haematological service is available. Joint replacement can reduce pain but patients are often left with a residual postoperative functional deficit.

RUPTURES OF THE EXTENSOR APPARATUS

Resisted extension of the knee may tear the extensor mechanism (Figure 20.34). The patient stumbles on a stair, catches his or her foot while walking or running, or may only be kicking a muddy football. In all these incidents, active knee extension is prevented by an obstacle. The precise location of the lesion varies with the patient's age. In the elderly the injury is usually above the patella; in middle life the patella fractures; in young adults the patellar ligament can rupture. In adolescents the upper tibial apophysis is occasionally avulsed; much more often it is merely 'strained'.

Tendon rupture sometimes occurs with minimal strain; this is seen in patients with connective-tissue disorders (e.g. SLE) and advanced rheumatoid disease, especially if they are also being treated with corticosteroids.

Rupture above the patella

Rupture may occur in the belly of the rectus femoris. The patient is usually elderly, or on long-term corticosteroid treatment. The torn muscle retracts and forms a characteristic lump in the thigh. Function is usually good, so no treatment is required.

Avulsion of the quadriceps tendon from the upper pole of the patella is seen in the same group of people, always follows a traumatic event and can be bilateral. Direct operative repair with sutures, usually augmented by passing through tunnels through the patella, is essential.

Rupture below the patella

This occurs mainly in young people. The ligament may rupture or may be avulsed from the lower pole of the patella (a sleeve fracture). Operative repair, again usually through tunnels in the patella, is necessary unless a sleeve fracture is minimally displaced, where splinting will suffice. Following treatment, pain and tenderness in the middle portion of the patellar ligament may occur in athletes; CT or ultrasonography will reveal an abnormal area. If rest fails to provide relief the paratenon should be stripped.

Partial rupture or avulsion sometimes leads to a traction tendinitis and calcification in the patellar ligament – the *Sinding-Larsen Johansson syndrome* (see below).

Osgood–Schlatter disease ('apophysitis' of the tibial tubercle)

In this common disorder of adolescence the tibial tubercle becomes painful and 'swollen'. Although often called osteochondritis or apophysitis, it is simply a traction injury of the apophysis into which part of the patellar tendon is inserted (the remainder is inserted on each side of the apophysis and prevents complete separation).

There is no history of injury and sometimes the condition is bilateral. A young adolescent complains of pain after activity, and of a lump (Figure 20.35). The lump is tender and its situation over the tibial tuberosity is diagnostic. Sometimes active extension of the knee against resistance is painful and X-rays may reveal fragmentation of the apophysis. An MRI scan confirms this and excludes other causes of pain.

Spontaneous recovery is usual but takes time, and it is wise to restrict such activities as cycling, jumping and soccer. Occasionally, symptoms persist and a separate ossicle in the tendon can be identified. In these rare situations surgical removal can be considered.

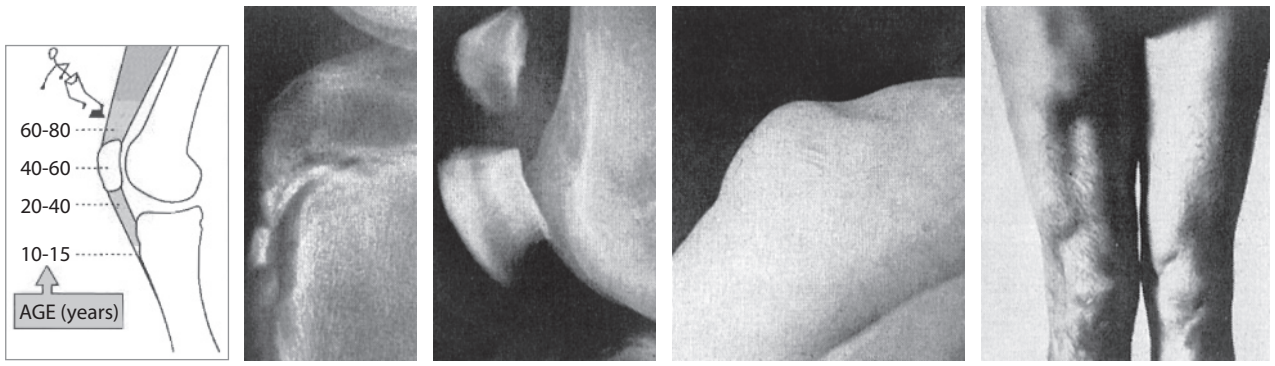
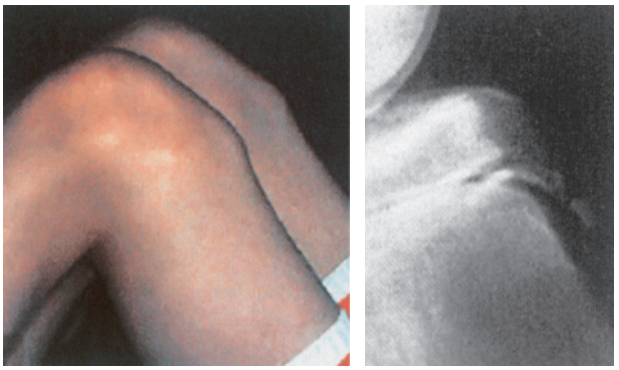


Figure 20.34 Extensor mechanism lesions These follow resisted action of the quadriceps; they usually occur at a progressively higher level with increasing age (a). (b) Osgood-Schlatter disease – the only one that usually does not follow a definite accident; (c) displaced fracture of patella; (d) ruptured quadriceps tendon (note the suprapatellar depression); (e) ruptured rectus femoris causing a lump with a hollow below.



(a) (b)

Figure 20.35 Osgood–Schlatter disease This boy complained of a painful bump below the knee. X-ray shows the traction injury of the tibial apophysis.

TENDINITIS AND CALCIFICATION AROUND THE KNEE

Calcification in the medial ligament

Acute pain in the medial collateral ligament may be due to a soft calcific deposit among the fibres of the ligament, although this is relatively uncommon. There may be a small, exquisitely tender lump in the line of the ligament. Pain is dramatically relieved by operative evacuation of the deposit.

Pellegrini–Stieda disease

X-rays sometimes show a plaque of bone lying next to the femoral condyle under the medial collateral ligament. Occasionally this is a source of pain. It is generally ascribed to ossification of a haematoma following a tear of the medial ligament, although a history of injury is not always forthcoming. No specific treatment is required.

Patellar 'tendinopathy' (Sinding-Larsen Johansson syndrome)

This condition was described independently by Sinding-Larsen in 1921 and Johansson in 1922. Following repetitive strain or a partial rupture of the patellar ligament the patient (usually a young athletic individual) develops a traction 'tendinitis' characterized by pain and point tenderness at the lower pole of the patella. Sometimes, if the condition does not settle, calcification appears in the ligament. CT or ultrasonography may reveal the abnormal area in the ligament. A similar disorder has been described at the proximal pole of the patella (jumper's knee).

The condition is comparable to Osgood–Schlatter disease and usually recovers spontaneously, although it may be very disabling during its prolonged course. If rest fails to provide relief over the longer term, injections, high frequency ultrasound therapy and very occasionally surgical decompression of the tendon with removal of the abnormal area may be required.

SWELLINGS OF THE KNEE

The knee is prone to a number of disorders which present essentially as 'swelling' (Figure 20.36); and, because it is such a large joint with a number of synovial recesses, the swelling is often painless until the tissues become tense. Malignancy must be excluded, although fortunately the majority of swellings are benign. An MRI scan is thus mandatory for the investigation of these conditions. They can be divided into four groups: *swelling of the entire joint* (Figure 20.36), *swellings in front of the joint* and *swellings behind the joint* (Figure 20.37) and *bony swellings.*

Acute swelling of the entire joint

POST-TRAUMATIC HAEMARTHROSIS

Swelling immediately after injury means blood in the joint. The knee is very painful and it feels warm, tense and tender. Later there may be a 'doughy' feel. Movements are restricted. X-rays are essential to see if there is a fracture; if there is not, suspect a tear of the anterior cruciate ligament (ACL).

If the joint is very tight, aspiration under aseptic conditions can relieve pain and help diagnosis. If a ligament injury is suspected, examination under anaesthesia is helpful and may indicate the need for operation; otherwise a crepe bandage is applied and the leg cradled in a back-splint. Quadriceps exercises are practised from the start. The patient may get up when comfortable, retaining the back-splint until muscle control returns. An MRI scan is then performed to identify the cause of the bleed (e.g. ACL damage).

BLEEDING DISORDERS

In patients with clotting disorders, the knee is the most common site for acute bleeds. If the appropriate clotting factor is available, the joint should be aspirated and treated as for a traumatic haemarthrosis. If the factor is not available, aspiration is best avoided; the knee is splinted in slight flexion until the swelling subsides.

ACUTE SEPTIC ARTHRITIS

Acute pyogenic infection of the knee is an acute emergency and requires prompt treatment to prevent systemic spread of the infection and to prevent local damage to the knee. The organism is usually *Staphylococcus aureus*, but in adults gonococcal infection can occur.

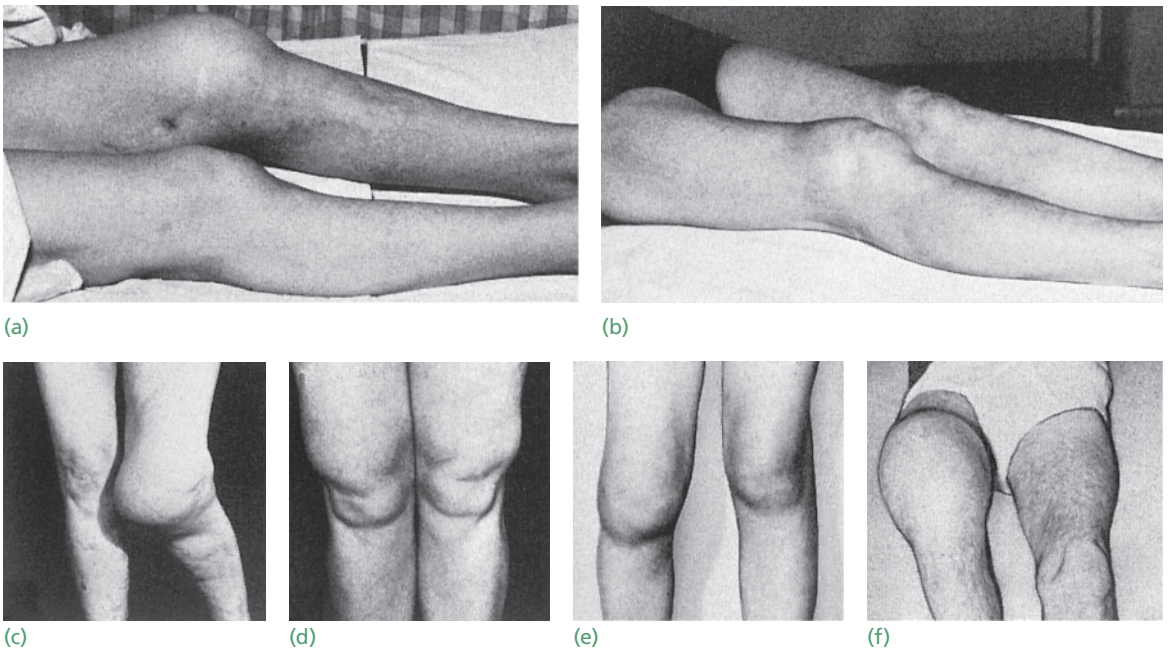


Figure 20.36 Swollen knees Some causes of swelling of the knee in the absence of trauma: (a) tuberculous arthritis; (b) rheumatoid arthritis; (c) Charcot's disease; (d) villous synovitis; (e) haemophilia; (f) malignant synovioma, considered whenever there is no obvious alternative diagnosis. Investigations should include Mantoux testing and synovial biopsy. The ideal is to start antituberculous chemotherapy before joint destruction occurs.

The joint is swollen, painful and inflamed. The patient reports extreme pain on movement of the joint, which can be confirmed during careful examination. Blood tests reveal an elevated white cell count and ESR. Aspiration reveals pus in the joint; fluid should be sent for bacteriological investigation, including anaerobic culture.

Treatment consists of systemic antibiotics and drainage of the joint – ideally by arthroscopy, with irrigation and complete synovectomy; if fluid reaccumulates, it can be aspirated through a wide-bore needle. As the inflammation subsides, movement is begun, but weight-bearing is deferred for 4–6 weeks.

TRAUMATIC SYNOVITIS

Injury stimulates a reactive synovitis; typically the swelling appears only after some hours, and subsides spontaneously over a period of days. There is inhibition of quadriceps action and the thigh wastes. The knee may need to be splinted for several days but movement should be encouraged and quadriceps exercise is essential. If the amount of fluid is considerable, its aspiration hastens muscle recovery. In addition, any internal injury will need treatment.

ASEPTIC NON-TRAUMATIC SYNOVITIS

Acute swelling, without a history of trauma or signs of infection, suggests *gout*, *pseudogout* or *inflammatory monarthropathy*. Aspiration will provide fluid which may look turbid, resembling pus, but it is sterile and microscopy (using polarized light) reveals the crystals. Treatment with anti-inflammatory drugs is usually effective while investigation as to the cause of the swelling begins.

CHRONIC SWELLING OF THE JOINT

The diagnosis can usually be made on clinical and X-ray/MRI examination. The more elusive disorders should be fully investigated by joint aspiration, synovial fluid examination, arthroscopy and synovial biopsy.

ARTHRITIS

The commonest causes of chronic swelling are *osteoarthritis* and *inflammatory arthritis*. Other signs, such as deformity, loss of movement or instability, may be present and X-ray examination will usually show characteristic features.

SYNOVIAL DISORDERS

Chronic swelling and synovial effusion without articular destruction should suggest conditions such as *synovial chondromatosis* and *pigmented villonodular synovitis*. The diagnosis will usually be obvious from an MRI scan and can be confirmed by arthroscopic synovial biopsy.

The most important condition to exclude is *tuberculosis*. There has been a resurgence of cases during

the last ten years and the condition should be considered seriously.

Swellings in front of the joint

PREPATELLAR BURSITIS ('HOUSEMAID'S KNEE')

The fluctuant swelling is confined to the front of the patella and the joint itself is normal. This is a bursitis due to recurrent friction between skin and bone and is seen mainly in carpet layers, paving workers, floor cleaners and miners who do not use protective knee pads. Treatment consists of firm bandaging, and kneeling is avoided; occasionally aspiration is needed. In chronic cases the lump is best excised.

Secondary infection (possibly due to foreign body implantation) results in a warm, tender swelling. Treatment is by rest, antibiotics and, if necessary, aspiration or excision.

INFRAPATELLAR BURSITIS ('CLERGYMAN'S KNEE')

The swelling is below the patella and superficial to the patellar ligament, being more distally placed than prepatellar bursitis; it used to be said that one who prays kneels more uprightly than one who scrubs! Treatment is similar to that for prepatellar bursitis. Occasionally the bursa is affected in gout.

OTHER BURSAE

Occasionally a bursa deep to the patellar tendon or the pes anserinus becomes inflamed and painful. Treatment is non-operative.

Swellings at the back of the knee

SEMIMEMBRANOSUS BURSA

The bursa between the semimembranosus and the medial head of gastrocnemius may become enlarged in children or adults. It presents usually as a painless lump behind the knee, slightly to the medial side of the midline and most conspicuous with the knee straight. The lump is fluctuant but the fluid cannot be pushed into the joint, presumably because the muscles compress and obstruct the normal communication. The knee joint is normal. Recurrence is common if excision is attempted and, as the bursa normally disappears in time, a waiting policy is the treatment of choice.

POPLITEAL 'CYST'

Bulging of the posterior capsule and synovial herniation may produce a swelling in the popliteal fossa. The lump, which is usually seen in older people, is in the midline of the limb and at or below the level of the joint. It fluctuates but is not tender. Injection of radio-opaque medium into the joint, and X-ray, will show that the 'cyst' communicates with the joint.

The condition was originally described by Baker, whose patients were probably suffering from tuberculous synovitis. Nowadays it is more likely to be caused by osteoarthritis, but it is still often called a 'Baker's cyst'. Occasionally the 'cyst' ruptures and the synovial contents spill into the muscle planes causing pain and swelling in the calf – a combination which can easily be mistaken for deep vein thrombosis.

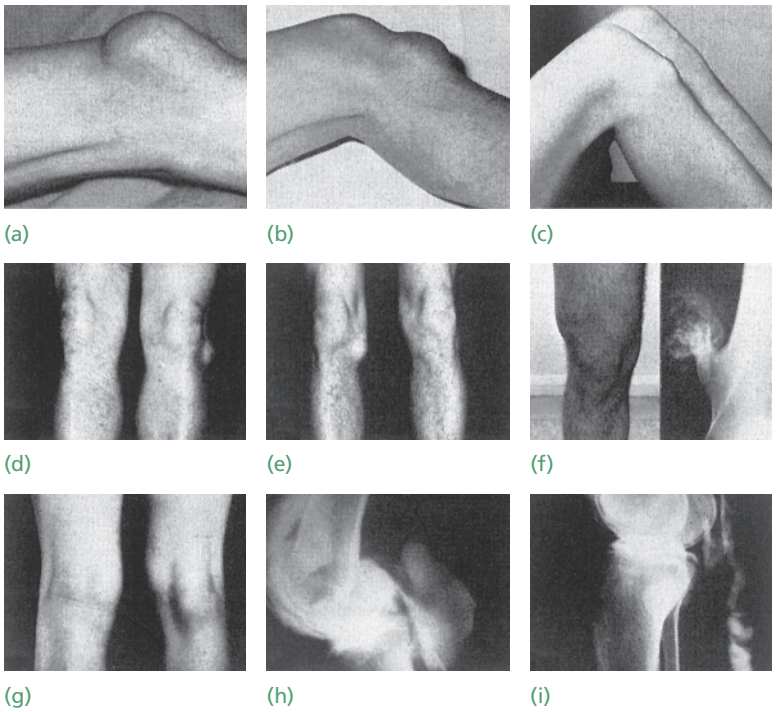


Figure 20.37 Lumps around the knee
In front: (a) prepatellar bursa; (b) infrapatellar bursa; (c) Osgood–Schlatter disease. On either side: (d) cyst of lateral meniscus; (e) cyst of medial meniscus; (f) cartilage-capped exostosis. Behind: (g) semimembranosus bursa; (h) arthrogram of popliteal cyst; (i) leaking cyst.

The swelling may diminish following aspiration and injection of corticosteroid; excision is not advised, because recurrence is common unless the underlying condition is treated.

POPLITEAL ANEURYSM

This is the commonest limb aneurysm and is sometimes bilateral. Pain and stiffness of the knee may precede the symptoms of peripheral arterial disease, so it is essential to examine any lump behind the knee for pulsation. A thrombosed popliteal aneurysm does not pulsate, but it feels almost solid.

Bony swellings around the knee

Because the knee is a relatively superficial joint, bony swellings of the distal femur and proximal tibia are often visible and almost always palpable. Common examples are cartilage-capped exostoses (osteochondromata) and the characteristic painful swelling of Osgood–Schlatter disease of the tibial tubercle (see above).

Malignancy, although rare, must be excluded, and an MRI scan is mandatory in the investigation of bony swellings around the knee that display any concerning features. Features that suggest malignancy are often referred to as 'red flags' and include disproportionate pain, sudden increase in size and a history of previous malignancy.

PRINCIPLES OF KNEE OPERATIONS

Arthroscopy

Arthroscopy is useful: (1) to establish or refine the accuracy of diagnosis, using the information to perform or not perform reconstructive surgery (e.g. performing a synovial biopsy or determining whether the ACL is deficient or not when there is a mismatch between MRI scan findings and patient symptoms); and (2) to perform certain operative procedures (e.g. meniscectomy, cartilage repair techniques, ligament reconstruction). Arthroscopy is not a substitute for clinical examination; a detailed history and meticulous assessment of the physical signs are indispensable preliminaries and remain the sheet anchor of diagnosis. Emerging evidence suggests that the indications for arthroscopic intervention are narrowing and that intervention rates may well reduce with time.

Technique

The patient is anaesthetized (though local anaesthesia may suffice for short procedures) and a thigh tourniquet usually applied. Through a tiny incision, a trocar and cannula is introduced; sometimes, saline is injected to distend the joint before it is punctured. Entry into the joint is confirmed when saline flows easily into the joint or, if the joint was distended previously, by the outflow when the trocar is withdrawn. A fibre-optic light source, camera and irrigation system are attached. The surgeon performs the procedure viewing the operative field on a screen while manipulating the instruments and camera with each hand ('triangulation'). All compartments of the joint are now systematically inspected; with special instruments and, if necessary, through multiple portals, biopsy, partial meniscectomy, patellar shaving, removal of loose bodies, synovectomy, ligament replacement and many other procedures are possible. Before withdrawing the instrument, saline is squeezed out. A firm bandage is applied; the arthroscopic portals are often small enough not to require sutures. Postoperative recovery is remarkably rapid with most cases performed as day-case surgery.

Complications

Intra-articular effusions and small haemarthroses are fairly common but seldom troublesome.

Reflex sympathetic dystrophy (which may resemble a low-grade infection during the weeks following arthroscopy) is sometimes troublesome. It usually settles down with physiotherapy and treatment with non-steroidal anti-inflammatory drugs; occasionally it requires more radical treatment. Infection and thromboembolic disease can occur but do so at a very low rate.

Ligament reconstruction

The collateral and cruciate ligaments and the knee capsule are important constraints which allow normal knee function; laxity or rupture of these structures, either singly or in combination, is often the source of recurrent episodes of 'giving way'. Although a significant proportion of such injuries are treated non-operatively, complete ruptures may require surgery where on-going instability reduces individuals' activity levels.

Surgery for ligament reconstruction includes the following:

- *Repair*, usually for collateral ligament mid-substance ruptures when they are found in combination with other multiligament injuries. This repair can be a simple end-to-end suture or can include graft augmentation.
- *Substitution*, usually for anterior cruciate ruptures: the semitendinosus and gracilis, usually in a four-bundle technique, can be carefully anchored to the femur and tibia, ensuring that stability is restored without loss of knee movement. Another method is to use an autologous graft from the patellar tendon.

- *Tenodesis*, using a variety of tendons which are passed through either bony or soft-tissue tunnels to 'check' the abnormal movement resulting from ligament rupture.

Osteotomy

Osteotomy above or below the joint used to be a popular method of treating arthritis of the knee, especially when articular destruction was more or less limited to one compartment and the knee had developed a varus or valgus deformity. With the development of joint replacement techniques, the operation gradually fell into disuse, or at best was seen as a temporizing measure to buy time for patients who would ultimately undergo some form of arthroplasty. However, improvements in technique and the introduction of operations for meniscal and articular cartilage repair have led to renewed interest in this procedure.

The rationale for osteotomy is based on both biomechanical and physiological principles. Malalignment of the limb results in excessive loading and stress in part of the joint and consequently increased damage to the articular cartilage in that area – the medial compartment if the knee is in varus and the lateral compartment in a valgus knee. As the articular surface is destroyed, the deformity progressively increases. Osteotomy and correction of deformity will improve the load-bearing mechanics of the joint. Furthermore, it will reduce the intraosseous venous congestion, and this may relieve some of the patient's pain.

Indications

Deformity of the knee Severe varus or valgus deformity (e.g. due to a growth defect, epiphyseal injury or a malunited fracture) may of itself call for a corrective osteotomy, and the operation may also prevent or delay the development of osteoarthritis.

Localized articular surface destruction Patients with unicompartmental osteoarthritis or advanced localized osteonecrosis, particularly when this is associated with deformity in the coronal plane, may benefit from an osteotomy which offloads the affected area. Provided the joint is stable and has retained a reasonable range of movement, this offers an acceptable alternative to a unicompartmental arthroplasty. Usually it is the medial compartment that is affected and the knee exhibits a varus deformity. By realigning the joint, load is transferred from the medial compartment to the centre or a little towards the lateral side. Slight over-correction may further offload the medial compartment but marked valgus should be avoided as this will rapidly lead to cartilage loss in the lateral compartment.

Published results suggest that the operation provides substantial improvements in pain and function over a 7–10-year period.

Intra-articular reconstructions The introduction of meniscal and articular cartilage reconstruction techniques has led to considerable interest in applying the favourable biomechanical effects of osteotomy to the younger patient who has a full-thickness chondral lesion or an absent meniscus. Similarly, osteotomy in conjunction with either simultaneous or staged cruciate ligament reconstruction appears to be beneficial in patients who have a combination of instability and pain from limb malalignment.

Technique

For sound biomechanical reasons, a varus deformity is best corrected by a *valgus osteotomy* at the proximal end of the tibia, whereas a valgus deformity should be corrected by a *varus osteotomy* at the femoral supracondylar level.

Angles must be accurately measured and the position of correction carefully mapped out on X-rays before starting the operation.

A high tibial valgus osteotomy can be performed either by removing a pre-determined wedge of bone based laterally and then closing the gap (*closing wedge technique*) or by opening a wedge-shaped gap on the medial side (*opening wedge technique*).

In the lateral closing wedge method the fibula must first be released either by dividing it lower down or by disrupting the proximal tibiofibular joint. The tibia is divided just above the insertion of the patellar ligament. Two transverse cuts are made, one parallel to the joint surface and another just below that, angled to create the desired laterally based wedge. The wedge of bone is removed and the fragments are then approximated and fixed in the corrected position either with staples or with compression pins. By 4–6 weeks the osteotomy should have started to unite.

An opening wedge valgus osteotomy on the medial side offers some advantages: the ability to adjust the degree of correction intra-operatively and the option to correct deformities in the sagittal plane as well as the coronal plane; it also makes it unnecessary to disrupt the tibiofibular joint. Advances in fixation methods have reduced the need to use a bone graft and typically there is no longer an extended period of restricted weight-bearing after surgery.

If a varus osteotomy is required – usually for active patients with isolated lateral compartment disease and valgus deformity of the knee – this is performed at the supracondylar level of the femur. The method most commonly employed is a medial closing wedge osteotomy, designed to place the mechanical axis at zero. The fragments should be firmly fixed with a blade-plate; in many cases postoperative cast immobilization

will also be needed. An alternative but less frequently used method to perform osteotomy is to deploy an Ilizarov circular external fixator and gradually dynamically correct the deformity over a period of time.

Results

High tibial valgus osteotomy, when performed for osteoarthritis, gives good results provided (1) the disease is confined to the medial compartment; and (2) the knee has a good range of movement and is stable. Relief of pain is good in 85% of cases in the first year but drops to approximately 60% after 5 years. More modern medial opening wedge osteotomy techniques can achieve satisfactory postoperative alignment in 93% of patients and survivorship rates of 94% at 5-year, 85% at 10-year, and 68% at 15-year follow-up, with conversion to total knee arthroplasty as the end point.

The clinical results of distal femoral varus osteotomy have been good in selected patients. Substantial improvements in pain and function can be expected in approximately 90% of patients.

Complications

Compartment syndrome in the leg This is the most important early complication of tibial osteotomy. Careful and repeated checks should be carried out during the early postoperative period to ensure that there are no symptoms or signs of impending ischaemia. Early features of compartment compression in the leg are sometimes mistaken for those of a deep vein thrombosis; this mistake should be avoided at all costs because the consequent delay in starting treatment could make the difference between complete recovery and permanent loss of function.

Peroneal nerve palsy Correction of a long-standing valgus deformity can stretch and damage the peroneal nerve.

Failure to correct the deformity Under- or overcorrection of the deformity are really failures in technique. With medial compartment osteoarthritis, unless a slight valgus position is obtained, the result is liable to be unsatisfactory. However, marked overcorrection is not only mechanically unsound but the cosmetic defect is liable to be bitterly resented by the patient.

Delayed union and non-union These complications can be avoided by ensuring that fixation of the bone fragments is stable and secure.

Arthrodesis

Arthrodesis of the knee has long been considered a demanding procedure that is subject to a variety of postoperative complications and often results in marginal or unacceptable outcomes. A stiff knee is a considerable disability; it makes climbing difficult and sitting in crowded areas distinctly awkward. Consequently, arthrodesis is not often performed and has typically been held in reserve as a final salvage procedure for patients with irretrievably failed total knee arthroplasties and other comparable conditions.

Indications

In the past – and even today in some parts of the world – the main indications for arthrodesis of the knee were (and are) *irremediable instability* due to the late effects of poliomyelitis and *painful loss of mobility* due to tuberculosis or chronic pyogenic infection. In countries with advanced medical facilities the commonest indication is *failed total knee replacement* (either septic or aseptic).

Contraindications

Contraindications include *severe general disability* because of age or multiple joint disease, especially if associated with problems in the ipsilateral hip or ankle; *amputation* or *knee fusion* of the opposite limb; and *persistent non-union* of a periarticular fracture or *massive periarticular bone loss*. Finally, *patient reluctance* may be an important factor, although a short period in a plaster cylinder before operation may convince the patient that a rigidly stiff leg is better than a painful and unstable knee.

Technique

A vertical midline incision is used. If the operation is for tuberculosis, the diseased synovium is excised; otherwise it is disregarded. The posterior vessels and nerves are protected and the ends of the tibia and femur removed by means of straight saw cuts, aiming to end with 15 degrees of flexion and 7 degrees of valgus as the position of fusion. Charnley's method, using thick Steinman pins inserted parallel through the distal femur and proximal tibia, and connecting these with compression clamps, was for many years the standard method. Nowadays, multiplanar external fixation is used or, if the joint is not infected, a long intramedullary nail which may be unlocked or locked.

Knee replacement

Indications

The main indication for knee replacement is pain, especially when this is combined with deformity and instability. Most replacements are performed for osteoarthritis or rheumatoid arthritis.

Types of operation

Total knee replacement (TKR) Most modern TKR designs are total condylar replacements with sacrifice of the anterior cruciate ligament and either retention (cruciate retaining – 'CR') or substitution (posterior stabilized – 'PS') of the posterior cruciate ligament.

In each case at operation all the articular surfaces are replaced – with metal on the femoral side, polyethylene on a metal tray on the tibial side (mono-block polyethylene tibial components may also be used) and typically polyethylene alone on the patella. It is important to ensure correct placement of the implants so as to reproduce the normal mechanics of the knee as closely as possible. The tibial and patellar components are fixed typically with cement, whereas the femoral component may be press-fitted but is more commonly cemented. Bone defects may be filled with bone graft, metal augmentation wedges or cement. It is important: (1) to overcome deformity (the knee should finally be about 7 degrees valgus); (2) to promote stability (by tailoring the bone cuts so that the collateral ligaments are equally tense in both flexion and extension); and (3) to permit rotation (otherwise cemented prostheses are liable to loosen). The development of suitable prostheses and instrumentation in recent years has led to vast improvements in technique, so the results are now similar to those of hip replacement.

Partial knee replacement In partial knee replacement the affected compartment is replaced, preserving the rest of the structures within the knee. The majority of partial knee replacements are performed for anteromedial osteoarthritis with fewer performed for lateral disease. It is essential to establish before surgery that the anterior cruciate ligament is intact and that the other compartments of the knee are functioning well. Following a successful operation, relief of pain, speed of recovery and restoration of function can be impressive. The revision rate of partial knee replacement is higher than of total knee replacement, although where surgeons perform the operation regularly revision rate may be reduced to that of TKR.

Highly constrained or hinged knee replacement Artificial knee joints with a high level of constraint or hinges (rotating or fixed) are used in complex reconstruction knee surgery when there is marked bone loss and severe instability involving the collateral ligaments. Their main clinical application is in complex revision surgery or following tumour excision.

Complications

General As with all knee operations (except arthroscopy) in which a tourniquet is used, there is a high incidence of deep vein thrombosis. Prophylaxis, either pharmacological (anticoagulants) or mechanical (foot pumps, compression stockings), is recommended.

Infection The methods of preventing and treating infection are similar to those used in hip replacement. For established and intractable infection, treatments by debridement and antibiotics, or by exchange replacement in one or two stages, are obvious possibilities.

Loosening Covert infection is only one cause of implant loosening. Aseptic loosening results from faulty prosthetic design, inaccurate bone shaping, incorrect placement of the implants or a combination of these factors. Revision surgery for loose prostheses must deal with the cause, be it malposition of the prosthesis, accumulation of wear debris or infection. A loose prosthesis can be recemented but unless the cause is dealt with, loosening will recur.

Patellar problems Although relatively uncommon, these can be very disabling. They include: (1) recurrent patellar subluxation or dislocation, which may need realignment; and (2) complications associated with patellar resurfacing, such as loosening of the prosthetic component, fracture of the remaining bony patella, and catching of soft tissues between the patella and the femur.

NOTES ON APPLIED ANATOMY

The knee joint combines two articulations, the tibiofemoral and the patellofemoral. The bones of the tibiofemoral joint have little or no inherent stability; this depends largely upon strong static and dynamic stabilizers such as ligaments and muscles. The patellofemoral joint is so shaped that the patella moves in a shallow path (or track) between the femoral condyles; if this track is too shallow, the patella readily dislocates; if its line is abnormal, the patellar articular cartilage is subject to excessive wear. One important function of the patella is to increase the power of extension; it lifts the quadriceps forward, thereby increasing its moment arm.

The quadriceps tendon is inserted into the upper pole of the patella. It is in line with the shaft of the femur, whereas the patellar tendon is in line with the shaft of the tibia. Because of the angle between them (the Q-angle) quadriceps contraction would pull the patella laterally were it not for the fibres of vastus medialis, which are transverse. This muscle is therefore important and it is essential to try to prevent the otherwise rapid wasting that is liable to follow any effusion.

The shaft of the femur is inclined medially, while the tibia is vertical; thus the normal knee is slightly valgus (average 7 degrees). This amount is physiological and the term 'genu valgum' is used only when the angle exceeds 7 degrees; significantly less than

this amount is genu varum. However, there is significance variability in physiological alignment and some patients' 'normal' is valgus while others have pronounced tibia vara and stand with relative varus.

During walking, weight is necessarily taken alternately on each leg. The line of body weight falls medial to the knee and must be counterbalanced by muscle action lateral to the joint (chiefly the tensor fascia femoris). To calculate the force transmitted across the knee, that due to muscle action must be added to that imposed by gravity; moreover, since with each step the knee is braced by the quadriceps, the force that this imposes also must be added.

Clearly, the stresses on the articular cartilage are (as they also are at the hip) much greater than consideration only of body weight would lead one to suppose. It is also obvious that a varus deformity can easily overload the medial compartment, leading to cartilage breakdown; similarly, a valgus deformity may overload the lateral compartment.

The interaction between the shapes of the articular surfaces, the ligaments that connect them and the muscle forces that act across the joint determine the pattern of movement of the joint. In relative terms the femur slides and rolls on the tibial surface. There are a number of characteristic movements: as the knee flexes there is considerable posterior movement of the tibiofemoral contact point in the lateral compartment, whereas in the medial compartment, under load, there is much less movement of the contact point, until high flexion angles. This differential movement between medial and lateral compartments explains the so-called terminal 'screw-home' mechanism. Disruption of the normal knee ligaments distorts joint movements as degrees of restraint are removed.

Situated as they are between these complexly moving surfaces, the fibrocartilaginous menisci are prone to injury, particularly during unguarded movements of extension and rotation on the weight bearing leg. The medial meniscus is especially vulnerable because, in addition to its loose attachments via the coronary ligaments, it is firmly attached at three widely separated points: the anterior horn, the posterior horn and to the medial collateral ligament. The lateral meniscus more readily escapes damage because it is attached only at its anterior and posterior horns and these are close to each other.

The main function of the menisci is to increase the contact area between femur and tibia. They play a significant part in stabilizing the joint and in weight transmission, and this applies at all angles of flexion and extension; as the knee bends they glide backwards, and as it straightens they are pushed forwards.

The deep portion of the medial collateral ligament, to which the meniscus is attached, is fan-shaped and blends with the posteromedial capsule. It is therefore not surprising that medial ligament tears are often associated with tears of the medial meniscus and of the posteromedial capsule. The lateral collateral ligament is situated more posteriorly and does not blend with the capsule; nor is it attached to the meniscus, from which it is separated by the tendon of popliteus.

The two collateral ligaments resist dynamic varus or valgus deformity of the joint of the extended knee. In addition, the medial ligament prevents the medial tibial condyle from subluxating forwards. Forward subluxation of the lateral tibial condyle, however, is prevented, not by the lateral collateral ligament but by the anterior cruciate. Only when the medial ligament and the anterior cruciate are both torn can the whole tibia subluxate forwards (giving a marked positive anterior drawer sign). Backward subluxation of the tibia is prevented by the powerful posterior cruciate ligament in combination with the arcuate ligament on its lateral side and the posterior oblique ligament on its medial side.

The cruciate ligaments are essential for stability of the knee. The anterior cruciate ligament prevents forward displacement of the tibia on the femur and, in particular, it prevents forward subluxation of the lateral tibial condyle (i.e. rotation), a movement that tends to occur if a person who is running twists suddenly. The posterior cruciate ligament prevents backward displacement of the tibia on the femur and its integrity is therefore important when progressing downhill.

FURTHER READING

Apley AG. The diagnosis of meniscus injuries: some new clinical methods. *J Bone Joint Surg* 1947; **29**: 78–84.

Bellemans J, Colyn W, Vandenneucker H, Victor J. The Chitranjan Ranawat award: is neutral mechanical alignment normal for all patients? The concept of constitutional varus. *Clin Orthop Relat Res* 2012; **470**: 45–53.

Brouwer RW, Raaij van TM, Bierma-Zeinstra SM, *et al.* Osteotomy for treating knee osteoarthritis. *Cochrane Database Syst Rev.* 2007 Jul 18; (**3**): CD004019.

Carr AJ, Robertsson O, Graves S, *et al.* Knee replacement. *Lancet* 2012; **379**(9823): 1331–40.

Insall JN, Salvati E. Patella position in the normal knee joint. *Radiology* 1971; **101**: 101.

Thorlund JB, Juhl CB, Roos EM, Lohmander LS. Arthroscopic surgery for degenerative knee: systematic review and meta-analysis of benefits and harms. *BMJ* 2015; **350**: h2747.

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The ankle and foot 21

Gavin Bowyer & Mike Uglow

CLINICAL ASSESSMENT

Symptoms

Adults with foot and ankle problems often present complaining of pain, swelling, deformity and impaired function. These will lead to difficulties with work, social and domestic activities. There are usually major clues to the pathology and diagnosis in the patient's history. Questions in the history should include those that flag up the possibility of neoplastic or generalized inflammatory disease and diabetes.

General questions include:

- Have you any pain or stiffness in your muscles, joints or back?
- Can you dress yourself completely without any difficulty?
- Can you walk up and down stairs without any difficulty?

Questions should also seek to assessing the impact of the condition on function and deciding on treatment in foot and ankle problems:

- What does this stop you doing?
- Does it interfere with work?
- Is your footwear restricted?
- Does it cause problems with hobbies, sports or domestic activities?

Pain over a bony prominence or a joint is probably due to some local disorder; ask the patient to point to the painful spot. Symptoms tend to be well localized to the structures involved, but vague pain across the forefoot (*metatarsalgia*) is less specific and is often associated with uneven loading and muscle fatigue. Often the main complaint is of shoe pressure on a tender corn over a toe joint or a callosity on the sole. Osteoarthritic pain at the first metatarsophalangeal (MTP) joint is often better in firm-soled shoes; hallux valgus/bunions will be exacerbated by close-fitting shoes; a functionally or mechanically unstable ankle often feels better in boots; metatarsalgia is worse in shoes with a higher heel. Morton's neuroma or a prominent metatarsal head feels like a marble or pebble in the shoe.

Deformity is sometimes the main complaint; the patient may abhor a 'crooked toe' or a 'twisted foot', even if it is not painful, and parents often worry about their children who are 'flat-footed' or 'pigeon-toed'. Elderly patients may complain chiefly of having difficulty finding shoes to fit – a common complaint among women with foot pathology in all age-groups.

Swelling is common, even in normal people, but it gains more significance if it is unilateral or strictly localized.

Instability of the ankle or subtalar joint produces repeated episodes of the joint 'giving way'. Ask about any previous injury (a 'twisted ankle').

Numbness and *paraesthesia* may be felt in all the toes or in a circumscribed field served by a single nerve or one of the nerve roots from the spine.

Signs with the patient upright

It is important to see the patient stand, as deformities will often be much better shown once the patient is weight-bearing. The patient, whose lower limbs should be exposed from the knees down, stands first facing the surgeon, then with his or her back to the surgeon (Figure 21.1). Ask the patient to rise up on tiptoes and then settle back on the heels. Note the posture of the feet throughout this movement. Normally the heels are in slight valgus while standing and inverted on tiptoes; the degree of inversion should be equal on the two sides, showing that the subtalar joint is mobile and the tibialis posterior functioning. Viewed from behind, if there is excessive eversion of one foot, the lateral toes are more easily visible on that side (the '*too-many-toes*' sign).

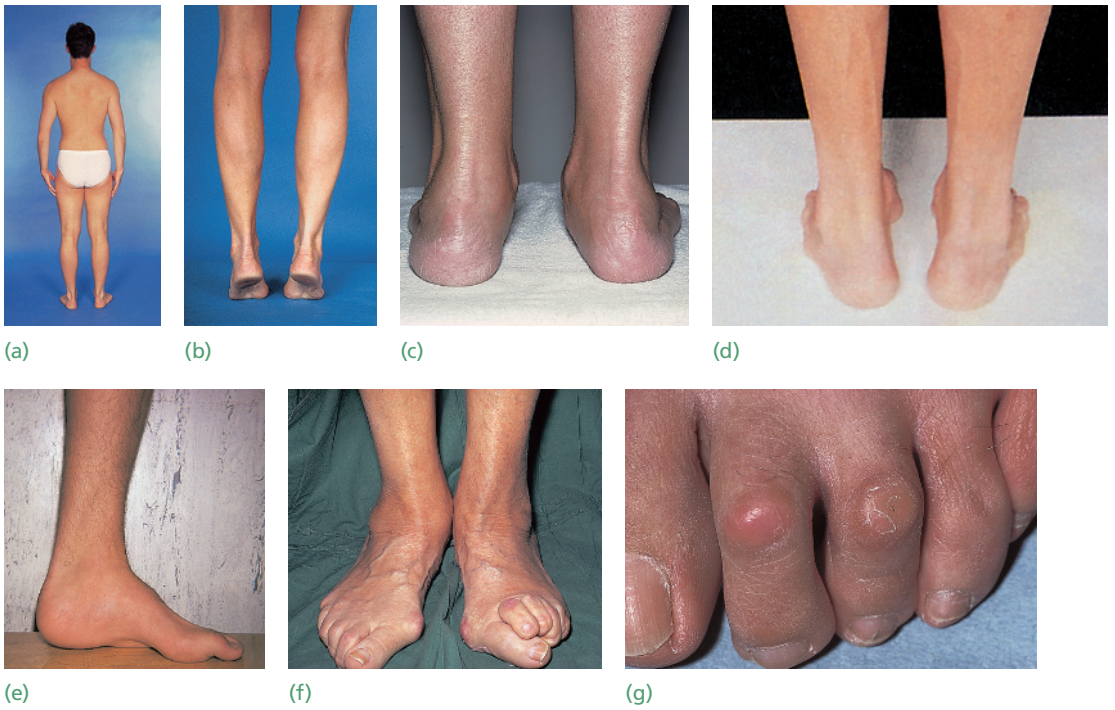


Figure 21.1 Examination with patient standing Look at the patient as a whole, first from in front and from behind. (a,b) The heels are normally in slight valgus and should invert equally when a patient stands on his/her toes. (c) This patient has flat feet (pes planus), while the patient in (d) has the opposite deformity, varus heels and an abnormally high longitudinal arch – pes cavus (e). From the front you can again notice (f) the dropped longitudinal arch in the patient with pes planus, as well as the typical deformities of bilateral hallux valgus and overriding toes. (g) Corns on the top of the toes are common.

Gait

Observing the gait also helps to identify dynamic problems and the effects of pathology from other lower limb joints. The patient is asked to walk normally. Note whether the gait is smooth or halting and whether the feet are well balanced. Gait is easier to analyse if concentrating on the sequence of movements that make up the walking cycle. It begins with heel-strike, then moves into stance, then push-off and finally swing-through before making the next heel-strike. The stance phase itself can be further divided into three intervals: (1) from heel-strike to flat foot; (2) progressive ankle dorsiflexion as the body passes over the foot; (3) ankle plantarflexion leading to toe-off (Figure 21.2).

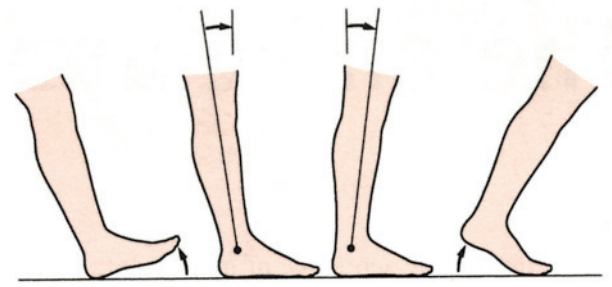


Figure 21.2 Gait – the three rockers of ankle-stance phase The first rocker begins with heel-strike – if the anterior compartment muscles are weak, a 'foot-slap' is noticeable; or if the ankle is in fixed equinus, this rocker may be absent altogether. In mid-stance, the centre of gravity of the body (and ground reaction force) moves from a position posterior to the ankle joint to anterior (second rocker). The third rocker produces an acceleration force that shifts the fulcrum of the pivot forwards to the metatarsal heads, just prior to toe-off.

Gait may be disturbed by pain, muscle weakness, deformity or stiffness. The position and mobility of each ankle is of prime importance. A fixed equinus deformity results in the heel failing to strike the ground at the beginning of the walking cycle; sometimes the patient forces heel contact by hyperextending the knee.

If the ankle dorsiflexors are weak, the forefoot may hit the ground prematurely, causing a 'slap'; this is referred to as foot drop (or drop foot). During swing-through the leg is lifted higher than usual so that the foot can clear the ground (a high-stepping gait).

Hindfoot and midfoot deformities may interfere with level ground contact in the second interval of stance; the patient walks on the inner or outer border of the foot.

Toe contact, especially of the great toe, is also important; pain or stiffness in the first MTP joint may prevent normal push-off.

Signs with the patient sitting or lying

A systematic approach to examination, following the *look*, *feel*, *move* steps, will lead to a diagnosis in the majority of cases.

Next the patient is examined lying on a couch, or it may be more convenient if he or she sits opposite the examiner and places each foot in turn on the examiner's lap.

Look

The heel is held square so that any foot deformity can be assessed. The toes and sole should be inspected for *skin changes*. The foot shows areas of overload by producing callosities, and there are often corresponding areas of wear and signs of overload on the footwear. Thickening and keratosis may be seen over the proximal toe joints or on the soles. Atrophic changes in the skin and toenails are suggestive of a neurological or vascular disorder, or commonly fungal infection of the nail.

Deformity may be in the ankle, the foot or toes. A foot that is set flat on the ground at a right angle to the tibia is described as *plantigrade*; if it is set in fixed plantarflexion (pointing downwards), it is said to be in *equinus*; a dorsiflexed position is called *calcaneus*. Common defects are a 'flat-footed' stance (*pes valgus*); an abnormally high instep (*pes cavus*); a downward-arched forefoot (*pes plantaris*); lateral deviation of the great toe (*hallux valgus*); fixed flexion of a single interphalangeal (IP) joint (*hammer toe*) or of all the toes (*claw toes*).

Swelling may be diffuse and bilateral, or localized; unilateral swelling nearly always has a surgical cause and bilateral swelling is more often 'medical' in origin (e.g. lymphoedema, heart failure, renal disease). Swelling over the medial side of the first metatarsal head (a *bunion*) is common in older women.

Corns are usually obvious; *callosities* must be looked for on the soles of the feet.

Feel

Pain and tenderness in the foot and ankle localize very well to the affected structures – the patient really does show us where the problem is. The skin temperature is assessed and the pulses are felt. Remember that one in every six normal people does not have a dorsalis pedis artery. If all the foot pulses are absent, feel for the popliteal and femoral pulses; the patient may need further evaluation by Doppler ultrasound.

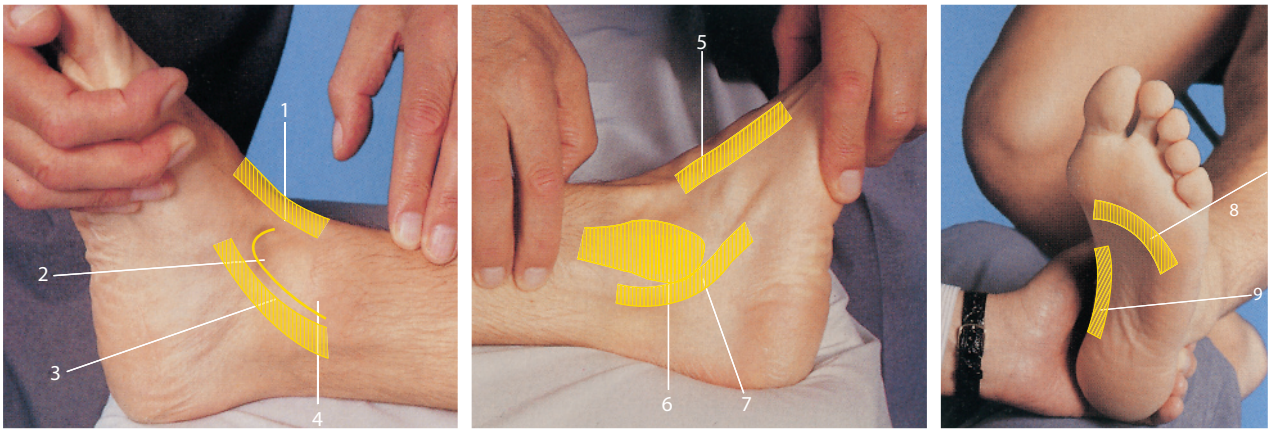
If there is tenderness in the foot, it must be precisely localized, for its site is often diagnostic (Figures 21.3 and 21.4). Any swelling, oedema or lumps must be examined.

Sensation may be abnormal; the precise distribution of any change is important. If a neuropathy is suspected (e.g. in a diabetic patient), test also for vibration sense, protective sensation and sense of position in the toes. The standard screening and monitoring test in the diabetic foot clinic is the 10 g monofilament test for sensation.

Move

The foot comprises a series of joints that should be examined methodically:

- *Ankle joint* – With the heel grasped in the left hand and the midfoot in the right, the ranges of plantarflexion (flexion) and dorsiflexion (extension) are estimated (Figure 21.5). Beware not to let the foot go into valgus during passive dorsiflexion as this will give an erroneous idea of the range of movement.



(a) (b) (c)

Figure 21.3 Foot – surface anatomy Medial aspect: (a) 1 tendon of tibialis anterior; 2 medial malleolus; 3 tendon of tibialis posterior; 4 sulcus behind medial malleolus; (b) 5 extensor tendons of toes; 6 lateral malleolus; 7 peroneal tendons curving behind the lateral malleolus; (c) 8 transverse metatarsal arch; 9 medial longitudinal arch.

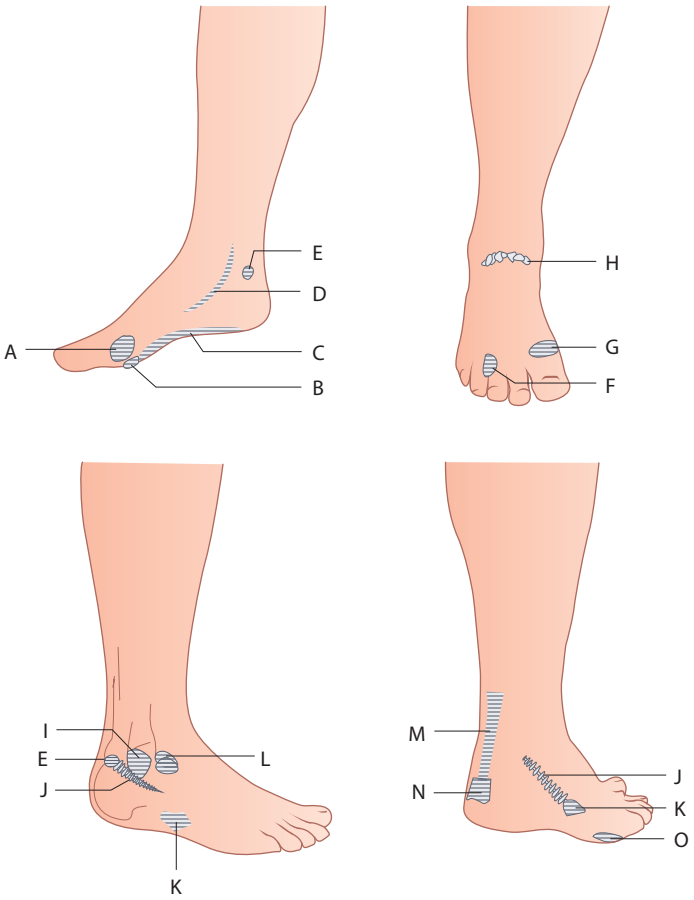


Figure 21.4 Where does it hurt? Where is it tender?

A Medial to first MTP joint – bunion
B Beneath first MTP joint – sesamoiditis
C Plantar fascia – plantar fasciitis
D Posterior to medial malleolus/line of tibialis posterior – tibialis posterior tendinitis or tear, and in planovalgus collapse of hindfoot
E Retrocalcaneal bursa – bursitis
F In third interspace – Morton's neuroma
G Dorsal to first MTP joint – OA, hallux limitus/rigidus
H Anterior ankle joint line – impingement from osteophytes in OA
I Bony tip/lateral malleolus – ankle fracture (Ottawa guidelines)
J Posterior/inferior to lateral malleolus – peroneal tenosynovitis or tear
K Base of fifth metatarsal – fracture/insertional problem with peroneus brevis
L Anterolateral angle of ankle joint – lateral gutter impingement in post-traumatic ankle with soft-tissue problems
M Achilles tendon – Achilles tendinitis/paratendinitis
N Achilles insertion – insertional tendinitis
O Beneath metatarsal heads – 'metatarsalgia'

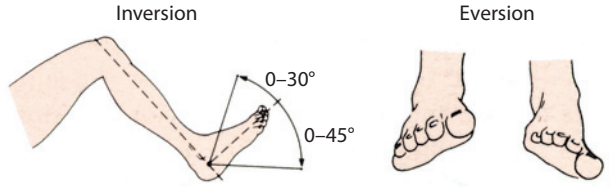


Figure 21.5 Normal range of movement All movements are measured from zero with the foot in the 'neutral' or 'anatomical' position: thus, dorsiflexion is 0–15 degrees and plantarfexion 0–40 degrees. Note that plantarflexion is a compound movement involving the ankle and the talonavicular joint, the latter contributing about 20 degrees to the movement – it is important to isolate the joints of interest when carrying out an assessment of their range. Inversion is about 30 degrees and eversion 15 degrees. Again these are compound movements combining supination and pronation at the hindfoot with midfoot movements.

- *Subtalar joint* – It is important to 'lock' the ankle joint when assessing subtalar inversion and eversion. This is done simply by ensuring that the ankle is plantigrade when the heel is moved. It is often easier to record the amount of subtalar movement if the patient is examined prone. Inversion is normally greater than eversion.
- *Midtarsal joint* – One hand grips the heel firmly to stabilize the hindfoot while the other hand moves the forefoot up and down and from side to side.
- *Toes* – The MTP and IP joints are tested separately. Extension (dorsiflexion) of the great toe at the MTP joint should normally exceed 70 degrees and flexion 10 degrees.

Stability

Stability is assessed by moving the joints across the normal physiological planes and noting any abnormal 'clunks'. Ankle stability should be tested in both coronal and sagittal planes, always comparing the two joints. Patients with recent ligament injury may have to be examined under anaesthesia.

Medial and *lateral stability* are checked by stressing the ankle first in valgus and then in varus. *Anteroposterior stability* is assessed by performing an anterior 'drawer test': the patient lies on the examination couch with hips and knees flexed and the feet resting on the couch surface; the examiner grasps the distal tibia with both hands and pushes firmly backwards, feeling for abnormal translation of the tibia upon the talus. Another way of doing this is to stabilize the distal tibia with one hand while the other grasps the heel and tries to shift the hindfoot forwards and backwards.

The same tests can be performed under X-ray and the positions of the two ankles measured and compared.

Muscle power

Power is tested by resisting active movement in each direction. The patient will be more cooperative if the movement required is demonstrated precisely. While the movement is held, feel the muscle belly and tendon to establish whether they are intact and functioning.

Shoes

Footwear often adds additional clues when examining the foot and ankle, providing valuable information about faulty stance or gait.

General examination

If there are any symptoms or signs of vascular or neurological impairment, or if multiple joints are affected, a more general examination is essential.

Imaging

There are practical problems with imaging in children, and babies in particular because: (1) babies tend not to keep still during examination; (2) their bones are not completely ossified and their shape and position may be hard to define.

X-rays

In the adult, the *standard X-ray* views of the ankle are anteroposterior (AP), mortise (an AP view with the ankle internally rotated 15–20 degrees) and lateral (Figure 21.6). Although the subtalar joint can be seen in a lateral view of the foot, medial and lateral oblique projections allow better assessment of the joint. These views are often used to check articular congruity after treatment of calcaneal fractures. The calcaneum itself is usually X-rayed in axial and lateral views, but a weight-bearing view is helpful in defining its relationship to the talus and tibia. X-ray under load, weight-bearing, is helpful in showing the coronal relationship of heel to tibia in stance. The foot, toes and intertarsal joints are well displayed in standing dorsoplantar and lateral views.

Stress X-rays complement the clinical tests for ankle stability. The patient should be completely relaxed; if the ankle is too painful, stress X-rays can be performed under regional or general anaesthesia. Both ankles should be examined, for comparison.

Computed tomography

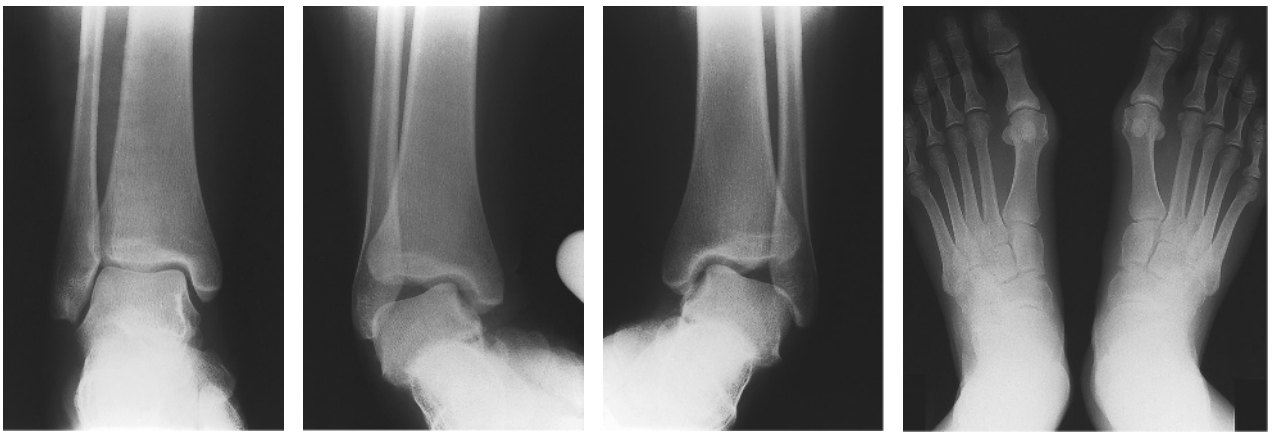
CT scans provide excellent coronal views and are important in assessing fractures and congenital bony coalitions.

Radioscintigraphy

Radioisotope scanning, though non-specific, is excellent for localizing areas of abnormal blood flow or bone remodelling activity; it is useful in the diagnosis of covert infection.

MRI and ultrasound

These methods are used to demonstrate soft-tissue problems, such as tendon and ligament injuries.



(a) (b) (c) (d)

Figure 21.6 X-rays (a) AP view of the ankle in a young woman who complained that after twisting her right ankle it kept giving way in high-heeled shoes. The X-ray looks normal; the articular cartilage width (the 'joint space') is the same at all aspects of the joint. The inversion stress view (b) shows that the talus tilts excessively; always X-ray both ankles for comparison and in this case the left ankle (c) does the same. She has generalized joint hypermobility, not a torn lateral ligament. (d) X-rays of the feet should be taken with the feet flat on the ground.

Pedobarography

A record of pressures beneath the foot can be obtained by having the patient stand or walk over a force plate; sensors in the plate produce a dynamic map of the peak pressures and the time over which these are recorded can be obtained. Although this is sometimes helpful in clinical decision making, or for comparing pre- and postoperative function, the investigation is used mainly as a research tool.

CONGENITAL DEFORMITIES

Congenital deformities of the foot are common. Many appear as part of a more widespread genetic disorder; only those in which the foot is the main (or only) problem are considered in this section. Isolated abnormalities of the toes are also dealt with elsewhere.

Talipes equinovarus (idiopathic club foot)

The term '*talipes*' is derived from *talus* (Latin = ankle bone) and *pes* (Latin = foot). Equinovarus is the most important talipes deformity with calcaneovalgus being more common and rarely causing problems. Occasionally spina bifida and some other neurological conditions can cause this deformity, so spinal examination is mandatory.

In the full-blown equinovarus deformity the heel is in equinus, the entire hindfoot in varus and the mid- and forefoot adducted and supinated. The first ray is flexed causing cavus. The abnormality is relatively common, the incidence ranging from 1–2 per thousand births; boys are affected twice as often as girls and the condition is bilateral in one-third of cases.

The exact cause is not known, although the resemblance to other disorders suggests several possible mechanisms. It could be a germ defect, or a form of arrested development. Its occurrence in neurological disorders and neural tube defects (e.g. myelomeningocele and spinal dysraphism) points to a neuromuscular disorder. Severe examples of club foot are seen in association with genetic conditions such as arthrogryposis, diastrophic dysplasia, Freeman–Sheldon syndrome and amniotic constriction rings. In some cases it is no more than a postural deformity caused by tight packing in an overcrowded uterus. These flexible deformities follow a particularly benign course.

Pathological anatomy

The key point is that the os calcis is internally rotated around the talocalcaneal ligament. This rotation forces the heel into equinus and varus and the connections of the forefoot with the hindfoot through the navicular and cuboid result in the forefoot being in an adducted and supinated position. The neck of the talus points downwards and deviates medially, whereas the body is rotated slightly outwards in relation to both the calcaneum and the ankle mortise. The posterior part of the calcaneum is held close to the fibula by a tight calcaneofibular ligament, and is tilted into equinus and varus.

The skin and soft tissues of the calf and the medial side of the foot are short and underdeveloped. If the condition is not corrected early, secondary growth changes occur in the bones; these are permanent. Even with treatment the foot is liable to be short, the calf may remain thin and a below-knee length discrepancy of less than 2 cm may occur.

Clinical features

The deformity is usually obvious at birth; the foot is both turned and twisted inwards so that in the worst cases the sole faces posteromedially. The clinical features have been classified by Pirani so that the severity can be assessed at birth and the progress of treatment can be monitored. The scoring system allocates 0, 0.5 or 1.0 for each of six clinical features:

- medial crease
- lateral border of the foot
- lateral head of talus
- posterior crease
- empty heel
- ankle dorsiflexion.

The foot is examined and held in the best position possible and a score assigned to each category. For example, the lateral border of the forefoot is visualized while the forefoot is abducted. No or minimal correction of the lateral curvature would score 1, partial correction would score 0.5 and complete straightening of the lateral border would score 0.

In a normal baby the foot can be abducted, everted and the ankle dorsiflexed. In club foot this manoeuvre meets with varying degrees of resistance and in severe cases the deformity is fixed (Figure 21.7).

The infant must always be examined for associated disorders such as congenital hip dislocation and spina bifida (Figure 21.7e). The absence of creases may suggest arthrogryposis; look to see if other joints are affected.

X-rays

X-rays are used mainly to assess progress after treatment in the older child and are rarely used in the initial

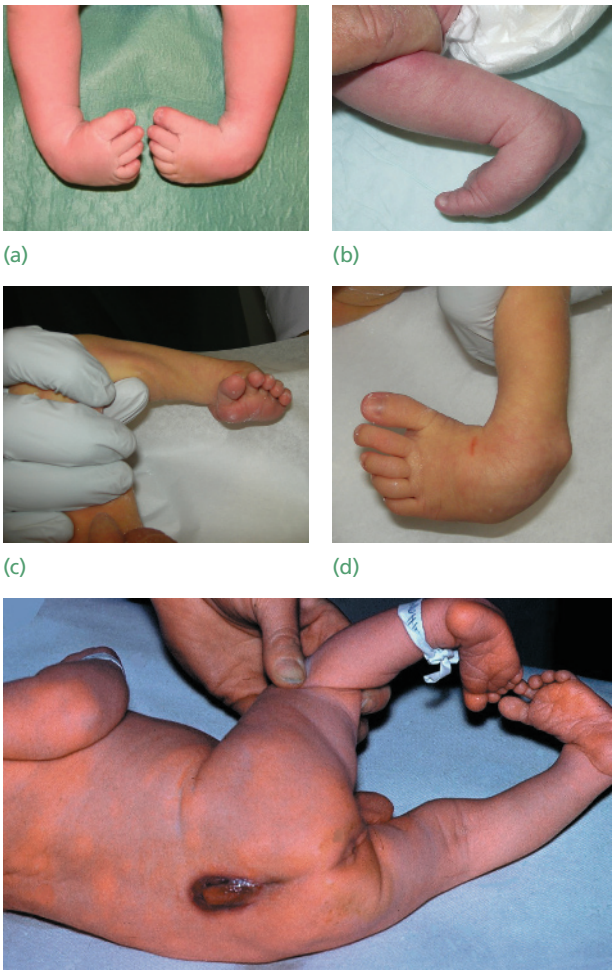


Figure 21.7 Talipes equinovarus (club foot) (a) True club foot is a fixed deformity, unlike (b) postural talipes, which is easily correctable by gentle passive movement. (c,d) With true club foot, the poorly developed heel is higher than the forefoot, which points downwards and inwards (varus). (e) Always examine the hips for congenital dislocation and the back for spina bifida (as in the case shown here).

assessment and management. The *anteroposterior film* is taken with the foot 30 degrees plantarflexed and the tube likewise angled 30 degrees perpendicular. Lines can be drawn through the long axis of the talus parallel to its medial border and through that of the calcaneum parallel to its lateral border; they normally cross at an angle of 20–40 degrees (*Kite's angle*) but in club foot the two lines may be almost parallel. Incomplete ossification makes it difficult to decide exactly where to draw these lines and this means that there is a considerable degree of interobserver variation.

The *lateral film* is taken with the foot in forced dorsiflexion. Lines drawn through the mid-longitudinal axis of the talus and the lower border of the calcaneum should meet at an angle of about 40 degrees. A measurement of less than 20 degrees shows that the calcaneum cannot be tilted up into true dorsiflexion (Figure 21.8); the foot may seem to be dorsiflexed but it may actually have 'broken' at the midtarsal level, producing the so-called *rocker-bottom deformity*.

Treatment

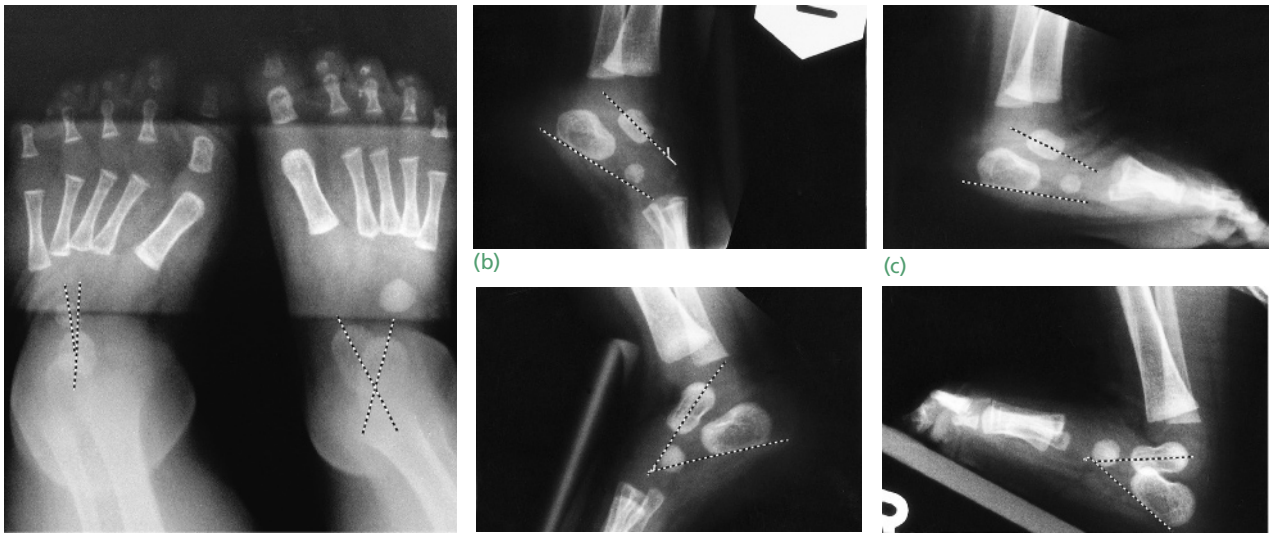
The aim of treatment is to produce and maintain a plantigrade, supple foot that will function well. Historically, there are several methods of treatment but conservative management with serial casting is now considered the most suitable. Relapse is common and continued bracing is required for the first 4 years of life to minimize this risk. Babies with associated neuromuscular disorders and other congenital conditions have a higher rate of recurrent deformities.

CONSERVATIVE TREATMENT

Treatment should begin early, preferably within the first 2 weeks after birth. This consists of manipulation repeated weekly, with the foot held in place with a plaster cast. The procedure is the brainchild and result of a lifetime of dedicated work to finesse the program by Dr Ignacio Ponseti, whose name is synonymous with the procedure.

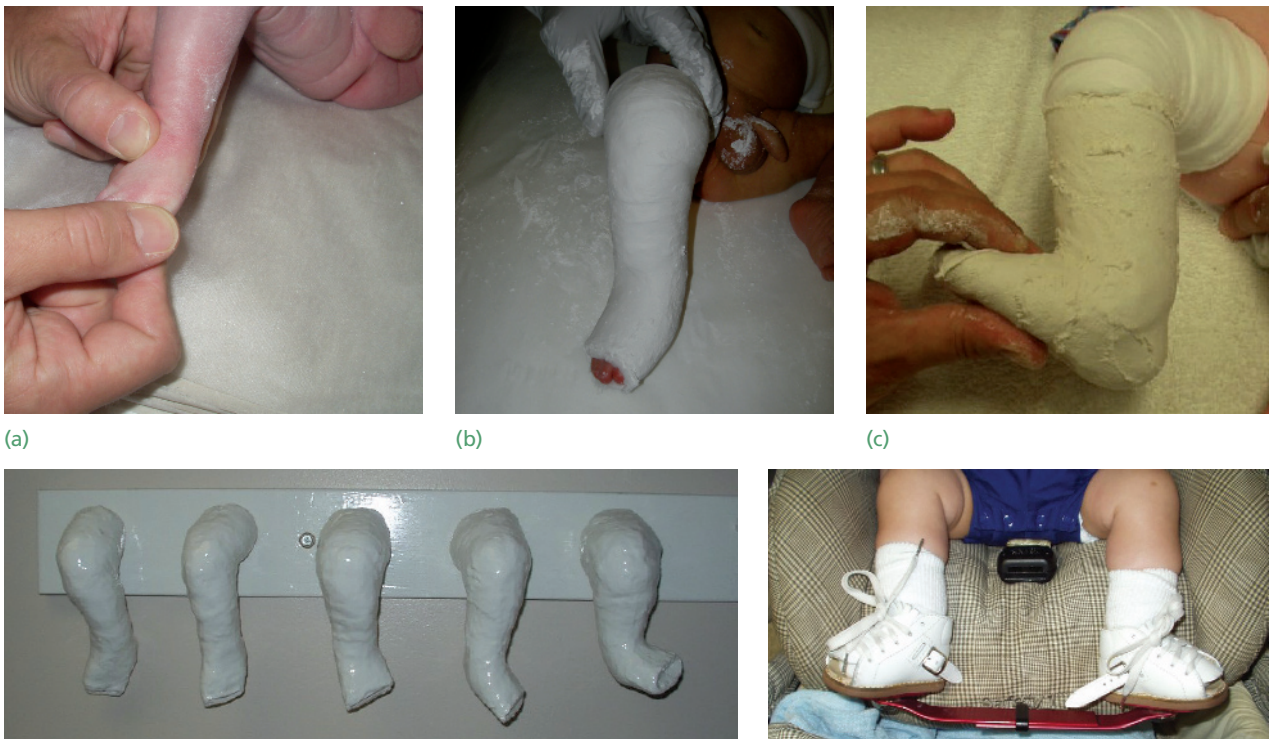
The three main components of the deformity are corrected in the following order. Firstly, the first metatarsal is elevated to correct the cavus and create a flat longitudinal medial border of the foot. Next, the forefoot is abducted in the plane of the metatarsals which rotates and everts the hindfoot and corrects the forefoot out of supination. This manoeuvre will also result in dorsiflexion of the os calcis as it rotates from underneath the talus. Finally, equinus is corrected by dorsiflexing the foot at the ankle joint. It is necessary in approximately 85% of cases to perform a percutaneous Achilles tenotomy in order to fully correct the equinus.

The key to correcting the club-foot deformity is using the lateral head of talus as the fulcrum of correction. Using one's thumb against the lateral talar head allows rotation of the entire calcaneopedal block around the talus. The forefoot must be 'overcorrected' to a position of 70 degrees of external rotation to ensure correct alignment of the os calcis. Care is needed to recognize a flexible foot with a very tight hindfoot because there is potential to cause a midfoot break through the midtarsal joint if this is not appreciated. Correction will typically require an average of five casts and, once corrected, the foot is retained in a further cast for 3 weeks. Following the complete correction of the club foot by serial casting, the deformity must be held using a foot abduction brace (FAB) to prevent relapse. This is retained for 3 months full-time (23 hours per day) and then night-time and nap-time for 4 years (Figure 21.9).



(a) (b) (c) (d) (e)

Figure 21.8 Talipes equinovarus – X-rays The left foot is abnormal. In the anteroposterior view (a) the talocalcaneal angle is 5 degrees, compared to 42 degrees on the right. In the lateral views, the left talocalcaneal angle is 10 degrees in plantarflexion (b) and 15 degrees in dorsiflexion (c). In the normal foot the angle is unchanged at 44 degrees, whatever the position of the foot (d,e).



(a) (b) (c) (d) (e)

Figure 21.9 Congenital talipes equinovarus – treatment Treatment consists of serial stretching and casting using Ponseti's method. (a) The fulcrum of correction is rotation around the lateral talar head. (b) A plaster cast is applied to hold the position achieved by stretching until (c) at least 15 degrees of dorsiflexion are achievable at the ankle. About 80% of feet will need a percutaneous Achilles tenotomy to achieve the corrected position. (d) A series of plaster casts showing the gradual correction from cast to cast. (e) To maintain the corrected position, boots on a bar (foot abduction brace) are worn full time for 3 months and at night only until the fourth birthday.

The relapse rates following Ponseti treatment have been clearly linked to the compliance of wearing the FAB, and skilled fitting is essential. As the child grows the boots will need to increase in size and the bar width must increase so that the internal distance from each heel matches the outer distance from shoulder to shoulder. The boot must be held at 70 degrees of abduction but for unilateral cases the unaffected foot is positioned at 30 degrees.

Atypical feet have been described by Ponseti and co-workers and must be recognized from the more usual idiopathic feet because a modification of the casting technique is required. Clinically, complex club feet were defined as having rigid equinus, severe plantar flexion of all metatarsals, a deep crease above the heel, a transverse crease in the sole of the foot, and a short and hyperextended first toe. The Achilles tendon was exceptionally tight and fibrotic up to the middle of the calf. During casting the forefoot will not correct into 70 degrees of abduction but advances to approximately 30 degrees only. In this position the os calcis is corrected under the talus, and this can be felt by careful digital palpation with the pulp of the index finger alongside the talar head with the fingertip pressing on the corrected anterior os calcis. There is midfoot cavus across the width of the foot and not just the first ray so dorsiflexion is achieved by elevation of the medial and lateral rays. An Achilles tenotomy is always required in these feet and occasionally may need to be performed earlier than usual with continued casting after it to ensure complete correction.

OPERATIVE TREATMENT

Surgery is rarely required for the early correction of the club-foot deformity. The rate of surgical intervention has fallen to below 5% from the 80% needed in the pre-Ponseti era. The objectives of club-foot surgery are: (1) the complete release of joint 'tethers' (capsular and ligamentous contractures and fibrotic bands); and (2) lengthening of tendons so that the foot can be positioned normally without undue tension. A detailed knowledge of the pathological anatomy is a sine qua non.

The tibialis posterior, flexor hallucis and longus tendons as well as the Achilles tendon are all Z lengthened. The talonavicular and subtalar joints are released, as is the posterior ankle joint. The peroneal sheath is released and the calcaneofibular ligament divided. The anterior third of the medial deltoid ligament and the talocalcaneal ligaments are retained to prevent overcorrection. The procedure can be performed through a posteromedial approach, through a curved posterior approach known as the 'Cincinatti', or through two incisions plantarmedial and posterolateral. The foot is put in a cast for 8–12 weeks and K-wires can be used to aid maintenance of correction.

LATE OR RELAPSED CLUB FOOT

Late presenters often have severe deformities with secondary bony changes, which are fortunately not at all common in the developed world but sadly more common in the developing world. The Ponseti programme has been run successfully across many developing areas of the world with very good success, and the rate of late presentation is reducing as a result. The first sign of relapse is usually a loss of dorsiflexion and, if allowed to continue, the foot will adopt the equinus and varus posture that it began with. The other common occurrence is the development of dynamic supination of the forefoot due to relative overactivity of the tibialis anterior muscle in the presence of weak peroneals. During the swing phase of gait the forefoot can be seen to supinate and a tendon transfer is necessary if this progresses. Most contemporary children's foot surgeons will transfer the whole tendon to the lateral cuneiform once the ossific nucleus has developed, usually between the ages of 3 and 4 years.

Some children with syndromic feet continue to relapse and treatment using the traditional open soft-tissue releases as described above may be required. Occasionally the lateral column will need to be shortened to correct severe adductus but, if the hindfoot remains in good position, the adduction can be treated over the age of 6 years by a double tarsal wedge osteotomy by transferring a wedge of bone from the cuboid to the medial column at the level of the cuneiforms.

Gradual correction using a circular external fixator (the Ilizarov method) has been reported in treating difficult relapsed cases and severe deformities with good results. Full corrections can be achieved even in feet severely scarred from previous surgery. The principles of Ponseti can be adapted to use with the Taylor spatial frame (TSF) as elucidated by Herzenberg. The procedure can be painful and long and is best reserved for these very difficult cases.

Despite initially successful casting and subsequent surgery, some teratological deformities may still recur. A deformed, stiff and painful foot in an adolescent is best salvaged by corrective extra-articular osteotomies where possible and using tendon transfers to balance the deforming forces. Arthrodesis is avoided if at all possible but in some cases this may be the only viable salvage option available. The goal of all the treatment options is to finish with a plantigrade, stable and pain-free foot by skeletal maturity.

METATARSUS ADDUCTUS

Metatarsus adductus describes a deformity affecting only the forefoot in relation to the hindfoot, and the key information is flexibility of the foot (Figure 21.10). A number of classifications have been

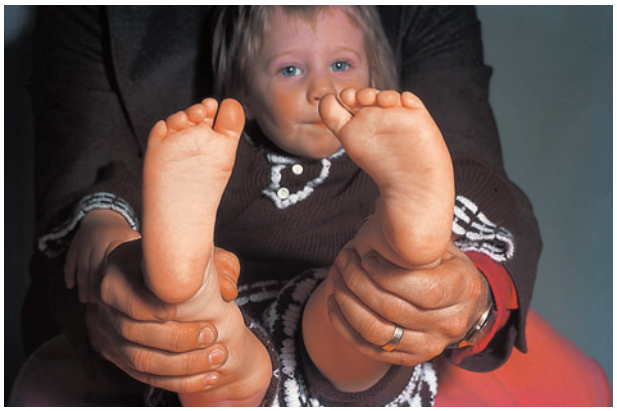


Figure 21.10 Metatarsus adductus In contrast to club foot, the deformity here is limited to the forefoot.

cited and all reflect the ability of the forefoot to overcorrect the midaxial line in flexible deformities compared to the stiff deformity where correction to the midaxial line is not possible. All cases of flexible metatarsus adductus resolve within 4 years. Those cases that are partially correctable are treated with serial casting, ideally between the ages of 6 and 12 months and have excellent long-term outcomes. There is only a 5% reported incidence of foot pain on strenuous activity as an adult.

Rigid deformities occur in children with certain rare conditions such as Aarskog syndrome and require serial casting. For those that fail to correct, surgery can be considered, consisting of either release of the abductor hallucis tendon, with or without release of the medial capsules of the navicular cuneiform and cuneiform metatarsal joints, in children under 4 years or corrective osteotomies through the cuneiforms in children 6 years and older. In very severe cases where metatarsal deformity exists, multiple osteotomies of the metatarsals can be performed.

Talipes calcaneovalgus

Calcaneovalgus is a relatively common deformity that presents in the newborn as an acutely dorsiflexed foot where the dorsum of the foot abuts the anterior border of the shin. There is a deep crease (or several wrinkles) on the front of the ankle, and the calcaneum juts out posteriorly. Unlike congenital vertical talus (which also presents as an acutely dorsiflexed foot), this deformity is flexible. In addition, the anterior creases in congenital vertical talus are located over the midfoot.

Calcaneovalgus is usually bilateral. This is a postural deformity, probably due to abnormal intrauterine positioning, and it often corrects spontaneously in the neonatal period. Severe deformities can occur which appear resistant to gentle stretching and occasionally require serial casts for correction.

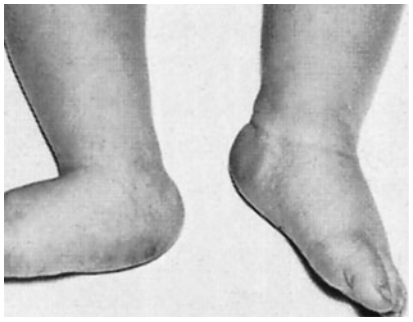
Rarely this foot deformity can be associated with spinal dysraphism so it is mandatory to examine the spine for abnormalities. The much rarer deformity of posteromedial bow of the tibia can be misinterpreted as a calcaneovalgus deformity to the unwary.

Congenital vertical talus

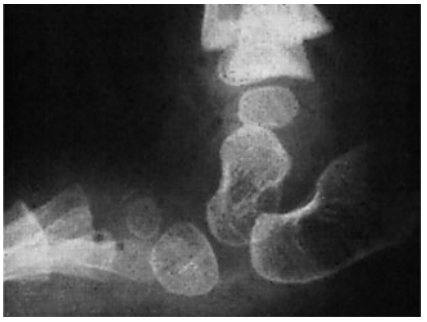
This rare condition is seen in infants, usually affecting both feet. Superficially it resembles other types of valgus foot, but the deformity is more severe; the medial arch is not only flat, it is the most prominent part of the sole, producing the appearance of a rocker-bottom foot (Figure 21.11). The hindfoot is in equinus and valgus and the talus points almost vertically towards the sole; the forefoot is abducted, pronated and dorsiflexed, with subluxation or dislocation of the talonavicular joint. Passive correction is impossible.

X-rays

The calcaneum is in equinus and the talus points into the sole of the foot, with the navicular dislocated dorsally onto the neck of the talus. It is important to repeat the lateral X-ray with the foot maximally plantarflexed; in congenital vertical talus the appearance will be unchanged, whereas in congenital oblique talus and flexible flat foot the dorsally subluxated navicular returns to the normal position.



(a)



(b)

Figure 21.11 Congenital vertical talus (a) The infant's foot is in marked valgus and has a rocker-bottom shape. The deformity is rigid and cannot be corrected. (b) X-ray shows the vertical talus pointing downwards towards the sole and the other tarsal bones rotated around the head of the talus.

Treatment

Serial casting using a so-called 'reverse Ponseti' technique followed by stabilization of the talonavicular joint with an open approach, to confirm that the joint is reduced and then held with a K-wire, together with an Achilles tenotomy has been shown to be very effective. For some resistant feet, open reduction in full may be required.

PES PLANUS AND PES VALGUS ('FLAT FOOT')

'Our feet are no more alike than our faces.' This truism from a *British Medical Journal* editorial sums up the problem of 'normally abnormal' feet. The medial arch may be normally high or normally low. The term 'flat foot' applies when the apex of the arch has reversed and lies plantar to the long axis of the medial border of the foot. The term 'collapsed arches' can be misleading because in some feet the arch has never developed and the foot has always been flat. In a flat-foot deformity, the medial border of the foot is in contact (or nearly in contact) with the ground; the heel becomes valgus and the foot pronates at the subtalar–midtarsal complex. The problems associated with flat foot differ in babies, children and adults and these three categories will therefore be considered separately.

FLAT FOOT IN CHILDREN AND ADOLESCENTS

Flat foot is a common complaint among children. Or rather their parents, grandparents, and assistants in the shoe shop – the children themselves usually don't seem to notice it!

Flexible flat-foot Flexible pes valgus appears in toddlers as a normal stage in development, and it usually disappears between 4 and 10 years of age, when medial arch development occurs; occasionally, though, it persists into adult life. The arch can often be restored by simply dorsiflexing the great toe (*Jack's test*), and during this manoeuvre the tibia rotates externally. Many of these children have ligamentous laxity and there may be a family history of both flat feet and joint hypermobility.

Stiff (or 'rigid') flat foot A deformity that cannot be corrected passively should alert the examiner to an underlying abnormality. *Congenital vertical talus* is dealt with earlier. In older children, conditions to be considered are: (1) *tarsal coalition*; (2) an *inflammatory joint disorder*; and (3) a *neurological disorder*.

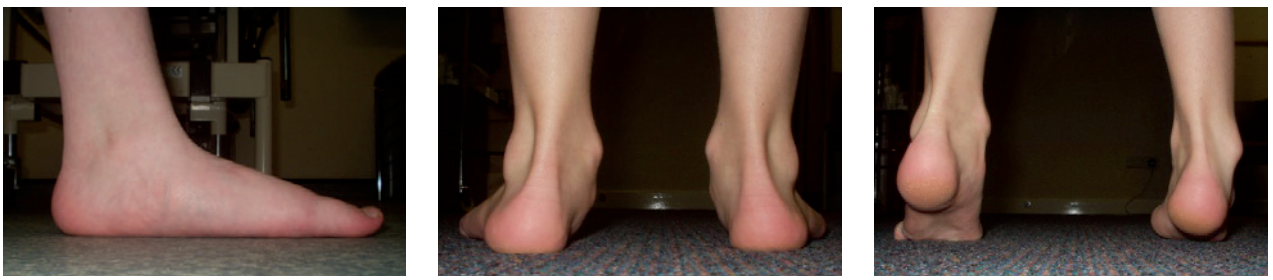
Compensatory flat foot This is a spurious deformity that occurs in order to accommodate some other postural defect. For example, a tight tendo Achillis (or a mild fixed equinus) may be accommodated by everting the foot; or if the lower limbs are externally rotated the body weight falls anteromedial to the ankle and the feet go into valgus – the Charlie Chaplin look.

Clinical assessment

Although there is usually nothing to worry about, the parents' concerns should not be dismissed without a proper assessment of the child. Enquire about neonatal problems and a family history.

Watch the child stand and note the position of the heels from behind. Are they in neutral or valgus, and do they invert when the child stands on tiptoe? The tiptoe test will confirm a mobile subtalar joint and functioning tibialis posterior tendon (Figure 21.12). Let the child walk: is the gait normal for the child's age? Are the heels set flat during the stance phase, or does the child have tight Achilles tendons?

Examine the foot and note its shape. In the neonate, the rare congenital vertical talus presents as a stiff, acutely dorsiflexed and very flat (almost rocker-bottom) foot. Palpate for tenderness: are there signs of inflammatory arthritis or infection? Test the movements in the ankle as well as the subtalar and



(a) (b) (c)

Figure 21.12 Mobile flat feet (a) Profound flattening of the medial border of the foot with (b) marked valgus heel alignment. On toe standing (c) the arch forms and the heels can be seen to invert indicating the subtalar joints are mobile.

midtarsal joints: a tight Achilles tendon may be 'constitutional' or part of a neuromuscular problem.

Try to correct the flat foot by gentle passive manipulation. Perform Jack's test (see earlier) to distinguish between a flexible and a stiff ('rigid') deformity.

The spine, hips and knees also should be examined. The clinical assessment is completed by a swift general examination for joint hypermobility and signs of neuromuscular abnormalities.

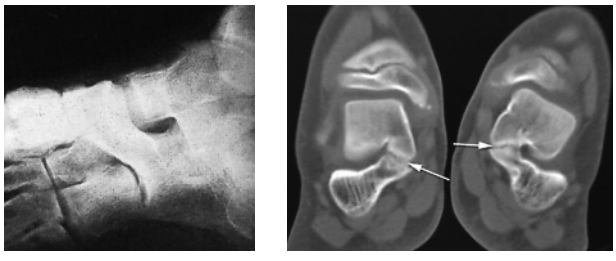
PERONEAL SPASTIC FLAT FOOT (TARSAL COALITION)

Older children and teenagers sometimes present with a painful, rigid flat foot in which the peroneal and extensor tendons are in spasm. X-rays and CT scans may show one or several of a variety of unions or partial unions between adjacent tarsal bones; the commonest two types are talocalcaneal and calcaneonavicular coalitions (Figure 21.13). The anomaly is inherited as an autosomal dominant condition and is present at birth but it becomes symptomatic only when the abnormal fibrous syndesmosis matures into a stiffer, cartilaginous synchondrosis that later ossifies to become a rigid bar.

The child, usually at puberty or during early adolescence, develops an increasingly stiff flat-foot deformity. Pain may be due to abnormal tarsal stress or even fracture of an ossified bar. The picture differs from that of the more common 'idiopathic' flat foot in that the deformity is more or less rigid, with spasm of the peroneal muscles.

Imaging

X-rays are unnecessary for asymptomatic, flexible flat feet. For painful or stiff flat feet, standing anteroposterior and lateral views as well as obliques may help to identify underlying disorders. On the lateral view, 'beaking' of the head of the talus suggests the presence of a tarsal coalition. Narrowing of the talocalcaneal joint, which is sometimes seen in talocalcaneal coalition, is easily mistaken for 'arthritis'. Calcaneonavicular coalitions, if ossified, can be easily seen in oblique views of the foot.



(a) (b)

Figure 21.13 Tarsal coalition (a) X-ray appearance of a calcaneonavicular bar. (b) CT image showing incompletely ossified talocalcaneal bars bilaterally (arrows).

CT scanning is the most reliable way of demonstrating tarsal coalitions and is very good for defining the pathological anatomy, as well as identifying dual coalitions.

MRI scanning may be helpful in a slightly younger child and when other causes of rigid flat foot are considered (e.g. inflammatory arthritis and infection of the hind- or midfoot).

Radioscintigraphy is rarely indicated due to the sensitivity of MRI scans.

Treatment

Physiological flat foot Young children with flexible flat feet require no treatment. Parents need to be reassured of its normality and told that the foot posture will usually correct itself between the ages of 5 and 10 years. If by then it does not fully correct, the foot will remain flat but function is unlikely to be impaired. Some parents will cite examples of other children who were helped by insoles or moulded heel-cups. These appliances may reduce symptoms and serve mainly to alter the pattern of weight bearing and hence that of shoe wear but do not influence foot growth; simply put, they are more effective in treating the shoes than the feet. There is no scientific evidence that any shoe, insole or orthotic influences foot growth; to the contrary there are longitudinal studies that show no change in a treated group of children with flat feet, compared to an untreated group. There is even evidence to suggest that shoes may inhibit natural development of the longitudinal arch of the foot.

Tight tendo Achilles Flat foot associated with a short tendo Achilles (FFF-STA) and restricted dorsiflexion at the ankle may benefit from tendon-stretching exercises and should be encouraged.

Accessory navicular This occurs in approximately 10% of feet and is usually asymptomatic. Sometimes the main complaint (with a flexible flat foot) is tenderness over an unusually prominent navicular on the medial border of the midfoot. X-rays may show an extra ossicle at this site – the accessory navicular (there are three recognized types). Symptoms are due to pressure (and possibly a 'bursitis') over the bony prominence (Type 3), or repetitive strain at the synchondrosis between the accessory ossicle and the navicular proper (Type 2). Type 1 rarely causes any symptoms at all.

Orthotics can be very helpful but must correct the heel alignment out of valgus which results in a secondary elevation of the medial longitudinal arch and offloads the accessory bone. If symptoms persist, the accessory bone can be shelled out from within the tibialis posterior tendon (Type 2). If the medial arch is significantly flat, the tibialis posterior

tendon can be used as a 'hitch' by reinserting it through a hole drilled in the navicular and suturing the loop with the foot held in maximum inversion (Kidner's operation).

Rigid flat foot (tarsal coalition) The initial treatment should always be conservative. Orthotics can be tried but need to be accommodative rather than corrective, the stiffer the subtalar joint becomes. If one tries to correct a stiff subtalar joint, the forces pass through to the ankle, which may be rendered painful as a result. If symptoms do not settle, operative treatment is needed. A calcaneonavicular bar can be resected without much difficulty through a lateral approach, and the operation may be performed at any stage through childhood. The sooner the joints are freed to move the more likely that a good longer-term outcome will be achieved. A section of the bar is removed and the gap filled with fat, a piece of muscle (e.g. extensor digitorum brevis) or bone wax to prevent recurrence. Talocalcaneal coalitions are more difficult to deal with and a CT scan is required to assess the surgical anatomy. A medial approach displacing the tibialis posterior and flexor digitorum longus tendons allows access to the sustentaculum tali and the medial facet of the subtalar joint, which is the typical location. If the coalition is confined to the medial facet or less than 50% of the total area of medial and posterior facet, resection will be possible with good expected outcomes. If the coalition is large or the cartilage of the posterior facet looks thin or worse, degenerate on CT scanning, then a poor outcome is likely and resection is not advisable. In these cases, if orthotics have failed, then changing the mechanics of the foot with a lateral column lengthening has been shown to be effective. Subtalar arthrodesis may be required to salvage a failure due to persistent pain or if notable degeneration has already occurred at presentation.

Flat foot in adults

As in children, the usual picture is of a flexible flat foot with no obvious cause. However, underlying disorders are common enough to always warrant a careful search for abnormal ligamentous laxity, tarsal coalitions, disorders of the tibialis posterior tendon, post-traumatic deformity, degenerative arthritis, neuropathy and conditions resulting in muscular imbalance.

Painful acquired flat foot often results from tibialis posterior dysfunction. Tibialis posterior tendon dysfunction affects predominantly women in later midlife. It is usually of insidious onset, affecting one foot much more than the other, and with identifiable systemic factors such as obesity, diabetes, steroids or surgery. There may be recollection of a minor episode of trauma, such as a twisting injury to the foot. The patient experiences aching discomfort in the line of the tibialis posterior tendon, often radiating up the inner aspect of the lower leg. The foot often feels 'tired'. As the tendon stretches out, the foot drifts into planovalgus, producing the typical acquired flatfoot deformity. As the tendon ruptures, the ache or pain will often improve, temporarily, but as the foot deformity then worsens, the plantar fascia becomes painful and there may be lateral hindfoot pain as the fibula starts to impinge against the calcaneum.

Pathology

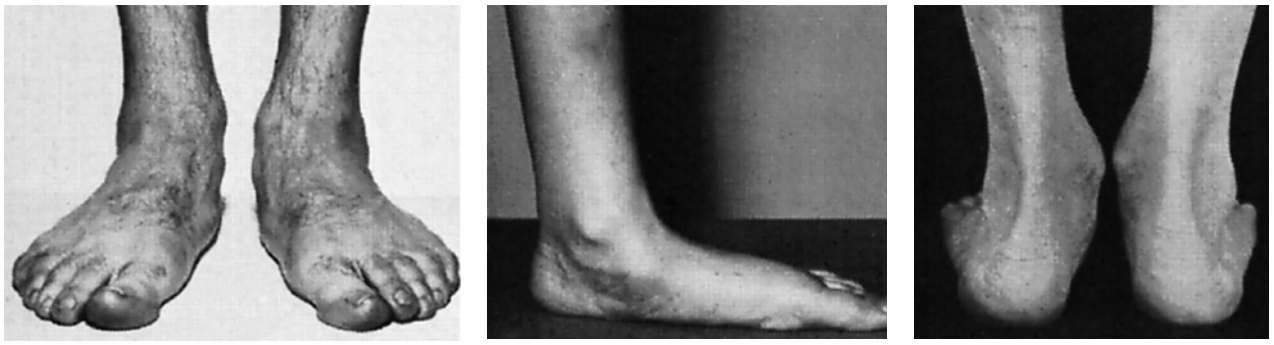
The tibialis posterior is a powerful muscle, with a short excursion of its tendon and a strong mechanical advantage as a foot inverter acting to help maintain the medial longitudinal arch of the foot. This tendon is probably inflamed more commonly and ruptures more frequently than the Achilles tendon. There is usually an initial tenosynovitis. The closely anatomically related spring ligament, the plantar calcaneonavicular ligament, is often also affected when the tibialis posterior tendon is inflamed. The spring ligament may become inflamed, rupture or partially rupture and lose its important structural integrity, helping to support the head of the talus and maintaining the medial arch of the foot. Tibialis posterior tendon elongation and rupture are probably related to an area of hypovascularity in the tendon. Once the tendon elongates, the pathology is then related to the loss of powerful hindfoot inversion, probably confounded by associated stretching of the related ligaments, in particular the spring ligament and the plantar fascia.

Examination

There is usually swelling and tenderness in the line of tibialis posterior, at and distal to the medial malleolus. The hindfoot collapse is best appreciated by viewing the patient from behind, when the valgus deformity of the heel is appreciated, and the forefoot abduction leads to 'too many toes' being seen from this position, compared to the contralateral foot (Figure 21.14). It is difficult for the patient to do a single leg heel raise, as the tibialis posterior cannot stabilize and invert the heel, impairing the heel-raise action of the Achilles tendon.

Imaging

Weight-bearing X-rays show the altered foot axes. The tendon and spring ligament can be assessed with ultrasound or magnetic resonance imaging (MRI) scans.



(a) (b) (c)

Figure 21.14 Flat foot in adults – clinical features (a) The medial arches have dropped and the feet appear to be pronated. (b) The medial border of the foot is flat and the tuberosity of the navicular looks prominent. (c) The heels are in valgus and the toes are visible lateral to the outer edge of the heel on the left side (the 'too-many-toes' sign).

Treatment

The key point is to recognize the condition. If it is in the early stages, then relative rest (sticks or crutches), support with a temporary insole, elasticated foot/ankle support and oral non-steroidal anti-inflammatory drugs (NSAIDs) may be effective. Whether or not to inject the tendon *sheath* with corticosteroid is contentious; but to inject the tendon itself is just plain wrong! These temporary measures may offer the opportunity to institute more permanent solutions, such as modification of weight and activity, and assessment for definitive orthotics.

ORTHOSES

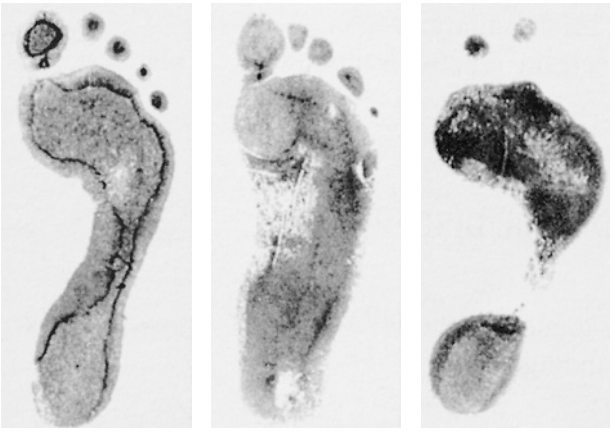
Functional foot orthoses (FFOs) have a role to play in the adult flexible but symptomatic flat foot. These appliances (usually called *orthotics*) are used to correct abnormal foot function or biomechanics and, in so doing, they also correct for abnormal lower extremity function; they are very much more than an 'arch support'.

Orthotics are useful in the treatment of a range of painful conditions of the foot and lower extremities, in particular first MTP joint arthritis, metatarsalgia, arch and instep pain, ankle pain and heel pain. Since abnormal foot function may cause abnormal leg, knee and hip function, orthotics can be used to treat painful tendinitis and bursitis conditions in the ankle, knee and hip, as well as exercise-induced leg pain ('shin splints'). Some types of FFOs are also designed to accommodate painful areas on the soles of the feet (like accommodative foot orthoses). Orthoses may be made of flexible, semi-rigid or rigid plastic or graphite materials. They are relatively thin and fit easily into several types of shoe. They are fabricated from a three-dimensional model of the foot or scanning the foot with a mechanical or optical scanner.

Assessment for orthotics can be performed by a podiatrist (Figure 21.15), who can also advise on whether the usual/intended footwear will accommodate such a device and offer the support needed for it to be effective. 'Off-the-shelf' insoles are cheaper, but there are several advantages to prescription foot orthoses: they are custom-made to precisely fit each foot, and to address specifically the component pathologies affecting the foot, and they are made in relatively rigid, durable materials with a minimal chance of discomfort or irritation to the foot and a greater potential to relieve pain.

PHYSIOTHERAPY

Local treatment of the associated inflammation with physiotherapy might be of benefit. Assessment of the hindfoot biomechanics by a podiatrist might help to prevent progression, and could offer protection to the contralateral side, which is often much less severely affected.



(a) (b) (c)

Figure 21.15 Footprints (a) The normal foot; (b) flat foot (the medial arch touches the ground); and (c) cavus foot (even the lateral arch barely makes contact).

SURGERY

If the condition does not improve with a few weeks of conservative treatment, or the patient presents several months after onset of the symptoms, then surgical intervention should be considered. Options include surgical decompression and tenosynovestomy, or reconstruction of the tendon. In addition, attention is paid to the spring ligament that might also require repair or reefing to improve the support for the talus at the apex of the medial longitudinal arch of the foot. These soft-tissue procedures are often combined with a calcaneal osteotomy to help to protect the tendon and improve the axis. If there is already degeneration in the hindfoot joints, then triple arthrodesis might be indicated (fusing the subtalar, calcaneocuboid and talonavicular joints – the ankle joint is not arthrodesed in this procedure, so foot plantarflexion and dorsiflexion are maintained).

PES CAVUS (HIGH-ARCHED FEET)

In pes cavus the arch is higher than normal, and often there is also clawing of the toes. The close resemblance to deformities seen in neurological disorders where the intrinsic muscles are weak or paralyzed suggests that all forms of pes cavus are due to some type of muscle imbalance. There are rare congenital causes, such as arthrogryposis, but in the majority of cases pes cavus results from an acquired neuromuscular disorder (see Box 21.1). A specific abnormality can often be identified; hereditary motor and sensory neuropathies and spinal cord abnormalities (tethered cord syndrome, diastematomyelia) are the commonest in Western countries but poliomyelitis is the most common cause worldwide. Occasionally the deformity follows trauma such as burns or a compartment syndrome resulting in Volkmann's contracture of the muscles in the sole.

Pathology

The toes are drawn up into a 'clawed' position, the metatarsal heads are forced down into the sole and the arch at the midfoot is accentuated. Often the heel is inverted and the soft tissues in the sole are tight. Under the prominent metatarsal heads callosities may form (Figure 21.16).

Clinical features

Patients usually present during middle childhood. Deformity may be noticed by the parents, grandparents, sports or dance teacher before there are any symptoms. There may be a past history of a spinal disorder, or a family history of neuromuscular defects. As a rule, both feet are affected, but asymmetric cavus can occur.

Pain may be felt under the metatarsal heads or over the toes where shoe pressure is most marked. Callosities appear at the same sites and walking tolerance is reduced. Enquire about symptoms of neurological disorders, such as muscle weakness and joint instability.

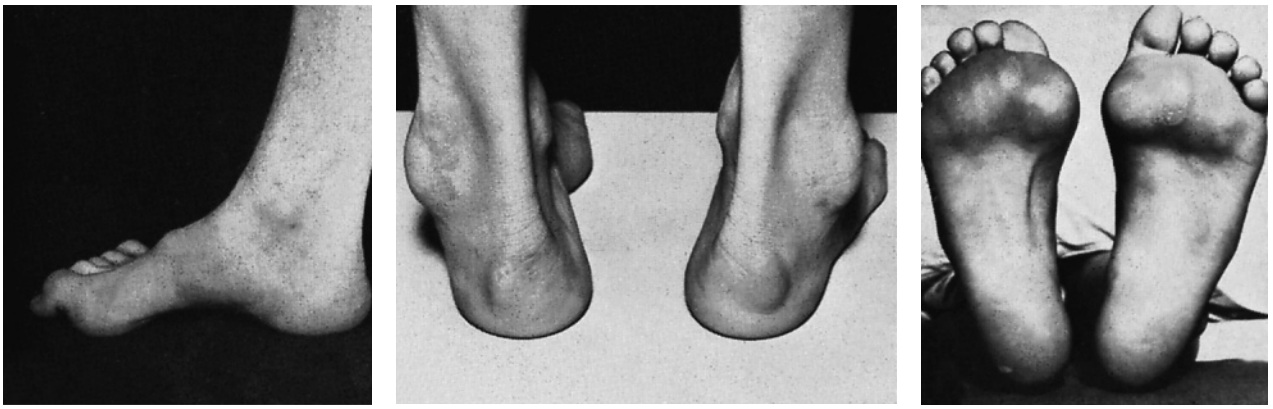
BOX 21.1 NEUROMUSCULAR CAUSES OF PES CAVUS

Muscular dystrophies
- Duchenne
- Becker

Neuropathies
- HMSN I
- HMSN II

Cord lesions
- Poliomyelitis
- Syringomyelia
- Diastomatomyelia
- Tethered cord

Cerebral disorders
- Cerebral palsy
- Friedreich's ataxia



(a) (b) (c)

Figure 21.16 Pes cavus and claw-toes (a) Typical appearance of 'idiopathic' pes cavus. Note the high arch and claw-toes. (b) This is associated with varus heels. (c) Look for callosities under the metatarsal heads.

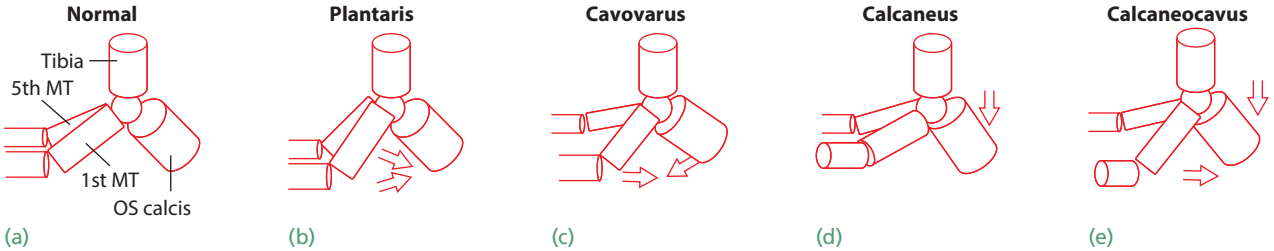


Figure 21.17 The tripod analogy for high-arched feet This simplifies understanding of the various types of pes cavus. (a) The calcaneum, first and fifth metatarsals of the foot are likened to the legs of a tripod. (b) When the first and fifth rays are drawn closer to the heel, a plantaris deformity is present. In a cavovarus deformity (c), the first ray alone is drawn towards the heel, which itself is in varus. In calcaneus (d), the heel is pushed plantarwards. Finally, a calcaneocavus deformity is present (e) when the heel is in calcaneus and the first ray is drawn in.

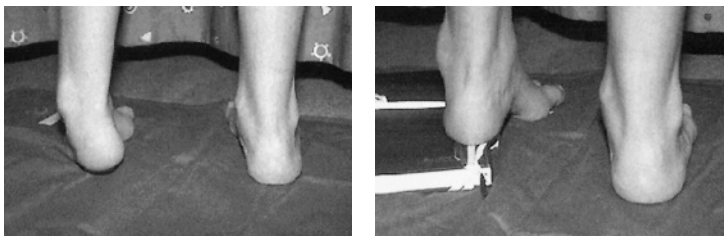


Figure 21.18 Coleman's block test This simple test is used on a high-arched foot to see if the heel is flexible. (a) Normal stance showing the varus position of the heel. (b) With the patient standing on a low block to permit the depressed first metatarsal to hang free, the heel varus is automatically corrected if the subtalar joint is mobile.

The overall cavus deformity is usually obvious; in addition the toes are often clawed and the heel may be varus. Closer inspection will show the components of the high arch; this is important because it leads to an understanding of the responsible deforming forces. Rang presented a tripod analogy that simplifies the problem. The foot is likened to a tripod of which the calcaneus, fifth metatarsal and first metatarsal form the legs. Combinations of deformities affecting one or more of these 'legs' produce the common types of high arch, namely plantaris, cavovarus, calcaneus and calcaneocavus (Figure 21.17).

The toes are held cocked up, with hyperextension at the MTP joints and flexion at the IP joints. There may be callosities under the metatarsal heads and corns on the toes. Early on, the toe deformities are 'mobile' and can be corrected passively by pressure under the metatarsal heads; as the forefoot lifts, the toes flatten out automatically. Later the deformities become fixed, with the MTP joints permanently extended. Mobility in the ankle and foot joints is important. In the cavovarus foot, the heel is inverted. *Coleman's block test* is useful to check if the deformity is reversible (Figure 21.18); if it is, this signifies that the subtalar joint is mobile. If the cavus deformity has been present for a long time, then movements of the ankle, subtalar and midtarsal joints are usually limited.

A neurological examination is important to try to identify a reason for the deformity. Disorders such as hereditary sensory and motor neuropathy and Friedreich's ataxia must always be excluded, and the spine should be examined for signs of dysraphism.

Imaging

Weight-bearing X-rays of the foot contribute further to the assessment of the deformity and the state of the individual joints. On the lateral view, measurement of the *calcaneal pitch* and *Meary's angle* help to determine the components of the high arch (Figure 21.19). In a normal foot the calcaneal pitch is between 10 and 30 degrees, whereas Meary's angle, formed by the axes of the talus and first metatarsal, is zero, i.e. these axes

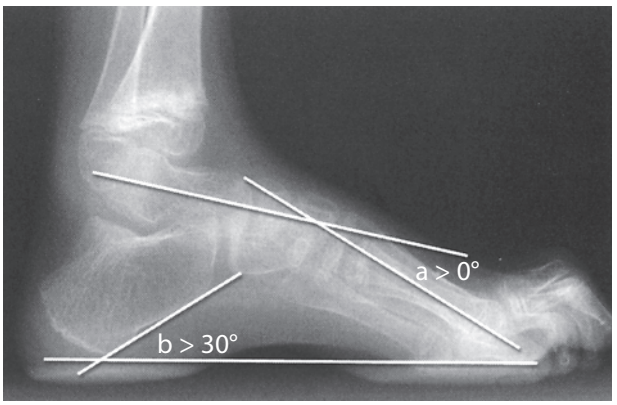


Figure 21.19 Weight-bearing X-rays in foot deformities Non-weight-bearing films are notorious for 'hiding' the true components of foot deformities. In standing lateral views, some measurements are useful in describing the type of high-arched foot: (a) the axes of the talus and first metatarsal are parallel in normal feet but cross each other in a plantaris deformity (Meary's angle); (b) the calcaneal pitch is greater than 30 degrees in calcaneus deformities.

are parallel. In a calcaneus deformity, the calcaneal pitch is increased; in a plantaris deformity, Meary's lines meet at an angle.

MRI scans of the spine will exclude a structural disorder, especially if this is more common than polio as a cause of high-arched feet in the region.

Treatment

In most physiological or non-progressive cases no treatment is required; apart from the difficulty of fitting shoes, the patient has few complaints.

Foot deformity In general, patients need treatment only if they have symptoms. However, the problem with high-arched feet is that it is often a progressive disorder that becomes more difficult to treat when the deformities are fixed; therefore treatment should start before the feet become stiff. Non-operative treatment in the form of custom-made shoes with moulded inserts may provide some relief but does not alter the deformity or influence its progression. Surgery is often needed and the type of procedure will depend on the child's age, underlying cause, site and flexibility of the individual deformities and type of muscle imbalance.

The aim of surgery is to provide a pain-free, plantigrade, supple but stable foot. The methods available are soft-tissue releases, osteotomies and tendon transfers. However, the deformity first needs to be corrected before a tendon transfer is considered; additionally, the transfer only works if the joints are mobile.

An equinus contracture is dealt with by lengthening of the tendo Achillis, and posterior capsulotomies of the ankle joint may need to be added in more severe cases. The varus hindfoot, if shown to be reversible by Coleman's block test, may not require any hind foot procedure as the deformity is in the forefoot. As the situation stiffens and the foot becomes partially correctable it may benefit from a superficial plantar medial release consisting of a release of the plantar fascia (the tight fascia acts as a contracted windlass on weight-bearing, accentuating the deformity), abductor hallucis and flexor hallucis brevis. For even more resistant feet the talonavicular joint is released in addition. However, if the subtalar joint is stiff and irreducible, then calcaneal osteotomy will be needed; two types are commonly used: (1) the lateral closing wedge (an opening wedge on the medial side is a comparable operation but is fraught with wound problems); (2) a lateral translation osteotomy (Figure 21.20).

Treatment of a calcaneocavus deformity (which is the least common type of high arch) differs according to the age of the child. In young children (who usually have a neurological problem) tendon transfers such as transferring the tibialis anterior through the interosseous membrane to the calcaneum may be combined with tenodesis of the ankle using the tendo Achillis. Older children may need crescentic calcaneal osteotomies, which will correct both varus and calcaneus deformities, or variations of a triple arthrodesis.

Midfoot deformities are usually cavus (plantarflexed first metatarsal) or plantaris (plantarflexed first and fifth metatarsals). The Jones tendon transfer helps elevate the depressed first metatarsal by using the extensor hallucis longus tendon as a sling through the neck of the first metatarsal (Figure 21.21). Often the peroneus longus is overactive and is partly responsible for pulling the first metatarsal down; some balance is restored by dividing this tendon on the lateral side of the foot and attaching the proximal end to the peroneus brevis, thereby removing the deforming force and improving the power of eversion simultaneously.

Occasionally the deformity affecting the first metatarsal is fixed, in which case a dorsal closing wedge osteotomy or plantar opening wedge osteotomy with graft of the medial cuneiform is needed. The apex of the deformity is at the cuneiform level and is preferred to the first metatarsal. A plantaris deformity is treated along similar lines for the first ray, and combined with a plantar fascia release if the deformity is mobile, but basal metatarsal osteotomies or even a wedge resection and arthrodesis across the midfoot are needed for rigid deformities.

In severe examples and in those who have either relapsed or who have responded poorly with soft-tissue releases and osteotomies, salvage surgery in the form of osteotomies and gradual correction using an external circular frame is possible. In severe cases a triple arthrodesis may be considered but produces a stiff and short but plantigrade and usually pain-free foot.

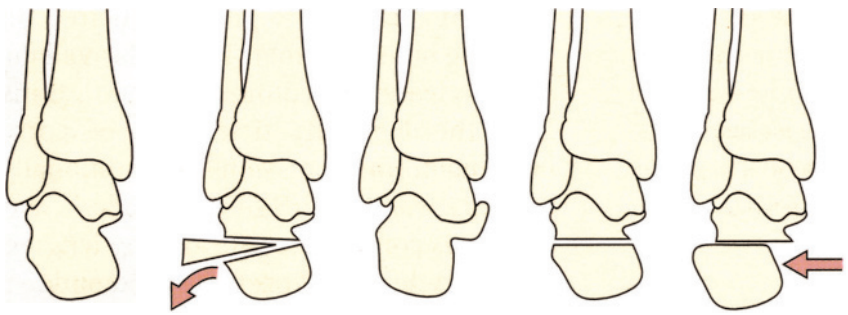
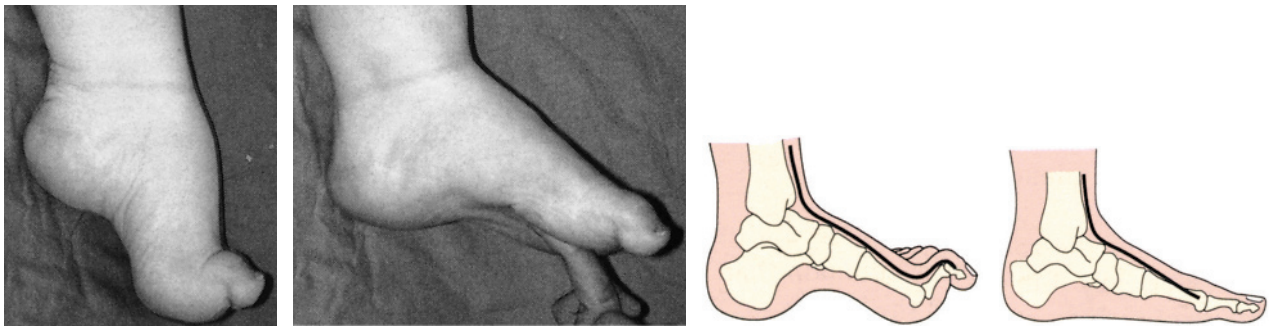


Figure 21.20 Treatment of pes cavus 1 In a normal foot (a), the point of contact of the heel is slightly lateral to the centre of the ankle, producing an eversion lever when weight is borne. In a varus heel (b) excising a wedge of bone from the lateral side, or (c) performing a lateral translation osteotomy.



(a) (b) (c) (d)

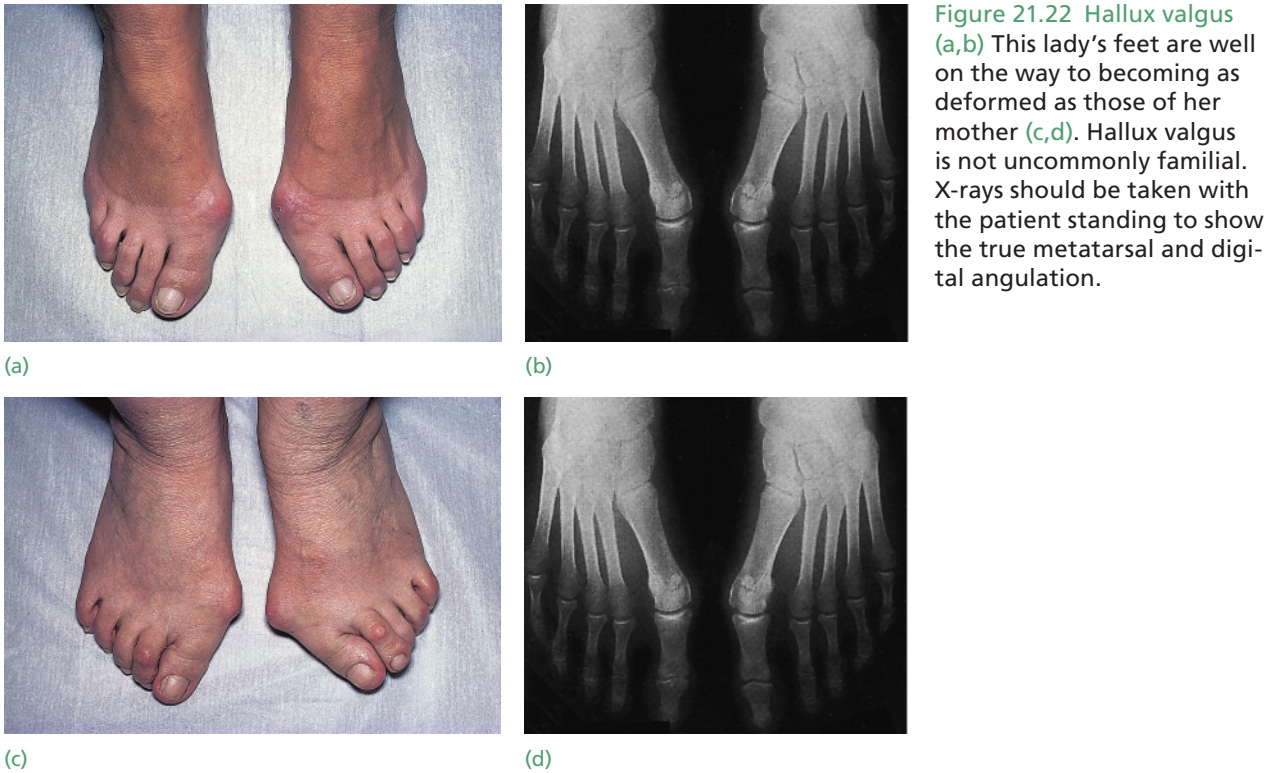
Figure 21.21 Treatment of pes cavus 2 (a,b) If the great toe is clawed and the first metatarsal depressed, reducing the subluxation at the metatarsophalangeal joint by simply elevating the neck of the metatarsal often reduces the severity of the cavus deformity. The surgical equivalent of this effect is (c,d) the Jones tendon transfer: the extensor halluces longus tendon is detached distally and transferred to the neck of the first metatarsal; the interphalangeal joint is then either fused or tenodesed.

Clawed toes Correction of a clawed first toe is by the Jones tendon transfer, which involves either a tenodesis or fusion of the IP joint. Clawing of the lesser toes is treated with a flexor tendon transfer to the extensor hood of each toe, and MTP joint capsulotomies if the toes are still passively correctable; however, if the deformities are fixed, proximal IP fusion is needed.

HALLUX VALGUS

Hallux valgus is the commonest of the foot deformities (and probably of all musculoskeletal deformities). In people who have never worn shoes the big toe is in line with the first metatarsal, retaining the slightly fan-shaped appearance of the forefoot. In people who habitually wear shoes the hallux assumes a valgus position; but only if the angulation is excessive is it referred to as 'hallux valgus' (Figure 21.22).

Splaying of the forefoot, with varus angulation of the first metatarsal, predisposes to lateral angulation of the big toe in people wearing shoes – more so in those who wear high-heeled shoes or shoes with a narrow toe box (for which read many women's shoes). *Metatarsus primus varus* may be congenital, or it may result from loss of muscle tone in the forefoot in elderly people. Hallux valgus is also common in rheumatoid arthritis, probably due to weakness of



(a) (b) (c) (d)

Figure 21.22 Hallux valgus (a,b) This lady's feet are well on the way to becoming as deformed as those of her mother (c,d). Hallux valgus is not uncommonly familial. X-rays should be taken with the patient standing to show the true metatarsal and digital angulation.

the joint capsule and ligaments. Heredity plays an important part; a positive family history is obtained in over 60% of cases.

Pathological anatomy

The elements of the deformity are lateral deviation and rotation of the hallux, together with a prominence of the medial side of the head of the first metatarsal (a bunion). Lateral deviation of the hallux may lead to overcrowding and deformity of the other toes and sometimes overriding of adjacent toes. When the valgus deformity exceeds 30-40 degrees, the great toe rotates into pronation so that the nail faces more medially and the sesamoid bones of flexor hallucis brevis are displaced laterally; in severe deformities the tendons of flexor and extensor hallucis longus bowstring on the lateral side, thus adding to the deforming forces. The contracted adductor hallucis and lateral capsule contribute further to the fixed valgus deformity.

Prominence of the first metatarsal head is due to subluxation of the MTP joint; increasing shoe pressure on the medial side leads to the development of an overlying bursa and thickened soft tissues, additional changes that combine to form the defining 'bunion' that eventually accompanies the great-toe deformity. When exposed at operation, the medial prominence looks like an exostosis (because of a deep sagittal sulcus on the head of the metatarsal) but there is usually no true exostosis.

In long-standing cases the MTP joint becomes osteoarthritic and osteophytes may then add to the prominence of the metatarsal head.

Clinical features

The commonest complaints are pain over the bunion, worries about cosmesis and difficulty fitting shoes. Often there is also deformity of the lesser toes and pain in the forefoot. With the patient standing, planovalgus hindfoot collapse may become apparent.

The great toe is in valgus and the bunion varies in appearance from a slight prominence over the medial side of the first metatarsal head to a red and angry-looking bulge that is tender. The MTP joint often retains a good range of movement, but in long-standing cases it may be osteoarthritic. Always check the circulation and sensation.

X-rays

Standing views will show the degree of metatarsal and hallux angulation. Lines are drawn along the middle of the first and second metatarsals and the proximal phalanx of the great toe; normally the intermetatarsal angle is less than 9 degrees and the valgus angle at the MTP joint less than 15 degrees. Any greater degree of angulation should be regarded as 'hallux valgus'.

Not all types of valgus deformity are equally progressive and troublesome. Based on the X-ray appearances, Piggott divided patients with hallux valgus into the following three types (Figure 21.23):

- *Type 1* – those in whom the MTP joint is normally centred but the articular surfaces, though congruent, are tilted towards valgus. Type 1 is a stable joint and any deformity is likely to progress very slowly or not at all.

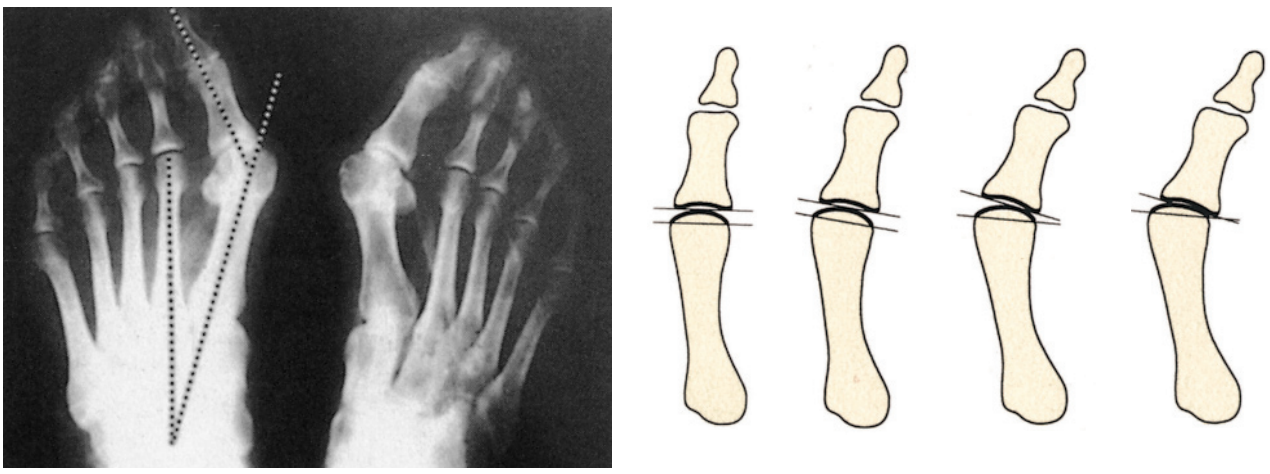


Figure 21.23 Hallux valgus – X-rays (a) The intermetatarsal angle (between the first and second metatarsals) as well as the metatarsophalangeal angle of the hallux are recorded. Piggott defined three types of hallux valgus, based on the position and tilt of the first MTP articular surfaces: in normal feet (b) the articular surfaces are parallel and centred upon each other. In congruent hallux valgus (c) the lines across the articular surfaces are still parallel and the joint is centred, but the articular surfaces are set more obliquely to the long axes of their respective bones. In (d) the deviated type of hallux valgus, the lines are not parallel and the articular surfaces are not congruent. In the subluxated type (e) the surfaces are neither parallel nor centred.

- *Type 2* – those in whom the articular surfaces are not congruent, the phalangeal surface being tilted towards valgus. This type is somewhat unstable and likely to progress.
- *Type 3* – those in whom the joint is both incongruent and slightly subluxated. Type 3 is even more unstable and almost certain to progress.

Treatment

ADOLESCENTS

Many young patients are asymptomatic, but worry over the shape of the toe and an anxious mother keen not to let the condition become as severe as her own will bring the patient to the clinic. It is wise to try conservative measures first, mainly because surgical correction in this age group carries a 20–40% recurrence rate. This consists essentially of encouraging the patient to wear shoes with wide and deep toe boxes, soft uppers and low heels – 'trainers' are a good choice. If X-rays show a type 1 (congruous) deformity, the patient can be reassured that it will progress very slowly, if at all. If there is an incongruous deformity, surgical correction will sooner or later be required.

There are a number of non-operative strategies that may be adopted to deal with the deformity and the resulting limitations, but none that will get rid of the bunion itself. Accommodating, comfortable shoes can help but are not acceptable for some patients (or professions). Lace-up or Velcro®-fastening shoes are better than slip-ons, and flat shoes are probably better than those with a raised heel.

Bunion pads (like a Polo/doughnut shape) can help to offload the tender bunion, but strapping and overnight splints are probably a waste of money with no quality research to support their use. Chiropody can help by taking care of the callosities and skin compromise.

Podiatrists may help to correct the foot biomechanics, but there is no good evidence that antipronatory orthoses are effective in the longer-term management of the bunion. Diabetic services often provide specialized footcare.

Operative treatment (See Figure 21.24 and 21.25) In the adolescent with *mild deformities*, where the hallux valgus angle is less than 25 degrees, correction can be obtained by either a soft-tissue rebalancing operation (see later) or by a metatarsal osteotomy. If the X-ray shows a congruent articulation, the deformity is largely bony and therefore amenable to correction by a distal osteotomy.

If the MTP articulation is incongruent, the deformity is in the joint and soft-tissue realignment is indicated. The tight structures on the lateral side (adductor

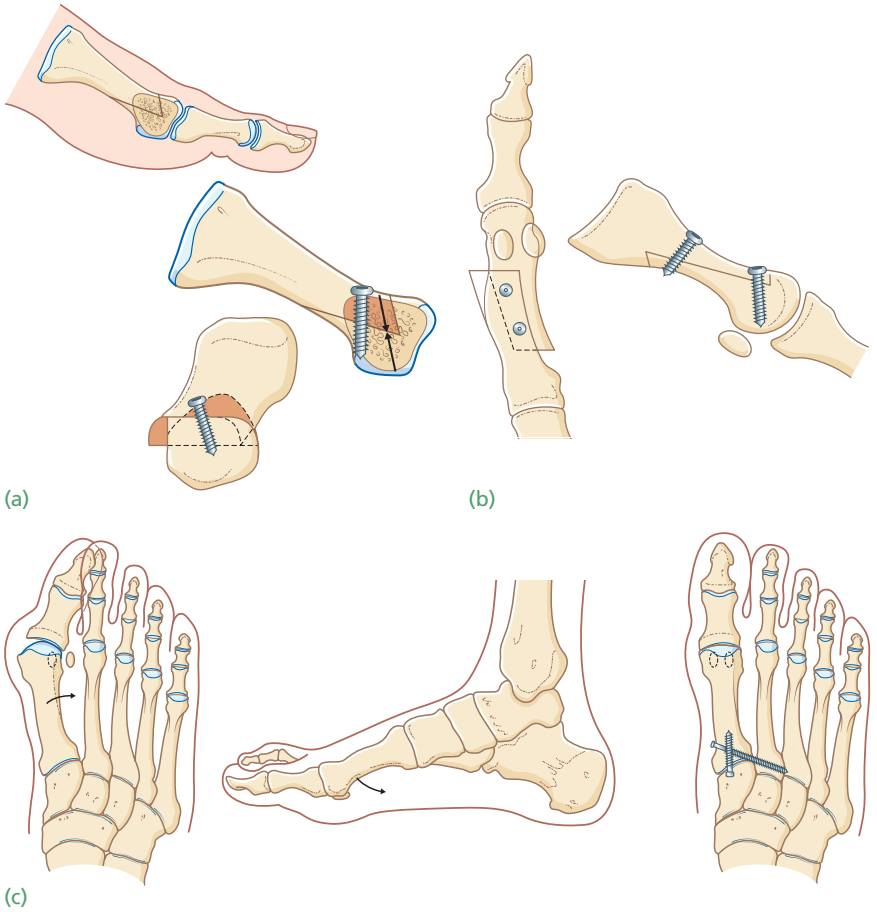
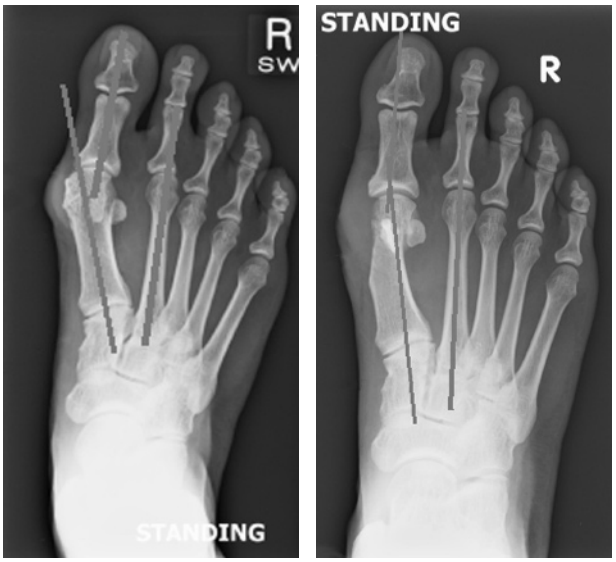


Figure 21.24 Hallux valgus – operative treatment (a) Distal metatarsal osteotomy – chevron; (b) double metatarsal osteotomy – scarf; (c) basal realignment and arthrodesis – Lapidus. These may be fixed in a variety of ways, and several modifications have been described.



(a) (b)

Figure 21.25 Hallux valgus – treatment (a) X-ray before operation. Note the valgus alignment of the great toe, the relative playing or increased angulation between 1st and 2nd metatarsals, and the lateral displacement of the sesamoids under the 1st metatarsal head. (b) X-ray after distal osteotomy. With fixation of the osteotomy, note the improved great toe alignment, less effective angulation between 1st and 2nd metatarsals, and improved position of the sesamoids.

hallucis, transverse metatarsal ligament, and lateral joint capsule) are released; the prominent bone on the medial side of the metatarsal head is pared down and the capsule on the medial side is reefed.

In *moderate and severe deformities* the hallux valgus angle may be greater than 30 degrees and intermetatarsal angle wider than 15 degrees. If the MTP joint is congruent, a distal osteotomy combined with a corrective osteotomy of the base of the proximal phalanx (Akin osteotomy) is recommended. For greater deformities, if the joint is subluxed, a soft-tissue adjustment is needed as well as a proximal metatarsal osteotomy. This basal osteotomy is carried out to reduce a wide intermetatarsal angle; care is needed not to injure an open physis or growth of the metatarsal will be stunted.

ADULTS

In the adult, when self-care is insufficient and the bunion is causing pain and difficulty with footwear, surgical options are appropriate. Recurrent infection and ulceration are also indications for operative treatment. The type of surgery proposed will depend on the level and extent of the deformity. This will usually comprise: (1) an osteotomy to realign the first metatarsal; (2) soft-tissue procedures to rebalance the joint.

A number of different osteotomy patterns have been described and named after their 'inventors' or the pattern of bone cut (chevron, scarf, etc.), or the part of the metatarsal that is osteotomized (distal usually if there is less deformity; proximal or basal for greater deformity). These procedures are reviewed in a paper by Robinson and Limbers (2005).

There is convincing evidence to show that a distal osteotomy is associated with reduced pain and increased ability to work in the medium to long term; the safety profile is good, with a less than 10% complication rate and with many procedures being performed as day-case operations and without plaster immobilization in the postoperative period. Patient satisfaction with bunion surgery is generally good, with 75% being satisfied with the outcome.

ELDERLY PATIENTS

Hallux valgus in the elderly is best treated by shoe modifications; where this fails, and in those whose functional demands are low, treatment by excision arthroplasty is usually successful. In the classic *Keller's operation*, the proximal third of the proximal phalanx, as well as the bunion prominence, are removed. This used to be the most common operation for hallux valgus but it has fallen into disuse because of the high rate of recurrent deformity and complications such as loss of control over great-toe movement, overload of the other metatarsals, metatarsalgia and dubious cosmetic improvement.

Complications

Recurrent deformity or failure to meet the patient's expectations are common problems in hallux valgus surgery. Up to 25% are dissatisfied in the longer term (5 years or more postoperatively), although most surgeons believe their own results are better than this!

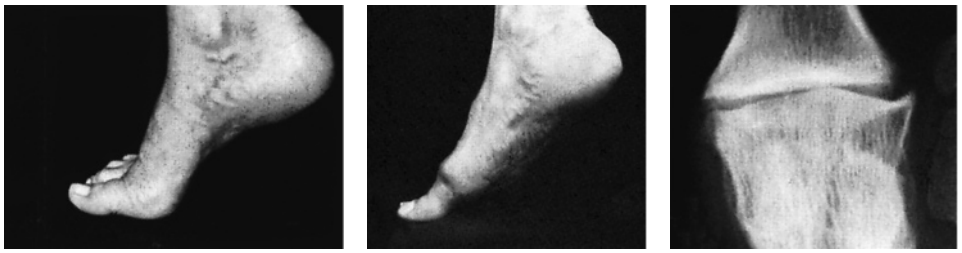
Infection and *ulceration* are particular problems in the diabetic foot and are an indication (rather than a contraindication) for surgery.

Transfer metatarsalgia may occur if the realignment or shortening of the first ray does not take account of the relative lengths of the lesser metatarsals, which then become prominent and overloaded; a metatarsal stress fracture sometimes occurs. Forefoot corrective surgery should strive to produce a balanced forefoot with appropriately distributed weight bearing.

Complex regional pain syndrome is a potential complication of all foot operations.

HALLUX RIGIDUS

'Rigidity' (or stiffness) of the first MTP joint occurs at almost any age from adolescence onwards. In young people it may be due to local trauma or osteochrondritis dissecans of the first metatarsal head. In older people it is usually caused by long-standing joint disorders such as gout, pseudogout or osteoarthritis (OA)



(a) (b) (c)

Figure 21.26 Hallux rigidus (a) In normal walking, the big toe dorsiflexes (extends) considerably. With rigidus (b), dorsiflexion is limited. (c) The usual cause is OA of the first MTP joint.

and is very often bilateral (Figure 21.26). In contrast to hallux valgus, men and women are affected with equal frequency. A family history is common.

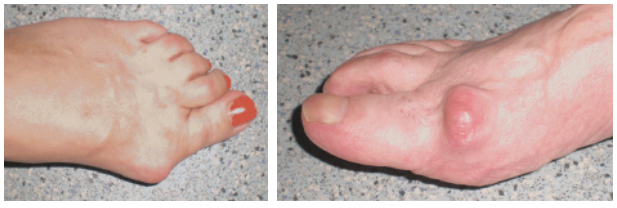
Clinical features

Pain on walking, especially on slopes or rough ground, is the predominant symptom. The patient eventually develops an altered gait, trying to offload the first MTP joint by transferring weight across to the lesser toes; there is also impaired power in toe-off during the gait cycle. The great toe is straight and often has a callosity under the medial side of the distal phalanx. The MTP joint feels knobbly; a tender dorsal 'bunion' (actually a large osteophyte) is diagnostic (Figure 21.27). Dorsiflexion is restricted and painful, and there may be compensatory hyperextension at the interphalangeal joint. The outer side of the sole of the shoe may be unduly worn – the result of rolling the foot outwards to avoid pressing on the big toe.

It is important to check the state of the other joints in the foot in order to rule out a polyarthropathy.

X-rays

The features are essentially those of OA: narrowing of the joint space, subchondral sclerosis and marginal osteophytes. There may be signs of recent or old osteochondritis ('squaring' of the metatarsal head).



(a) (b)

Figure 21.27 'Bunions' Compare the two types of 'bunion': (a) medial bunion in hallux valgus – what the surgeon will refer to as a bunion. (b) Dorsal prominence of an osteophyte in hallux rigidus – what referral letters from GPs will often refer to as a bunion.

Treatment

NON-OPERATIVE TREATMENT

If the condition is not interfering with activity, it can be left alone and the patient can be reassured. Intermittent attacks of pain can be relieved by an intra-articular injection of corticosteroid and local anaesthetic. If, however, the condition is painful and restricting of activity, the risks of long-term NSAIDs must be balanced against those of surgical intervention. Some orthotic devices will offload or reduce movement at the first MTP joint, but these are usually full-length insoles and relatively bulky – they may not fit in a smart shoe (at least not when the foot is in it as well!) A rocker-soled shoe can abolish pain by allowing the foot to 'roll' without the necessity for dorsiflexion at the MTP joint, but many people are unwilling to wear such shoes.

OPERATIVE TREATMENT

Pain at the first MTP joint that is intrusive or limits activity should be an indication for referral. In limited arthritic disease, simply removing the dorsal osteophyte (*cheilectomy*) might be effective and may be coupled with an extension osteotomy in the proximal phalanx, to alter the load-bearing region of the articulation.

If the joint is more arthritic, then a fusion or *arthrodesis* offers a good chance of returning the patient to function, walking comfortably without a limp. The joint should be fused in 10 degrees of valgus and 10–15 degrees of dorsiflexion in relation to the sole of the foot, or with about 5–10 mm clearance between the line of the sole of the foot and the pulp of the great toe. Too little dorsiflexion will cause pain during toe-off and too much will result in the toe pressing against the shoe upper. Female patients may be concerned that they will be unable to wear shoes with a higher heel if the toe is fused, but in fact the majority are able to wear footwear that can include moderate heels.

Arthroplasty is more controversial. Keller's operation (an *excisional arthroplasty*), carries a high risk of complications and seldom brings improvement in function; the procedure is no longer recommended. *Interposition*

arthroplasty has from time to time been popular and can provide excellent pain relief, especially in patients with advanced OA. A simple capsular arthroplasty is probably the safest. Silicone implants were often used in the past, but silicone-related complications were common and the operation is no longer recommended for hallux rigidus. Metallic implants have fared better (in experienced hands) but these also produce variable long-term results. The majority of implant designs have not performed well in long-term follow-up studies.

DEFORMITIES OF LESSER TOES

The commonest deformities of the lesser toes are 'claw', 'hammer' and 'mallet' (Figure 21.28). These terms are often used interchangeably, leading to confusion.

- *Claw toe* is characterized by hyperextension at the MTP joint and flexion at both IP joints.
- *Hammer toe* is an acute flexion deformity of the proximal IP joint only; in severe examples there may be some extension at the MTP joint. The distal IP joint is either straight or hyperextended.
- *Mallet toe* is a flexion deformity of the distal IP joint.

Claw toes

The IP joints are flexed and the MTP joints hyperextended. This is an 'intrinsic-minus' deformity that is seen in neurological disorders (e.g. peroneal muscular atrophy, poliomyelitis and peripheral neuropathies) and in rheumatoid arthritis. Usually, however, no cause is found. The condition may also be associated with pes cavus.

Clinical features

The patient complains of pain in the forefoot and under the metatarsal heads. Usually the condition is bilateral and walking may be severely restricted. At first the joints are mobile and can be passively corrected; later the deformities become fixed and the MTP joints subluxed or dislocated. Painful corns may develop on the dorsum of the toes and callosities under the metatarsal heads. In the most severe cases the skin ulcerates at the pressure sites.

Treatment

FLEXIBLE DEFORMITY

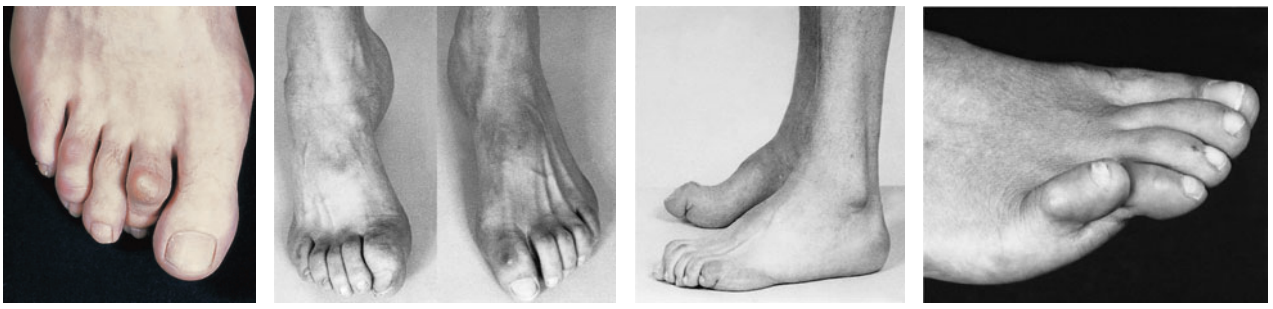
So long as the toes can be passively straightened the patient may obtain relief by wearing a metatarsal support or by having a transverse metatarsal bar fitted to the shoe. A daily programme of intrinsic muscle exercises is important. If these measures fail to relieve discomfort, an operation is indicated. 'Dynamic' correction is achieved by transferring the long toe flexors to the extensors. The operation at one stroke removes a powerful IP flexor and converts it to a MTP flexor and IP extensor.

FIXED DEFORMITY

When the deformity is fixed, it may be either accepted and accommodated by special footwear or treated by one of the following operations: interphalangeal arthrodesis, joint excision or amputation.

Interphalangeal arthrodesis If there is no joint disease, proximal IP arthrodesis and dorsal capsulotomy of the MTP joints permits active flexion of the MTP joints by the long flexors. This is sometimes combined with transfer of the extensor hallucis longus to the first metatarsal, thus removing a deforming force while retaining the muscle as a forefoot stabilizer.

Joint excision Fixed claw deformities, usually associated with destruction of the MTP joints (e.g. in rheumatoid arthritis), can be dealt with by a range of excision arthroplasties of the MTP joints – preferably removal of only the bases of the proximal phalanges and trimming of the metatarsal heads. This can usually be achieved through two longitudinal incisions on the dorsum of the foot. If the great toe is affected, a modified Keller's



(a) (b) (c) (d)

Figure 21.28 Disorders of the lesser toes (a) Hammer-toe deformity. (b,c) Claw toes. This patient suffered from peroneal muscular atrophy, a neurological disorder causing weakness of the intrinsic muscles and cavus feet. (d) Overlapping or 'curly' fifth toe.

operation is performed. The base of the proximal phalanx is excised and the plantar pad (which is often displaced in these deformities) is returned to its normal position beneath the metatarsal head; the space between the metatarsal and phalanx is then filled by suturing the long extensor tendon to the flexor.

Amputation Toes that are severely contracted, dislocated and ulcerated are worse than none. If the circulation is satisfactory and the patient is willing to accept the appearance, amputation of all ten toes is a useful palliative operation.

Hammer toe

The proximal IP joint is fixed in flexion, while the distal joint and the MTP joint are extended. The second toe of one or both feet is commonly affected, and hyperextension of the MTP joint may go on to dorsal dislocation. Shoe pressure may produce painful corns or callosities on the dorsum of the toe and under the prominent metatarsal head.

The cause is obscure: the similarity to boutonnière deformity of a finger suggests an extensor dysfunction, a view supported by the frequent association with a dropped metatarsal head, flat anterior arch and hallux valgus. A simpler explanation is that the toe was too long or the shoe too short.

Treatment

Operative correction is indicated for pain or for difficulty with shoes. The toe is shortened and straightened by excising the joint. An ellipse of tissue (including the corn and the underlying extensor tendon) is removed and the proximal IP joint is entered; the articular surfaces are nibbled away and the raw ends of the proximal and middle phalanges are brought together with the toe almost straight. The position is held by a longitudinally placed K-wire, which is retained for 6 weeks. An alternative (and some would say preferable) operation is simple excision of the head of the proximal phalanx, or excision of both articular surfaces, without formal arthrodesis; this has been described with a range of combined soft-tissue procedures. The toe is splinted for a few weeks to allow healing in the corrected position.

If the MTP joint is dislocated, a dorsal capsulotomy and elongation of the extensor tendon may be necessary; the toe is held in position by driving the K-wire more proximally, or by inserting a second wire.

Mallet toe

In mallet toe it is the distal IP joint that is flexed. The toenail or the tip of the toe presses into the shoe, resulting in a painful callosity.

If conservative treatment (chiropody and padding) does not help, operation is indicated. The distal IP joint is exposed, the articular surfaces excised and the toe straightened; flexor tenotomy may be needed. A thin K-wire is inserted across the joint and left in position for 6 weeks.

Fifth toe deformities

Overlapping fifth toe

This is a common congenital anomaly (Figure 21.28d). If symptoms warrant, the toe may be straightened by a dorsal V/Y-plasty, reinforced by transferring the flexor to the extensor tendon. Tight dorsal and medial structures may have to be released. The toe is held in the overcorrected position with tape or K-wire for 6 weeks. Severe deformities or relapses may need a transfer of the long extensor tendon beneath the proximal phalanx to the abductor digiti minimi.

Cock-up deformity

The MTP joint is dislocated and the little toe sits on the dorsum of the metatarsal head. Operative treatment is usually successful: through a longitudinal plantar incision, the proximal phalanx is winkled out and removed; the wound is closed transversely, thus pulling the toe out of the hyperextended position.

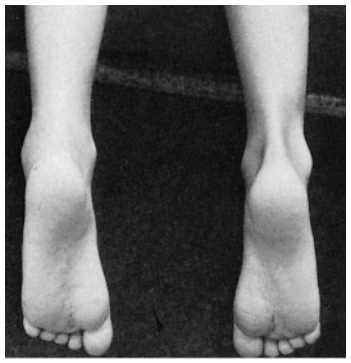
Tailor's bunion

An irritating or painful bunionette may form over an abnormally prominent fifth metatarsal head. If the shoe cannot be adjusted to fit the bump, the bony prominence can be trimmed, taking care not to sever the tendon of the fifth toe abductor. If the metatarsal shaft is bowed laterally (as is often the case), it can be straightened by performing either a distal osteotomy or a varus correction at the base of the metatarsal.

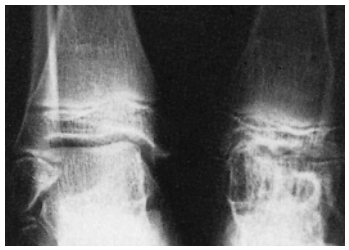
TUBERCULOUS ARTHRITIS
(see also Chapter 2)

Tuberculous infection of the ankle joint begins as a synovitis or as an osteomyelitis and, because walking is painful, may present before true arthritis supervenes. The ankle is swollen and the calf markedly wasted; the skin feels warm and movements are restricted. Sinus formation occurs early.

X-rays show regional osteoporosis, sometimes a bone abscess and, with late disease, narrowing and irregularity of the joint space (Figure 21.29).



(a)



(b)

Figure 21.29 Tuberculous arthritis of the ankle in a child (physes are open) (a) The swelling of the left ankle is best seen from behind; (b) shows regional osteoporosis and joint destruction.

Treatment

In addition to general treatment (Chapter 2) a removable splint is used to rest the foot in neutral position. If the disease is arrested early, the patient is allowed up without weight-bearing in a calliper, gradually taking more weight and then discarding the calliper altogether. Following arthritis, weight-bearing is harmless, but stiffness is inevitable and usually arthrodesis is the best treatment.

RHEUMATOID ARTHRITIS
(see also Chapter 3)

The ankle and foot are affected almost as often as the wrist and hand. Early on there is synovitis of the MTP, intertarsal and ankle joints, as well as of the sheathed tendons (usually the peronei and tibialis posterior). As the disease progresses, joint erosion and tendon dysfunction prepare the ground for increasingly severe deformities (Figure 21.30).

Forefoot

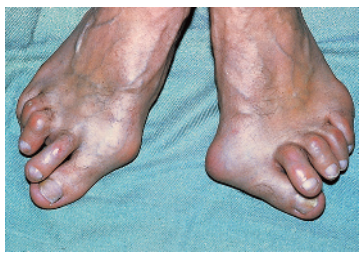
Pain and swelling of the MTP joints are among the earliest features of rheumatoid arthritis. Shoes feel uncomfortable and the patient walks less and less. Tenderness is at first localized to the MTP joints; later the entire forefoot is painful on pressing or squeezing. With increasing weakness of the intrinsic muscles and joint destruction, the characteristic deformities appear: a flattened anterior arch, hallux valgus, claw toes and prominence of the metatarsal heads in the sole (patients say it feels like walking on pebbles). Subcutaneous nodules are common and may ulcerate. Dorsal corns and plantar callosities also may break down and become infected. In the worst cases the toes are dislocated, inflamed, ulcerated and useless.

X-rays show osteoporosis and periarticular erosion at the MTP joints. Curiously – in contrast to the situation in the hand – the smaller digits (fourth and fifth toes) are affected first.

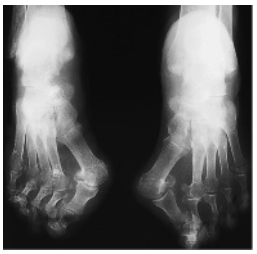
Treatment

During the stage of synovitis, corticosteroid injections and attention to footwear may relieve symptoms; operative synovectomy is occasionally needed. Once deformity is advanced, treatment is that of the claw toes and hallux valgus. Sometimes specially made shoes will accommodate the toes in relative comfort. If this does not help, the most effective operation is excision arthroplasty in order to relieve pressure in the sole and to correct the toe deformities. For the hallux, an alternative is MTP fusion.

Forefoot surgery is more likely to succeed if the hindfoot is held in the anatomical position. It is therefore important to treat the foot as a whole and attend also to the proximal joints.



(a)



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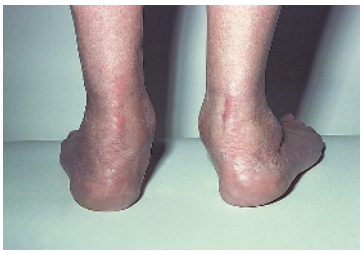


(c)

Figure 21.30 Rheumatoid arthritis (a,b) Forefoot deformities are similar to those in non-rheumatoid feet but more severe. They are due to a combination of joint erosion and tendon attrition. (c) Swelling and deformity of the hindfoot due to a combination of arthritis and tenosynovitis. In this case, both the ankle and the subtalar joints are affected.

Ankle and hindfoot

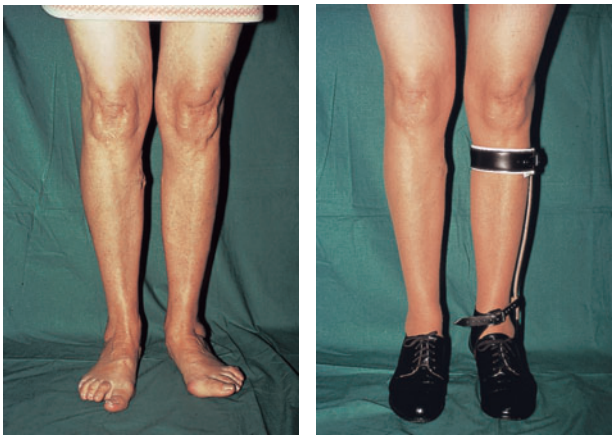
The earliest symptoms are pain and swelling around the ankle. Walking becomes increasingly difficult and, later, deformities appear. On examination, swelling and tenderness are usually localized to the back of the medial malleolus (tenosynovitis of tibialis posterior) or the lateral malleolus (tenosynovitis of the peronei). Less often the ankle swells (joint synovitis) and its movements are restricted. Inversion and eversion may be painful and limited; subtalar erosion is common. In the late stages the tibialis posterior may rupture (all too often this is missed), or become ineffectual with progressive erosion of the tarsal joints, and the foot gradually drifts into severe valgus deformity (Figure 21.31).

X-rays show osteoporosis and, later, erosion of the tarsal and ankle joints. Soft-tissue swelling may be marked.

Treatment

In the stage of synovitis, splintage is helpful (to allow inflammation to subside and to prevent deformity) while waiting for systemic treatment to control the disease. Initially, tendon sheaths and joints may be injected with methylprednisolone, but this should not be repeated more than two or three times because of the risk of tendon rupture. A lightweight below-knee calliper with an inside supporting strap restores stability and may be worn almost indefinitely.

If the synovitis does not subside, operative synovectomy is advisable. Frayed tendons cannot be repaired and, although tendon replacement is technically feasible, progressive erosion of the hindfoot joints will countervail any improvement this might achieve.



(a) (b)

Figure 21.31 Rupture of tibialis posterior tendon (a) This patient with rheumatoid arthritis suddenly developed a painful valgus foot on the left. (b) The deformity was well controlled by a lightweight orthosis, and operative repair was unnecessary.

In the very late stage, arthrodesis of the ankle and tarsal joints can still restore modest function and abolish pain. The place of arthroplasty is not yet firmly established.

Seronegative arthropathies

The seronegative arthropathies are dealt with in Chapter 3. These conditions are similar to rheumatoid arthritis, but there are differences in the pattern of joint involvement, the severity of the changes and the soft-tissue features.

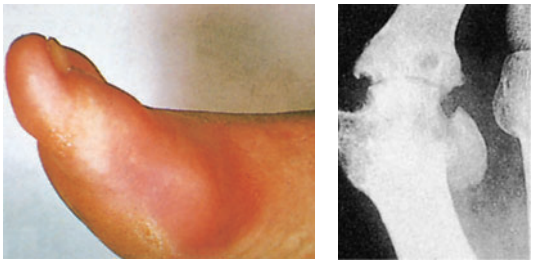
The clinical features are often asymmetrical and the ankle and hindfoot tend to be more severely affected than the forefoot. However, in psoriatic arthritis the toe joints are sometimes completely destroyed. An inflammatory reaction around the insertions of tendons and ligaments is a feature of the spondyloarthropathies. This appears in the foot as plantar fasciitis and Achilles tendinitis. Splintage and local injection of triamcinolone are helpful.

Gout (see also chapter 4)

Swelling, redness, heat and exquisite tenderness of the MTP joint of the great toe ('podagra') is the epitome of gout (Figure 21.32). The ankle joint, or one of the toes, may be similarly affected – especially following a minor injury. The condition may closely resemble septic arthritis, but the systemic features of infection are absent. The serum uric acid level may be raised.

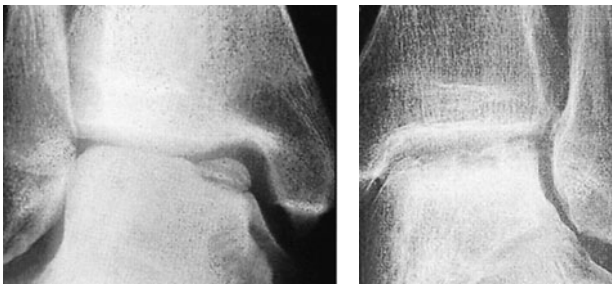
Treatment with anti-inflammatory drugs will abort the acute attack of gout; until the pain subsides the foot should be rested and protected from injury.

Chronic tophaceous gout Tophi may appear around any of the joints. The diagnosis is suggested by the characteristic X-ray features and confirmed by identifying the typical crystals in the tophus. Treatment may require local curettage of the bone lesions.



(a) (b)

Figure 21.32 Gout (a) The classical image of gout in the big toe. An inflamed 1st MTP joint. (b) X-ray showing large erosions due to tophi at the first metatarsal head.



(a) (b)

Figure 21.33 Osteochondritis dissecans
(a) Osteochondritis dissecans at the common site, the anteromedial part of the articular surface of the talus. (b) More extensive lesions can lead to secondary OA of the ankle.

Plantar fasciitis Pain under the heel due to plantar fasciitis is another manifestation of gout, though the association may be hard to prove in any particular case.

Osteochondritis dissecans of the talus

Unexplained pain and slight limitation of movement in the ankle of a young person may be due to a small osteochondral fracture of the upper surface of the talus, though the injury may have been forgotten.

X-rays taken at appropriate angles to produce tangential views of the talar surface show the small bony separation (no more than a few millimetres in diameter) at either the anteromedial or posterolateral part of the superior surface of the talus (Figure 21.33). *MRI* is also helpful and the lesion may be visualized directly by *arthroscopy*.

Treatment depends on the degree of cartilage damage. As long as the articular cartilage is intact, it is sufficient to restrict activities. Once it is softened, arthroscopic drilling may be helpful. A loose fragment may need to be removed, but often the symptoms are insufficient to warrant intervention.

Atraumatic osteonecrosis of the talus (see also Chapter 6)

Osteonecrosis of the talus is a well-recognized complication of trauma (dislocation or fracture of the neck of the talus). Atraumatic osteonecrosis, though less common than its counterpart in the femoral head, is associated with the same group of systemic disorders as the latter (hypercortisonism, alcoholism, systemic lupus erythematosus, Gaucher's disease, sickle-cell disease, etc.) and is often one of multiple sites affected.

Patients complain of pain, which is often aggravated by weight bearing, and gradually increasing restriction of movement.

X-rays and MRI show the typical features of osteonecrosis, almost always involving the posterolateral part of the talar dome. Lesions can be staged according to Ficat's radiographic classification (see Chapter 6). For purposes of treatment, it is important to distinguish between 'pre-collapse' and 'collapse' of the talar dome.

Treatment

Conservative treatment is sometimes effective; the ankle is more forgiving than the hip and patients may cope for some years on simple analgesics and restricted weight bearing. If symptoms persist and interfere significantly with function, *operative treatment* may be needed. During the pre-collapse phase, core decompression is worth trying as a first approach. If this fails, ankle arthrodesis is indicated.

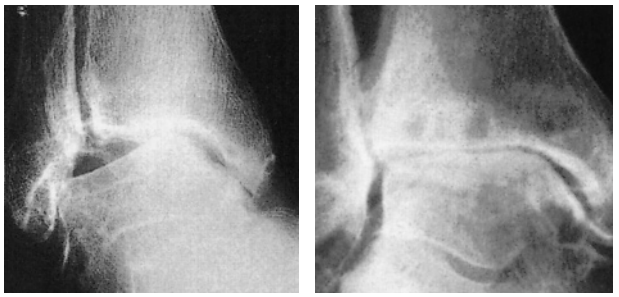
Ankle osteoarthritis (see also Chapter 5)

OA of the ankle is less common than OA of the hip or knee; when it does occur, it is almost always secondary to some underlying disorder: a malunited fracture, recurrent instability, osteochondritis dissecans of the talus, avascular necrosis of the talus or repeated bleeding with haemophilia. Sometimes, however, the ankle is involved in generalized OA and crystal arthropathy (see Chapter 4).

Clinical features

The presentation is usually with pain and stiffness localized to the ankle, particularly noticed at 'start-up', when first standing up from rest. Patients often indicate the site of pain as being transversely across the front of the ankle. The ankle is usually swollen, with palpable anterior osteophytes and tenderness along the anterior joint line. Dorsiflexion (extension) and plantarflexion at the ankle are often restricted. If heel inversion and eversion movements are restricted, suspect subtalar joint involvement. Gait is often antalgic, offloading the affected leg; the foot is often turned outwards as the patient walks through on the affected ankle, to compensate for the loss of ankle movement.

X-rays show the typical features of OA; the predisposing disorder is almost always easily detected (Figure 21.34).



(a) (b)

Figure 21.34 OA (a) The obvious malalignment that followed an old injury has led to OA. (b) In this ankle the narrowed joint space and subarticular cysts are characteristic of OA; the cause is not clear, though it may have been trauma.

Treatment

NON-OPERATIVE TREATMENT

When the condition flares up, minor, generally non-intrusive symptoms can be managed with analgesia or NSAIDs. Relative rest of the joint might be achieved with the use of a walking stick; weight loss might be appropriate. Activity such as walking, cycling and swimming can be encouraged.

Physiotherapy can be helpful in improving the range of movement, correcting gait and ensuring correct use of walking aids. An ankle support or brace may help.

OPERATIVE TREATMENT

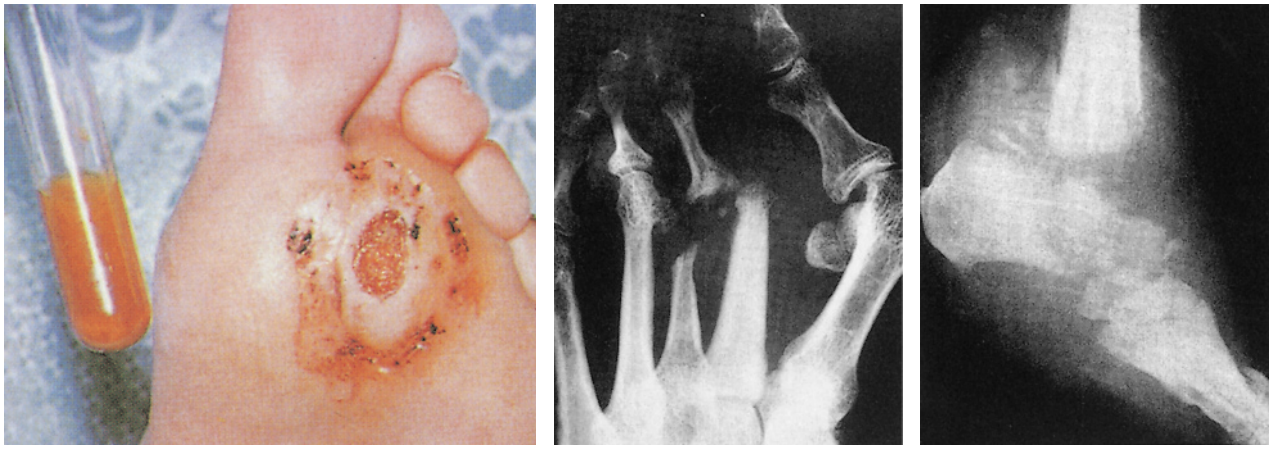
Ankle arthritis that is interfering with the activities of daily living and limiting work, social or domestic function warrants consideration for operative treatment. Depending on the severity of the condition, ankle surgery such as arthroscopic or open removal of anterior osteophytes (cheilectomy) might be offered, and consideration may be given to ankle arthrodesis; the ideal position for fusion is at zero in the sagittal plane (the foot therefore plantigrade) and 5 degrees of valgus. Total ankle arthroplasty is not as well established as hip and knee arthroplasty, but encouraging results are being reported.

DIABETIC FOOT

The complications of long-standing diabetes mellitus often appear in the foot, causing chronic disability. More than 30% of patients attending diabetic clinics have evidence of peripheral neuropathy or vascular disease and about 50% of lower-extremity amputations in England are in people with diabetes; only about one-third of these amputees go on to regain the level of ambulation that they had pre-amputation.

Factors affecting the foot in diabetes are interrelated and include: (1) a predisposition to peripheral vascular disease; (2) damage to peripheral nerves; (3) reduced resistance to infection; and (4) osteoporosis. These problems are more common in those who have been diabetic for a decade or more, and those in whom glycaemic control is poor.

Peripheral vascular disease Atherosclerosis affects mainly the medium-sized vessels below the knee. The patient may complain of claudication or ischaemic changes and ulceration in the foot (Figure 21.35). The skin feels smooth and cold, the nails show trophic changes and the pulses are weak or absent. Doppler studies should corroborate the clinical findings. Superficial ulceration occurs on the toes, with deep ulceration typically under the heel; unlike neuropathic ulcers, these are painful and tender. Digital vessel occlusion may cause dry gangrene of one or more toes; proximal vascular



(a) (b) (c)

Figure 21.35 The diabetic foot (a) Ulceration in a patient with poorly controlled diabetes. (b,c) Despite the severe changes in these two patients with diabetic neuropathy, the feet were relatively painless.

occlusion is less common but more serious, sometimes resulting in extensive wet gangrene.

Peripheral neuropathy Early on, patients are usually unaware of the abnormality, but clinical tests will discover loss of vibration and joint position sense and diminished temperature discrimination in the feet. Symptoms, when they occur, are mainly due to sensory impairment: symmetrical numbness and paraesthesia, dryness and blistering of the skin, superficial burns and skin cracks or ulceration due to shoe scuffing or localized pressure. Motor loss usually manifests as claw toes with high arches and this, in turn, may predispose to plantar ulceration.

Neuropathic joint disease 'Charcot joints' occur in less than 1% of diabetic patients, yet diabetes is the commonest cause of a neuropathic joint in Europe and America (leprosy and tertiary syphilis being the other common causes worldwide). The midtarsal joints are the most commonly affected, followed by the MTP and ankle joints. There is usually a provocative incident, such as a twisting injury or a fracture, following which the joint collapses relatively painlessly. Diagnosis is made principally on clinical grounds, supported by the radiological findings.

X-rays show marked and fairly rapid destruction of the articular surfaces. These changes are easily mistaken for infection but the simultaneous involvement of several small joints and the lack of systemic signs point to a neuropathic disorder. Joint aspiration and microbiological investigation will also help to exclude infection.

In late cases there may be severe deformity and loss of function. A rocker-bottom deformity from collapse of the midfoot is diagnostic and disastrous. The foot is further at risk once there is collapse and the development of bony prominences that become weight-bearing, increasing the risk of ulceration and secondary infection.

Osteoporosis There is a generalized loss of bone density in diabetes. In the foot the changes may be severe enough to result in insufficiency fractures around the ankle or in the metatarsals.

Infection Diabetes, if not controlled, is known to have a deleterious effect on white cell function. This, combined with local ischaemia, insensitivity to skin injury and localized pressure due to deformity, makes sepsis an ever-recurring hazard. It is unusual for infection to occur unless there is a breach in the skin, usually an ulcer.

Management

The orthopaedic surgeon will usually be one member of a multidisciplinary team comprising a physician (or endocrinologist), surgeon, chiropodist and orthotist. The best way to prevent complications is to insist on regular attendance at a diabetic clinic, full compliance with medication, examination for early signs of vascular or neurological abnormality, advice on foot care and footwear, and a high level of skin hygiene.

Examination in the specialist diabetic foot clinic for early signs of neuropathy should include the use of Semmes-Weinstein hairs – the 10 g monofilament test for skin sensitivity. A biothesiometer may be used for testing vibration sense. Peripheral vascular examination is enhanced by using a Doppler ultrasound probe. Ulcers must be swabbed for infecting organisms; frequently, multiple bacterial types are isolated (anaerobes make a regular appearance). More importantly, it is essential to obtain tissue samples from deep within the area of sepsis when treating diabetic foot infections. This is particularly important when there is bony involvement, in osteomyelitis.

X-ray examination may reveal periosteal reactions, osteoporosis, cortical defects near the articular margins and osteolysis – often collectively described as 'diabetic osteopathy'. Great care is needed with nail trimming; skin cracks should be kept clean and covered and ulcers should be treated with local dressings and antibiotics if necessary. Occasionally, septicaemia calls for admission to hospital and treatment with intravenous antibiotics. Ischaemic changes need the attention of a vascular surgeon who can advise on ways to improve the local blood supply. Arteriography may show that bypass surgery is feasible. Dry gangrene of the toe can be allowed to demarcate before local amputation; severe occlusive disease with wet gangrene may call for immediate amputation.

Indolent neuropathic ulcers require patient dressing and, if infected, antibiotic treatment. Total contact casts may avoid the need for prolonged inpatient stays or bed rest. If a bony 'high spot' is identified, it should be trimmed or excised. Custom-made shoes with total contact insoles must follow the successful healing of these ulcers to avoid recurrence.

Insufficiency fractures should be treated, if possible, without immobilizing the limb; or, if a cast is essential, it should be retained for the shortest possible period.

Neuropathic joint disease is a major challenge. Arthrodesis is fraught with difficulty, not least a very poor union rate, and sometimes is simply not feasible. 'Containment' of the problem in a weight-relieving orthosis may be the best option.

Orthopaedic surgery has a potential role to play in stabilizing the collapsing foot as well as removing bony prominences that would leave the foot vulnerable to localized pressure overload and ulceration, and hence infection.

Bone or joint infection is an ever-present risk and should be borne in mind in the differential diagnosis of insufficiency fractures and neuropathic joint erosion, particularly in diabetics.

DISORDERS OF THE TENDO ACHILLIS

ACHILLES TENDINITIS

Athletes, joggers and hikers often develop pain and swelling around the tendo Achillis, due to local irritation of the tendon sheath or the paratenon.

Pathology

The condition usually affects the 'watershed' area about 4 cm above the insertion of the tendon, an area where the blood supply to the tendon is poorer than elsewhere. The tendon sheath or the flimsy tissue around it may become inflamed. In a minority of cases the changes appear at the tendon insertion, or there may be inflammation of the retrocalcaneal bursa just above the calcaneum and deep to the tendon; anatomical deformity of the posterior part of the calcaneum may contribute to the pathogenesis.

Clinical features

The condition may come on gradually, or rapidly following a change in sporting activity (or a change of sports footwear). Less commonly there is a history of direct trauma to the Achilles tendon. The area above the heel may look inflamed and function is inhibited because of pain in the heel-cord, especially at push-off. The tendon feels thickened in the watershed area about 4 cm above its insertion. In chronic cases an ultrasound scan may be helpful in confirming the diagnosis and excluding split tears or cysts within the tendon and impending rupture. If the onset is very sudden, suspect tendon rupture (see below).

Treatment

If the condition starts acutely, it will often settle within about 6 weeks if treated appropriately. Referral for early physiotherapy is important. In the interim, advice on rest, ice, compression and elevation (RICE) and the use of an NSAID (oral or topical) are helpful.

When the symptoms improve, stretching exercises, followed by a muscle-strengthening programme, should be advised. The use of a removable in-shoe heel-raise might be helpful. If there is a planovalgus hindfoot, correction with orthotics will often bring about improvement and reduce the risk of recurrence. When the onset is insidious and treatment is started late, symptoms will be prolonged and may last for 9 months or longer.

Operative treatment is seldom necessary but, if symptoms fail to settle with physiotherapy, surgery may be appropriate – even more so if there is suspicion of an acute (or missed) tendon rupture. This will involve some type of 'decompressive' operation. Treatments such as radiofrequency coblation or extracorporeal shockwave lithotripsy are now showing some promise.

Potential pitfalls

Injection with corticosteroids should be avoided. Tendon rupture is a real risk and could well give rise to litigation.

Do not diagnose 'partial rupture' of the Achilles tendon; this should be entertained only if there is clearly some discontinuity of the tendon on ultrasound scan.

ACHILLES TENDON RUPTURE

A ripping or popping sensation is felt, and often heard, at the back of the heel. This most commonly occurs in sports requiring an explosive push-off: squash, badminton, football, tennis, netball. The patient will often report having looked round to see who had hit them over the back of the heel, the pain and collapse are so sudden.

The typical site for rupture is at the vascular watershed about 5 cm above the tendon insertion onto the calcaneum (Figure 21.36). The condition is often

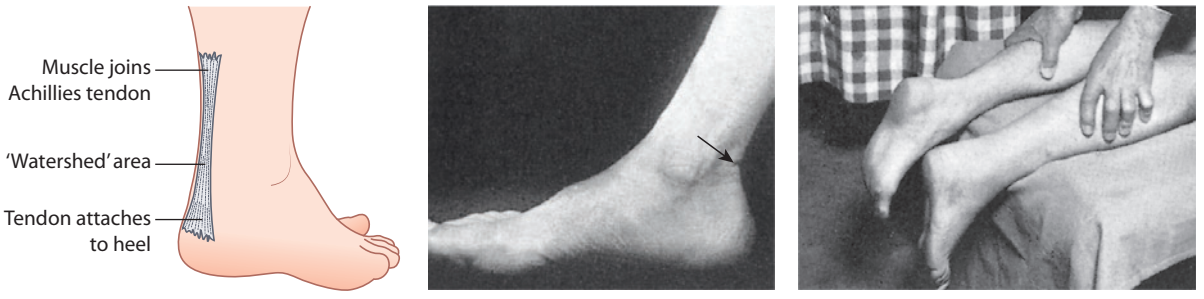


Figure 21.36 Tendo Achillis (a) The soleus may tear at its musculotendinous junction, but the tendo Achillis itself ruptures about 5 cm above its insertion, the so-called 'watershed area'. (b) The depression seen in this picture at the site of rupture (indicated by the arrow) later fills with blood. (c) Simmonds' test: both calves are being squeezed but only the left foot plantarflexes – the right tendon is ruptured.

associated with poor muscle strength and flexibility, failure to warm up and stretch before sport, previous injury or tendinitis and ill-advised corticosteroid injection.

Examination

Plantarflexion of the foot is usually inhibited and weak (although it may be possible, as the long flexors of the toes are also ankle flexors). There is often a palpable gap at the site of rupture; bruising comes out a day or two later. The calf squeeze test (Thompson's or Simmond's test) is diagnostic of Achilles tendon rupture: normally, with the patient prone, if the calf is squeezed, the foot will plantarflex involuntarily; if the tendon is ruptured, the foot remains still. Clinical assessment is often sufficient.

Ultrasound scans must be used to confirm or refute the diagnosis.

Differential diagnosis

Incomplete tear A complete rupture is often mistaken for a partial tear (which is rare). The mistake arises because, if a complete rupture is not seen within 24 hours, the gap is difficult to feel; moreover, the patient may by then be able to stand on tiptoe (just), by using his or her long toe flexors.

Tear of soleus muscle A tear at the musculotendinous junction causes pain and tenderness halfway up the calf. This recovers with the aid of physiotherapy and raising the heel of the shoe.

Treatment

If the patient is seen early, the ends of the tendon may approximate when the foot is passively plantarflexed. If so, a plaster cast or special boot is applied with the foot in equinus; rehabilitation and physiotherapy regimes vary, but it is probably safe, and may be better for eventual tendon strength, to commence physiotherapy within 4–6 weeks. A shoe with a raised heel should be worn for a further 6–8 weeks. The 're-rupture rate' has classically been quoted as about 10%, but early intervention with active rehabilitation seems to have significantly reduced this risk.

Operative repair is associated with an earlier return to function, better tendon and calf muscle strength and a lower re-rupture rate, at least when compared to traditional immobilization techniques. Supported rehabilitation and physiotherapy are commenced early (within a week or two of repair). There are, however, risks associated with operative tendon repair, including wound healing problems and sural nerve neuroma. For ruptures that present late, reconstruction using local tendon substitutes (e.g. flexor hallucis longus tendon) or strips of fascia lata is still possible.

PARALYZED FOOT

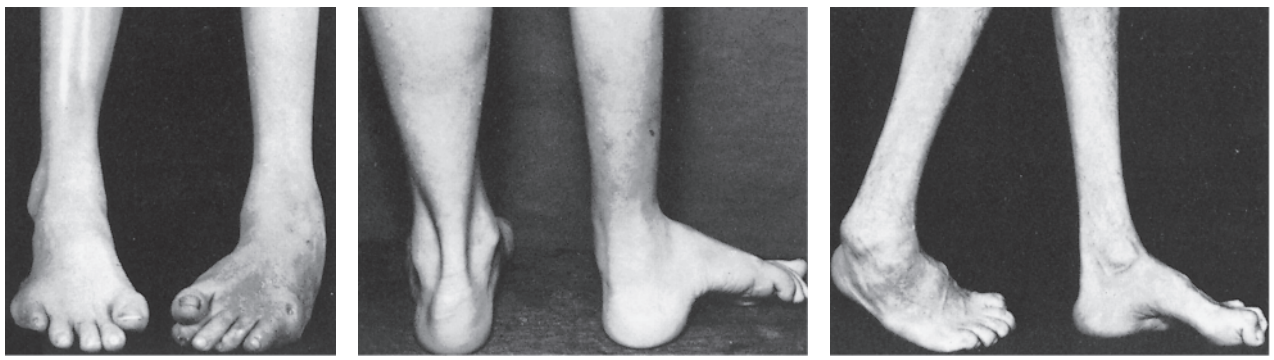
Weakness or paralysis of the foot may be symptomless, or may present in one of three characteristic ways. The patient may: (1) complain of difficulty in walking; (2) 'catch his toe' on climbing stairs (due to weak dorsiflexion); or (3) stumble and fall (due to instability).

Clinical features

See Figure 21.37.

Upper motor neuron lesions Spastic paralysis may occur in children with cerebral palsy or in adults following a stroke. Muscle imbalance usually leads to equinus or equinovarus deformity. The reflexes are brisk but sensation is normal. The entire limb is usually abnormal; sometimes both lower limbs are affected.

Lower motor neuron lesions Poliomyelitis was (and in some parts of the world still is) a common cause of foot paralysis. If all muscle groups are affected, the foot is flail and dangles from the ankle; if knee



(a) (b) (c)

Figure 21.37 The paralyzed foot (a) In spina bifida – the small ulcer is an indication of insensitive skin. (b) Poliomyelitis and (c) peroneal muscular atrophy, in both of which sensation is normal.

extension is also weak, the patient cannot walk without a calliper. With unbalanced weakness, the foot develops fixed deformity; it may also be smaller and colder than normal, but sensation is normal. Other lower motor neuron disorders such as spinal cord tumours, peroneal muscular atrophy and severe nerve root compression are rare causes of foot weakness or deformity.

Peripheral nerve injuries The sciatic, lateral popliteal or peroneal nerve may be affected. The commonest abnormalities are drop foot and weakness of peroneal action. Postoperative or postimmobilization drop foot may be due to pressure on the lateral popiteal or on the peroneal nerve as the leg rolls into external rotation. In addition to motor weakness there is an area of sensory loss. Unless the nerve is divided, recovery is possible but may take many months.

NOTE: 'Peroneal nerve lesion' is sometimes diagnosed after a hip operation. Beware! This is more often due to injury of the peroneal portion of the *sciatic nerve*.

Treatment

The weakness may need no treatment at all, or only a drop-foot splint.

Drop foot due to nerve palsy can be treated by transferring the tibialis posterior through the interosseous membrane to the midtarsal region. Spastic paralysis is treated by tendon release and transfer, but great care is needed to prevent overaction in the new direction. Thus, a spastic equinovarus deformity may be converted to a severe valgus deformity by transferring the tibialis anterior to the lateral side; this is avoided if only half the tendon is transferred.

Fixed deformities must be corrected first, before doing tendon transfers. If no adequate tendon is available to permit dynamic correction, the joint may be reshaped and arthrodesed; at the same time muscle rebalancing (even of weak muscles) is necessary, otherwise the deformity will recur.

PAINFUL ANKLE

Except after trauma or in rheumatoid arthritis, persistent pain around the ankle usually originates in one of the periarticular structures or the talus rather than in the joint itself. Conditions to look for are chronic ligamentous instability, tenosynovitis of the tibialis posterior or peroneal tendons, rupture of the tibialis posterior tendon, osteochondritis dissecans of the dome of the talus or avascular necrosis of the talus.

Tenosynovitis Tenderness and swelling are localized to the affected tendon, and pain is aggravated by active movement – inversion or eversion against resistance. Local injection of corticosteroid usually helps.

Rupture of tibialis posterior tendon Pain starts quite suddenly and sometimes the patient gives a history of having felt the tendon snap. The heel is in valgus during weight bearing; the area around the medial malleolus is tender and active inversion of the ankle is both painful and weak. In physically active patients, operative repair or tendon transfer using the tendon of flexor digitorum longus is worthwhile. For poorly mobile patients, or indeed anyone who is prepared to put up with the inconvenience of an orthosis, splintage may be adequate (see Figure 21.31).

Osteochondritis dissecans of the talus Unexplained pain and slight limitation of movement in the ankle of a young person may be due to a small osteochondral fracture of the dome of the talus. Tangential X-rays will usually show the tiny fragment. MRI is also helpful and the lesion may be visualized directly by arthroscopy. If the articular surface is intact, it is sufficient to simply restrict activities. If the fragment has separated, it may have to be removed.

Avascular necrosis of the talus The talus is one of the preferred sites of 'idiopathic' necrosis. The causes are the same as for necrosis at other more common sites such as the femoral head. If pain is marked, arthrodesis of the ankle may be needed.

PAINFUL FEET

'My feet are killing me!' This complaint is common but the cause is often elusive. Pain may be due to: (1) mechanical pressure (which is more likely if the foot is deformed or the patient obese); (2) joint inflammation or stiffness; (3) a localized bone lesion; (4) peripheral ischaemia; (5) muscular strain – usually secondary to some other abnormality. Remember, too, that local disorders may be part of a generalized disease (e.g. diabetes or rheumatoid arthritis), so examination of the entire patient may be indicated. Specific foot disorders that cause pain are considered later.

Posterior heel pain

Two common causes of heel pain are *traction 'apophysitis'* and *calcaneal bursitis* (Figure 21.38).

Traction 'apophysitis' (Sever's disease) This condition usually occurs in boys aged about 10 years.

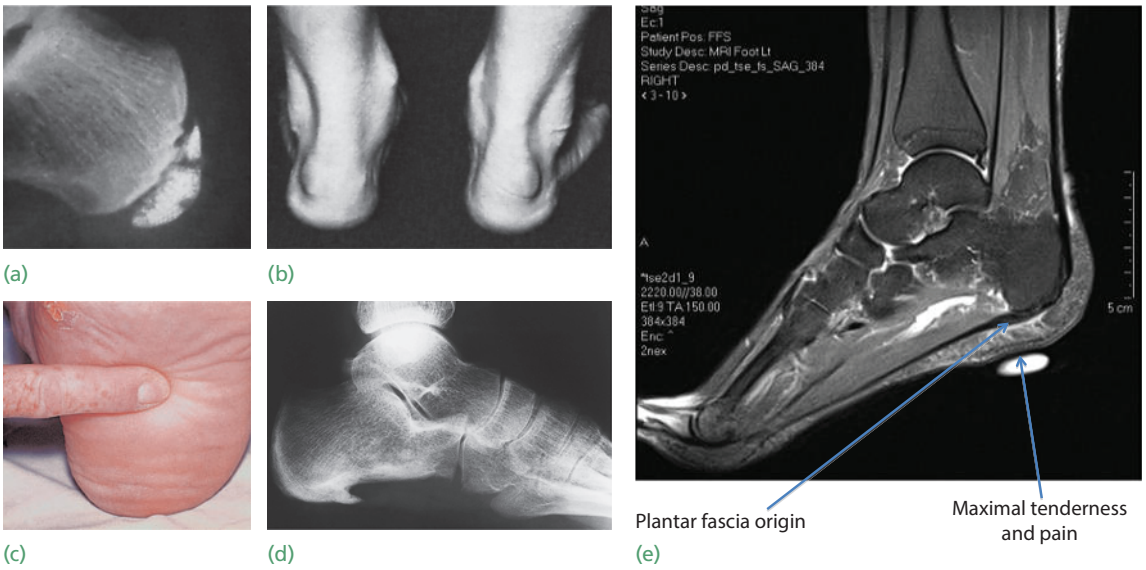


Figure 21.38 Painful heel (a) Sever's disease – the apophysis is dense and fragmented. (b) Bilateral 'heel bumps'. (c) The usual site of tenderness in plantar fasciitis. (d) X-rays in patients with plantar fasciitis often show what looks like a spur on the undersurface of the calcaneum. In reality this is a two-dimensional view of a small ridge corresponding to the origin of flexor digitorum brevis, close to the plantar fascia. It is doubtful whether the 'spur' is responsible for the pain and local tenderness in most cases. (e) MRI shows the thickened origin of the plantar fascia, just superficial to the origin of flexor digitorum brevis – the bright oil capsule placed over the heel shows the site of maximum pain and tenderness.

It is not a 'disease' but a mild traction injury. Pain and tenderness are localized to the tendo Achillis insertion. The X-ray report usually refers to increased density and fragmentation of the apophysis, but often the painless heel looks similar. The heel of the shoe should be raised a little and strenuous activities restricted for a few weeks.

Calcaneal bursitis Older girls and young women often complain of painful bumps on the backs of their heels. The posterolateral portion of the calcaneum is prominent and shoe friction causes retrocalcaneal bursitis. Symptoms are worse in cold weather and when wearing high-heeled shoes (hence the use of colloquial labels such as 'winter heels' and 'pumpbumps'). Treatment should be conservative – attention to footwear (open-back shoes are best) and padding of the heel. Operative treatment – removal of the bump or dorsal wedge osteotomy of the calcaneum – is feasible but the results are unpredictable; despite the reduction in the size of the bumps, patients often continue to experience discomfort, potentially added to by an operation scar.

Inferior heel pain

Calcaneal bone lesions Any bone disorder in the calcaneum can present as heel pain: a stress fracture, osteomyelitis, osteoid osteoma, cyst-like lesions and Paget's disease are the most likely. X-rays usually provide the diagnosis.

Plantar fasciitis

This is an annoying and painful condition that limits function. There is pain and tenderness in the sole of the foot, mostly under the heel, with standing or walking. The condition usually comes on gradually, without any clear incident or injury, but sometimes there is a history of sudden increase in sporting activity, or a change of footwear, sports shoes or running surface. There may be an associated tightness of the Achilles tendon. The pain is often worse when first getting up in the morning, with typical hobbling downstairs, or when first getting up from a period of sitting – the typical start-up pain and stiffness. The pain can at times be very sharp, or it may change to a persistent background ache as the patient walks about. The condition can take 18–36 months or longer to resolve, but it is generally self-limiting, given time.

Pathology

The plantar fascia or aponeurosis is a dense fibrous structure that originates from the calcaneum, deep to the heel fat pad, and runs distally to the ball of the foot, with slips to each toe. The plantar fascia stiffens and becomes less pliable with age. The fascia is

probably not actually inflamed in this condition, at least not beyond the first week or two of onset. There may be micro-tears in the fascia, and the fascia thickens. The term 'plantar fasciitis' is apt in some cases, as the condition is sometimes associated with inflammatory disorders such as gout, ankylosing spondylitis and Reiter's disease, in which enthesopathy is one of the defining pathological lesions.

Clinical features

There is localized tenderness, usually at the medial aspect beneath the heel and sometimes in the midfoot. This is essentially a clinical diagnosis. If there are features suggesting an inflammatory disease (seronegative arthropathy), blood tests may be indicated. An ultrasound scan shows the thickening, and sometimes the Doppler test shows increased local blood flow and neovascularization, but this investigation is not indicated in every case.

Imaging

A plain lateral *X-ray* can help to exclude a stress fracture, and will often show what looks like a bony spur on the undersurface of the calcaneum. The 'spur' is, in fact, a bony ridge that looks sharp and localized in the two-dimensional X-ray image; it is an associated, not a causative, feature in plantar fasciitis. Patients, and sometimes doctors, can become fixated on the idea of a spur of bone causing the symptoms by digging into the plantar fascia and cannot conceive of how the condition could possibly resolve while the spur remains – but it can and does get better.

MRI can be helpful in excluding a calcaneal stress fracture, which is an important differential diagnosis.

Treatment

NON-OPERATIVE TREATMENT

Relative rest and NSAIDs, orally or topically, can be helpful in settling. An analysis of causative factors (footwear, sports and exercise factors) can help the patient to overcome the condition. There is an important role for the patient in managing the condition, with stretching exercises and massage; self-help advice sheets are available.

Patients might expect (or dread!) an injection into the plantar fascia, and they are right to be apprehensive. There is no convincing research to support this, and there is evidence to show that it can lead to rupture of the plantar fascia (which will often immediately ease the symptoms, but leads to a painful flat foot and impairs sporting function).

A physiotherapist can help to educate the patient about the condition and its likely progress, and can emphasize the need for a regular stretching regime for 8–12 weeks, supplemented with local massage (e.g. with a foot roller, golf ball or frozen water bottle). Local manual treatments from the physiotherapist can help, as can the use of taping and a cushioned heel pad.

Night splints have been tried, to keep the foot up in a plantigrade position overnight, preventing stiffening in the Achilles tendon and plantar fascia; there is logic in this but no clear evidence for its efficacy, and trials have been hampered by poor compliance. Podiatric assessment of the hindfoot biomechanics may identify predisposing factors such as planovalgus hindfoot alignment, which can be corrected with orthotics.

OPERATIVE TREATMENT

Patients may lose heart and demand that something be done. However, there is no reliable surgical procedure for this condition. Limited fasciotomy to release part of the plantar fascia can help in some cases, but there is a significant risk of complications including worsening of the condition. Where the driving force behind the condition is tightness of the gastrocnemius muscle, a surgical recession of that muscle can be of benefit. Promising new interventions include shockwave lithotripsy and localized radiofrequency (coblation) therapy, but these have yet to be fully tested in rigorous and large-scale studies.

Potential pitfalls

It is important not to miss a manifestation of a systemic disease such as an inflammatory arthropathy (often seronegative), a peripheral neuropathy (usually diabetic) or a stress fracture.

If a corticosteroid steroid injection is used, it should be done cautiously with a small dose into a limited area, and after appropriate warnings to the patient. Excising a 'spur' is usually a vain endeavour.

Differential diagnosis

Painful fat pad Chronic pain and tenderness directly over the fat pad under the heel sometimes follows a direct blow to the area (e.g. in a fall from a height). The condition is also seen in athletes and has been attributed variously to separation of the fat pad from the bone, loss of its normal shock-absorbing effect and atrophy. Non-specific 'inflammation' has also been blamed. Treatment is palliative: wearing soft-soled shoes or shock-absorbing heel cups, foot baths and anti-inflammatory agents.

Nerve entrapment Entrapment of the first branch of the lateral plantar nerve has been reported as a cause of heel pain. The commonest complaint is pain after sporting activities. Characteristically, tenderness is maximal on the medial aspect of the heel, where the

small nerve branch is compressed between the deep fascia of abductor hallucis and the edge of the quadratus plantae muscle. Diagnosis is not easy, because the symptoms and signs may mimic those of plantar fasciitis.

Treatment, in the first instance, is conservative: a long trial (6–8 months) of shock-absorbing orthoses, foot baths, anti-inflammatory preparations and one or two corticosteriod injections. Only if these measures fail to give relief should surgical decompression of the nerve be considered.

Pain over the midfoot

In children, pain in the midtarsal region is rare: one cause is *Kohler's disease* (osteochondritis of the navicular) (Figure 21.39a). The bony nucleus of the navicular becomes dense and fragmented. The child, under the age of 5, has a painful limp and a tender, warm thickening over the navicular. Usually no treatment is needed as the condition resolves spontaneously. If symptoms are severe, a short period in a below-knee plaster helps. A comparable condition occasionally affects middle-aged women (*Brailsford's disease*); the navicular becomes dense, then altered in shape, and later the midtarsal joint may degenerate.

In adults, especially if the arch is high, a ridge of bone sometimes develops on the adjacent dorsal surfaces of the medial cuneiform and the first metatarsal (the '*overbone*'). A lump can be seen, which feels bony and may become bigger and tender if the shoe presses on it (Figure 21.39c,d). If shoe adjustment fails to provide relief, the lump may be bevelled off.

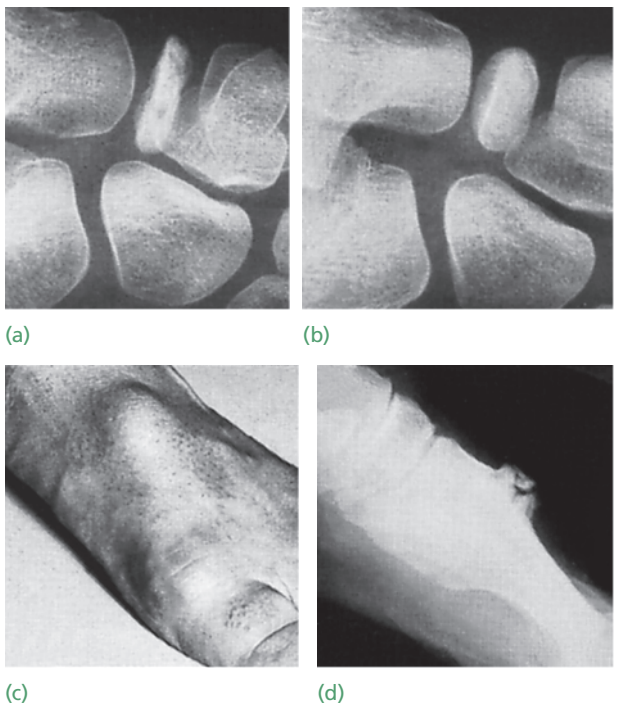


Figure 21.39 Pain over the midfoot (a) Köhler's disease compared with (b) the normal foot. (c,d) The bump on the dorsum of the foot due to OA of the first cuneiform metatarsal joint.

Generalized pain in the forefoot

Metatarsalgia Generalized ache in the forefoot is a common expression of foot strain, which may be due to a variety of conditions that give rise to faulty weight distribution (e.g. flattening of the metatarsal arch, or undue shortening of the first metatarsal), or merely the result of prolonged or unaccustomed walking, marching, climbing or standing. These conditions have this in common: they give rise to a mismatch between the loads applied to the foot, the structure on which those loads are acting, and the muscular effort required to maintain the structure so that it can support those loads.

Aching is felt across the forefoot and the anterior metatarsal arch may have flattened out. There may even be callosities under the metatarsal heads.

Treatment involves: (1) dealing with the mechanical disorder (correcting a deformity if it is correctable, supplying an orthosis that will redistribute the load, fitting a shoe that will accommodate the foot); and (2) performing regular muscle strengthening exercises, especially for the intrinsic muscles that maintain the anterior (metatarsal) arch of the foot. A good 'do-it-yourself' exercise is for the patient to stand barefoot on the floor, feet together, and then drag their body forwards by repeatedly crimping the toes to produce traction upon the floor. Ten minutes a day should suffice.

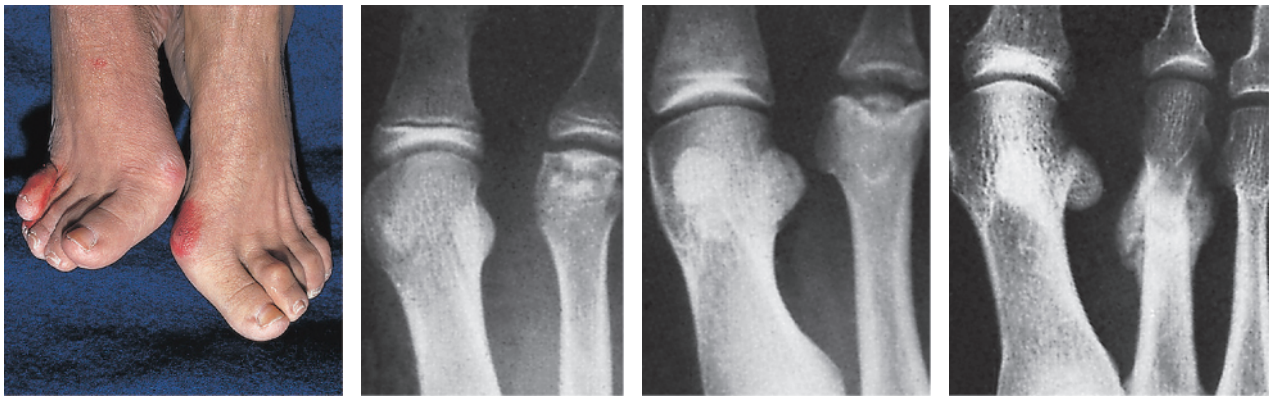
Pain in the metatarsophalangeal joints Inflammatory arthritis (e.g. rheumatoid disease) may start in the foot with synovitis of the MTP joints. Pain in these cases is associated with swelling and tenderness of the forefoot joints and the features are almost always bilateral and symmetrical.

Localized pain in the forefoot

Whereas metatarsalgia involves the entire forefoot, localized pain and tenderness is related to a specific anatomical site in the forefoot and could be due to a variety of bone or soft-tissue disorders: 'sesamoiditis', osteochondritis of a metatarsal head (Freiberg's disease), a metatarsal stress fracture or digital nerve entrapment (Morton's disease).

Sesamoiditis

Pain and tenderness directly under the first metatarsal head, typically aggravated by walking or



(a) (b) (c) (d)

Figure 21.40 Pain in the forefoot (a) Long-standing deformities such as dropped anterior arches, hallux valgus, hammer toe, curly toes and overlapping toes (all of which are present in this patient) can cause metatarsalgia. Localized pain and tenderness suggest a more specific cause. (b,c) Stages in the development of Freiberg's disease. (d) Periosteal new-bone formation along the shaft of the second metatarsal, the classic sign of a healing stress fracture. The radiological signs usually lag behind the onset of symptoms by 2–3 weeks.

passive dorsiflexion of the great toe, may be due to sesamoiditis. This term is a misnomer: symptoms usually arise from irritation or inflammation of the peritendinous tissues around the sesamoids – more often the medial (tibial) sesamoid, which is subjected to most stress during weight-bearing on the ball of the foot.

Acute sesamoiditis may be initiated by direct trauma (e.g. jumping from a height) or unaccustomed stress (e.g. in new athletes and dancers).

Chronic sesamoid pain and *tenderness* should signal the possibility of sesamoid displacement, local infection (particularly in a diabetic patient) or avascular necrosis.

Sesamoid chondromalacia is a term coined by Apley to explain changes such as fragmentation and cartilage fibrillation of the medial sesamoid. X-rays in these cases may show a bipartite or multipartite medial sesamoid, which is often mistaken for a fracture.

Treatment In the usual case, treatment consists of reduced weight-bearing and a pressure pad in the shoe. In resistant cases, a local injection of methylprednisolone and local anaesthetic often helps; otherwise the sesamoid should be shaved down or removed, taking great care not to completely interrupt the flexor halluces brevis tendon, in which the sesamoid is contained.

Freiberg's disease (osteochondritis; osteochondrosis)

Osteochondritis (or osteochondrosis) of a metatarsal head is probably a type of traumatic osteonecrosis of the subarticular bone in a bulbous epiphysis (akin to osteochondritis dissecans of the knee). It usually affects the second metatarsal head (rarely the third) in young adults, mostly women.

The patient complains of pain at the MTP joint. A bony lump (the enlarged head) is palpable and tender and the MTP joint is irritable. X-rays show the head to be flattened and wide, the neck thick and the joint space apparently increased (Figure 21.40).

Treatment If discomfort is marked, a walking plaster or moulded sandal will help to reduce pressure on the metatarsal head. If pain and stiffness persist, operative synovectomy, debridement and trimming of the metatarsal head should be considered. Pain relief is usually good and the range of dorsiflexion is improved.

Stress fracture

Stress fracture, usually of the second or third metatarsal, occurs in young adults after unaccustomed activity or in women with postmenopausal osteoporosis. The dorsum of the foot may be slightly oedematous and the affected shaft feels thick and tender. The X-ray appearance is at first normal but later shows fusiform callus around a fine transverse fracture. Long before X-ray signs appear, a radioisotope scan will show increased activity.

Treatment Treatment is either unnecessary or consists simply of rest and reassurance.

Interdigital nerve compression (Morton's metatarsalgia)

Morton's metatarsalgia is a common problem, with neuralgia affecting a single distal metatarsal interspace, usually the third (affecting the third and fourth toes), sometimes the second (affecting the second and third toes), rarely others. The patient typically complains of pain on walking, with the sensation of walking on a pebble in the shoe, or of the sock being rucked-up under the ball of the foot. The pain is worse in tight footwear and often has to be relieved by removing the footwear

and massaging the foot. Activities that load the forefoot (running, jumping, dancing) exacerbate the condition, which often consists of severe forefoot pain and then a reluctance to weight-bear. In Morton's metatarsalgia the pain is typically reproduced by laterally compressing the forefoot while also compressing the affected interspace – this produces the pathognomic Mulder's click as the 'neuroma' displaces between the metatarsal heads.

This is essentially an entrapment or compression syndrome affecting one of the digital nerves, but secondary thickening of the nerve creates the impression of a 'neuroma'. The lesion, and an associated bursa, occupy a restricted space between the distal metatarsals, and are pinched, especially if footwear also laterally compresses the available space.

Treatment A step-wise treatment programme is advisable. Simple offloading of the metatarsal heads by using a metatarsal dome insole and wider fitting shoes may help. If symptoms do not improve with these measures, a steroid injection into the interspace will bring about lasting relief in about 50% of cases. Surgical intervention is often successful; the nerve should be released by dividing the tight transverse intermetatarsal ligament; this can be done through either a dorsal longitudinal or a plantar incision; most surgeons will also excise the thickened portion of the nerve. This improves matters in about 80–90% of patients, although it is not uncommon for patients still to have some residual symptoms or restrictions of footwear.

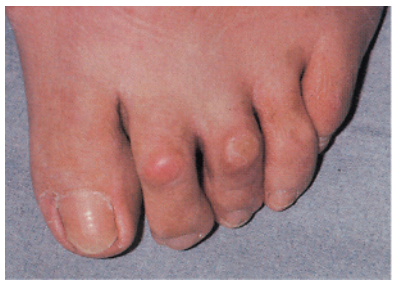
Tarsal tunnel syndrome

Pain and sensory disturbance in the medial part of the forefoot, unrelated to weight bearing, may be due to compression of the posterior tibial nerve behind and below the medial malleolus. Sometimes this is due to a space-occupying lesion such as a ganglion, haemangioma or varicosity. The pain is often worse at night and the patient may seek relief by walking around or stamping the foot. Paraesthesia and numbness may follow the characteristic sensory distribution, but these symptoms are not as well defined as in other entrapment syndromes. The diagnosis is difficult to establish but nerve conduction studies may show slowing of motor or sensory conduction.

Treatment To decompress the nerve, it is exposed behind the medial malleolus and followed into the sole; sometimes it is trapped by the belly of adductor halluces arising more proximally than usual.

SKIN DISORDERS

Painful skin lesions (Figure 21.41) are important for two reasons: (1) they demand attention in their own right; (2) postural adjustments to relieve pressure may give rise to secondary problems and metatarsalgia.



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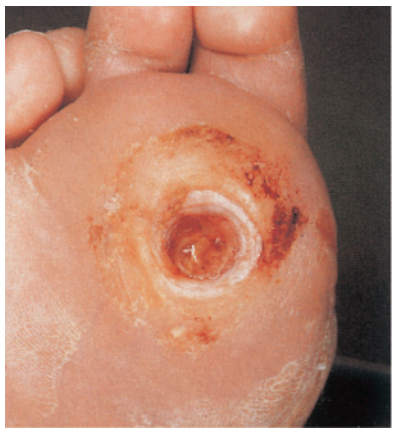

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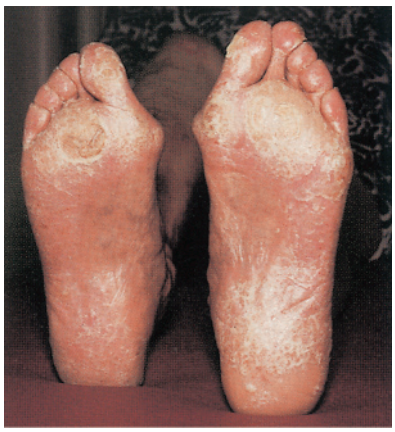

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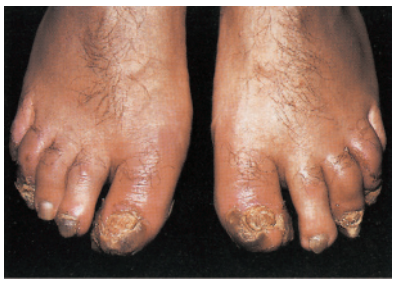


(d)

Figure 21.41 Skin lesions (a) Corns. (b) Callosities in a patient with claw toes and a 'dropped' anterior metatarsal arch. (c) A typical pressure ulcer in a patient with long-standing diabetic neuropathy. (d) Keratoderma blenorrhagica, a complication of Reiter's disease.

Corns and calluses

These are hyperkeratotic lesions that develop as a reaction to localized pressure or friction.

Corns are fairly small and situated at 'high spots' in contact with the shoe upper: the dorsal knuckle of a claw toe or hammer toe, or the tip of the toe if it impinges against the shoe. Soft corns also appear on adjacent surfaces of toes that rub against each other. Treatment consists of paring the hyperkeratotic skin, applying felt pads that will prevent shoe or toe pressure, correcting any significant deformity (if necessary by operation) and attending to footwear.

Calluses are more diffuse keratotic plaques on the soles – either under prominent metatarsal heads or under the heel. They are seen mainly in people with 'dropped' metatarsal arches and claw toes, or varus or valgus heels. Treatment is much the same as for corns: it is important to redistribute foot pressure by altering the shoes, fitting pressure-relieving orthoses and ensuring that the shoes can accommodate the malshaped feet. Surgical treatment for claw toes may be needed.

Plantar warts

Plantar warts resemble calluses but they tend to be more painful and tender, especially if squeezed. They can be distinguished from calluses by paring down the hyperkeratotic skin to expose the characteristic papillomatous 'core', which is seen to be dotted with fine blood vessels. These are viral lesions but it is usually local pressure that renders them painful.

Treatment This can be frustrating as plantar warts are difficult to eradicate. Salicylic acid plasters are applied at regular intervals, and smaller lesions may respond to cryosurgery. Surgical excision is avoided as this usually leaves a painful scar at the pressure site.

Foreign body 'granuloma'

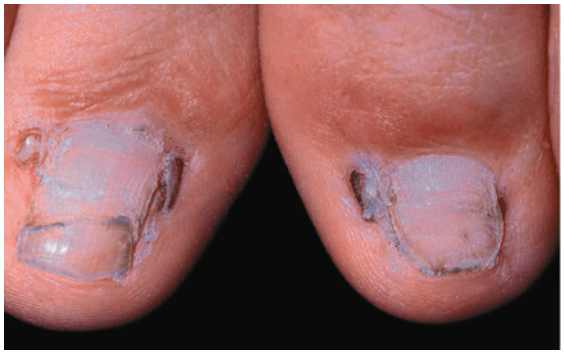
The sole is particularly at risk of penetration by small foreign bodies (usually a thorn, a splinter or a piece of glass), which may give rise to a painful lump resembling a wart or callus. This diagnosis should always be considered if the 'callosity' is situated in a non-pressure area. X-rays may help to detect the foreign body.

Treatment Remove the object; the reactive lesion heals quickly.

TOENAIL DISORDERS

The toenail of the hallux may be *ingrown*, *overgrown* or *undergrown* (Figure 21.42).

Ingrown toenails The nail burrows into the nail groove; this ulcerates and its wall grows over the nail, so the term 'embedded toenail' would be better. The patient is taught to cut the nail square, to insert pledgets of wool under the ingrowing edges and to keep the feet clean and dry at all times. If these measures fail, the portion of germinal matrix that is responsible for the 'ingrow' should be ablated, either by operative excision or by chemical ablation



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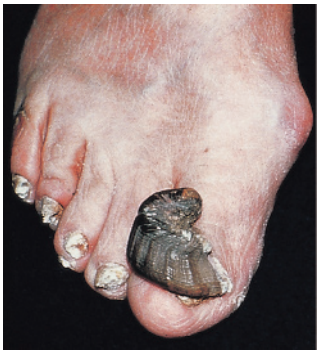

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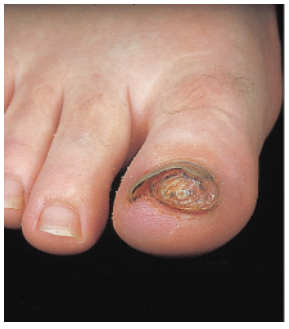

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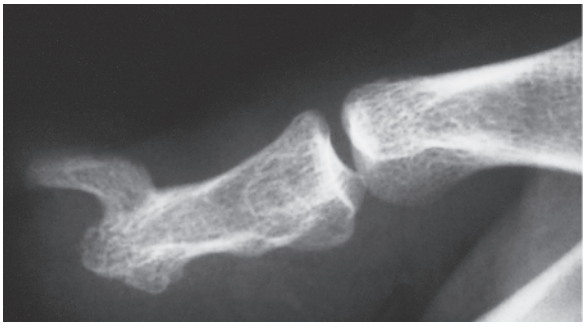

(d)

Figure 21.42 Toenail disorders (a) Ingrown toenails. (b) Overgrown toe-nail (onychogryposis). (c,d) Exostosis from the distal phalanx, pushing the toenail up.

with phenol. The phenol is applied to the exposed matrix with a cotton bud for 3 minutes and then washed off with alcohol, which neutralizes the caustic effect. Rarely is it necessary to remove the entire nail or completely ablate the nail bed.

Overgrown toenails (onychogryposis) The nail is hard, thick and curved. A chiropodist can usually make the patient comfortable, but occasionally the nail may need excision.

Undergrown toenails A subungual exostosis grows on the dorsum of the terminal phalanx and pushes the nail upwards. The exostosis should be removed if the nail deformity is causing problems, usually with footwear.

NOTES ON APPLIED ANATOMY

The ankle and foot function as an integrated unit, and together they provide stable support, proprioception, balance and mobility.

Ankle

The ankle fits together like a tenon and mortise; the tibial and fibular parts of the mortise are bound together by the inferior tibiofibular ligament, and stability is augmented by the collateral ligaments. The medial ligament fans out from the tibial malleolus to the talus, the superficial fibres forming the deltoid ligament. The lateral ligament has three thickened bands: the anterior and posterior talofibular ligaments and, between them, the calcaneofibular ligament.

Tears of these ligaments may cause tilting of the talus in its mortise. Forced abduction or adduction may disrupt the mortise altogether by (1) forcing the tibia and fibula apart (diastasis of the tibiofibular joint); (2) tearing the collateral ligaments; (3) fracturing the malleoli.

Foot

The footprint gives some idea of the arched structure of the foot. This derives from the tripodial bony framework between the calcaneum posteriorly and the first and fifth metatarsal heads. The medial arch is high, with the navicular as its keystone; the lateral arch is flatter. The anterior arch formed by the metatarsal bones thrusts maximally upon the first and fifth metatarsal heads and flattens out (spreading the foot) during weight bearing; it can be pulled up by contraction of the intrinsic muscles, which flex the MTP joints.

Movements

The ankle allows movement in the sagittal plane only (plantarflexion and dorsiflexion). Adduction and abduction (turning the toes towards or away from the midline) are produced by rotation of the entire leg below the knee; if either is forced at the ankle, the mortise fractures. Pronation and supination occur at the intertarsal and tarsometatarsal joints; the foot rotates about an axis running through the second metatarsal, the sole turning laterally (pronation) or medially (supination) – movements analogous to those of the forearm. The combination of plantarflexion, adduction and supination is called inversion; the opposite movement of dorsiflexion, abduction and pronation is eversion.

Inversion and eversion are necessary for walking on rough ground or across a slope. If the joints at which they occur are arthrodesed in childhood, a compensatory change may occur at the ankle so that it becomes a ball-and-socket joint.

Foot positions and deformities

A downward-pointing foot is said to be in equinus; the opposite is calcaneus. If only the forefoot points downwards, the term 'plantaris' is used. Supination with adduction produces a varus deformity; pronation with abduction causes pes valgus. An unusually high arch is called pes cavus. Many of these terms are used as if they were definitive diagnoses when, in fact, they are nothing more than Latin translations of descriptive anatomy.

FURTHER READING

Apley AG. Open sessamoid. *Proc R Soc Med* 1966; **59**: 120.

Banta J, Sutherland DH, Wyatt M. Anterior tibialis transfer to os calcis with Achilles tenodesis for calcaneal deformity in myelomeningocoele. *J Paediatr Orthop* 1981; **1**: 125–30.

Berndt AL, Harty M. Transchondral fractures (osteochondritis dissecans) of the talus. *J Bone Joint Surg* 1959: **41-A**: 988–1020.

Bowyer GW. The role of the orthopaedic surgeon in diabetic foot complications. In: Shearman CP (ed.) *Management of Diabetic Foot Complications*. London: Springer-Verlag, 2015, pp. 155–63.

Caroll NC. Technique of plantar fascia release and calcaneocuboid joint release in clubfoot surgery. In: Simons GW (ed.) *The Clubfoot*. New York: Springer-Verlag, 1994, pp. 246–52.

Chong A, Nazarian N, Chandrananth J, *et al.* Surgery for the correction of hallux valgus: minimum five-year results with a validated patient reported outcome tool and regression analysis. *Bone Joint J* 2015; **97-B**: 208–14.

Cholmeley JA. Elmslie's operation for the calcaneus foot. *J Bone Joint Surg* 1953; **35B**: 46–9.

Coleman WC, Brand PW, Birke JA. The total contact cast. A therapy for plantar ulceration on insensitive feet. *J Am Podiatry Med Assoc* 1984; **74**: 548–52.

Coughlin MJ, Shurnas PS. Hallux rigidus: demographics, etiology, and radiographic assessment. *Foot Ankle Int* 2003; **24**: 731–43.

Crawford A, Marxen J, Osterfeld D. The Cincinatti incision: A comprehensive approach for surgical procedures of the foot and ankle in childhood. *J Bone Joint Surg* 1982; **64A**: 1355–8.

Delanois RE, Mont MA, Yoon TR, *et al.* Atraumatic osteonecrosis of the talus. *J Bone Joint Surg* 1998; **80A**: 529–36.

Dobbs MB, Purcell DB, Nunley R, Morcuende JA. Early results of a new method of treatment for idiopathic congenital vertical talus. *J Bone Joint Surg Am* 2006; **88**: 1192–200.

Duncan RD, Fixsen JA. Congenital convex pes valgus. *J Bone Joint Surg* 1999; **81B**: 250–4.

Eidelman M, Katzman A, Bor N, *et al.* Treatment of residual clubfoot deformities with the Taylor Spatial Frame using a Ponseti sequence. Poster presented at European Paediatric Orthopaedic Society 26th Annual Meeting. Sorrento, Italy, 2007.

Evans D. Relapsed clubfoot. *J Bone Joint Surg* 1961; **43B**: 722–33.

Gage JR. *Gait Analysis in Cerebral Palsy.* New York: MacKeith Press, 1991.

Herzenberg JE, Carroll NC, Christofersen MR, *et al.* Clubfoot analysis with three-dimensional computer modeling. *J Paediatr Orthop* 1988; **8**: 257–62.

Lapidus PW. Transplantation of the extensor tendon for correction of the overlapping fifth toe. *J Bone Joint Surg* 1942; **24**: 555–9.

Morcuende JA, Dolan LA, Dietz FR, Ponseti IV. Radical reduction in the rate of extensive corrective surgery for clubfoot using the Ponseti method. *Pediatrics* 2004; **113**: 376.

Mosca VS, Bevan WR. Tarsal coalitions and the calcaneal lengthening osteotomy: the role of deformity correction. *J Bone Joint Surg Am* 2012; **94**: 1584–94.

Ogden JA. Skeletal Injury in the Child, 3rd ed. New York: Springer-Verlag, 24: 1119–20.

Piggott H. The natural history of hallux valgus in adolescence and early adult life. *J Bone Joint Surg* 1960; **42B**: 749–60.

Pirani S, Outerbridge HK, Sawatzky B, *et al.* A reliable method of clinically evaluating a virgin clubfoot evaluation. In: Proceedings of the 21st SICOT World Congress, Sydney, Australia, 1999.

Pohl M, Nicol RO. Transcuneiform and opening wedge medial cuneiform osteotomy with closing wedge cuboid osteotomy in relapsed clubfoot. *J Pediatr Orthop* 2003 Jan-Feb; **23**(1): 70–3.

Ponseti IV. Treatment of congenital club foot. *J Bone Joint Surg* 1992; **74A**: 448–54.

Ponseti IV, Becker JR. Congenital metatarsus adductus: The results of treatment. *J Bone Joint Surg Am* 1966; **48**: 702–11.

Ponseti IV, Zhivkov M, Davis N, *et al.* Treatment of the complex idiopathic clubfoot. *Clin Orthop Relat Res* 2006 Oct; 451: 171–6.

Rang M. High arches. In: Wenger DR, Rang M (eds), *The Art and Practice of Children's Orthopaedics.* New York: Raven Press, 1993, pp. 168–79.

Rao U, Joseph B. The influence of footwear on the prevalence of flat foot: A survey of 2300 children. *J Bone Joint Surg* 1992; **74-B**: 525–7.

Robinson AHN, Limbers JP. Modern concepts in the treatment of hallux valgus. *J Bone Joint Surg* 2005; **87B**: 1038–45.

Rose GK, Welton EA, Marshall T. The diagnosis of flat foot in the child. *J Bone Joint Surg* 1985; **67B**: 71–8.

Samilson RL. Proscentic osteotomy of the os calcis for calcaneocavus feet. In: Bateman JE (ed.), *Foot Science.* Philadelphia: WB Saunders, 1976, p. 18.

Shearman CP. Foot complications in diabetes: the problem. In: Shearman CP (ed.), *Management of Diabetic Foot Complications.* London: Springer-Verlag, 2015, pp. 1–7.

Thacker MM, Scher DM, Sala DA, *et al.* Use of the foot abduction orthosis following Ponseti casts: is it essential? *J Pediatr Orthop* 2005 Mar-Apr; **25**(2): 225–8.

Tindall AJ, Steinlechner CW, Lavy CB, *et al.* Results of manipulation of idiopathic clubfoot deformity in Malawi by orthopaedic clinical officers using the Ponseti method: a realistic alternative for the developing world? *J Pediatr Orthop* 2005 Sep-Oct; **25**(5): 627–9.

Turco V. Surgical correction of the resistant clubfoot. One stage posteromedial release with internal fixation; a preliminary report. *J Bone Joint Surg* 1971; **53A**: 477–97.

Wenger DR, Mauldin D, Speck G, *et al.* Corrective shoes and inserts as treatment for flexible flatfoot in infants and children. *J Bone Joint Surg Am* 1989; 71: 800–10.

Section 3

Trauma

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The management of major injuries

22

David Sutton & Max Jonas

INTRODUCTION

Aetiology of major trauma

Injury is one of the top three causes of death for people between the ages of 5 and 44 worldwide, so it has a particular impact on young people. Nearly a third of the global 5.8 million deaths from injuries are the result of violence and nearly a quarter the result of road traffic crashes (Figure 22.1). Injuries are a growing problem; injuries resulting from violence and road traffic crashes are predicted by the World Health Organization (WHO) to rise in rank when compared to other causes of death, with road traffic crash deaths expected to become the fifth leading cause of death by 2030.

In the UK, the highest rate of accidental deaths in 2010 was due to falls, replacing transport accident deaths. Deaths from trauma in all road user groups fell by 51% between 2003 and 2014 to 1775 annually, with 45% of the deaths occurring in car occupants (Figure 22.2). In terms of distance travelled, pedestrians, bicyclists and motorcyclists are particularly vulnerable road users (Figure 22.3). Road traffic deaths and serious injuries show a peak incidence in young people between the ages of 17 (age of learning to drive) and 23 (Figure 22.4).

Mortality subsequent to major trauma is dependent on a number of factors, of which the economic

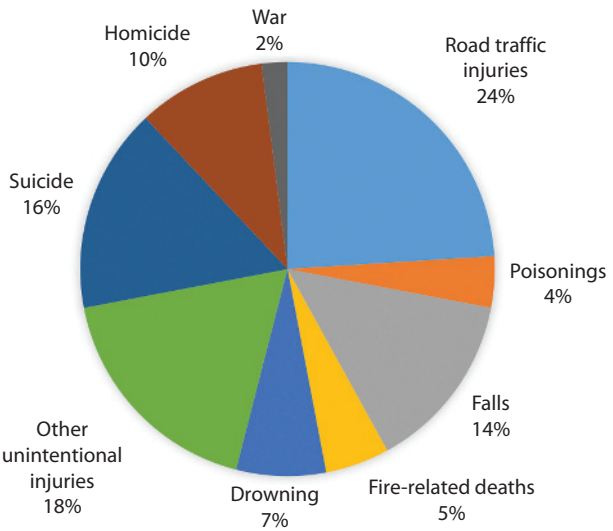


Figure 22.1 Global percentage of deaths due to injury (2014) (Source: WHO Global Health Estimates, 2014.)

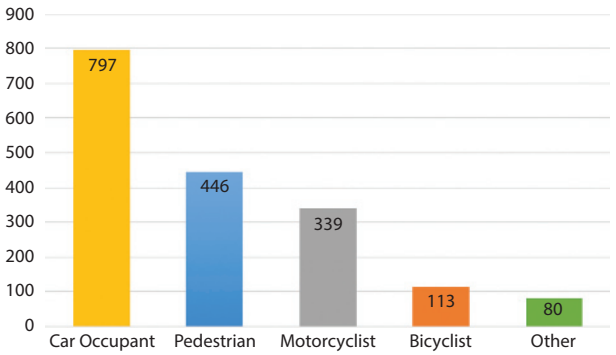


Figure 22.2 Fatalities in reported road accidents by road user type (Source: Department of Transport Data, UK, 2014.)

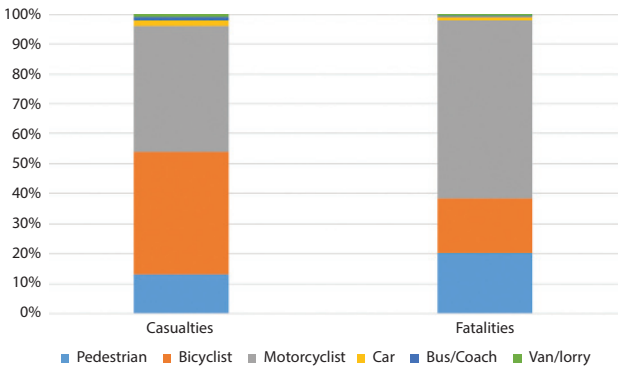


Figure 22.3 Casualty and fatality rates per billion passenger miles by road user type (Source: Department of Transport Data, UK, 2014.)

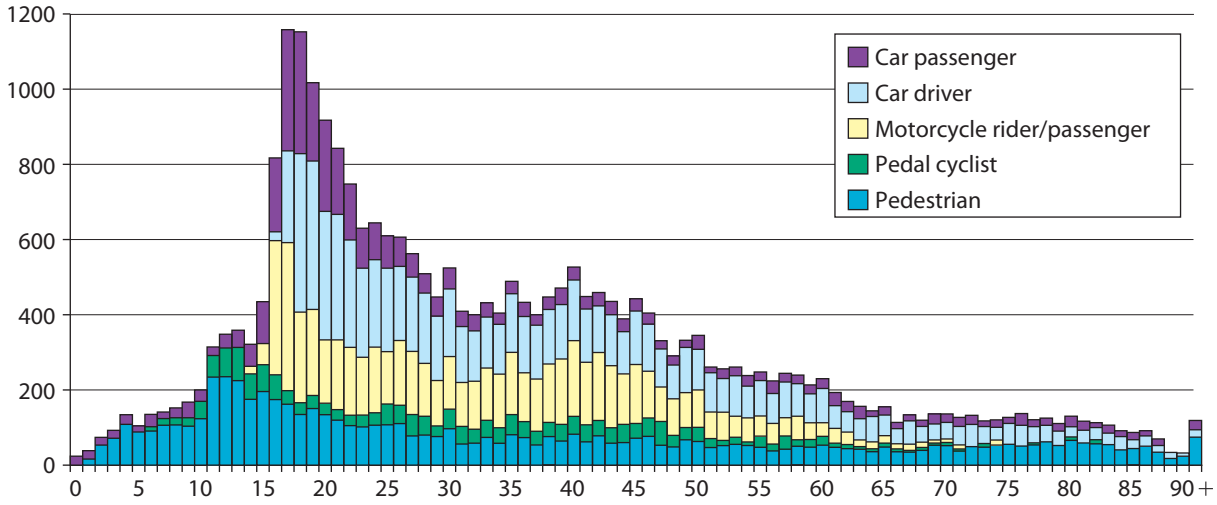


Figure 22.4 Deaths and serious injuries by road user type and age (Source: Department of Transport Data, UK, 2007.)

level of a nation is a major determinant. The 2004 WHO report cites mortality rates for seriously injured adults, i.e. those with an injury severity score (ISS) of 9 or higher. The ISS will be described in greater detail in a subsequent section. The overall mortality rate, including pre-hospital and in-hospital deaths, is 35% in high-income nations, but rises to 55% in middle-income economies and 63% in low-income economies. More seriously injured patients (ISS 15–24) reaching hospital show a six-fold increase in mortality in rural areas of low-income economies, when compared with those benefiting from a developed emergency medical service within a high-income economy.

For every death from trauma, three other victims of trauma will suffer permanent disability. As well as causing personal tragedy, this represents an enormous drain on a nation's healthcare economy; timely and effective management of major injuries can reduce both morbidity and mortality, as has been demonstrated in the UK. Since the introduction of the UK Major Trauma Centres and the Trauma Networks in in 2012, the Trauma Audit and Research Network (TARN) has reported a 63% improvement in the odds of a major trauma patient surviving their injury.

Mode of death

Deaths as a result of trauma have classically been shown to follow a trimodal pattern, described by Trunkey in 1983, with three waves following the injury. Some 50% of fatally injured casualties died from non-survivable injuries immediately, or within minutes after the accident; 30% survived the initial trauma, but died within 1–3 hours; the remaining 20% died from complications at a late stage during the 6 weeks after injury (Figure 22.5).

This trimodal pattern has more recently been shown to have moved towards a bimodal pattern similar to that seen in combat deaths, with a significant drop in the third, delayed deaths peak and with most deaths occurring shortly after admission.

The initial mortality peak is usually due to non-survivable, central nervous system injury or cardiovascular disruption. The severe nature of the injuries, the immediate nature of the deaths and the likely location in the pre-hospital environment means that very few of these casualties can be saved. Preventative measures that are effective in reducing the incidence of major traumatic incidents may lead to a reduction in the number of these deaths. However, a small proportion die as a result of early airway obstruction and external haemorrhage and these deaths can be prevented by immediate first-aid measures. A significant proportion of head-injured casualties who die on the

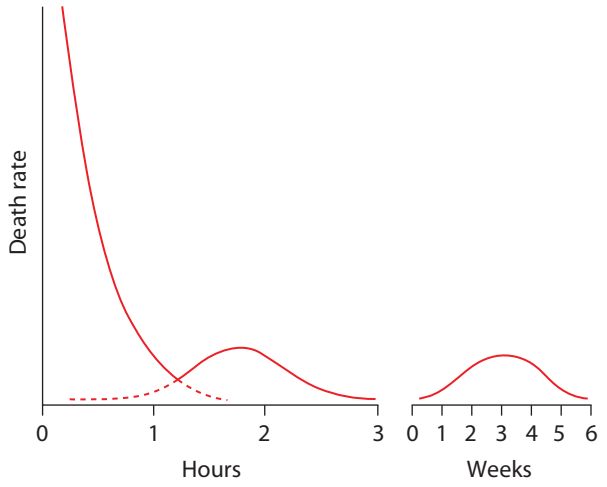


Figure 22.5 Death following trauma The trimodal pattern of mortality following severe trauma.

scene succumb not to the primary brain injury, but to secondary brain injury, caused by the hypoxia and hypercarbia associated with airway obstruction and respiratory dysfunction.

The second peak of deaths during the first few hours after injury is most often due to hypoxia and hypovolaemic shock. A significant proportion of these deaths can be avoided with an effective emergency medical service (EMS), as has been demonstrated in the UK since 2012; hence, this period has been called 'the golden hour'.

The third peak in the cumulative mortality rate within the 6 weeks following injury was largely due to multisystem failure and sepsis. This peak has reduced significantly in the United States and is not seen in contemporary UK practice, possibly because trauma management is now more effective, or because the UK trauma population has never conformed to the three-peak model.

Sequence of management

In developed healthcare systems, an effective EMS is available to initiate management at the scene of the injury and transfer the casualty rapidly to hospital. Immediate first-aid manoeuvres, such as opening the airway and controlling external haemorrhage with direct pressure, are life-saving interventions that require minimal equipment and training.

More complex treatment requires specialist equipment and expert training not always available at the scene and rapid transfer to a medical centre is mandatory. However, EMS medical teams can deliver advanced management to entrapped casualties. Such treatment is difficult to deliver in vehicles and aircraft and a balance has to be drawn between delaying to give treatment on scene and transferring an unstable casualty to a suitable medical centre.

In sophisticated healthcare systems, casualties are taken to the nearest hospital offering comprehensive Emergency Department management. This may mean bypassing the nearest hospital in favour of a more prolonged transfer to a Major Trauma Centre if the casualty's injuries are severe enough to warrant it. Treatment is centred on evaluation, resuscitation and stabilization. This phase merges into definitive care in the operating theatre, with control of airway, ventilation and surgical management of haemorrhage; these early interventions are known as damage-limitation surgery and anaesthesia. Musculoskeletal injuries are initially stabilized, followed by definitive treatment.

Level 2 or 3 critical care may be required to minimize complications and prevent third-phase deaths, and prolonged rehabilitation may be necessary to address the needs of casualties with brain injuries and complex musculoskeletal damage.

Guidelines for trauma management have changed rapidly; in UK practice, management is predicated on ATLS® (Advanced Trauma Life Support) principles, NICE (National Institute for Health and Care Excellence) guidelines and contemporary practice based on expert and consensus views. There are some significant differences between these three sets of guiding principles:

ATLS® is a tried and tested system that remains relevant as a structured resuscitation template; however, there are significant changes in key areas such as airway and shock management which are now standard in UK Major Trauma Centres.

The UK NICE trauma guidelines are based on Level 1 evidence, with recommendations based on significant improvements in outcomes based on randomized, controlled trials. This level of evidence is difficult to demonstrate given the variables of trauma care and the complexities of performing research in this setting.

Contemporary UK practice incorporates ATLS® and NICE guidance but also includes interventions which have been demonstrated to be effective in practice (e.g. military experience in conflicts) but have not yet been statistically proven. An example of this is the use of haemostatic dressings.

PRE-HOSPITAL MANAGEMENT

Essential elements include the following:

1 Organization
2 Safety on scene – including personal protection
3 Immediate actions and triage
4 Assessment and initial management
5 Extrication and immobilization
6 Transfer to hospital
7 Helicopters and air ambulances.

Organization

Provision of a pre-hospital EMS depends on economic resources and varies from no provision in rural, low-income countries to sophisticated services linked to hospital care in developed economies. The EMS in most countries is based on ambulances crewed by emergency medical technicians (EMTs) or paramedics with support from pre-hospital emergency medicine doctors varying between countries. The most integrated system is probably the French Services de l'Aide Medical Urgente (SAMU): all emergencies are triaged by a control room team, which includes a doctor, and an appropriate response is mounted. For major cases, intervention is provided by Services Mobile d'Urgence et de Reanimation (SMUR) teams – hospital-based medical teams with sophisticated equipment and

access to a range of transport including helicopters. SMUR teams can deliver an advanced level of care on scene with rapid transfer to an appropriate hospital and European experience is that a doctor-led pre-hospital service leads to a 2.8-fold improvement in mortality for seriously injured patents. However, the service is very expensive and demands a high number of experienced medical staff. In the UK, medical support in the pre-hospital environment comes primarily from Helicopter Emergency Medical Service (HEMS) doctors working with charitably funded air ambulances.

Safety on scene

In the UK, pre-hospital emergency medicine (PHEM) is now recognized as a sub-specialty of critical care medicine, which requires specialist training and the use of professional equipment and personal protection. The scene of a traumatic incident is invariably hazardous, and the immediate priority for a doctor on scene is personal safety; if this is neglected, the doctor can become a casualty rather than a rescuer. Some hazards are obvious, such as unstable wreckage, jagged metal debris and fire. However, there are concealed hazards that can injure the unwary. Undeployed airbags can be triggered, and a variety of toxic chemicals can be released, such as battery acid.

All members of pre-hospital medical teams should therefore be equipped with personal protective equipment (PPE) and clothing appropriate to the incident (Figure 22.6), and this should be deployed before the scene is entered. The safety of the immediate scene will normally be the responsibility of the fire service, with police controlling the incident overall. Nations' differing EMS will have their own specific regulations covering the specification of PPE for doctors working in the pre-hospital environment. As a rule, PPE must protect the head, eyes, hands, feet, limbs and body to an appropriate extent against physical, chemical, thermal and acoustic risks. Full chemical, biological, radiological and nuclear protection is a specialist requirement rarely applicable to doctors outside a military setting.

(a) (b)

Figure 22.6 Medical personal protective equipment (PPE) (a) Inadequate PPE; (b) correct PPE.

Immediate actions and triage

The initial action of a doctor arriving on scene is to establish safety – personal safety, scene safety and casualty safety. Contact should be made with the officers commanding medical, fire and police emergency services for a situation report and direction to casualties on a priority basis. Communications should be established. In the event of multiple casualties, priorities are determined by triage.

Triage is a system of medical sorting originating from the Napoleonic battlefields to identify casualties in an order of priority for evacuation and treatment. In trauma management, triage is used when the number of casualties is greater than can be managed simultaneously by the medical personnel available. There are two stages applicable in the pre-hospital environment: a *triage sieve* and a *triage sort*.

The triage sieve is a quick and uncomplicated system based on simple clinical observation of a casualty's ability to walk, breathe and maintain peripheral perfusion. It can be performed by trained, but non-clinical, personnel. Current UK practice, as advised by the National Ambulance Resilience Unit (NARU), is to perform the triage sieve in pairs and institute basic medical interventions during the triage; these are limited to application of tourniquets and homeostatic dressings for catastrophic haemorrhage, opening the airway and placing unconscious, but breathing, casualties into the recovery position. The triage sort requires a degree of clinical training and uses physiological measurements to score casualties and place them into priority groups. Both triage systems place casualties into four colour-coded priority categories, as shown in Box 22.1.

In the event of an overwhelming number of casualties, an expectant category can be used. This identifies casualties whose injuries suggest that survival is unlikely, enabling medical resources to be prioritized to those more likely to survive. In the event of improved resources, expectant casualties are re-categorized as Priority 1.

BOX 22.1 CASUALTY PRIORITY CATEGORIES

Priority 1	Immediate
Priority 2	Urgent
Priority 3	Delayed
Priority 4	Dead

The category of a casualty does not necessarily dictate the order of evacuation or treatment; for example, the 'walking wounded' and uninjured (Priority 3) may be evacuated first ('reverse triage').

Assessment and initial management

Once safety, command, communications and priorities have been established, patients can be given individual attention. This calls for a structured approach predicated on *awareness, recognition and management (ARM).*

Awareness

Awareness of the environment, pattern of damage to a vehicle and the nature of the incident can help the attending doctor predict the likely injuries and facilitate their early recognition. For example, ejection from a vehicle or death of another occupant of the same vehicle increases the likelihood of serious injury. Particular impaction patterns and intrusion of wreckage into the passenger compartment can suggest specific injuries; a bull's-eye fracture of a windscreen from inside a car indicates impaction of the passenger's head against the windscreen and likely head, maxillofacial and neck injuries. Entrapment in a fire is associated with smoke inhalation and possible inhalational burns.

Recognition

Recognition of injuries is based on a rapid and systematic questioning and examination of the casualty. An immediate assessment is made of the airway, breathing and circulation – the 'ABC' of trauma assessment. An instant assessment can be made by questioning the patient and eliciting a verbal response; the ability to speak means that the brain is being perfused with oxygenated blood and hence the patient has a patent airway, is breathing and has an adequate circulation. Head injury leading to loss of consciousness is the most common cause of airway obstruction and consequent hypoxia and hypercarbia; lack of response to command or painful stimulus indicates a significant level of coma. Access to an entrapped casualty may be extremely limited, but an assessment can usually be made of the airway and breathing, presence of peripheral pulses and peripheral perfusion, head, chest, abdomen, pelvis and limbs. This initial assessment guides immediate management and the urgency of extrication and transfer to hospital.

Management

Management of injuries is prioritized on treating the most immediately life-threatening injuries first, traditionally following the ABCDE sequence. The exception to this is the casualty suffering severe, external, peripheral haemorrhage. Military experience has shown that bleeding from limb wounds is a leading cause of combat casualty deaths, a significant proportion of which are avoidable. This has led to the development of a C-ABC sequence, where C stands for catastrophic haemorrhage. Life-threatening, external bleeding is controlled, and then the usual ABC sequence is followed.

As casualties with airway obstruction succumb within minutes, securing a patent airway is always a priority. Once the airway is open, the casualty must be oxygenated and ventilated if breathing is not adequate. Further circulatory compromise is addressed primarily by control of external haemorrhage; an intravenous cannula should be placed, but fluid administration is limited and titrated against the maintenance of a central pulse, if blood is not immediately available.

During this immediate management phase, the assumption is always made that damage to the cervical and thoracolumbar spine may have occurred. The stability of the cervical spine must be protected at all times until the neck can be cleared from the risk of injury. Stabilization has traditionally been achieved by two methods: manual, in-line immobilization, or securement with head blocks, head straps and a rigid cervical collar. However, the evidence for the efficacy of cervical collars is weak and they can potentially cause harm, for example by raising the intracranial pressure in head-injured patients. Their routine use is therefore being questioned and it is likely that there will be a move to using head blocks and head straps, with collars used for extrication only. The thoracolumbar spine is protected by immobilization with straps on a scoop stretcher or other extrication device. Long spinal boards should be used only for extrication and not for prolonged immobilization or transfer and conveyance, as they can rapidly cause tissue damage from pressure effects.

AIRWAY

The airway is opened initially with the 'bare hands' manoeuvres of chin lift and jaw thrust; the head should not be extended and should be kept in a neutral position. If blood, saliva or vomit is present in the airway, suction should be used. If 'bare hands' techniques are not adequate, an oropharyngeal (OP) airway or nasopharyngeal (NP) airway should be carefully placed to prevent the posterior aspect of the tongue obstructing the pharynx. NP airways are particularly useful in casualties with obstructing airways who have retained enough of a gag reflex to resist OP airways; however, they should be used cautiously in casualties with clinically apparent, basal skull fractures. If these manoeuvres are unsuccessful, there is a range of supraglottic devices which can be inserted

in difficult situations. The i-gel®, a second-generation, supraglottic airway device, is now in routine use with UK ambulance services.

Definitively securing the airway with intubation or cricothyroidotomy is very difficult in entrapped casualties. Without the use of anaesthetic drugs and muscle relaxants, casualties can only be intubated when jaw tone and protective reflexes have disappeared immediately prior to cardiac arrest. The survival rates of intubation in this situation are, not surprisingly, very poor. However, intubation with a rapid sequence anaesthetic induction (RSI) remains the gold standard for securing the airway in trauma casualties, as it offers reliable protection from airway leaks and aspiration. Prolonged attempts at intubation should not be made without effective pre-oxygenation and ventilation; casualties do not die from not being intubated, they die from hypoxia and hypercarbia. Accumulating evidence suggests that only practitioners with an appropriate level of anaesthetic training should be attempting RSI and intubation (Association of Anaesthetists Great Britain and Ireland draft guidance *AAGBI Guidelines: Safer pre-hospital anaesthesia 2016*). The UK National Institute for Health and Care Excellence (NICE) February 2016 guideline (*Major trauma: assessment and initial management February 2016*) recommends RSI for airway securement and suggests a 45-minute timescale from initial call to emergency services to intubation.

BREATHING

Once the airway is opened and secure, an assessment of the casualty's breathing is made. If breathing is clearly adequate, oxygen is administered from a high-flow, non-rebreathing, reservoir mask. With a flow rate of 15 L/minute, approximately 85% oxygen is delivered; there is no place for lower concentrations of oxygen in this situation. If there is any doubt that breathing is adequate, then ventilation must be supported with a bag–valve–mask (BVM) assembly. This should have a reservoir attached with oxygen flow of 15 L/minute. BVM ventilation is a difficult skill even in ideal situations, but chances of success can be improved with a two-person technique; one person holds the mask in place over the face with both hands and pulls the jaw up into the mask to open up the airway, while the second squeezes the bag. Care must be taken not to extend the neck and potentially compromise the cervical spine. Insufflation of air into the stomach must be avoided as it increases the risk of aspiration of stomach contents and can make ventilation of the lungs difficult if the stomach becomes grossly distended.

Adequacy of oxygenation should be judged by clinical assessment of lip colour to detect cyanosis, or use of a pulse oximeter. Adequacy of ventilation can be judged by clinical assessment of chest expansion and breath sounds, or use of a chemical or electronic end-tidal carbon dioxide ($EtCO_2$) monitoring (capnography) if a supraglottic airway device or tracheal tube is in place. Continuous, waveform capnography should be used to monitor ventilation in both spontaneously breathing and ventilated casualties and is considered mandatory in pre-hospital emergency anaesthesia.

Absence of breath sounds indicates a pneumothorax or haemothorax and, when associated with hyper-resonance, surgical emphysema or deviation of the trachea and cardiovascular collapse, a tension pneumothorax. A tension pneumothorax is an immediately life-threatening injury, traditionally treated in the first instance by decompression with a large-bore (14-gauge) intravenous cannula through the second intercostal space (ICS) in the midclavicular line or the 4/5 ICS in the anterior axillary line (alternative in obesity, etc.). This converts the tension pneumothorax into a simple pneumothorax and so should only be used on one side in unintubated casualties; definitive treatment of a simple pneumothorax in a spontaneously breathing casualty is to insert a wide-bore chest drain in the fifth intercostal space, anterior to the midaxillary line, with the drain being connected to a Heimlich-type valve. However, if the casualty is breathing and stable with a simple pneumothorax, rapid transfer to hospital is preferable. Decompression with an open thoracostomy is the preferred technique to decompress a tension pneumothorax if the expertise is available. Open or sucking pneumothoraces should be covered with a specialist, valved dressing or an occlusive dressing; be aware that a tension pneumothorax can subsequently develop.

Positive-pressure ventilation is likely to accelerate the conversion of a simple pneumothorax into a tension pneumothorax. If the casualty is intubated and ventilated and a pneumothorax suspected, a simple, open thoracostomy is made in the fifth intercostal space, anterior to the midaxillary line. This allows a tension pneumothorax to decompress; however, the lung can still be inflated as the casualty is being ventilated and so this procedure can be performed bilaterally if indicated. A thoracostomy is made by making a 3 cm horizontal incision immediately above the sixth rib, just anterior to the midaxillary line, dissecting the subcutaneous tissues with large, straight Spencer Wells forceps until the chest cavity is entered. A finger is used to open up the thoracostomy and ensure no vital structures are felt.

CIRCULATION

Severe, external haemorrhage should be aggressively controlled by packing the wound with haemostatic gauze (e.g. Celox™), applying pressure with a compression dressing (e.g. Olaes® modular bandage), and use of a tourniquet such as the C-A-T™ (Combat Application Tourniquet). If specialist equipment is

not available, use simple dressings and direct pressure. Tranexamic acid (1 g) should be administered at the earliest opportunity and the casualty kept warm with insulating blankets and self-heating blankets to prevent exacerbating bleeding through hypothermia.

Vascular access should be secured with a wide-bore cannula sited in a large vein, or an intraosseus cannula sited in the humeral head or tibia (e.g. EZ-IO® intraosseus vascular access system). Administration of intravenous fluids for patients with active bleeding should be restricted in the pre-hospital environment; rapid infusion of large volumes of fluids has been shown to increase mortality, and the 2016 NICE guidance in the UK is to titrate fluids against the presence of a central pulse, with a crystalloid solution such as Ringer's lactate or Hartmann's compound sodium lactate being the preferred fluid (large, infused volumes of 0.9% sodium chloride solution can be associated with the development of a hyperchloraemic acidosis and should be avoided).

Pre-hospital blood transfusion is increasingly available in the UK, with two units of O-negative packed red blood cells typically being carried on HEMS air ambulances. Some UK HEMS teams now also carry two units of plasma, given with the blood to replace clotting factors and avoid raising the haemoglobin inappropriately. Prothrombin complex concentrate can also be used in bleeding patients known to be anticoagulated (increasingly common with cardiac stenting and stroke management, etc.).

Severe, unresponsive shock is likely to be the result of uncontrollable bleeding externally or into the chest, abdomen, pelvis and multiple long bones (embodied in the aperçu 'onto the floor and four more'). Loss of cardiac output can also be due to tension pneumothorax or cardiac tamponade. Cardiac tamponade is most commonly associated with penetrating trauma of the chest within the nipple lines anteriorly or scapulae posteriorly. The use of ultrasonography is increasingly common in UK pre-hospital practice for the identification of pneumothorax, tamponade and intra-abdominal bleeding. However, its accurate use depends on a high level of expertise and regular usage and so should not replace clinical evaluation.

Severe shock leading to pulseless electrical activity (PEA) or asystolic cardiac arrest is an indication for bilateral thoracostomies and/or clam-shell opening of the chest and incision of the pericardium. These manoeuvres will treat the reversible causes of trauma cardiac arrest – hypoxia, hypovolaemia, tension pneumothorax and cardiac tamponade – and may precede intubation, ventilation and intravenous cannulation in this dire, pre-mortem situation.

DISABILITY

The casualty is quickly assessed for neurological disability using the Glasgow Coma Scale (GCS) (see Table 22.1) and assessment of pupillary size and asymmetry.

Extrication and immobilization

More complex management is often impractical in an entrapped casualty, and so extrication of the patient becomes a priority. This should be done with due care and attention paid to spinal protection, usually using spinal boards or other rigid immobilization devices. Fractured limbs should be splinted in an anatomical position to preserve neurovascular function. Analgesia may be necessary to extricate an injured casualty, and this can be achieved with inhalational or intravenous agents.

The initial manoeuvre in the extrication process is manual immobilization of the cervical spine. This can be done from behind the casualty (typically in seated casualties entrapped in vehicles with a rescuer in the rear of the vehicle), or from the front and side if access is limited. A stiff cervical collar may be sized and fitted at this stage, but manual immobilization is still mandatory until the cervical spine can be protected with head blocks and taping.

Further immobilization and extrication may be impossible until wreckage has been cleared enough to enable an extrication device to be positioned under the casualty. Managing wreckage is a specialist skill that is the province of the Fire and Rescue crews; however, the pre-hospital doctor should be familiar with the techniques used to advise how extrication can be managed without causing additional injury to the casualty. Common manoeuvres in road vehicle wreckage are removal of glass and doors, a dashboard roll to lift the dashboard off trapped limbs, and removal of the roof by cutting through the A, B and C pillars. The seat can then be carefully flattened, and a long spinal board slid under the casualty from the rear of the vehicle, minimizing movement of the spinal column. If a casualty is deteriorating fast, the rescue crews should be advised and a rapid extrication carried out.

Limb fractures and joint dislocations should be reduced and the limb returned, if possible, to its anatomical position with gentle traction and straightening. This may require procedural sedation. Note that some injuries such as posterior hip dislocations may prevent an anatomical alignment, and the limb must not be forced. The limb should then be splinted with traction, gutter or vacuum splints as appropriate. This reduces pain and haemorrhage, and minimizes neurovascular damage. Femoral traction splints such as the Kendrick Traction Device (KTD™) are effective for midshaft femur fractures. The traction reduces the fracture, and the fusiform compression of the fracture haematoma reduces further bleeding. Procedural sedation with opioids and ketamine may be required to effectively apply femoral traction splints. A unilateral, closed, femoral fracture can cause a 1.5 L blood loss – 30% of the adult blood volume and enough to

cause significant shock without other injury. Unilateral traction devices are a temporary measure only; if they are left on the patient, they can cause pressure areas and should therefore be swapped for formal splintage or traction on arrival in the Emergency Department.

Open-book pelvic fractures may cause uncontrollable retroperitoneal bleeding. Blood loss can be minimized by stabilizing and reducing the fracture, using specialist pelvic compression devices (pelvic binders). These should be sited around the level of the greater trochanters – they are ineffective if too low or too high and can be used with a figure-of-eight bandage around the feet to internally rotate the legs. If a formal pelvic binder is not available, a sheet wrapped round the patient at the same level (greater trochanters) can be used. Reduction of the pelvis is achieved via either a windlass mechanism or weights hung off the tails of the sheet. Tying the knees and ankles together may also help.

Analgesia may be necessary to extricate an injured casualty. In the absence of a pneumothorax, this can be administered by inhalation with Entonox, a 50:50 mixture of nitrous oxide and oxygen, delivered via a breath-actuated regulator valve and mask or mouthpiece. Single use methoxyflurane inhalers can also be used. Parenteral analgesics should only be given intravenously, and titrated cautiously against effect. Other routes of administration are very unpredictable, especially in shocked casualties. Pure opioid agonists such as morphine, diamorphine and fentanyl are most effective, but it should be noted that there is a wide variation in response between individuals, and care should be taken not to cause respiratory depression by overdosage. Ketamine is a very useful drug that is a powerful analgesic in doses of 0.1–0.5 mg/kg intravenously and a general anaesthetic in doses of 2–4 mg/kg. The advantage of ketamine is that it does not cause respiratory depression and the casualty's airway is more predictably maintained. Doses and administration times of all drugs given should be noted.

Transfer to hospital

Delayed or prolonged transfer to hospital is associated with poor outcomes, and every effort should be made to minimize the on-scene times for injured casualties. There is a balance between 'scoop and run' and 'stay and play' management. The airway must be secured, and life-threatening chest injuries (e.g. tension pneumothorax) and catastrophic external haemorrhage dealt with before transfer commences. Prolonged attempts at complex management on scene are disadvantageous and should be limited to life-saving interventions where possible.

The appropriate method of transport should be chosen, with helicopters offering some advantage for long-distance transfers or rescue from remote and rough terrain. The appropriate destination hospital should be chosen for the casualty's likely injuries, and this may mean bypassing a small unit that does not have the appropriate facilities, in favour of direct conveyance to a Level 1 or Major Trauma Centre. In the UK, casualties are assessed using a triage tool, with the recommendation that the more seriously injured (Level 1 trauma) are conveyed directly to a Major Trauma Centre if within 60 minutes transfer time. The receiving medical team should be directly advised of the estimated time of arrival (ETA) and the identified injuries, enabling a trauma team to be standing by.

During the transfer, the casualty's vital signs should be monitored clinically and with available equipment. Conscious casualties should be constantly assessed by speaking to them, and a decrease in conscious level detected early. ECG and pulse should be continuously monitored, blood pressure measured with a non-invasive blood pressure (NIBP) monitor, and oxygen saturations measured if peripheral perfusion allows. Waveform capnography should be used for gauging adequacy of ventilation for both spontaneously breathing and ventilated casualties.

The casualty's airway must be maintained at all times, as should oxygenation and ventilation. Oxygen saturations should be maintained above 95% if possible, and ventilated casualties have their $EtCO_2$ maintained at a low normal level (4.0–4.5 kPa). Haemorrhage is controlled with direct pressure and specialist dressings and crystalloid fluids titrated intravenously to maintain a palpable, central pulse if blood is not immediately available.

If the patient deteriorates en route, the medical attendant must decide whether to attempt resuscitation while on the move, to stop and resuscitate or to make a run for the nearest appropriate hospital. This decision will depend on the nature of the intervention required and the ETA at the different hospitals. Casualties should not be taken to hospitals not designated as being able to manage trauma as a delay in non-expert hands can be deleterious.

Contemporaneous records are almost impossible to maintain during a transfer, but electronic equipment can usually download a paper or electronic record. If not, notes should be made as soon as possible after arrival at the hospital. On arrival, the pre-hospital medical attendant should remain part of the resuscitation team until an effective handover can be made.

Helicopters and air ambulances

A helicopter emergency medical service (HEMS) offers significant advantages but is expensive to run. London's Air Ambulance data show that the primary life-saving benefit is the rapid delivery of advanced resuscitation skills to the scene. The most essential

life-saving skill is advanced airway management and this requires an anaesthetically trained doctor who can perform a rapid sequence anaesthetic induction (RSI) and manage tracheal intubation in difficult circumstances. International data show that, as a result of these interventions, there is a reduction of 15% in death from head injuries, and a reduction of between 5 and 7 days in intensive care stays.

However, the availability of appropriately trained doctors is variable; many international and some UK air ambulances are crewed by paramedics or EMTs only, and this reduces the effectiveness of the service to less advanced life support and rapid delivery and evacuation of casualties to an appropriate facility. A common standard for response times in the UK and Europe is 12 minutes from call-out to arrival. This ability to transport casualties quickly over large distances also means that smaller, less well-equipped and well-staffed hospitals can be bypassed in favour of large, specialist Level 1 or Major Trauma Centres.

A wide variety of helicopters are used internationally for HEMS work, ranging from aircraft such as the Eurocopter EC135 and MD902 Explorer to the larger EC145 and AW169, or even the very large Sikorsky S92 used for maritime search and rescue. (Figure 22.7). A feature common to all HEMS is that the helicopter is twin-engined for safety and flexibility of flight paths. As costs rise dramatically with increased size of the helicopter, HEMS aircraft are a compromise. With the exception of military and Coastguard craft, the size is usually restricted.

Cramped cabin space and poor patient access in these helicopters greatly restrict the patient interventions possible during flight. The aircraft are noisy and vibration is considerable, so monitoring the patient's condition is difficult. These factors make it essential that the patient is stabilized and immobilized prior to transfer; the airway must be secured and protected, ventilation maintained, haemorrhage controlled and intravenous access for fluid administration preserved. Monitoring should be reliable, and the ECG, blood pressure, oxygen saturations and capnography observed.

Safety is paramount for doctors working with helicopters, and all personnel should be trained and familiar with safety guidelines. The helicopter should not be approached or exited until directed by the crew, and this should be done from within a 45-degree arc either side of the nose of the aircraft. If asked to disembark while the rotor blades are revolving, personnel must exit to the front of the aircraft only on instruction from the pilot and be aware that the rotor disc droops as it slows and may come below head height, especially uphill, when landing on an incline or in windy conditions.

HOSPITAL MANAGEMENT

Upon reaching hospital, the following are important in hospital management:

1 Organization
2 Trauma teams
3 Assessment and management – the ATLS® concept
4 Initial assessment and management
5 Definitive, systemic management.

Organization

The aim of any integrated EMS is to 'get the right patient to the right hospital in the right amount of time' (Trunkey). Regional services were set up in the USA in 1973, with three levels of hospital designated as able to manage trauma to differing levels, with Level 1 trauma centres able to manage all degrees of trauma. However, the development and integration of this system was patchy and the expense of such a system prevents full development in many countries.

In the UK, a trauma network system was set up in 2012 after a series of reports highlighted the unacceptably high trauma mortality rates. The system divided hospitals into three groups:

- *Receiving Hospitals* – hospitals not able to take any trauma
- *Trauma Units (TUs)* – hospitals able to manage single system trauma which is not immediately life-threatening (Level 2)
- *Major Trauma Centres (MTCs)* – large, regional hospitals able to manage all levels of polytrauma (Level 1), with all relevant specialties on site.


(a)


(b)

Figure 22.7 HEMS helicopter interior (a) Eurocopter EC135 T3 air ambulance. (b) Interior of EC135 showing attendants seat and patient stretcher.

UK ambulance crews use a 'trauma unit bypass tool' algorithm to determine which patients meet the requirements for major trauma centre care and casualties that meet these criteria are taken directly to the MTC, if the centre is within 60 minutes' conveyance time. The use of a national network of air ambulances has enabled patients to be taken directly to the MTCs without any time penalty. Audit of the outcomes in 2015 showed a 63% improvement in trauma survival over the 3 years from the Trauma Network implementation and improved survival rates have been backed up by a meta-analysis of US and Canadian trauma centres.

Regionalized trauma systems are now operational in many countries, including the USA, Canada and Australia, and across Europe. However, in many healthcare economies, the majority of available hospitals will not have all the specialist staff and facilities to manage major injuries adequately. Each hospital must therefore have standard operating procedures (SOPs) for assessing, managing and, if indicated, transferring trauma casualties, depending on the facilities available.

The common standard for trauma resuscitation within hospitals is Advanced Trauma Life Support (ATLS®). In the UK, the National Institute for Health and Care Excellence (NICE) has issued a set of guidelines for the assessment and management of major trauma (February 2016) which strictly adhere to high quality evidence. There is some dissonance between the ATLS® and NICE guidelines, and contemporary UK practice is based on a combination of these recommendations, with the additional, pragmatic use of specialist techniques and equipment.

Trauma teams

Casualties who have survived their initial trauma and reach hospital alive need rapid assessment and appropriate resuscitation to avoid their dying in the hours following injury. Crucial to the effective management of seriously injured casualties is the immediate availability of appropriately trained and experienced doctors and healthcare professionals, and this need has led to the development of the trauma team concept.

The team is led by a senior doctor, with advanced trauma skills, whose base specialty is less important than his or her training and experience. The trauma team is preferably activated by the pre-hospital practitioners according to a set of standard criteria and should therefore be awaiting the casualty as they arrive at the hospital. Team members in the UK would normally include the following personnel:

- Emergency Department physician
- physician anaesthetist and anaesthetic technician
- Emergency Department nurses
- radiographer
- surgeon from appropriate specialties, usually general surgery and orthopaedics
- intensive care specialists
- appropriate specialists – e.g. paediatric, obstetric, ear, nose and throat (ENT), maxillofacial, neurosurgical, plastic surgery, cardiothoracics can be called in as required once a specialist problem is identified.

The development of emergency medicine as a distinct specialty and the increasing availability of experienced and senior emergency medicine and anaesthetic doctors, with sophisticated trauma imaging availability on a 'round-the-clock' basis, has enabled the immediate management of trauma casualties without the need for immediate specialist surgical skills; once the initial assessment and imaging has been completed, the appropriate specialist surgeon can be called in or stood by in the operating theatre for definitive surgical management of specific injuries. It is important that transfer to an operating theatre is not carried out until the relevant specialties are aware.

Trauma teams should function in an appropriate environment and most hospitals will have a resuscitation room with all required equipment immediately available. Personal protective equipment including gowns, gloves and eye protection must be available. A sophisticated resuscitation room will have anaesthetic delivery systems, equipment and drugs for airway management, intravenous fluid and rapid administration systems for shock management, and a variety of surgical packs for specific interventions such as chest drain insertion. Patient trolleys should be compatible with the taking of X-rays, and the X-ray equipment can be built onto an overhead gantry. Ultrasound imaging equipment should be available for central venous cannulation and extended focused assessment sonography in trauma (eFAST). Both the environment and intravenous fluids should be warmed to minimize hypothermia.

The goal is to perform a rapid assessment of the patient, identify and manage the immediately life-threatening injuries and get the patient into the CT scanner within 15 minutes for a trauma CT to identify the injuries that have been sustained. The patient can then be further resuscitated and transferred for definitive care. In some centres, casualties can be admitted direct to the CT scanner from the scene by the EMS team, bypassing the Resuscitation Room, with resuscitation from the field continuing in the scanner.

The ATLS® concept

Major musculoskeletal injuries can be dramatic and distracting, but it is rare for them to be immediately life-threatening in the absence of catastrophic haemorrhage. The classic mistake when treating

trauma is to focus on the attention-grabbing compound fracture, and miss the obstructing airway, which is far more likely to cause a 'golden hour' death. Hence the most immediately life-threatening injuries should always be treated first. However, although this principle has been known for generations, in the stress of the moment a logical sequence may not be followed unless the treating doctor is trained and practised. To meet this need, a number of training systems have been developed over the years, of which the best known is the Advanced Trauma Life Support Program for Doctors (ATLS®) developed by the American College of Surgeons Committee on Trauma.

ATLS® originates from 1976, when James Styner, an orthopaedic surgeon, crashed his light aircraft in rural Nebraska with his wife and four children on board. His wife was killed instantly and three of his four children sustained critical injuries. Having arrived at the nearest hospital, Styner found that the care delivered to his family was inadequate and inappropriate, and this stimulated him to initiate a trauma care training programme that became ATLS®. The course has since become an internationally recognized standard and is currently taught in more than 40 countries worldwide. The current iteration is the 10th edition of ATLS®, which is currently being rolled out. It was introduced in the US in March 2017 and courses will commence in the UK in January 2018.

The ATLS® course is based on validated teaching techniques, and it uses a system of core content lectures and practical skill stations to develop skills that are practised and finally tested in simulated trauma scenarios. The system taught is based on a three-stage approach:

1 *Primary survey and simultaneous resuscitation* – a rapid assessment and treatment of life-threatening injuries.
2 *Secondary survey* – a detailed, head-to-toe evaluation to identify all other injuries.
3 *Definitive care* – specialist treatment of identified injuries.

The primary and secondary surveys constitute the initial assessment and management, which leads to the definitive care of the casualty following transfer if required.

The intention of ATLS® is to train doctors who do not manage major trauma on a regular basis, but it is applicable to any trauma situation as an underlying system on which to base management of an injured casualty and it acts as a common language for those involved in the care of trauma patients. The sequence is taught assuming one non-specialist doctor supported by one nurse, working on a single casualty, but the various components can be performed simultaneously if a team is available. The training is didactic, but the use of specialist skills (e.g. anaesthetic) should not be excluded. Although the course is updated on a 4-yearly basis, there is an inevitable time lag, and fast-developing areas such as imaging may introduce changes to local trauma management not found in current ATLS® courses. There are also national and local variations in practice that need to be taken into account; for example, the 9th edition ATLS® practice differed significantly from contemporary UK practice as recommended by NICE. These are discussed later in this chapter – however, the ATLS concept and structured approach have stood the test of time and remain the most widely recognized basis for trauma management internationally.

Initial assessment and management

The initial assessment and management is part of a sequence leading to the transfer and definitive care of a casualty. During the primary and secondary surveys, a number of monitoring and investigative adjuncts are used alongside clinical examination as given in Figure 22.8 and the accompanying Box 22.2.

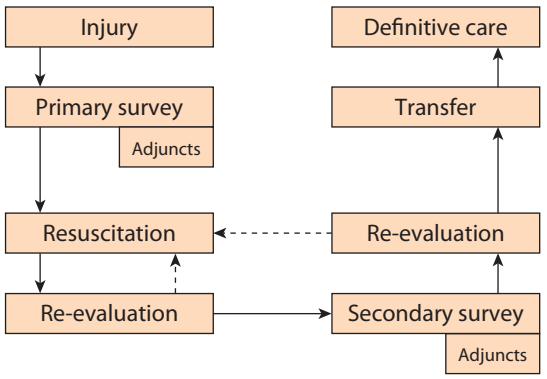
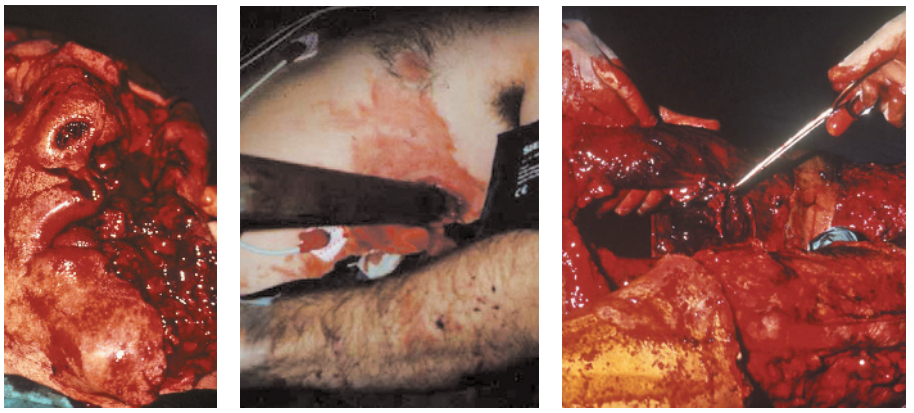


Figure 22.8 Algorithm of ATLS initial assessment and management

BOX 22.2 ADJUNCTS TO PRIMARY SURVEY

Vital signs
ECG
Pulse oximetry
End-tidal carbon dioxide
Arterial blood gases
Urinary output
Urethral catheter (unless contraindicated)
Nasogastric tube (unless contraindicated)
Chest X-ray
Pelvic X-ray



(a) (b) (c)

Figure 22.9 Triage priorities
(a) *Priority 1 – Airway:* severe face and neck wounds.
(b) *Priority 2 – Breathing:* severe chest wounds.
(c) *Priority 3 – Circulation:* severe bleeding and shock.

The ABCs

The underlying principle of ATLS is to identify the most immediately life-threatening injuries first and start resuscitation. As a general rule, airway obstruction kills in a matter of minutes, followed by respiratory failure, circulatory failure and expanding intracranial mass lesions. This likely sequence of deterioration has led to the development of the trauma '*ABCs*', a planned sequence of management predicated on treating the most lethal and time-critical injuries first. Throughout this sequence, the assumption is made (until proven otherwise) that there may be an unrecognized and unstable cervical spine injury. Hence, the sequence is:

A **A**irway with cervical spine protection
B **B**reathing
C **C**irculation with haemorrhage control
D **D**isability or neurological status
E **E**xposure and **E**nvironment – remove clothing, keep warm.

As previously described, catastrophic haemorrhage may be controlled before the airway, designated by the C-ABC sequence; however, death is ultimately caused by cerebral anoxia, regardless of whether the anoxia is a result of airway obstruction, respiratory failure or shock. Hence, the goal of resuscitation is to preserve the perfusion of the brain with oxygenated blood.

Triage

Triage, as described in the pre-hospital section of this chapter, is medical sorting to prioritize multiple casualties for resuscitation, and is used when the number of casualties outstrips the available resources. The initial two phases of triage, usually pre-hospital, are the *sieve* and the *sort*, to group casualties into the four priority groups of immediate, urgent, delayed or dead. Within the ATLS® system, multiple casualties are triaged systemically by rapidly assessing each patient's ABCs. Those with the most immediately life-threatening injuries are treated first (Figure 22.9); these are injuries of the:

Airway:	Actual or impending obstruction	Priority 1
Breathing:	Hypoxia or ventilatory failure	Priority 2
Circulation:	External haemorrhage or shock	Priority 3

Primary survey and resuscitation

During the primary survey, life-threatening conditions are identified and resuscitation started simultaneously, again following the ABCDE sequence.

The *Awareness Recognition Management (ARM)* system enables the treating doctor to focus rapidly on the likely problems; for example:

A*wareness* – A head injury is the most likely cause of unconsciousness and obstructed airway in trauma casualties. Coma therefore alerts the practitioner to the likelihood of airway obstruction.
R*ecognition* – An obstructed airway is recognized by looking, listening and feeling for the diagnostic signs.
M*anagement* – The airway is established with simple 'bare hands' manoeuvres, airway adjuncts, advanced airway interventions or surgical airway techniques.

As each stage in the ABCs is completed, the casualty is re-evaluated for deterioration or improvement; on completion of the breathing assessment, the airway is re-examined and the airway and breathing reassessed before moving on to the circulation, etc.

A – AIRWAY AND CERVICAL SPINE CONTROL

The cervical spine is stabilized immediately on the basis that an unstable injury cannot initially be ruled out. There are two techniques for this:

- manual, in-line immobilization
- cervical collar, head supports and strapping.

Simultaneously, the airway is examined for obstruction by *looking*, *listening* and *feeling* for signs such as

respiratory distress, use of accessory muscles of respiration, decreased conscious level and lack of detectable breath on hand or cheek. The airway is supported initially by lifting the chin or thrusting the jaw forward from under the angles of the mandible. Secretions and blood are carefully suctioned, and OP or NP airways used to hold the tongue forward. If these simple manoeuvres are unsuccessful, the options are supraglottic airway devices, tracheal intubation or surgical airway. All these techniques can be performed without extending the neck.

B – BREATHING

A clear airway does not mean the casualty is breathing adequately to enable peripheral tissue oxygenation. As soon as the airway is secured, the chest must be exposed and examined by *looking*, *listening* and *feeling*. Adequate and symmetrical excursion, bruising, open wounds and tachypnoea are looked for, and the chest is auscultated for abnormal or absent breath sounds, which indicate a pneumothorax or haemothorax. The trachea is palpated in the suprasternal notch to detect the deviation caused by a tension pneumothorax and the chest is percussed for the hyper-resonance of a tension pneumothorax or the dullness of a haemothorax. Surgical emphysema is recognized on palpation by its characteristic crackling and popping and suggests tension pneumothorax or bronchial injury.

A tension pneumothorax must be treated immediately if the diagnostic signs of absent breath sounds, hyper-resonance, surgical emphysema and deviated trachea are found. Initial management is decompression with a large bore cannula placed in the second intercostal space in the midclavicular line or fifth intercostal space anterior to the midaxillary line (or preferably by thoracostomy if appropriate expertise available) followed by chest drain placement. If there is any doubt as to the adequacy of the casualty's breathing and oxygenation, ventilation should be started with a reservoir BVM assembly using high-flow oxygen. Any trauma casualty who has required intubation must be ventilated.

C – CIRCULATION WITH HAEMORRHAGE CONTROL

The circulation is assessed by *looking* for external bleeding and the visible signs of shock such as pallor, prolonged capillary refill and decreased conscious level. The heart is *auscultated* to detect the muffled sounds of cardiac tamponade, and poor perfusion assessed by *feeling* for clammy and cool skin. The peripheral and central pulses are palpated to detect tachycardia and diminished or absent pulse pressure.

External bleeding is controlled by pressure, and two 14-gauge cannulae sited for administration of IV fluids and blood. The use of tranexamic acid and tourniquets feature in the ATLS® 10th edition. Blood samples can be drawn from the cannulae for baseline diagnostic tests and transfusion cross-matching. The 9th edition ATLS® fluid guidelines advises giving 2 L of warmed, crystalloid intravenous fluids alhough this is likely to be reduced to 1 L in the 10th edition; this differs from contemporary UK practice where crystalloid infusion is restricted, titrated against a central pulse and early transfusion with warmed, packed red blood cells and fresh frozen plasma advocated.

D – DISABILITY

The key element of assessing a patient's neurological status is the Glasgow Coma Score (GCS) (Table 22.1). This score records eye opening and the best motor and verbal responses, giving a score of between 15 for normal responses and 3 for no responses. Repeat GCS scoring can track deterioration in the conscious level, and indicate the need for elective intubation and ventilation. It is much more precise than the AVPU score (Aware, Verbally responsive, Pain responsive, and Unresponsive). The classic pitfall of intoxication should be considered, but a lowered GCS is assumed to be secondary to a cerebral injury until proven otherwise.

The pupils are examined for any difference in size indicating possible raised, intracerebral pressure (ICP), and unresponsive pupils, fixed at midpoint, which can indicate serious brain damage.

Table 22.1 Glasgow Coma Score (GCS)

Response	Score
Eye opening	
Spontaneous	4
On command	3
On pain	2
Nil	1
Best motor response	
Obeys	6
Localizes pain	5
Normal flexor	4
Abnormal flexor	3
Extensor	2
Nil	1
Verbal response	
Orientated	5
Confused	4
Words	3
Sounds	2
Nil	1

E – EXPOSURE AND ENVIRONMENT

The patient should have all clothing removed to enable a full examination of the entire body surface area to take place. This will require log-rolling to examine the posterior aspects and allow removal of any glass or debris. The casualty should be kept warm to maintain body temperature as close to 37 °C as possible, and all fluids and ventilated gases should be warmed. A hypothermic patient becomes peripherally shut down and acidotic and, if shivering, has greatly increased oxygen demands. Severe hypothermia (<32 °C) accompanying trauma is associated with a mortality incidence approaching 100%.

Adjuncts to primary survey

A number of monitoring and diagnostic adjuncts are used to supplement the primary survey and resuscitation, in addition to vital signs monitoring and haematological assays (see Box 22.2 and Table 22.2):

- *Electrocardiographic (ECG) monitoring* – is used to monitor heart rate and detect arrhythmias and ischaemic changes.
- *Pulse oximetry* – measures arterial oxygen saturations (SaO_2) and monitors peripheral tissue perfusion. Note that this is unreliable in low-output states, hypothermia and with motion artifact.
- *End-tidal carbon dioxide monitoring ($EtCO_2$; capnography)* – gives an estimation of arterial carbon dioxide partial pressure in intubated and ventilated patients, allowing optimization of lung ventilation. It also confirms tracheal intubation and alerts the practitioner to a drop in cardiac output.
- *Arterial blood gases (ABGs)* – allows quantification of arterial oxygen and carbon dioxide partial pressures with acid–base balance. This will also give the haemoglobin, sodium, potassium and glucose levels.
- *Urethral catheter* – allows measurement of hourly urine output (unless insertion contraindicated, e.g. in the case of a ruptured urethra).
- *Nasogastric tube* – decompresses the stomach and helps prevent aspiration (unless insertion contraindicated, e.g. because of a basal skull fracture).

Table 22.2 Palpable pulses at different blood pressures

Pulses palpable	Likely systolic blood pressure
Carotid, femoral, radial	>80 mmHg
Carotid, femoral	>70 mmHg
Carotid	>60 mmHg
No pulse	<60 mmHg

- *Chest X-rays* – are important for diagnosis of life-threatening chest injuries such as pneumothorax, which will require early treatment.
- *Pelvic X-rays* – enable a fractured pelvis to be diagnosed, which will alert to the likelihood of retroperitoneal bleeding.

NOTE: *Lateral cervical spine X-rays* do not exclude fractures or unstable necks and so do not alter management; if required, they can be performed as part of the secondary survey.

Increasingly, *ultrasonography* and *CT* are used as adjuncts during or immediately after the primary survey and have supplanted early use of chest and pelvic X-rays in some centres.

Secondary survey

The secondary survey is a detailed, head-to-toe evaluation to identify all injuries not recognized in the primary survey. It takes place after the primary survey has been completed, if the patient is stable enough and not in immediate need of definitive care; it may, in fact, take place after surgery, or on the intensive care unit (ICU). The importance of the secondary survey is that relatively minor injuries (e.g. small joint dislocations and fractures) can be missed during the primary survey and resuscitation but can cause long-term morbidity if overlooked.

The components of the secondary survey are:

- the history
- physical examination
- 'tubes and fingers in every orifice'
- neurological examination
- further diagnostic tests
- re-evaluation.

THE HISTORY

The patient's account of their trauma and their past medical history should be recorded. Particularly important is to establish whether the trauma was subsequent to a medical collapse: did the patient suffer a myocardial infarct causing a car crash, or was the infarct a result of hypovolaemia? With the increasing proportion of the elderly in developed societies, more patients are receiving chronic treatment for hypertension, etc., which can have a profound effect on their response to hypovolaemia. An example of this is a combination of beta-blockers and angiotensin-converting enzyme (ACE) inhibitors, which cause a profound drop in blood pressure if the patient's cardiac output is minimally compromised. A useful mnemonic is AMPLE: allergies; medications; past illnesses; last meal; events and environment.

EXAMINATION

Examination follows a logical sequence from the head down to the extremities, including a log-roll to ensure that all the body surfaces are examined. The guiding injunctions are *look*, *listen* and *feel*.

Head The head is examined for contusions, lacerations and clinically detectable fracture. The eyes and ears are examined for local damage, and examined internally with an ophthalmoscope/otoscope for signs of bleeding, etc. Bleeding from the ears can indicate a basal skull fracture. The GCS should be repeated.

Face The face is examined for signs of fractures with a consequent risk of airway obstruction – contusion, laceration, deformity, malocclusion of teeth and crepitus. Cerebrospinal fluid issuing from the nose (rhinorrhoea) is indicative of a basal skull fracture.

Neck All aspects of the neck are examined for contusions, lacerations, swelling, tenderness and a step in the cervical spine indicative of fracture/dislocation. Minor-looking contusions over the anterior neck can be indicative of underlying damage to the laryngeal and tracheal structures, which are associated with airway obstruction.

Chest The chest is inspected for deformity, contusions such as the classic 'seat belt' sign and open, possibly penetrating, wounds. A stethoscope is used to auscultate the lungs, comparing left and right apices and bases to identify the loss of breath sounds, indicating a pneumothorax. Feel for tenderness and crepitus due to fractured ribs and sternum, which may also be associated with underlying lung and heart contusions. Percussion can reveal the hyper-resonance of a tension pneumothorax, and the dullness of a haemothorax.

Abdomen The abdomen is inspected for contusions and wounds and auscultated for the absence of bowel sounds indicative of visceral damage. Palpation primarily detects rigidity and tenderness in the conscious patient, and percussion can identify gastric distension, but these are unreliable in many trauma casualties. The early use of specialist imaging such as ultrasound and computed tomography (CT) is indicated. Discrete areas such as the perineum, rectum and vagina should not be forgotten, and must be examined for bleeding, contusions, lacerations, etc.

Pelvis The key indicators of pelvic fracture are unequal leg lengths and pain or crepitus on palpation of the pelvis. If these signs are positive, a pelvic fracture is indicated, with the risk of profound haemorrhage. The examination should not be repeated and the pelvis does not need to be 'sprung', which risks dislodging a clot and causing further bleeding.

Limbs All four limbs are examined for contusions, deformity and pallor. Pain and crepitus on palpation are indicative of underlying fracture or dislocation, and this examination should not be repeated if positive. Distal pallor and absence of pulses suggest a vascular injury, and sensory loss, neurological damage. X-rays that include the joint above and below the injury site are indicated.

Neurology A rapid neurological assessment is carried out to detect lateralizing signs, loss of sensation and motor power, and abnormality of reflexes. Levels of sensory loss should be carefully documented to enable deterioration or improvement to be quantified. X-rays and CT may be indicated to detect spinal fractures.

IMAGING

Imaging techniques are developing rapidly and changing practice.

X-rays Chest and pelvis X-rays may still be performed in the primary survey, but cervical spine radiographs have a high incidence of false-negative results which limits their use. The incidence of spinal cord injury without obvious radiographic abnormality (SCIWORA) is around 10% of all spinal injuries, and is more common in children.

Computed tomography CT scans have in the past had the drawback of sending an unstable casualty for a lengthy procedure in a remote radiology department, which is too dangerous. However, modern spiral CT scanners are fast and, if located adjacent to the Emergency Department, a whole-body trauma CT can be completed in minutes. The patient can be scanned after an initial clinical evaluation, as part of the primary survey, and returned to the resuscitation room for continued management. The risk of patient instability may therefore be outweighed by the benefit of a CT scan in enabling accurate diagnosis, and this technique is becoming a gold standard.

Magnetic resonance imaging MRI is not usually available as an emergency procedure and it is not safe with an unstable casualty. However, its ability to identify soft-tissue injuries is of use in diagnosing SCIWORA; removal of spinal precautions may not be safe until an MRI has excluded unstable spinal injuries.

Ultrasound Scanning is often helpful, particularly for diagnosing intra-abdominal bleeding. Extended focused assessment with sonography in trauma (eFAST) has supplanted diagnostic peritoneal lavage and is also used to detect cardiac tamponade and pneumothorax; however, it will not reliably enable diagnosis of specific visceral injuries. Though it remains a quick and useful Emergency Department adjunct, it does not provide the diagnostic information of CT.

Pain management

Pain management has in the past been underemphasized, due to concerns about masking surgical signs and the risks of sedation and respiratory depression. However, in expert hands, there are various techniques that can be used in the hospital setting.

INTRAVENOUS ANALGESIA

This is the most commonly preferred technique, with morphine being the usual drug. Morphine is a pure agonist opioid and should be diluted and titrated against patient response as there is a wide variation in effect between individuals. It also provides a degree of mental detachment and euphoria useful in the trauma patient, but it has the side effects associated with opioids of respiratory depression, sedation, hypotension, nausea and dysphoria. Being a pure agonist, its effects can be reversed with naloxone. Respiratory depression can be reversed while preserving analgesia with the respiratory stimulant doxapram. Partial agonists such as buprenorphine should be avoided as they are not fully reversed by naloxone. An antiemetic such as cyclizine or ondansetron should be given with morphine to minimize nausea. Fentanyl, a potent, short-acting, rapid-onset synthetic opioid, is increasingly used. Ketamine is useful in patients where preservation of respiratory drive is paramount and in patients where opioid analgesia has not controlled the pain. The analgesic dose is 0.1–0.5 mg/kg.

INHALATIONAL ANALGESIA

Nitrous oxide/oxygen 50:50 mix (Entonox) is useful for short-term analgesia when moving patients or aligning fractures. However, nitrous oxide diffuses into air-filled closed cavities such as a pneumothorax, and it will expand the volume by a factor of four, potentially causing an undrained pneumothorax to tension. Disposable, methoxyflurane inhalers can be used in place of Entonox and can be used safely in the presence of a suspected pneumothorax.

NERVE BLOCKS

Nerve blocks can be used with great effect in some limb injuries, but these should be administered only after discussion with an orthopaedic surgeon due to the risk of masking a compartment syndrome. Femoral nerve blocks are technically straightforward and can be used for midshaft femur, anterior thigh and knee injuries.

Intra-hospital and inter-hospital transfer

Few hospitals enjoy the luxury of having the Emergency Department, radiology, operating theatres and ICUs all in the same location, and so transfer of seriously injured casualties is inevitable at some point. Transfer is indicated when the patient's needs exceed what can be delivered with the resources immediately available. The transfer may be between units within the same hospital, from a small hospital to a larger facility (e.g. to a Level 1/Major Trauma Centre), or to a specialist unit (e.g. burns, neurosurgical or cardiothoracic). Even the shortest transfer within a hospital is fraught with hazard as monitoring and resuscitation are difficult on the move, and so any transfer must be carefully planned. A number of questions should be answered before a transfer is initiated: *When? Where? Who? What way? With?*

WHEN?

When to transfer the casualty is determined by the condition of the individual and the urgency of definitive care. Patient outcome is directly related to time from injury to definitive care, so delays should be minimized. However, transferring partially assessed and unstable patients is dangerous, and so transfer is not usually contemplated or indicated until the primary survey and resuscitation have been completed. Ideally, the patient should be stable when transferred, but this may not be possible (e.g. if bleeding is severe and radiological or surgical control is required). Definitive care may be so urgent that intervention is required before the secondary survey is reached (e.g. for evacuation of an expanding intracerebral bleed). Transfer should not be delayed for investigations such as cervical spine X-ray, which will not change management. However, it is crucial that the ABCs are assessed and addressed; the airway should be secured and protected, the patient must be oxygenated and ventilated optimally, and resuscitation for shock should be commenced even if it cannot be fully addressed without the need for interventions that can only happen following transfer.

WHERE?

Where to transfer the casualty to is determined by the definitive care required and the best facility available that can offer that care. Multiply injured patients may have injuries requiring input from differing surgical specialties such as neurosurgery and general surgery; in this situation, the definitive care surgeons must decide on the priorities, having assessed the patient. The advantage of Level 1/Major Trauma Centres is that the facilities to treat multiply injured patients with multiple systems affected are on the same site. The back of the head should always be examined as injuries at the back of the head may sometimes be missed (Figure 22.10). In life-threatening circumstances (e.g. with expanding intracerebral and intra-abdominal bleeds), the patient may require simultaneous management of both injuries.

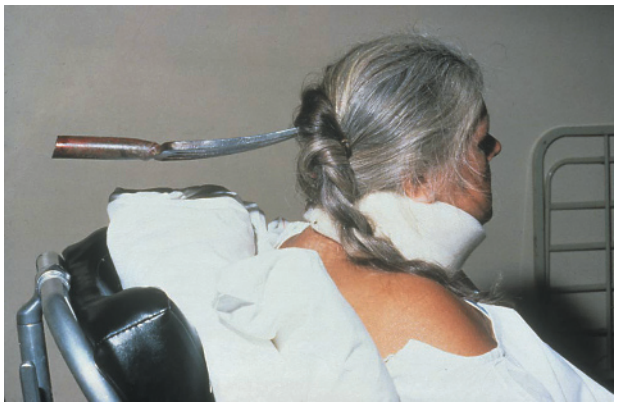


Figure 22.10 The head Failure to examine the back of the head may result in missed injuries!

WHO?

Who conducts the transfer is determined by the staff available. The transferring physician should have an appropriate set of critical care competencies including advanced airway skills – this is not a job for the nearest junior doctor. Transfer should be authorized by the senior doctor with responsibility for the patient, and an appropriate team of doctors, nurses, technicians or paramedics should accompany the patient. The referring doctor should have direct communication with the receiving doctor and transfer team, who should be briefed on the patient's condition, destination, ETA and changes in condition en route.

WHAT WAY?

The way in which the transfer is achieved depends on factors such as whether the transfer is between hospitals or within units of the same facility. The casualty must be secured and spinal stabilization in place if spinal injury cannot be excluded. Spinal boards are not appropriate for transferring patients due to the risk of pressure-induced tissue necrosis, and closely fitting cervical collars can raise intracerebral pressure. The casualty should be transferred on an appropriate trolley with head blocks in place and a medical kit with equipment for ABC interventions must be carried. Full monitoring to include ECG, NIBP/intra-arterial BP, SaO_2 and $EtCO_2$ should be in place. For transfers between hospitals, an appropriate form of transport must be available.

WITH?

The casualty should be transferred with a full set of paperwork to include patient identity and documentation of the full initial assessment; it is particularly important to note whether the secondary survey has been carried out, with any injuries duly noted. If the urgency of the transfer has taken precedence over the secondary survey, then this should be highlighted so that the survey can be completed after the initial, life-saving, definitive care. Results of all blood tests and investigations such as X-rays must accompany the patient.

Definitive care

Definitive care describes the specialist care required to manage the injuries identified during the initial assessment and subsequent investigations. This may be specialist surgery to address a particular problem (e.g. neurosurgical evacuation of an intracerebral bleed), or critical care management on an ICU to provide systemic support (e.g. oxygenation and ventilation of patients with severe lung contusions).

Systemic management

Accurate and effective management of a casualty with multiple injuries depends on a logical progression of examination, moving through the systems in a sequence most likely to identify the most immediately life-threatening injuries first. Using the ARM system described earlier helps structure the approach:

Awareness – Use the history and accident mechanism to predict likely injuries and anticipate problems.

Recognition – Examine the patient logically using the look–listen–feel sequence to identify the physical signs of injury.

Management – Having identified injuries, implement the most effective and life-saving interventions first.

Systemic management may progress simultaneously in a hospital location with a trauma team; in the absence of a team, work through the systems following the ABCDE format. The exception to this would be control of catastrophic haemorrhage preceding airway management – C-ABCDE.

A – Airway and cervical spine

Management of the airway in all forms can be implemented while protecting the cervical spine. Until the airway is both secured and protected, this is best done by in-line immobilization, as use of a stiff cervical collar makes intubation difficult. Conventionally, in-line immobilization is performed with the practitioner standing at the head of the casualty, holding the head on both sides with the hands and maintaining it in a neutral position, in line with the neck and torso. This can make airway management difficult, with the in-line immobilizer squatting awkwardly to one side. An alternative and more effective stance is for the immobilizer to stand to one side of the casualty's shoulder and immobilize the head from below.

Once the airway is secured and protected, the trinity of stiff collar, head blocks and tape should be implemented. However, the routine use of stiff, cervical collars is increasingly being questioned and may cease to be routine. Whatever techniques are used, the cervical spine should be immobilized at all times until an unstable injury is excluded. This may require CT or MRI scanning and may occur after definitive care.

AIRWAY – AWARENESS

Head injury This is by far the most common cause of airway compromise in the trauma patient. As the level of consciousness decreases, so does muscle tone, and the pharynx collapses around the glottis, obstructing the airway. In the supine position, the tongue drops backwards, plugging the glottis anteriorly. Airway obstruction can be sudden or insidious, and partial or complete, but it will result in damaging hypoxia and hypercarbia, which are particularly dangerous in a casualty with a head injury.

Maxillofacial trauma Disruption of the facial bones allows the face to fall back, compressing and obstructing the pharynx. This is associated with soft-tissue swelling and bleeding, which further obtund the airway. Typically, these patients need to sit up to allow the face to fall away from the pharynx and open up the airway (Figure 22.11).

Neck trauma Penetrating or blunt-force trauma results in haemorrhage and swelling, which compresses, distorts and obstructs the upper airway. This can progress rapidly and make tracheal intubation impossible and surgical airway difficult.

Laryngeal trauma Blunt force trauma from impact to the anterior neck (on a car steering wheel, for example) can disrupt the larynx and fracture the cartilaginous structures, leading to immediate or incipient airway obstruction. Signs can be subtle; contusion over the larynx with a hoarse voice, coughing of bright red blood and surgical emphysema should alert the practitioner to the likelihood of sudden airway obstruction. Around 3% of chest traumas are associated with laryngeal injury.

Inhalational burns Inhaling super-heated air burns the airway and can result in rapid development of swelling and airway obstruction. Signs such as facial burns, smoke staining and singed nasal hair suggest an inhalational burn, requiring early and expert intubation.

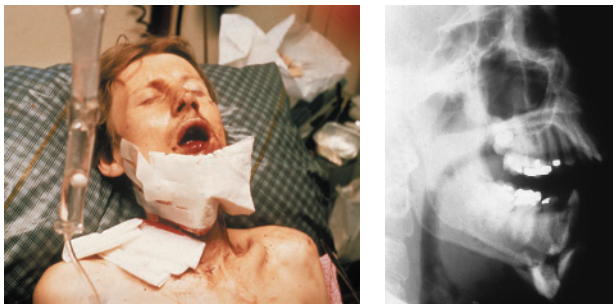


Figure 22.11 Mandibular fracture Patient with a mandibular fracture showing the characteristic position to maintain the airway.

AIRWAY – RECOGNITION

Airway obstruction and respiratory failure may be obvious (to an experienced clinician), but early signs can sometimes be subtle and need systematic examination to detect:

Look

- Agitation, aggression, anxiety – suggest hypoxia.
- Obtunded conscious level – suggests hypercarbia.
- Cyanosis – blue discoloration of nail beds and lips caused by hypoxaemia due to inadequate oxygenation.
- Sweating – increased autonomic activity.
- Use of accessory muscles of ventilation; casualty classically sitting forward splinting chest and using neck and shoulder muscles to aid breathing. May also display flared nostrils.
- Tracheal tug and intercostal retraction – caused by exaggerated intrathoracic pressure swings.

Listen

- Noisy breathing – collapsing pharyngeal muscles obstruct airway leading to snoring sounds.
- Stridor – air flow through an obstructing upper airway changes from laminar to turbulent, resulting in the typical hoarse wheeze of stridor – a sinister sign, as even minimal further reduction in the airway lumen can result in critical airway obstruction.
- Hoarse voice (dysphonia) – functional damage to larynx.
- Absence of noise – may indicate complete airway obstruction or apnoea.

Feel

- Feel for passage of air through mouth and nose with palm of hand; very sensitive for detecting air flow.
- Palpation of the trachea in the suprasternal notch will detect the deviation associated with a tension pneumothorax.

AIRWAY – MANAGEMENT

A range of manoeuvres is available to secure a patent airway, ranging from 'bare hands' techniques to a surgical airway. All these techniques can be performed without extending the head and potentially compromising an unstable cervical spine. The classical

anaesthetic 'sniffing the early morning air' position (head extended and neck flexed) should not be used in the trauma patient. Bare hands techniques and the use of pharyngeal airways are used together to pull the pharyngeal tissues and tongue off the posterior pharyngeal wall and away from the glottis, opening up the airway.

Supraglottic airway devices provide more reliable airway maintenance, but only intubation or the surgical airway will provide a definitive airway that is both secured and protected.

All the non-surgical airway manoeuvres described are applicable to children, but they require some modification in technique to accommodate their anatomical and physiological differences. Surgical airways are not recommended in children under 12 years of age, as the cricoid cartilage can be damaged, leading to tracheal collapse.

Chin lift The chin is lifted forwards with the practitioner positioned at the casualty's head or side, using one hand. This pulls the jaw and pharyngeal structures forward off the posterior pharyngeal wall and glottis, and opens up the airway (Figure 22.12).

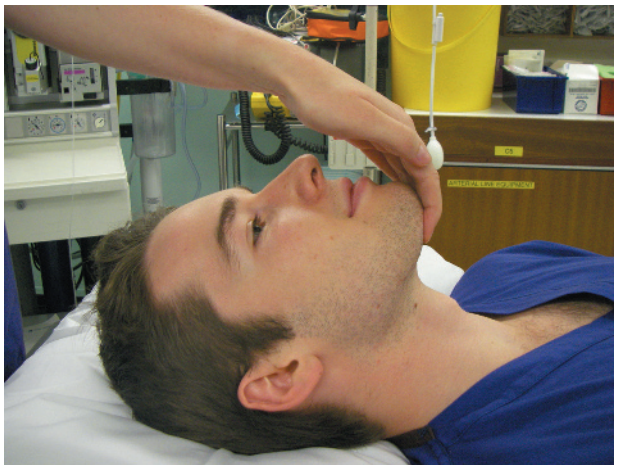


Figure 22.12 Chin lift

Jaw thrust This is a more assertive manoeuvre that is effective in patients with small jaws or thick necks, or who are edentulous. From the casualty's head, the thenar eminences are rested on the casualty's maxillae (assuming no obvious fracture), and the four fingers positioned under the angles of the mandible. Using the thenar eminences to provide a counterpoint on the maxillae, the mandible is lifted up and forwards to open up the airway as with the chin lift. Considerable pressure can be exerted without displacing the head on the neck, and the manoeuvre can be combined with application of a BVM assembly for ventilation of the lungs (Figures 22.13 and 22.14).

Release of chin lift and jaw thrust almost inevitably results in loss of the airway, and progression to airway adjuncts will be required to free up the practitioner.

Oropharyngeal (OP) airway The OP airway is a curved and flattened, hard, plastic tube with a proximal flange, which is shaped and sized to hold the tongue and pharynx off the posterior pharyngeal wall (Figure 22.15). They are available in a range of sizes from neonate to large adult (Figure 22.16); selection of the correct size is important, as the pharyngeal tissues will collapse across the end of too small a device, while one too large will risk impinging on the glottis. The correct size is selected by lining up the OP airway alongside the patient's jaw; the flange to tip length of the OP airway should match the distance from the corner of the patient's mouth to the external auditory canal.

The OP airway is inserted above the tongue, initially with the concave aspect upwards. As the tip passes over the tongue, the OP airway is rotated so the concave aspect slides over the tongue, and slipped into the pharynx until the flange rests on the incisors.

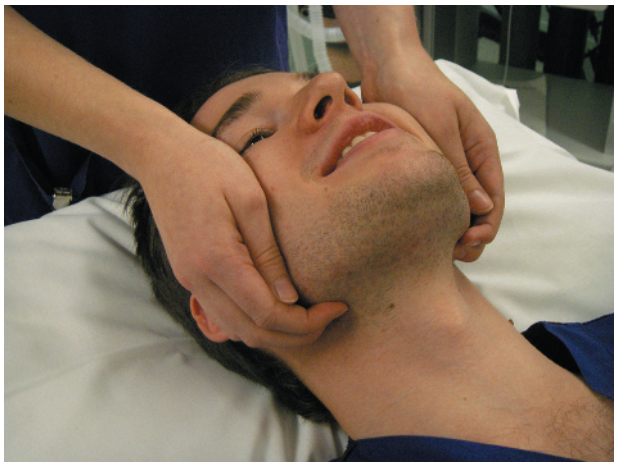


Figure 22.13 Jaw thrust

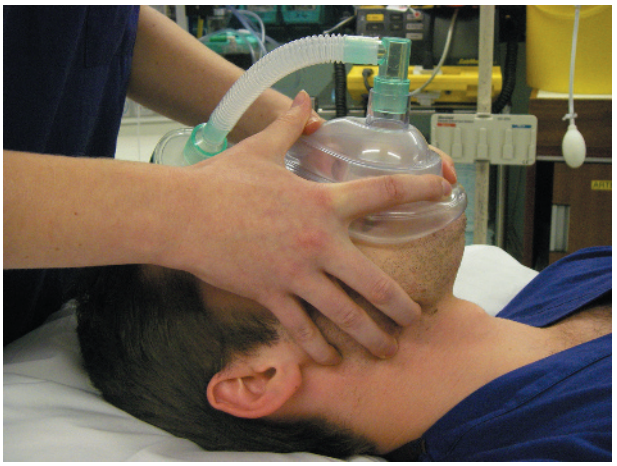
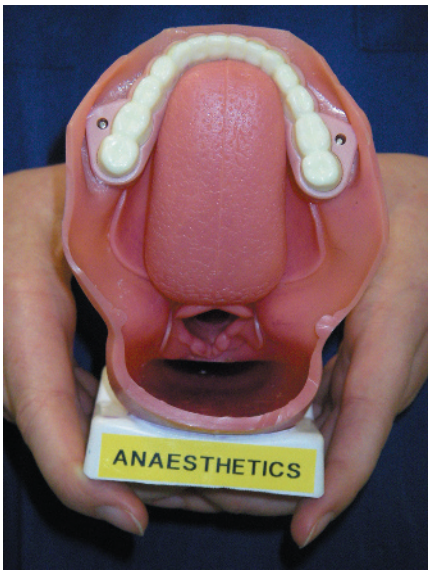


Figure 22.14 Jaw thrust with O_2 mask



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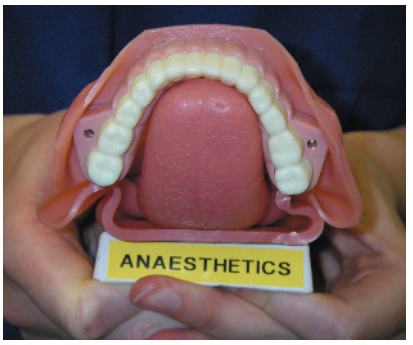

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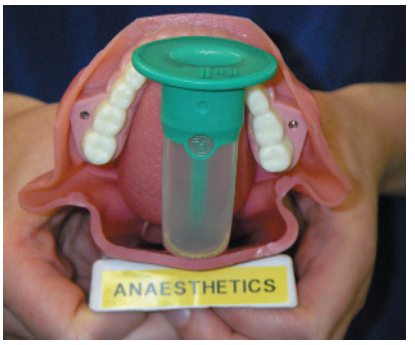


(c)

Figure 22.15 Pharyngeal airways preventing the tongue from falling back across the glottis (a) Open airway. (b) Obstructed airway – collapse of pharynx and tongue across glottis. (c) Airway secured with oropharyngeal airway.

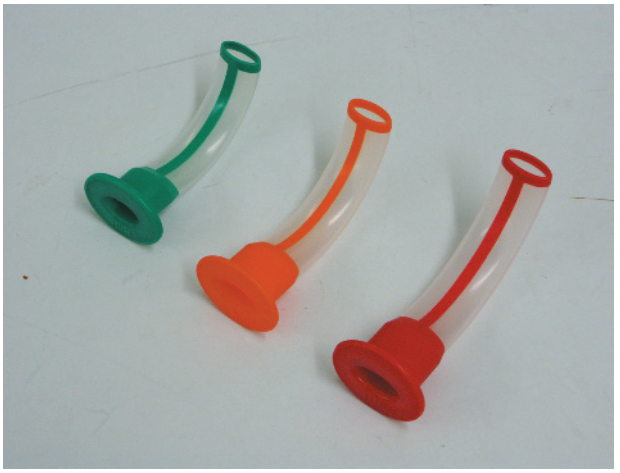


Figure 22.16 Oropharyngeal airway (Guedel)

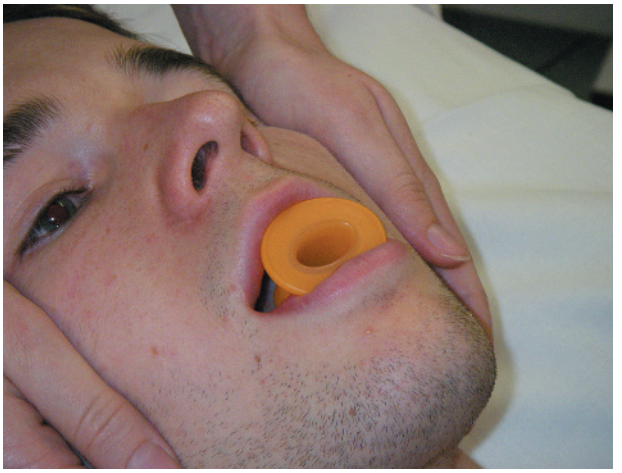


Figure 22.17 Oropharyngeal airway – correct position

A correctly sized OP airway should neither project up beyond the teeth nor disappear into the buccal cavity (Figure 22.17).

Use of the OP airway may need to be combined with chin lift or jaw thrust to maintain a patent airway, as they should only be used in obtunded patients with absent gag reflexes.

Nasopharyngeal (NP) airway The NP airway is a soft plastic tube with a smooth, distal bevel and a proximal flange. Some makes have a safety pin to insert through the flange to prevent the NP airway disappearing into the nose. It is supplied in a number of internal diameter sizes, and should be selected according to the approximate size of the casualty's little finger (Figure 22.18). The NP airway is lubricated with aqueous jelly, and inserted along the floor of the nasal cavity into the nasopharynx (Figure 22.19). The NP airway should not be inserted up the nose as this risks haemorrhage from the mucosa and turbinates, further compromising the airway, and also introduces the possibility of entering the cranial cavity through a basal skull fracture.

NP airways are particularly useful as they can be tolerated by responsive casualties with obstructing airways. They also provide access to suction the nasopharynx with a soft suction catheter.

Oropharyngeal suction Secretions and blood should be cleared with a specialist pharyngeal sucker such as the Yankauer. Care should be taken not to damage the soft tissues and, as a general rule, the sucker should not be passed further than can be seen. Suction of the oronasopharynx with a Yankauer sucker, under direct vision using a laryngoscope, is effective in the obtunded patient.

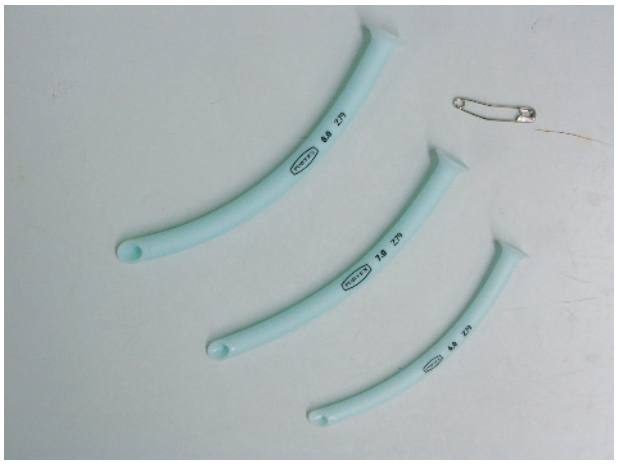


Figure 22.18 Nasopharyngeal airway

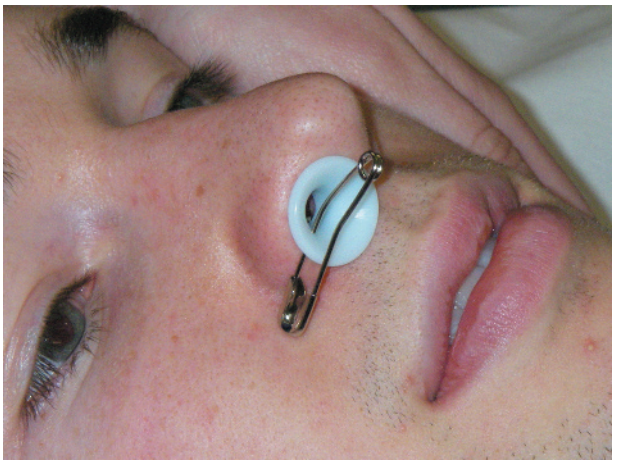


Figure 22.19 Nasopharyngeal airway – correct position

Supraglottic airway devices These are devices that function between an OP airway and a tracheal tube; contemporary use is largely limited to the laryngeal mask airway (LMA) or second-generation devices such as the i-gel®. Since their introduction in the 1980s, the supraglottic airway has found an international role for resuscitation and trauma airway management, with the advantages that it is more effective than other airway devices but does not require the skill and training required for successful tracheal intubation. Consideration of their use is recommended in the UK NICE 2016 trauma guidelines.

International evidence suggests that intubation performed by practitioners without anaesthetic training can be detrimental to patient survival and in the UK the ambulance service regulatory body (Joint Royal Colleges Ambulance Service Liaison Committee, JRCALC) has removed tracheal intubation as a core paramedic skill, instead recommending the use of supraglottic airway devices.

The LMA is available in a range of sizes from neonatal to large adult; for adult use, a size 3 will fit small women, size 4 larger women and smaller men, and size 5 larger men. The device consists of a cuffed distal portion shaped to fit into the oropharynx over the glottis (Figure 22.20). The cuff is inflated with air to fit snugly against the pharynx, but it does not seal as does a tracheal tube cuff, and hence it does not reliably protect the airway. The LMA is lubricated and inserted over the tongue with the open end of the cuffed distal portion positioned anteriorly. The device is slipped around the oropharynx until it is snugly positioned over the glottis, and the cuff inflated according to the size of the device (#3 20 mL; #4 30 mL; #5 40 mL).

Second-generation supraglottic airway devices such as the i-gel® are increasingly used in preference to the LMA. They have the advantage of having an integrated, gel-filled, supraglottic cushion which does not require inflation. The device incorporates a bite block and has a distal tip orifice which sits above the oesophagus and enables passage of a suction tube.

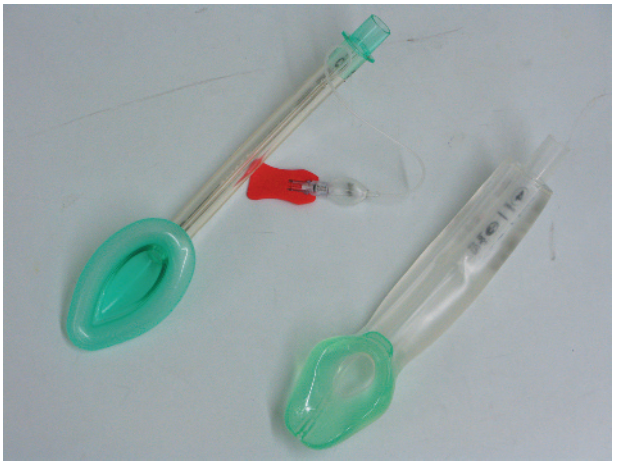


Figure 22.20 Supraglottic airways

As a supraglottic airway device does not provide a definitive and protected airway, consideration should be given to its being replaced with a tracheal tube at the earliest opportunity when appropriately trained and skilled practitioners are available.

Tracheal intubation Orotracheal intubation is the preferred method for securing and protecting the compromised airway in the trauma patient. However, it is a difficult procedure with minimal survival rates in unanaesthetized, trauma casualties; unanaesthetized casualties can normally only be intubated when protective reflexes are absent, allowing a view of the vocal cords on laryngoscopy. Lack of reflexes to this degree is associated with terminally deep levels of coma, when casualties are at the point of death.

Casualties requiring a definitive airway should therefore be identified early, and expert assistance sought from an anaesthetist or critical care physician. The indications for orotracheal intubation are:

- apnoea
- inability to maintain the airway by other means
- need to protect the airway from aspiration of blood and stomach contents
- impending airway obstruction, e.g. inhalational burn, expanding neck haematoma, facial fractures
- closed head injury with GCS below 8
- inability to maintain adequate oxygenation and ventilation with a face mask or BVM assembly.

Nasotracheal intubation has a poor success rate with a high incidence of complications such as nasal haemorrhage and is no longer routinely recommended.

Trauma tracheal intubation should be performed with a rapid sequence induction (RSI) anaesthetic; after pre-oxygenation, anaesthesia is rapidly induced with an intravenous agent, cricoid pressure applied to hold the oesophagus closed and prevent passive reflux of stomach contents, the patient paralyzed with a rapid-onset muscle relaxant and a tracheal tube placed under direct vision with use of a laryngoscope. The tracheal tube cuff is inflated until no leak is detected, and the cricoid pressure is not released until the anaesthetist confirms the tracheal tube is secure. Intubating bougies should be routinely used in anticipation of a difficult intubation, and specialist laryngoscopes such as the elevating tip McCoy should be available. Tracheal placement of the tube is confirmed with capnography.

A variety of disposable and reusable videolaryngoscopes are now available and can be used in patients predicted to have a difficult airway.

This procedure should not be performed by any practitioner without the necessary training and experience in anaesthetic techniques, as injudicious use of muscle relaxants can lead to immediate loss of the airway and a 'can't intubate, can't ventilate' scenario. Non-RSI trained practitioners should use second-generation supraglottic airways and not attempt drug-assisted, tracheal intubation.

All intubated trauma patients should be ventilated, as it is unlikely that they would be able to maintain adequate oxygenation and ventilation spontaneously.

Needle cricothyroidotomy Needle cricothyroidotomy is the insertion of a needle through the cricothyroid membrane into the trachea to allow jet insufflation of the lungs with oxygen. It is used in emergency 'can't intubate, can't ventilate' situations to buy time while expert assistance is sought or a definitive surgical airway prepared. Oxygenation is achievable but ventilation limited, so carbon dioxide accumulates and the $EtCO_2$ rises.

Complications of needle cricothyroidotomy and jet insufflation are commonly misplacement, surgical emphysema and barotrauma. It should be attempted only if intubation and other airway maintenance techniques have failed, but it has increasingly been supplanted by immediate surgical airway. It is recommended for use in young children, where surgical airway is contraindicated.

Surgical airway Surgical airway is the insertion of a tracheal or tracheostomy tube, through an incision in the cricothyroid membrane, into the trachea. It is used in emergency situations when orotracheal intubation has been attempted and failed and will both secure and protect the airway. Adequate ventilation is just as achievable as with orotracheal intubation, and 100% oxygen can be delivered. Once the 'can't intubate, can't ventilate' situation has been identified, surgical airway should be performed as an emergency procedure. The preferred technique is to insert a small bore (e.g. 6 mm) tracheal tube over an intubating bougie into the trachea, through a transverse incision in the cricothyroid membrane.

A surgical airway can be a difficult procedure in casualties with challenging anatomy, and complications can be serious; this procedure should be used only if orotracheal intubation and supraglottic airway have been attempted and failed. Complications include haemorrhage, damage to laryngeal structures, false passage formation, misplacement of the tracheal tube, surgical emphysema and barotrauma.

AIRWAY – TAKE-HOME MESSAGE

Author/proof-reader – colour for summary Take-home message heading is intentional!

- Whatever the means of airway management used, the goal is to secure and protect the airway.
- The focus should be on oxygenation and ventilation, not intubation.
- Casualties die from hypoxia and hypercarbia, not failure of intubation.

B – Breathing and chest injuries

Of severely injured patients admitted to hospital in the UK, JRCALC data indicates that 20% have chest injuries and thoracic trauma is a significant cause of mortality. However, the majority of chest injuries are not fatal and do not require specialist surgical intervention.

BREATHING/CHEST INJURY – AWARENESS

The proportion of penetrating to blunt chest injuries varies between countries, and between rural and urban environments. Only 10% of blunt chest injuries and 20% of penetrating injuries require thoracotomy. Non-surgical management centres on supportive treatment of contused lungs and the insertion of

chest drains. However, with blunt trauma, the force of impact and energy transfer to the lung parenchyma should alert the clinician to the likelihood of severe intrathoracic damage and the potential for progressive cardiopulmonary problems.

Early recognition and management of immediately life-threatening injuries in the primary survey is imperative, with early imaging repeated as necessary. Potentially life-threatening injuries are sought during the secondary survey, and sophisticated imaging modalities such as CT and MRI may be indicated. Major chest injuries will require urgent referral to a specialist thoracic or cardiothoracic surgeon and a surgeon capable of immediate thoracotomy must be available in hospitals designated as receiving major trauma cases.

BREATHING/CHEST INJURY – RECOGNITION

The patient's chest, neck and abdomen must be fully exposed to allow assessment of the chest. Examination should be systematic:

Look

- Respiratory rate – tachypnoea is indicative of hypoxia.
- Shallow, gasping or laboured breathing – suggests respiratory failure.
- Cyanosis – indicates hypoxia.
- Plethora and petechiae – suggest asphyxia and chest crushing.
- Paradoxical respiration; 'pendulum' breathing with asynchronization between chest and abdomen, resulting in a see-saw motion – indicates respiratory failure or structural damage.
- Unequal chest inflation – suggestive of pneumothorax or flail chest.
- Bruising and contusions – indicate significant energy transfer and consequent underlying lung contusion and potential hypoxia (e.g. 'seat belt' sign).
- Penetrating chest injuries – potential for pneumothorax and open, sucking pneumothorax.
- Distended neck veins – increased venous pressure secondary to a tension pneumothorax or cardiac tamponade.

Listen

- Absence of breath sounds – indicates apnoea or tension pneumothorax.
- Noisy breathing/crepitations/stridor/wheeze – suggests a partially obstructed airway, blood and secretions in airways, tracheal or bronchial damage.
- Reduced air entry unilaterally – indicates a pneumothorax, haemothorax or haemopneumothorax, and flail chest.

Feel

- Tracheal deviation – indicative of tension pneumothorax, shifting the mediastinum. (**Note:** the trachea is felt inferiorly in the suprasternal notch; do not confuse it with the larynx, which is extrathoracic and hence does not shift.)
- Tenderness – suggests significant chest wall contusion and/or fractured ribs.
- Crepitus/instability – indicates underlying fractured ribs.
- Surgical emphysema (crackling and popping, 'bubble wrap', crepitus feel to subcutaneous tissues on palpation, due to presence of air forced into tissues under pressure) – tension pneumothorax, ruptured bronchi or trachea and fractured larynx.

BREATHING/CHEST INJURY – MANAGEMENT

Immediate management is to stabilize the cervical spine, control catastrophic limb haemorrhage, secure the airway, administer oxygen at high flow and ventilate the lungs if breathing is absent or inadequate. It is vital to rapidly identify and manage immediately life-threatening chest injuries during the primary survey (Box 22.3), as positive-pressure ventilation of the lungs can cause a rapid deterioration; a simple pneumothorax can be converted to a tension pneumothorax, and a tension pneumothorax will increase in pressure, leading to sudden collapse and cardiac arrest. Hence, if a patient is intubated and ventilated, signs of a pneumothorax must immediately be sought and, if present, decompressed and drained. Potentially life-threatening injuries can then be identified during the secondary survey (see Box 22.4).

TENSION PNEUMOTHORAX

A tension pneumothorax is the build-up of air under pressure in the pleural cavity, leading to compression and collapse of the underlying lung. The resultant ventilation–perfusion mismatch leads to hypoxia. However, the life-threatening, terminal event is a shift of the mediastinum away from the affected side, kinking the great vessels and obstructing venous return to the heart. This results in a deadly combination of hypoxia and loss of cardiac output, with a pulseless electrical activity (PEA) cardiac arrest.

BOX 22.3 IMMEDIATELY LIFE-THREATENING CHEST INJURIES (PRIMARY SURVEY)

1 Tension pneumothorax
2 Open pneumothorax (sucking chest wound)
3 Massive haemothorax
4 Cardiac tamponade
5 Flail chest
6 Disruption of tracheobronchial tree

Diagnosis should usually be clinical, not radiological, if the casualty is unstable and the clinician should look specifically for the three cardinal signs:

- absent breath sounds – on the side of the pneumothorax
- deviated trachea – away from the side of the tension pneumothorax
- hyper-resonance – on the side of the pneumothorax.

Surgical emphysema is an additional sign, highly suggestive of a tension pneumothorax or tracheobronchial tree injury. The neck veins may also be distended, as venous return is obstructed; however, this may not be readily visible, and it is unreliable with concurrent hypovolaemia. There is an argument for radiological diagnosis if this is immediately available in the resuscitation room and the patient is not exhibiting cardiovascular compromise (Figure 22.21); a tension pneumothorax can be mimicked by other conditions such as endobronchial intubation with distal lung collapse.

Immediate management has classically been decompression (needle thoracocentesis) of the tensioning pneumothorax by insertion of a 14-gauge cannula into the pleural cavity through the second intercostal space, in the midclavicular line. However, this is unreliable, and the relatively short 50 mm intravenous cannulae commonly used may not penetrate a thick chest wall in muscular or obese casualties; the use of the lateral approach is becoming a preferred option. The 2016 NICE guidelines recommend the use of open thoracostomy rather than needle decompression, if expertise is available.

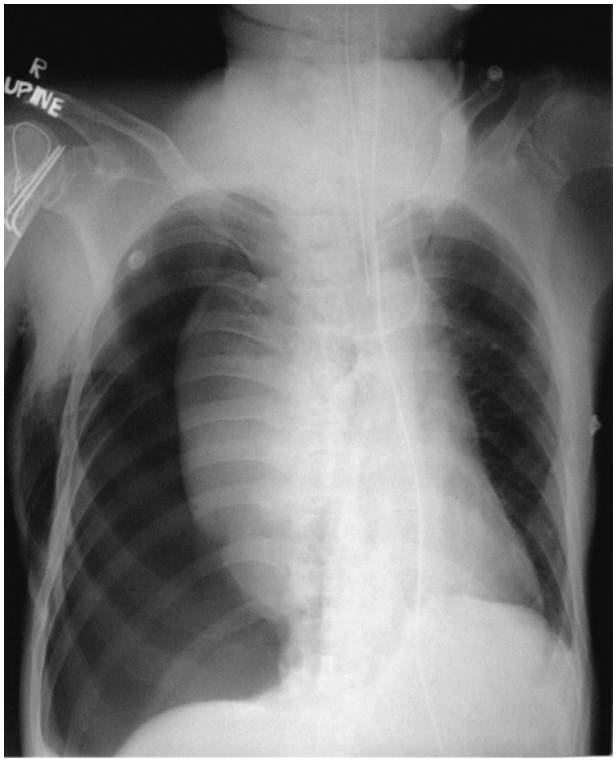


Figure 22.21 Right-sided tension pneumothorax

Needle decompression or open thoracostomy should not be performed if the only sign elicited is reduced or absent breath sounds, as there are associated complications such as misplacement and damage to the underlying lung. If indicated, these manoeuvres will convert a tension pneumothorax into a simple pneumothorax, which will in turn need draining to allow the lung to reinflate. Neither technique should be performed bilaterally in spontaneously breathing patients as both lungs may collapse; if the patient has been intubated and is ventilated, bilateral decompression is acceptable, as the positive pressure will enable the lungs to be ventilated.

OPEN PNEUMOTHORAX (SUCKING CHEST WOUND)

An open wound in the chest wall will immediately result in a simple pneumothorax as intrathoracic pressure equilibrates with atmospheric pressure. If the defect is greater than some two-thirds of the diameter of the trachea (which has a diameter of 20–25 mm), air is preferentially drawn into the pleural cavity rather than into the lungs via the trachea. This causes paradoxical respiration, where the lung deflates on inspiration, with resulting hypoventilation and hypoxia. If a flap valve effect occurs, the intrapleural pressure will rise with each breath, leading to a tension pneumothorax.

Specific immediate management is the application of an occlusive dressing or specialist valved dressing. A chest drain should be sited at the earliest opportunity and the patient may need intubating and ventilating.

MASSIVE HAEMOTHORAX

The chest cavity presents an enormous space in which blood can potentially accumulate following both blunt and penetrating chest injury (one of the four of 'bleeding onto the floor and four more'). Around one-third of the patient's blood volume (1500 mL) can rapidly accumulate, leading to a combination of hypoxia and shock. Smaller haemothoraces are usually due to lung parenchymal tears, fractured ribs and/or minor venous injuries and are self-limiting. Massive bleeds are usually due to arterial damage, which is more likely to require surgical repair and/or pulmonary lobectomy.

Diagnosis is based on the presence of hypoxia, reduced chest expansion, absent breath sounds and/or dullness to chest percussion, and hypovolaemic shock. Supine chest percussion may not demonstrate dullness, and supine X-rays may not reveal moderate haemothoraces.

Management is by open thoracostomy followed by insertion of a chest drain, correction of hypovolaemia, tranexamic acid and blood transfusion. If the

total volume of blood initially drained is greater than 1500 mL, the bleeding continues at 200 mL/hour, or the patient remains haemodynamically unstable, surgical referral and thoracotomy are indicated. The signs can be confused with a traumatic, diaphragmatic hernia on the left and so great care must be taken with thoracostomies and chest drains.

Smaller volumes of blood causing haemothorax are more difficult to diagnose and may require eFAST or CT imaging. Treatment is with a chest drain.

CARDIAC TAMPONADE

Cardiac tamponade is the accumulation of blood within the pericardium, restricting the ability of the heart to fill, and resulting in a progressive loss of cardiac output leading to PEA cardiac arrest. It is associated with penetrating rather than blunt trauma, especially stab wounds between the nipple lines or scapulae, and gunshot wounds.

Clinical *diagnosis* can be difficult, as the signs can be subtle and difficult to elicit in the trauma room. The three classic diagnostic criteria constitute Beck's Triad:

1 distended neck veins due to elevated venous pressure
2 muffled heart sounds
3 fall in arterial blood pressure.

Reliable diagnosis may require sophisticated imaging. No change is seen on standard chest X-rays, but CT scanning, MRI scanning, eFAST and transoesophageal echocardiogram (TOE) can all be used to confirm the diagnosis.

Management has two components: relieving the pressure within the pericardium by draining the accumulated blood, and stopping the source of the bleeding to prevent reaccumulation. Since the bleeding is likely to come from the heart, immediate surgical repair to the myocardium may be required, and surgical assistance should be sought early.

Classically, aspiration of blood from the pericardium has been achieved by needle pericardiocentesis, which should be viewed as a diagnostic procedure rather than curative. This technique may not be effective as blood clots cannot be aspirated and pericardotomy via a clamshell thoracotomy is becoming the definitive technique for managing suspected cardiac tamponade following penetrating chest trauma and witnessed cardiac arrest associated with blunt chest trauma.

FLAIL CHEST

Massive impact to the chest wall can result in multiple rib fractures, and this is more common in older people who have less flexible rib cages. The multiple fractures, particularly if anterior and posterior, can result in a loss of the structural integrity of the chest wall, and a segment can 'float'; as the patient inspires, the flail segment is sucked in and the lung cannot inflate (paradoxical respiration). This results in hypoxia and ventilatory compromise. However, the force required to cause this injury inevitably causes a severe, underlying lung contusion, and this is often the more significant cause of the hypoxia. The associated severe pain further compromises the respiratory function, and respiratory failure can ensue.

Diagnosis is by clinical examination, chest X-rays to reveal the fractures and lung contusion, and arterial blood gases to quantify the hypoxia.

Management is initially supportive with administration of oxygen and analgesia. Advanced pain-relieving methods such as epidurals may be required. Profound hypoxia may require that patients are intubated and ventilated until the contusion has adequately resolved and pain can be controlled. Intravenous fluids may need to be restricted to avoid overload and worsening hypoxia. There is a move towards fixation of fractured ribs or costochondral disruption that causes respiratory compromise. This is currently usually performed in the post-acute phase if there is difficulty weaning the patient from supportive ventilation in order to improve and speed up rehabilitation.

DISRUPTION OF TRACHEOBRONCHIAL TREE

Major disruption of the tracheobronchial tree can result in a bronchopleural fistula; the disrupted trachea or bronchus allows an air leak into the pleura which, if large enough, will not allow inflation of the lung, even with a large-bore chest drain *in situ*.

Diagnosis is made by the presence of a persistent pneumothorax, pneumomediastinum, pneumopericardium or air below the deep fascia of the neck, often in patients who have suffered a deceleration injury.

Immediate management with tracheal intubation may not be successful, as the air leak may prevent inflation of either lung. In this situation, endobronchial intubation of the opposite lung or use of a bronchial blocker may be required before adequate lung ventilation can be achieved, and this may need the services of a thoracic anaesthetist.

BOX 22.4 POTENTIALLY LIFE-THREATENING CHEST INJURIES (SECONDARY SURVEY)

1 Simple pneumothorax
2 Haemothorax
3 Pulmonary contusion
4 Tracheobronchial tree injury
5 Blunt cardiac injury
6 Traumatic aortic disruption
7 Traumatic diaphragmatic injury
8 Mediastinal traversing wounds

SIMPLE PNEUMOTHORAX

A simple pneumothorax results from air entering the pleural cavity, causing collapse of the lung with resulting ventilation–perfusion mismatch and hypoxia. As the air is at atmospheric pressure, and there is no one-way valve effect, no mediastinal shift develops, and cardiac output is maintained. The cause is usually a lung laceration, which can follow both blunt and penetrating chest trauma or thoracic spine fracture-dislocations.

Diagnosis is made during the primary or secondary survey, primarily by the absence or reduction of breath sounds. Hyper-resonance may not be demonstrated and a chest X-ray may be required to confirm the pneumothorax. If the pneumothorax is stable, definitive treatment with a chest drain can be deferred to the secondary survey. However, a simple pneumothorax can develop into a tension pneumothorax at any time, and so a high index of suspicion should be maintained.

Intubation and ventilation in the presence of a pneumothorax predisposes to the development of a tension pneumothorax and so chest drains should immediately be placed. Anaesthesia with a nitrous oxide-based anaesthetic will increase the air space by a factor of four, and can therefore cause rapid tensioning, as can air transport at altitude. In these situations, chest drains should be placed prophylactically.

Chest drain insertion is a procedure with the potentially dangerous complication of visceral damage, and trochars should not be used for the introduction of chest drains in this setting. The appropriate technique is:

1 Confirm the correct side on the chest X-ray.
2 Identify the fifth intercostal space, just anterior to the midaxillary line on the affected side.
3 Prepare the skin with 2% chlorhexidine in 705 isopropyl alcohol or alcoholic iodine.
4 Infiltrate the skin and subcutaneous tissues with lignocaine if the patient is aware.
5 Make a 2–3 cm horizontal incision through the skin, just above the sixth rib (to avoid the intercostal vessels below the fifth rib).
6 Bluntly dissect through the subcutaneous tissues with straight forceps, and puncture the parietal pleura with the tips.
7 Insert your gloved little finger through the incision into the chest cavity and sweep the finger around to ensure the cavity is empty and your incision is above the diaphragm (no viscus is felt).
8 Grasp the tip of an appropriately sized thoracostomy tube between the tips of the forceps and introduce through the incision into the chest cavity; unclamp the forceps and slide the tube posteriorly along the inside of the chest wall.
9 Attach the tube to an underwater drain or Heimlich valve and observe for tube fogging and underwater bubbling.
10 Suture the chest drain in place and apply a dressing.
11 Check lung reinflation with a chest X-ray.

The important steps are illustrated in Figure 22.22.

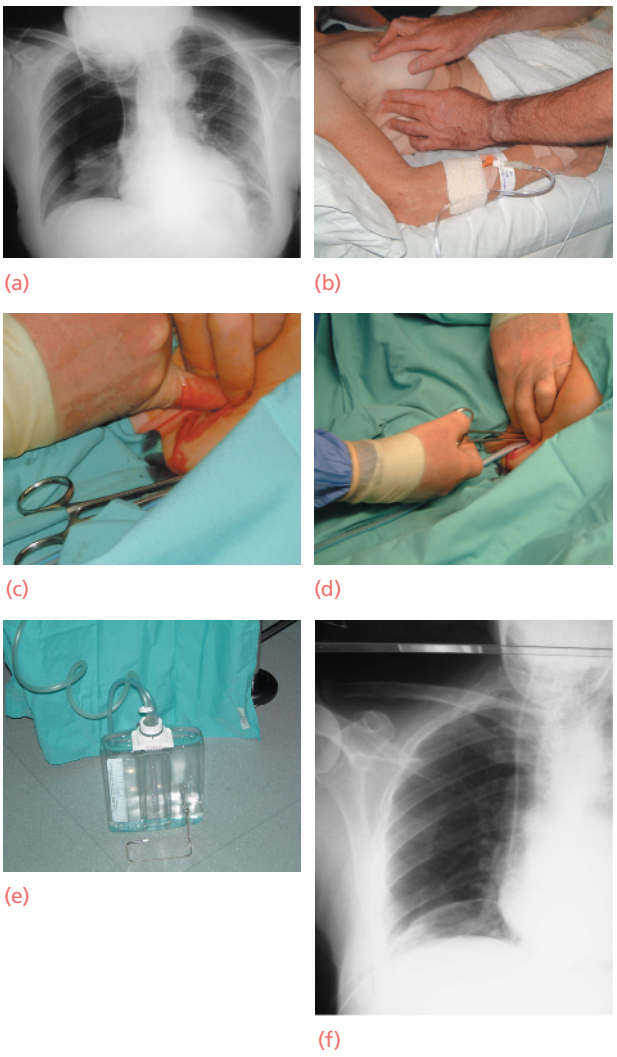


Figure 22.22 Chest drain insertion sequence (a) Chest X-ray to confirm the correct side. (b) Identify the fifth intercostal space, just anterior to the midaxillary line on the affected side. (c) Insert a gloved little finger through the incision into the chest cavity and finger-sweep to ensure the cavity is empty and the incision is above the diaphragm (no viscus is felt). (d) Grasp the tip of an appropriately sized thoracostomy tube between the tips of the forceps and introduce it through the incision into the chest cavity. Unclamp the forceps and slide the tube posteriorly along the inside of the chest wall. (e) Attach the tube to an underwater drain or Heimlich valve and observe for tube fogging and underwater bubbling. (f) Check lung reinflation with chest X-ray.

HAEMOTHORAX

Haemothoraces are primarily caused by lung lacerations or damage to intercostal and internal mammary vessels. They are normally self-limiting and rarely require operative intervention. Diagnosis can be difficult, as clinical signs may not be present and chest X-rays unreliable. The diagnosis is often coincidental with chest drain insertion for suspected pneumothorax, but most commonly on FAST or CT scanning. Treatment is with a large-calibre, intercostal drain; if more than 1500 mL of blood are drained initially, or drainage continues at 200 mL/hour or faster, thoracoscopy should be considered.

PULMONARY CONTUSION

Pulmonary contusion is the commonest potentially life-threatening chest injury, occurring in 20% of casualties with an injury severity score (ISS) of >15. Mortality is in the range of 15–20%, and 40–60% of patients will require ventilating. Blunt force trauma to the chest wall, or crushing injury, will contuse the underlying lung, which then becomes oedematous and haemorrhagic, with subsequent collapse and consolidation. This causes a ventilation–perfusion mismatch and hypoxia, dependent on the extent of the contusion and limitation of the patient's ventilation by pain. About half of these patients will develop bilateral acute respiratory distress syndrome (ARDS), a systemic inflammatory response to the injury.

Pulmonary contusion may not be associated with obvious rib fractures, particularly in children and teenagers with pliable rib cages. The initial chest X-ray may not reveal the extent of the contusion, which can develop over the following 48 hours. The *diagnosis* should be made taking into account the mechanism of injury and the degree of hypoxia revealed by oximeter saturation readings and arterial blood gas estimations.

Treatment is with supportive measures and oxygen administration. Patients with severe hypoxia despite inspired oxygen (e.g. PaO_2 < 8.5 kPa or SaO_2 < 90%) should be considered for elective ventilation. Pre-existing pulmonary disease should be taken into account.

TRACHEOBRONCHIAL TREE

Tracheobronchial tree injuries are rare but can easily be overlooked, as signs can be subtle. Some 3% of chest-crushing injuries are associated with upper airway injuries, but most tracheobronchial tree injuries are within 2 cm of the carina. Patients frequently present with haemoptysis, surgical emphysema and a simple or tension pneumothorax. The pneumothorax may be resistant to reinflation with a chest drain, and a post-drain and persistent air leak suggests the presence of a bronchopleural fistula. CT and MRI imaging may confirm the diagnosis, but bronchoscopy may be required.

Treatment is initially with one or more large chest drains that may need a high-volume/low-pressure pump to allow lung reinflation. Persistent bronchopleural fistulae may require operative intervention. Major tracheobronchial injuries are immediately life-threatening, and management is described above.

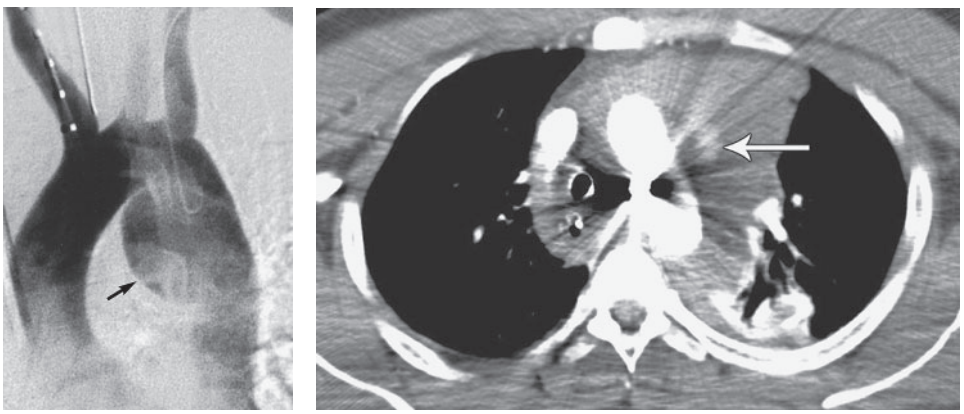
BLUNT CARDIAC INJURY

Blunt cardiac injury follows a direct blow to the anterior chest and is associated with a fractured sternum. This can result in myocardial contusion or, more rarely, chamber rupture and valvular disruption. The myocardial damage can result in hypotension due to myocardial dysfunction, conduction abnormalities, and dysrhythmias. Sudden onset of dysrhythmias can result in death from ventricular fibrillation.

Management is supportive, and the patient should be monitored closely for a minimum of 24 hours, following which the risk of sudden dysrhythmias diminishes substantially.

TRAUMATIC AORTIC DISRUPTION

Blunt aortic injury is a deceleration injury commonly following high-speed road traffic crashes (RTCs) and falls from a height. Up to 15% of deaths from road vehicle collisions are a result of damage to the thoracic aorta. Most injuries occur in the proximal thoracic aorta, where the relatively mobile aortic arch can move against the fixed descending aorta near the ligamentum arteriosum (Figure 22.23).



(a) (b)

Figure 22.23 Ruptured aorta (a) Angiogram showing a rupture of the arch of the aorta. (b) CT scan showing the haematoma around the rupture.

Complete transection or rupture is immediately fatal, but the haematoma can be contained by the adventitial layer of the aortic wall, enabling the patient to survive to reach hospital.

Specific clinical signs and symptoms are often absent, and the mechanism of injury should provoke a high index of suspicion. *Diagnosis* is aided by chest X-ray findings, classically of a widened mediastinum (note that an anteroposterior (AP) film will magnify a normal width mediastinum), loss of the aortic knuckle and deviation of the trachea to the right. Although angiography has been the gold standard diagnostic tool, CT scanning has an accuracy approaching 100% and is highly specific for detecting the injury.

Initial management is supportive, but a contained haematoma may rupture if the patient is hypertensive. Blood pressure should therefore be controlled in patients with suspected blunt aortic injury until CT scanning has excluded the injury. Once the injury is confirmed, the blood pressure must be controlled with drugs such as beta blockers or calcium antagonists until the patient can be taken to the operating theatre for definitive cardiothoracic repair. Endovascular repair is possible for some blunt aortic injuries.

TRAUMATIC DIAPHRAGMATIC INJURY

Traumatic rupture of the diaphragm is associated with blunt and penetrating trauma to the abdomen. Blunt trauma is usually the result of a lateral or frontal vehicular collision, with distortion of the chest wall, shearing of the diaphragm and compressive rise in intra-abdominal pressure. Rupture is more common (in survivors) on the left side, probably because the severity of injury required to cause a right-sided rupture above the protective liver is more usually fatal. The injury is rarely found in isolation, and is associated with other chest, abdominal and pelvic injuries.

Diaphragmatic ruptures associated with penetrating trauma are usually due to gunshot and stab injuries, and they result in a smaller tear with less visceral tissue protruding through the diaphragm.

Signs and symptoms can be subtle and the injury missed, only becoming apparent years later as the herniation develops. The standard chest X-ray may only show an elevated but indistinct hemidiaphragm; however, the appearance of bowel gas or a nasogastric tube within the chest will help confirm the diagnosis. Contrast studies via a nasogastric tube, CT and MRI scanning are all useful adjuncts. Diaphragmatic rupture and visceral herniation may be mistaken for a haemothorax on the plain chest X-ray; however, the insertion of a finger into the chest during chest drain insertion may reveal the presence of stomach or bowel loops (hence the need to avoid the use of sharp trochars for chest drain insertion to prevent visceral injury).

Initial *management* is supportive with careful assessment and management of the ABCs. Careful chest drain insertion is advisable prior to transfer or anaesthesia. Definitive treatment is surgical – the diaphragmatic rupture can be repaired during a trauma laparotomy but may require a thoracotomy or thoracoabdominal approach.

MEDIASTINAL TRAVERSING WOUNDS

Penetrating objects that cross the mediastinum may cause damage to the lungs and to the major mediastinal structures (the heart, great vessels, tracheobronchial tree and oesophagus). The *diagnosis* is made by careful examination of the chest, backed up by chest X-ray and trauma CT imaging. The significant clinical finding is an entrance wound in one hemithorax and an exit wound or radiologically visible missile in the other. Bullets and shrapnel can tumble, so the trajectory is unpredictable. The presence of fragments adjacent to the mediastinum on X-ray should raise suspicion of a traversing injury.

Patients with symptomatic, haemodynamically unstable mediastinal traversing wounds should be assumed to have an ongoing haemothorax, tension pneumothorax or cardiac tamponade.

Initial *management* is ABC resuscitation with bilateral chest drains, prior to definitive surgical management. Stable patients should undergo extensive investigation with ultrasound, trauma CT, angiography, oesophagoscopy and bronchoscopy as indicated, and an early consultation with a cardiothoracic surgeon should be arranged. Stable patients should be continually re-evaluated as they can suddenly deteriorate and require urgent surgical intervention; 50% of patients with mediastinal traversing wounds are haemodynamically unstable on presentation, with a doubled mortality of 40% over those who are stable.

BREATHING/CHEST INJURY – TAKE-HOME MESSAGE

- The primary goal in management of traumatic chest injuries is to rapidly identify and manage the six immediately life-threatening injuries within the primary survey.
- The eight potentially life-threatening injuries should be sought within the primary and secondary surveys, and they may require sophisticated imaging to diagnose.
- Oxygenation, ventilation and shock management are paramount.
- Only 15% of chest injuries require operative intervention.

C – Circulation and shock

For the healthcare professional ‘shock’ is not the commonly reported emotional condition in someone witnessing a disturbing incident. It can be broadly

defined as circulatory failure, or inadequate perfusion of the tissues and organs with oxygenated blood.

Untreated, or inadequately treated, shock leads to organ damage and ultimately death from multi-organ failure. Recognition of shock, diagnosis of the cause and subsequent management are therefore important steps in the resuscitation and care of the seriously ill or traumatized patient. The C for circulation follows the A for airway and B for breathing, but in the presence of catastrophic, external bleeding from limb wounds, control of the bleeding takes precedence. This is the C-ABC sequence, and it holds true in a hospital environment if the airway and catastrophic limb bleed cannot be managed simultaneously by the trauma team.

CIRCULATION AND SHOCK – AWARENESS

There are five main types of shock that can be grouped into two pathogenic groups:

- *vasoconstrictive* – hypovolaemic and cardiogenic shock
- *vasodilative* – septic, neurogenic and anaphylactic shock.

The majority of patients presenting with shock following a major injury will be suffering from hypovolaemic shock; however, any patient can present with a combination of types of shock.

Hypovolaemic shock Hypovolaemic shock results from a loss of volume within the circulation; it may be due to whole blood loss from haemorrhage, or plasma and fluid loss from burns or severe medical conditions. As the circulating blood volume decreases, compensatory mechanisms are triggered to preserve blood pressure and vital organ perfusion. These mechanisms can maintain systolic blood pressure up to around 30% blood loss in a fit patient. Above this, compensation increasingly fails until unconsciousness, followed by death at around 50% blood loss.

Early compensatory mechanisms are tachycardia and peripheral vasoconstriction with a narrowed pulse pressure. (Vasoconstriction raises the diastolic blood pressure, bringing it closer to the systolic, e.g. 120/60 → 120/90.) Further compensations include tachypnoea, shift of fluid from tissues into circulation and reduced urine output.

Some injuries mimic hypovolaemic shock, classically tension pneumothorax and cardiac tamponade; the low-output state follows obstruction to the venous return and cardiac output, respectively. Peripheral vasoconstriction is not a feature of these conditions in the absence of hypovolaemia, unlike cardiogenic shock and the veins remain full.

Cardiogenic shock Cardiogenic shock results from a decrease in myocardial contractility, and hence a reduction in stroke volume and cardiac output. This classically follows myocardial infarction or severe ischaemia, but it can follow trauma damage to the myocardium from blunt or penetrating injury (e.g. fracture of the sternum). The disproportionate vasoconstriction is due not to hypovolaemia but to an outpouring of catecholamines and the profound autonomic stimulus, which can put further strain on the heart by causing vasoconstriction and increasing afterload. Trauma patients may present with cardiogenic shock if the cardiac event precedes and indeed causes the traumatic event.

Septic shock This results from the entry of toxins into the circulation, which poison the vasoconstrictive mechanisms within the blood vessels. These toxins usually come from infection, or are released from within the bowel secondary to bowel damage caused by ischaemia. The profound vasodilatation that results dramatically reduces afterload; even with a normal circulating blood volume and raised cardiac output, the patient's blood pressure falls and the pulse pressure widens (e.g. 110/70 → 90/30). Oxygen consumption increases and, despite the high cardiac output, tissue perfusion and oxygenation are reduced, and organ damage results. The toxins can also damage the myocardium and cause capillary leakage, complicating the presentation with elements of cardiogenic and hypovolaemic shock.

Neurogenic shock Neurogenic shock is produced by high spinal cord injury, which disrupts the sympathetic nerves controlling vasoconstriction. The peripheral vasculature relaxes and becomes profoundly dilated, reducing pre-load and afterload. Even with a raised cardiac output, the patient cannot maintain an adequate blood pressure and shock ensues. Neurogenic shock is not caused by an isolated head injury and is different from 'spinal shock', which is a temporary flaccidity following spinal damage. Since neurogenic shock is always related to traumatic spinal cord damage, it is likely to coexist with a degree of hypovolaemia from associated trauma.

Anaphylactic shock This is a type of allergic reaction. Exposure to an antigen to which an individual has previously been sensitized triggers off a cascade reaction. The mast cells degranulate and release large quantities of histamine into the bloodstream. Other vasoactive substances are released, and profound vasodilatation is caused. Massive capillary leakage results in sudden oedema, which with loss of fluid into the bowel causes hypovolaemia. (1 mm depth of oedema across the body surface equates to a 1.5 L fluid loss.) This picture is complicated by other effects such as bronchospasm.

Anaphylaxis can be triggered by many common antigens such as shellfish or peanuts. Of particular significance to the hospital practitioner are allergies to

drugs, particularly to penicillin antibiotics (which are now commonly given to open fracture patients immediately or very soon after the injury is recognized), chlorhexidene and latex.

CIRCULATION AND SHOCK – RECOGNITION

Recognition of shock is relatively easy in the late stages when signs of underperfusion are obvious. Earlier stages of shock present with more subtle signs that require careful patient examination to elucidate; for example, the systolic blood pressure may not drop significantly until 30% of the patient's blood volume has been lost. Hypovolaemic shock passes through a number of clinical stages as blood loss increases, with increasingly apparent signs (Table 22.3). (Adult blood volume is approximately 7% of ideal body weight, or 5 L for a non-obese man weighing 70 kg.) It should be remembered, however, that the development and progression of shock is a continuum.

Blood loss of greater than 50% (>2500 mL) results in loss of consciousness, pulse and blood pressure, and finally respiration, causing a hypovolaemic, PEA, cardiac arrest.

The values shown in Table 22.3 relate to adults and children over the age of 12. Younger children compensate more effectively to a greater degree of blood loss, but they deteriorate very rapidly when they finally decompensate. The pulse rate is a good indicator of shock level in children, as is the respiratory rate; tables showing normal parameters for children at different ages are available.

Conversely, hypovolaemia is not tolerated well in older patients ('silver trauma') who are commonly taking a variety of cardiovascular drugs such as angiotensin inhibitors (ACEIs), sartans and beta blockers. Blood pressure and cardiac output are lost at a much earlier stage of shock and may need urgent support with inotropes, chronotropes and vasoconstrictors.

Recognition of shock therefore depends on a rapid clinical assessment of the patient, with measurement of the appropriate vital signs. The *look*, *listen*, *feel* sequence should be applied to identify the signs of hypovolaemic shock; blood pressure and pulse alone are not adequate.

Look and listen

- Peripheral/central cyanosis and pallor
- Sweating
- Tachypnoea and respiratory distress
- Change in mental status – anxiety, fear, aggression, agitation
- Depressed level of consciousness or unconsciousness.

Feel

- Peripheral perfusion poor – cool, clammy, shut down
- Capillary refill time > 2 seconds – note this is unreliable in cold and frightened patients

Table 22.3 Stages of shock

Blood volume loss		No change	Observations
Percentage loss	Equivalent volume in 70 kg male		
<15%	<750 mL	Blood pressure Pulse pressure Capillary refill	Minimal tachycardia <100 bpm Skin pallor possible
15–30%	750–1500 mL	Systolic blood pressure	↓ Peripheral perfusion with cool, pale, clammy skin ↑ Capillary refill > 2 seconds Tachycardia > 100 bpm ↓ Pulse pressure as diastolic BP rises Increased respiratory rate (tachypnoea) of 20–30 bpm Subtle mental status changes: anxiety, fear, aggression
30–40%	1500–2000 mL		Marked tachycardia > 120 bpm Measurable fall in systolic blood pressure from patient's normal, e.g. <100 mmHg Thready peripheral pulses Flat/empty veins Marked tachypnoea > 30 bpm Significant mental status changes: agitated ++ Dropping urine output
>40%	>2000 mL		Severe tachycardia > 140 bpm Moribund, decreased conscious level Significant drop in systolic blood pressure, e.g. <70 mmHg Impalpable peripheral pulses, weak central pulses Respiratory distress Central and peripheral cyanosis Minimal urine output

- Pulse rate and character – tachycardia and thready pulse
- Loss of pulses – radials, then femorals, then carotids as severity of shock increases
- Blood pressure – initially a raised diastolic with narrowed pulse pressure, then drop in systolic and diastolic, and finally an unrecordable blood pressure.

Observation of these factors will usually enable an assessment to be made of the presence and level of shock, and the likely degree of blood loss. This will act as a guide to whether volume replacement is indicated, and if so how much.

Hypovolaemic shock that remains unresponsive to treatment is likely to be due to continued bleeding into the body cavities or potential spaces, and evidence of this should be sought. Diagnosis may be helped by trauma imaging such as eFAST or CT. A useful reminder of where to look is the catchy slogan: 'bleeding onto the floor and four more' (i.e. external bleeding and chest, abdomen, pelvis/retroperitoneum, long bones). Bear in mind, though, that there are other forms of shock that need to be excluded.

Cardiogenic shock can mimic many of the signs of hypovolaemic shock. The history will give a good indication of the likely cause. The veins tend to be full in cardiogenic shock, and cyanosis more profound. There may be other diagnostic signs present such as pulmonary oedema.

Septic, neurogenic and anaphylactic shock are characterized by vasodilatation as opposed to vasoconstriction. The veins tend to be full and the peripheral pulses easily palpable and bounding. Peripheral perfusion may be good, with warm and flushed peripheries, but the skin may be mottled or cyanosed with sepsis.

CIRCULATION AND SHOCK – MANAGEMENT

Control of the airway (with cervical spine control), optimal oxygenation and ventilation are prerequisites to shock management. Immediate management of haemorrhagic shock depends on control of the bleeding and administration of limited volumes of intravenous fluids and blood to restore intravascular volume and haematocrit.

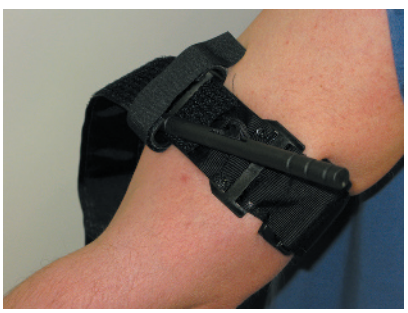
Control of haemorrhage This is achieved by direct pressure on the bleeding wounds with appropriate dressings, and elevation of the affected part where practicable. Continuing developments from military experience have led to the introduction of additional measures to control external and limb bleeding. Wounds can be packed with a dressing, and a circumferential bandage applied around and over the packed wound. Specialist bandages have been designed for this purpose, such as the *Olaes™ Modular Bandage*. This incorporates a gauze bandage for wound packing, with a plastic cup to compress into the packed wound beneath a circumferential, self-adhering, elastic bandage.

Tourniquets have been developed for controlling peripheral limb haemorrhage, with devices such as the *Combat Application Tourniquet (C-A-T™)* (Figure 22.24). The C-A-T™ is a single-handed device that uses a windlass system with a free-moving internal band to provide circumferential pressure around the extremity. Once it is tightened and the bleeding stopped, the windlass is applied tightly (to avoid acting as a venous tourniquet only and increasing blood loss) and locked in place. A Velcro® strap is then applied for further securing of the windlass during casualty evacuation. Once correctly applied, the distal limb is ischaemic and perfusion must be restored within 2 hours, to avoid lasting ischaemic damage, but the tourniquet should not be loosened or removed until a surgeon is available to definitively repair the injury. Two tourniquets should be applied to the lower limb and one to the arm.

Haemostatic dressings are useful for emergency control of arterial and venous haemorrhage. Chitosan (derived from crushed shellfish exoskeleton) granules or impregnated gauze such as *Celox™* are now commonly used in UK HEMs.

Clamping of bleeding points is difficult and can damage vessels; this should remain the province of the experienced surgeon who is capable of definitive repair.

Fracture of the pelvis can result in devastating retroperitoneal haemorrhage; this can be reduced by compressing the pelvis to approximate the bleeding fracture sites and reduce the potential volume of



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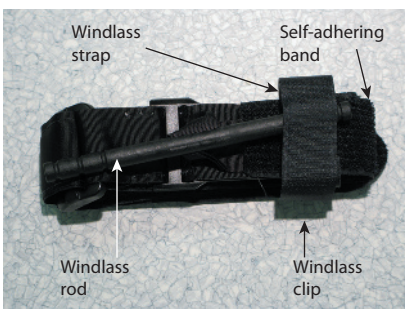

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Figure 22.24 The C-A-T™ tourniquet (a) Tourniquet in use. (b) Tourniquet components.

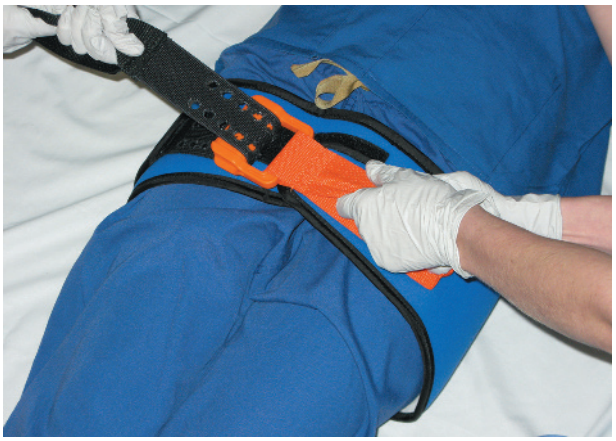


Figure 22.25 SAM Sling™ ratcheted compression belt in use

the pelvis. Compression can be achieved manually, with a towel or blanket passed under the patient and tightened from both sides above the pelvis, or preferably with a specialized pelvic binder such as the *SAM Sling*™ (Figure 22.25). Pelvic binders should be applied at the level of the greater trochanters; above or below this level, they become less effective. They can be used in association with a figure-of-eight bandage around the feet or tying the knees together to internally rotate the legs and should not be removed until the patient is in an environment where any resulting worsening of the degree of shock can be appropriately managed.

Resuscitative endovascular balloon occlusion of the aorta (REBOA) is a recently introduced technique for managing non-compressible torso haemorrhage (NCTH). It has been used both in the pre-hospital environment and in Level 1/Major Trauma Centres. REBOA involves the temporary occlusion of the aorta, using a percutaneously deployed intravascular balloon, usually inserted via the femoral artery. The aim is to improve blood pressure and maintain cerebral perfusion until surgical control of the bleeding has been achieved. However, improvements in mortality using REBOA have yet to be proved.

Tranexamic acid, an anti-fibrinolytic, is now given in the dose of 1 g IV as soon as possible in patients with major trauma and active or suspected active bleeding.

Peripheral venous cannulation Intravenous access must be secured at the earliest opportunity; this can be very difficult in later stages of shock. The size of the cannula is important because of its effect on flow, which is directly proportional to the fourth power of the radius of the cannula (Poiseuille's Law); halving the radius of a cannula reduces the flow rate by a factor of 16 (Figure 22.26). Flow is also reduced as cannula length is increased.

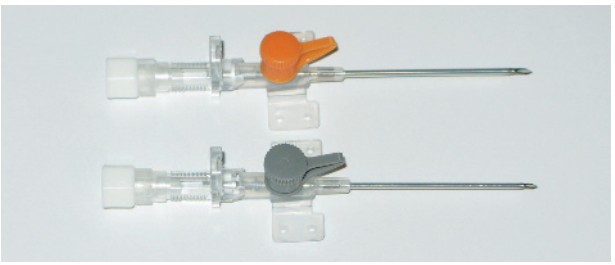


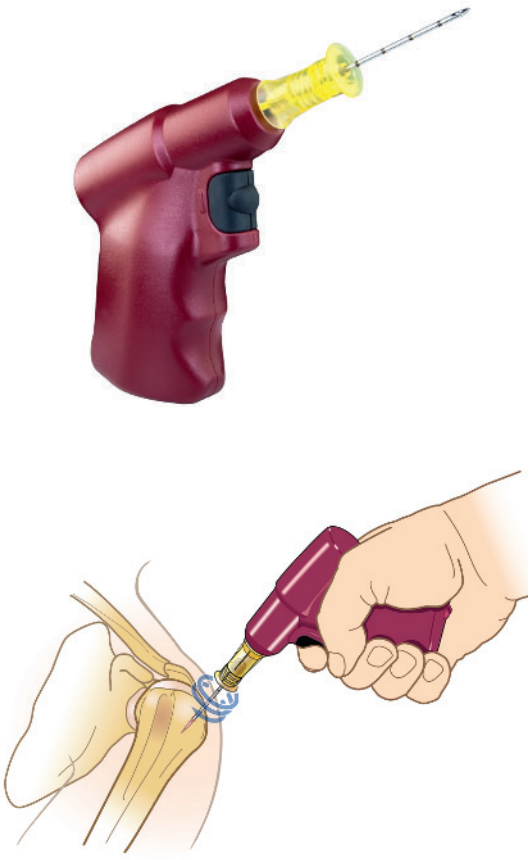
Figure 22.26 Cannulae A 16-gauge cannula (grey tap) has a 20% smaller diameter but 40% less flow than a 14-gauge cannula (orange tap).

Clearly it is difficult, if not impossible, to keep up with major haemorrhage without a minimum of two short large-bore cannulae. Hence, the ATLS® guideline for in-hospital trauma cannulation is insertion of two cannulae, minimum size 16-gauge, but preferably 14-gauge, into large peripheral veins, typically in the antecubital fossae. Catastrophic haemorrhage may require the insertion of a large-bore, 8-gauge resuscitation cannulae into a large vein such as the femoral vein and use of a rapid infusor to deliver a massive transfusion of blood and plasma under pressure.

Central venous cannulation This is an option reserved for those with appropriate expertise and equipment; it can be difficult and carries a significant risk of life-threatening complications (pneumothorax and arterial damage most commonly). In the UK, the use of two-dimensional (2D) ultrasound imaging is strongly recommended in the routine siting of a central venous pressure (CVP) line. Access to the internal jugular can be difficult in a trauma patient, especially if he or she is immobilized with a stiff cervical collar and head blocks in place. The subclavian approach has the highest incidence of complications; femoral cannulation is a safer option than either central approach and a cannula can often be sited in the femoral vein, medial to the femoral artery.

Intraosseus cannulation Intraosseus cannulation is now an accepted technique in both children and adults, using specialist equipment (Figure 22.27). Most commonly employed in the UK is the EZ-IO® system, using a hand-held electric drill to drive a specialist cannula through the cortex of the tibia or humeral head into the marrow cavity. Response time to drug administration is close to IV administration, and entire resuscitations can be performed through intraosseus cannulae, including all anaesthetic drugs and fluids. Due to the resistance to flow, fluids should be given as boluses using a 50 mL syringe.

Fluid administration Fluid administration has long been a controversial issue. The traditional ATLS approach for trauma circulation resuscitation, based on military experience, is to site two large-bore



(a)

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Figure 22.27 Intraosseous cannulation (a) EZ-IO System intraosseous vascular access hand-held power driver. (b) Intraosseus needle insertion humeral head.

intravenous cannulae and administer an initial bolus of 2 L of warmed Ringer's lactate or Hartmann's solution. This is certainly successful in improving perfusion in bleeding patients, but it is now not recommended for pre-hospital use where haemorrhage cannot be surgically controlled and blood is not available for transfusion. Casualties bleeding to a class 3 or 4 shock can reach a steady state as the blood pressure drops to a point where active bleeding may cease. Restoring vascular volume with crystalloids or colloids can restore the blood pressure to a point where bleeding resumes; further administration of clear fluids repeats the cycle until the haemoglobin level drops below a point where adequate oxygen can be carried. Cardiac arrest and death then result from anaemic hypoxia.

Consequently, in the UK, the NICE 2016 guidelines recommend titrating volume resuscitation to maintain a palpable, central pulse using crystalloids, not colloids. Large volumes (>2 L) of normal saline 0.9% can cause a hyperchloraemic acidosis, and a lactated, balanced electrolyte solution such as Ringer's lactate or Hartmann's is preferable. In hospital and where available in the pre-hospital environment, haemorrhagic shock should be treated with blood and fresh frozen plasma in a 1:1 ratio, titrated against response, with optimum organ and peripheral tissue perfusion the goal. Blood pressure, pulse rate, peripheral perfusion and CVP are all used to assess response. All fluids should be warmed to prevent hypothermia-induced coagulation failure. Serial measurement of metabolic acidosis parameters such as bicarbonate, base deficit and lactate levels can be used to gauge adequate response to fluid therapy. More sophisticated methods such as oesophageal Doppler and arterial waveform analysis are also used in the critical-care setting.

Platelets and other specialist blood products such as cryo-precipitate may be required in continued bleeding and should be given as guided by laboratory coagulation results and haematology advice.

These guidelines refer to resuscitation of bleeding patients only. Most other forms of shock will respond initially to IV fluids pending accurate assessment and diagnosis. However, hypotension and vasoconstriction in elderly casualties without evidence of major trauma should raise a high index of suspicion for cardiogenic shock; in this situation, infusion of even small volumes of fluid can overload the circulation and cause collapse and cardiac arrest.

CIRCULATION AND SHOCK – TAKE-HOME MESSAGE

In patients suffering from haemorrhagic, hypovolaemic shock the source of the bleeding must be identified and surgically or radiologically controlled. The priorities for restoring and maintaining adequate circulation are as follows:

- Control external and pelvic bleeding, give tranexamic acid and keep the patient warm.
- Administer restricted volume resuscitation with crystalloids until blood is available.
- Transfuse blood and plasma in a 1:1 ratio – replace blood with blood.
- Turn off the tap – call a surgeon to achieve definitive control early.

D – Disability – head injury

The immediate management of the seriously head-injured patient is designed to prevent secondary injury and to provide the neurosurgeon with a live patient who has some hope of recovery. A significant number of fatalities from head injury are caused by the secondary and not the primary injury; prevention of this secondary brain injury is facilitated by following the ABC principles set out in ATLS®.

HEAD INJURIES – AWARENESS

In the UK, severe head injuries account for more than 50% of trauma-related deaths, and these usually

follow RTCs, assaults and falls. Injury patterns differ between countries; in the UK patients experience predominantly closed injuries, with a peak incidence in males between the ages of 16 and 25 years. A second peak occurs in the elderly, with a high incidence of chronic subdural haematomas.

Only 10% of head-injured patients presenting at Emergency Departments have a severe injury. The injuries can be classified into three groups based on the GCS:

Mild (80%)	GCS 13–15
Moderate (10%)	GCS 9–12
Severe (10%)	GCS 3–8

Investigation, management and outcomes depend on the severity of the injury; however, this is a continuum, and the classification given earlier is only a guideline. Even mild head injuries can be associated with prolonged morbidity in the form of headaches and memory problems; only 45% are fully recovered 1 year later. With moderate head injuries, 63% of patients remain disabled 1 year after the trauma, and this rises to 85% with severe injuries.

A knowledge of anatomy and pathophysiology is needed to understand and anticipate the development of a head injury.

The scalp comprises five layers of tissue, remembered using the mnemonic SCALP: skin, connective tissue, aponeurosis, loose areolar tissue, and periosteum. It has a generous blood supply and serious scalp lacerations can result in major blood loss and shock if bleeding is not controlled.

The skull is composed of the cranial vault and the base. The vault has an inner and outer table of bone and is particularly thin in the temporal regions, although protected by the temporalis muscle. The base of the skull is irregular, which may contribute to accelerative injuries. The floor of the cranial cavity has three distinct regions: the anterior, middle and posterior fossae.

The meninges cover the brain and consist of three layers:

- *Dura mater* – a tough, fibrous layer, firmly adherent to the inner skull
- *Arachnoid mater* – a thin, transparent layer, not adherent to the overlying dura and so presenting a potential space. Cerebrospinal fluid (CSF) is contained and circulates within this space
- *Pia mater* – a thin, transparent layer, firmly adherent to the underlying surface of the brain.

The brain itself is divided into three main structures:

- *Cerebrum* – composed of right and left hemispheres, divided into:
  - frontal lobes – emotions, motor function, speech
  - parietal lobes – sensory function, special orientation
  - temporal lobes – some memory and speech functions
  - occipital lobes – vision
- *Cerebellum* – coordination and balance
- *Brainstem* – composed of three main structures:
  - midbrain – reticular activating system (alertness)
  - pons – relays sensory information between cerebrum and cerebellum
  - medulla – vital cardiorespiratory centres.

The midbrain passes through a large opening in the tentorium, a fibrous membrane that divides the middle and posterior fossae. The third cranial nerve, which controls pupillary constriction, also runs through this opening, and is vulnerable to pressure damage if the cerebral hemispheres swell. This results in pupillary dilatation, an early sign of a significant rise in intracerebral pressure.

Pathophysiology The skull is in effect an enclosed, bony box containing the brain, blood vessels and the CSF. The intracerebral pressure (ICP) is normally maintained at approximately 10 mmHg, and is a balance of brain tissue, intravascular and CSF volumes. Traumatic damage to the brain can cause swelling of the brain tissue itself, and bleeds from arteries and veins into the extradural space, subdural space or brain substance (intracerebral bleed) increase the intracerebral volume and raise the ICP. If the ICP is sustained at above 20 mmHg, permanent brain damage can result, with poor outcomes; this is the secondary brain injury. There is only limited capacity for intracranial compensation for a rise in ICP, and this is largely achieved by a reduction in CSF volume (Monro-Kellie doctrine). Once pressure compensation has reached its limits, the ICP rises rapidly in a breakaway exponential.

As the pressure rises, the conscious level decreases and the GCS falls. The medial part of the temporal lobe (the uncus) herniates through the tentorial notch, compressing the third cranial nerve and the midbrain pyramidal tracts. This usually results in pupillary dilatation on the side of the injury, and hemiplegia on the opposite side. Pressure changes in the medulla cause a sympathetic discharge, with a rise in blood pressure and reflex bradycardia. With further pressure rise, cerebral blood flow is compromised, and it ceases terminally when the ICP rises above the mean arterial pressure (MAP). Ultimately, the cerebellar tonsil is forced into the foramen magnum, resulting in a loss of vital cardiorespiratory function; this is known as brainstem or brain death, and it is a terminal event.

Mechanism of brain injury Brain injury can be blunt or penetrating. The *primary brain injury* occurs at the time of the trauma, and results from sudden distortion and shearing of brain tissue

within the rigid skull. The damage sustained may be focal, typically resulting from a localized blow or penetrating injury, or diffuse, typically resulting from a high-momentum impact. Sudden acceleration or deceleration can cause a contra-coup injury, as the brain impacts on the side of the skull away from the impact. High-velocity missile penetrating injuries will also cause a diffuse and severe brain injury as the resultant pressure wave moves across the brain. The *secondary brain injury* is pressure related, and is caused by swelling within the brain, causing a rise in ICP as described earlier. This is compounded by hypoxia, hypercarbia and hypotension.

Severity of brain injury The Glasgow Coma Score is a well-tested and objective score for assessing the severity of brain injury: 13–15 is mild; 9–13 is moderate; 8 or less is severe.

Morphology of brain injury *Skull fractures* (Figure 22.28) are seen in the cranial vault or skull base; they may be linear or stellate, and open or closed. The significance of a skull fracture is in the energy transfer to the brain tissue as a result of the considerable force required to fracture the bone. Open skull fractures may tear the underlying dura, resulting in a direct communication between the scalp laceration and the cerebral surface, which may be extruded as ICP rises.

Basal skull fractures are caused by a blow to the back of the head, or rapid deceleration of the torso with the head unrestrained, as in high-speed vehicular crashes. These fractures are rare, occurring in 4% of severe head injuries, but they can cause severe damage and are a cause of death in front-end collisions and motor sport crashes. There are key physical signs pathognomic of basal skull fracture:

- periorbital ecchymosis (bruising – 'raccoon' or 'panda' eyes)
- retroauricular ecchymosis (Battle sign – bruising behind ears)
- otorhinorrhea (CSF leakage from nose and ears)
- VIIth and VIIIth cranial nerve dysfunction (facial paralysis and hearing loss).

Basal skull fractures are not always visible on X-ray or CT, but blood in the sinus cavities and the clinical signs described above should suggest their presence.

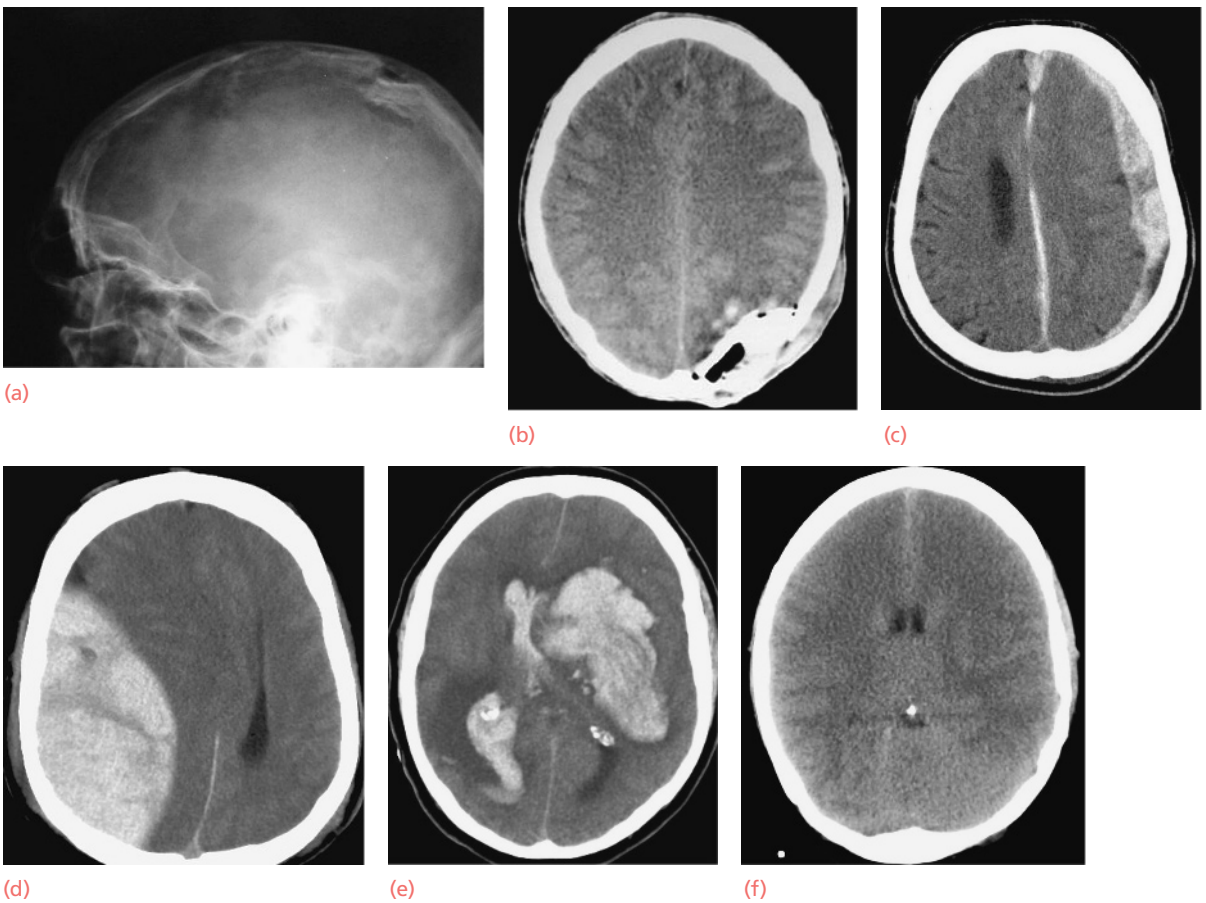


Figure 22.28 Fractured skull – imaging (a) X-ray showing a depressed fracture of the skull. (b–f) CT scans showing various injuries: (b) a fracture; (c) a subdural haematoma; (d) an extradural haematoma and compression of the left ventricle; (e) an intracerebral haematoma; (f) diffuse brain injury with loss of both ventricles.

Diffuse brain injury is due to axonal disruption of the neurones and varies from minor, resulting in mild concussion, to severe, resulting in an ultimately fatal hypoxic and ischaemic insult to the brain.

Extradural (epidural) haematomas are relatively uncommon, occurring in 0.5% of all brain-injured patients, and 9% of those who are comatose. The haematoma is contained outside the dura but within the skull and is typically biconvex or lenticular in shape. They are commonly located in the temporal or temporoparietal region, and usually result from a middle meningeal artery bleed caused by a fracture.

Subdural haematomas are more common, and constitute 30% of severe brain injuries. They usually result from tearing of cortical surface vessels and normally cover the entire surface of the hemisphere. Underlying brain damage is usually much more severe due to the greater energy transfer.

Contusions and intracerebral haematomas are fairly common (20–30% of severe brain injuries). The majority occur in the frontal and temporal lobes. Inoperative contusions can evolve into haematomas requiring surgical evacuation over a period of hours or days, and repeat CT scanning within 24 hours may be indicated.

HEAD INJURIES – RECOGNITION

Initial recognition of a head injury takes place in the primary survey as part of the ABCDE sequence. The airway, cervical spine, breathing and circulation must all be assessed and resuscitation commenced before the brief neurological assessment takes place, as these measures will prevent the development of a secondary brain injury. The AVPU score is an instant and useful assessment but the level of consciousness should be assessed accurately at this point, using the GCS. The pupils are assessed for equality, diameter and response to light.

As there is a 5–10% association between cervical spine fracture and head injury, the assumption is made that the neck is unstable until proved otherwise. As the cervical spine X-ray does not rule out a fracture, immobilization should remain in place until the neck is cleared clinically or with further imaging such as CT.

A more thorough assessment of the neurological status takes place during the secondary survey. The GCS and pupils are re-evaluated, lateralizing signs are looked for, and the upper and lower limb motor and sensory function evaluated. If the patient is stable, further imaging may be indicated, and a number of guidelines exist to aid the decision.

CT scanning is the primary examination of choice for patients with a clinically important brain injury (National Institute for Health and Clinical Excellence). Modern, fast, spiral CT scanners are increasingly available adjacent to Emergency Departments, enabling rapid trauma CTs in the course of minutes. All patients suffering a severe head injury require an urgent CT scan. The Royal College of Surgeons of England advises that specific indications for a head CT are:

- GCS <13 on first Emergency Department assessment
- GCS <15 2 hours after initial assessment
- suspected open or depressed skull fracture
- clinical basal skull fracture
- post-traumatic seizure
- focal neurological deficit
- >1 episode of vomiting
- amnesia of events >30 minutes before impact
- post-injury amnesia if:
  - age >65 years
  - associated with coagulopathy
  - due to a dangerous mechanism of injury (pedestrian versus motor vehicle, ejection from motor vehicle, fall from height >1 m).

HEAD INJURIES – MANAGEMENT

The management of head injuries depends on the severity, as assessed by the clinical examination, GCS and CT scan. Patients with a *mild head injury* should be admitted and monitored, with frequent neurological observations. Should there be any deterioration, CT scanning is indicated, and referral to the local neurosurgical unit is necessary. Discharge is when a complete neurological recovery has been made and provided the patient can be supervised at home by a responsible adult.

Patients sustaining *moderate head injuries* will need CT scanning and discussion with a neurosurgeon to decide on the need for transfer and definitive care. Other indications for neurosurgical referral, regardless of imaging findings, include:

- persistent coma after initial resuscitation (GCS <8)
- unexplained confusion >4 hours
- post-admission deterioration in GCS
- progressive, focal neurological signs
- seizure without full recovery
- definite or suspected penetrating injury
- CSF leak.

Patients with *severe head injuries* will require immediate resuscitation as described previously. The cervical spine must be immobilized while the airway is secured; this will require a competent, rapid sequence induction (RSI) of anaesthesia, and an anaesthetist must be involved early. Once the airway is secured and protected with a tracheal tube, the oxygenation and ventilation must be optimized. Hypoxia and hypercarbia must be avoided, but overventilation is equally damaging, as cerebral

blood flow is compromised. Ventilation must be monitored with capnography, and the minute volume adjusted to maintain a low-normal $EtCO_2$ (4.5 kPa). Oxygen saturation levels should be maintained above 95% and sequential arterial blood gas estimations made to ensure the oxygen partial pressure is maintained in the normal range (> 13 kPa) as far as is possible.

The circulation should be monitored to maintain intravascular filling within an appropriate range. Overfilling will worsen cerebral oedema, but hypovolaemia will result in persistent shock. If traumatic brain injury is the dominant condition in the presence of shock, fluid administration should be less restrictive. Central venous pressure should be monitored, and arterial pressures kept within a normal range for that patient, with reference to the ICP. This requires expert critical care skills, and patients with a severe brain injury must be managed in an appropriate critical care unit.

The rapid administration of intravenous mannitol at a dose of 0.5 mg/kg, or hypertonic saline, may be indicated to reduce ICP, and this should be given following discussion with the neurosurgeon that the patient has been referred to. It can be a useful holding measure if signs of rising ICP (e.g. a dilated pupil) develop prior to or during transfer to a specialist centre.

Patients with significant head injuries in units without neurosurgical capability will require transfer, on discussion with the neurosurgeons. An expanding, intracerebral haematoma will need to be evacuated within 4 hours of injury to prevent serious and permanent secondary brain injury.

HEAD INJURIES – TAKE-HOME MESSAGE

- Head-injured patients require early assessment and recognition of their brain injury.
- A severe blow to the head causes a primary brain injury.
- Hypoxia and hypercarbia cause cerebral swelling and a secondary brain injury.
- Secondary brain injury should be minimized by optimal oxygenation, ventilation and blood pressure management.

Abdominal injuries

The abdomen is difficult to assess in the multiply injured trauma patient, especially when the patient is unconscious. The immediately life-threatening injury is bleeding into the abdominal cavity, and this is one of the 'onto the floor and four more' areas into which lethal volumes of blood may be lost or sequestered. The abdomen is therefore examined in the primary survey as part of the circulation assessment.

ABDOMINAL INJURIES – AWARENESS

Abdominal injuries may be blunt or penetrating. Unrecognized abdominal injury is a cause of avoidable death after blunt trauma and may be difficult to detect. A direct blow from wreckage intrusion or crushing from restraints can compress and distort hollow viscera, causing rupture and bleeding. Deceleration causes differential movement of organs, and the spleen and liver are frequently lacerated at the site of supporting ligaments. In patients requiring laparotomy following blunt trauma, the organs most commonly injured are:

- spleen (40–55%)
- liver (35–45%)
- small bowel (5–10%)
- retroperitoneum (15%).

The mechanism of injury should lead to a high index of suspicion; for example, flexion lap-belt injuries in car crashes can rupture the duodenum, with retroperitoneal leakage and subtle signs (Figure 22.29). Early imaging and exploratory laparotomy may be required.

Penetrating injuries between the nipples and the perineum may cause intra-abdominal injury, with unpredictable and widespread damage resulting from tumbling and fragmenting bullet fragments. High-velocity rounds transfer significant kinetic energy to the abdominal viscera, causing cavitation and tissue destruction. Gunshot wounds most commonly involve the:

- small bowel (50%)
- colon (40%)
- liver (30%)
- abdominal vasculature (25%).

Stab wounds injure adjacent abdominal structures. Small wounds may result from thin-bladed knives that have penetrated deep and damaged several structures, with the most commonly injured being:

- liver (40%)
- small bowel (30%)
- diaphragm (20%)
- colon (15%).

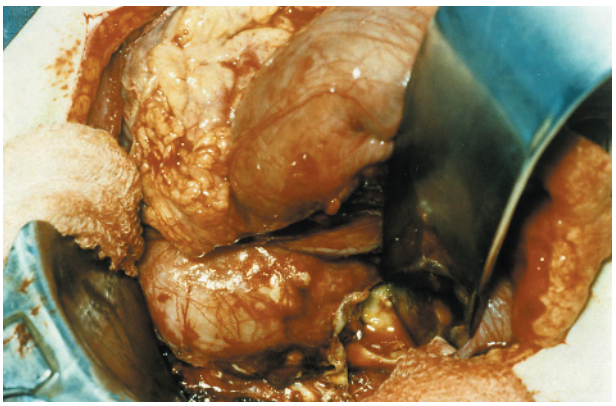


Figure 22.29 Abdominal injury Ruptured duodenum following flexion lap belt injury.

ABDOMINAL INJURIES – RECOGNITION

The abdomen is initially examined during the primary survey to determine if shock is due to an abdominal injury. A history from the patient, bystanders and paramedics is important, as the mechanism of injury can be identified and injuries predicted.

Examination of the abdomen follows the '*look, listen, feel*' format. The patient must be fully exposed, and the anterior abdomen should be inspected for wounds, abrasions and contusions.

The flanks and posterior abdomen and back should be examined, and this may require log-rolling to both sides. Auscultation is difficult in a noisy resuscitation room, but it may reveal absence of bowel sounds caused by free intraperitoneal blood or gastrointestinal fluid. Percussion and palpation may reveal tenderness or peritonism. The genitalia and perineum should be examined, and a rectal examination performed during the log-roll.

Early imaging is indicated; eFAST examination will reveal the presence of intraperitoneal fluid and can be performed in the resuscitation room; the technique has a high specificity but low sensitivity. Presence of fluid is an indication for laparotomy. CT scanning requires the patient to be stable, but is a much more effective diagnostic tool and all adults with major blunt trauma and suspected multiple injuries should have a whole body CT. Whole body CT should not be routinely used in children under the age of 16 due to the radiation exposure involved.

ABDOMINAL INJURIES – MANAGEMENT

Initial management of an abdominal injury is to manage shock as described in circulation management. External bleeding is controlled with direct pressure, wound packing or haemostatic dressings. Intravenous access is established with two large-bore cannulae and crystalloids administered judiciously to maintain the central pulse until blood and plasma are available. Confirmation of bleeding into the abdomen is an indication for tranexamic acid and immediate laparotomy and imaging other than eFAST may not be possible with an unstable patient. Other indications for laparotomy include:

- unexplained shock
- rigid silent abdomen
- evisceration
- radiological evidence of intraperitoneal gas
- radiological evidence of ruptured diaphragm
- gunshot wounds.

A naso- or orogastric tube should be passed in all multiple trauma patients; this should be passed orally in the presence of facial and basal skull fractures. A urinary catheter should be passed unless urethral bleeding or other signs of urethral injury such as genital bruising are present.

Laparotomy is the definitive management and the province of the surgeon; the general principle at initial operation is to perform damage control surgery:

- control haemorrhage with ligation of vessels and packing
- remove dead tissue
- control contamination with clamps, suturing and stapling devices
- lavage the abdominal cavity
- close the abdomen without tension.

Second-look laparotomy at 24–48 hours may be indicated to allow:

- removal of packs
- removal of dead tissue
- definitive treatment of injuries
- restoration of intestinal continuity
- closure of musculofascial layers of the abdominal wall.

The patient will require supportive critical care, and may require ventilation on an ICU until after the second-look laparotomy.

ABDOMINAL INJURIES – TAKE-HOME MESSAGE

- Abdominal injuries are difficult to assess in the multiply injured patient.
- The immediate threat to life is bleeding into the peritoneal cavity.
- Shock should be treated effectively.
- Early imaging with eFAST and CT should be performed.
- Early consultation with a surgeon should be facilitated – diagnostic or definitive laparotomy may be required.

Musculoskeletal injuries

In the absence of catastrophic bleeding, musculoskeletal injuries are not immediately life-threatening. They are, however, limb threatening and potentially life-threatening. Definitive management is detailed elsewhere in this book, so this section will merely put these injuries into the context of the overall management of a severely injured casualty.

PELVIC FRACTURES

Awareness The pelvis and retroperitoneum constitute one of the 'onto the floor and four more' spaces into which blood can be sequestered to a level resulting in non-responsive shock. A haemorrhaging fracture of the pelvis therefore becomes a life-threatening emergency and should be considered in every patient with a serious abdominal or lower limb injury. Potential causes are RTCs, falls from a height or crush injuries.

Recognition The pelvis is examined in the primary survey as part of the C–circulation assessment, once the airway and breathing have been assessed and the cervical spine immobilized. Significant signs are swelling and bruising of the lower abdomen, thighs, perineum, scrotum or vulva, and blood at the urethral meatus. The pelvic ring should be gently palpated for tenderness; however, the pelvis should not be compressed for crepitus, as this can dislodge a clot from the fracture site and provoke further bleeding. If tenderness is elicited, the examination should not be repeated.

If the patient is stable, a trauma CT scan will give more detailed information, and also provide information on intra-abdominal and retroperitoneal bleeding. Alternatively, an AP X-ray should be obtained during the primary survey, and in most cases this will enable a preliminary diagnosis of pelvic fracture to be made. If images are taken with a pelvic binder *in situ*, an AP X-ray of the pelvis with the binder off should be taken as part of the secondary survey to exclude an unstable pelvic fracture that has been reduced by the binder and hence was not apparent on the first images.

Management The immediate management of a pelvic fracture resulting in shock is to control the bleeding and restore volume as described previously. There are a number of pelvic binders available to wrap around the pelvis and apply compression to approximate the bleeding fracture sites and allow clot formation. If these are not available, manual approximation can be used; this can be facilitated with a sheet wrapped around the pelvis and twisted anteriorly.

Once in place, the pelvic compression devices should not be removed until interventions such as embolization, external fixation or pelvic packing are available. Developments in interventional radiology and angiography have enabled embolization to be used to control haemorrhage from a fractured pelvis.

PELVIC FRACTURES – TAKE-HOME MESSAGE

- Pelvic fractures can result in life-threatening haemorrhage and should be recognized and managed as part of the circulation assessment during the primary survey.
- Pelvic compression devices should be used to minimize bleeding, and a rapid surgical referral should be made for definitive management.

SPINAL INJURIES

Vertebral column injury, with or without neurological damage, must be considered in all patients with multiple injuries. A missed spinal injury can have devastating consequences. Immediate management therefore focuses on immobilization, recognition and referral for definitive care.

Awareness Spinal injuries can be stable or unstable, an unstable injury being one where there is a significant risk of fracture displacement and neurological sequelae. The mechanisms of injury are traction (avulsion), direct injury and indirect injury. Direct injuries are penetrating wounds usually associated with firearms and knives. Indirect injuries are the most common and are typically the result of falls from a height or vehicular accidents where there is violent free movement of the neck or trunk. There is an association between cervical spinal damage and injuries above the clavicles, and some 5% of head-injured patients have an associated spinal injury; 10% of those with a cervical spine fracture have a second, non-contiguous spinal fracture. Regional occurrences of spinal injuries are approximately:

- cervical (55%)
- thoracic (15%)
- thoracolumbar junction (15%)
- lumbosacral (15%).

Spinal fractures with spinal cord transection also disrupt the sympathetic nerve supply and cause distal vasodilatation. A high spinal transection will therefore cause neurogenic shock – this is vasodilatory shock and is characterized by hypotension, a low diastolic blood pressure, widened pulse pressure, warm and well-perfused peripheries and bradycardia. However, neurogenic shock can be complicated by hypovolaemic shock in multiply injured patients.

Recognition The spinal column and neurological function are examined in the secondary survey, with immobilization maintained throughout. While the head is immobilized manually, and the patient log-rolled, the cervical spine and vertebral column from neck to sacrum are examined for:

- bruising, contusions and ecchymosis
- penetrating injury
- swelling or 'bogginess'
- tenderness on palpation
- step or misalignment between vertebrae.

A rectal examination is performed to assess anal tone. A neurological examination is carried out to identify loss of sensory and motor function.

If the casualty is conscious, has no neck pain, has no distracting painful injury, is not intoxicated and has not received any analgesia, the cervical spine can be examined and a fracture clinically excluded. Head blocks, cervical collar and tape are removed, and the patient is taken through a full range of active movements (i.e. patient's voluntary movement). If there is neither pain nor neurological symptoms on movement, the cervical spine can be cleared.

X-rays are of limited use in the resuscitation phase as they do not reliably exclude unstable fracture-dislocations. Because they do not alter initial

management, they are no longer routinely used. If the cervical spine has not been cleared clinically or it is not possible to do so, precautions should be maintained until CT or MRI scans have been reviewed by a radiologist and pronounced clear.

Management Initial management follows the ATLS® ABCDE sequence. The cervical spine must be immobilized at all times; deterioration of neurological function of even one myotome can cause a devastating change in subsequent motor function. However, only 5% of multiply injured patients have cervical spine injuries, in contrast to the high percentage of patients with compromised airways; this is particularly significant with head injuries. In high spinal transections, the patient's respiratory function may be compromised, leading to ventilatory failure. The airway must be maintained without causing neck flexion or extension, and secured and protected with careful anaesthetic induction and intubation. This can be successfully done with specialist laryngoscopes such as the McCoy (lever-activated, flexing tip to lift the epiglottis), or video-laryngoscopes, in conjunction with an intubating catheter. The procedure should be carried out by an experienced anaesthetist.

Oxygenation and ventilation is optimized, monitoring SaO_2 and $EtCO_2$. The neurogenic shock will require judicious use of intravenous fluids and may need circulatory support with vasoconstrictors and chronotropes.

The spinal fracture and neurological deficits are managed by immobilization and referral to a spinal surgeon.

SPINAL INJURIES – TAKE-HOME MESSAGE

- Spinal injuries should be identified during the secondary survey and managed according to the ABCs.
- Immobilization is crucial throughout, and ventilatory and circulatory failure must be recognized and managed.
- Injuries should be excluded clinically, or with CT and MRI, as soon as possible.

LONG-BONE INJURIES

Long-bone injuries can be spectacular, but should not distract from the injuries compromising the airway, breathing or circulation. They are limb-threatening but not immediately life-threatening, and in the absence of catastrophic bleeding they can be addressed in the secondary survey.

Awareness Musculoskeletal injuries occur in 85% of patients sustaining blunt trauma. Major injuries signify significant force applied to the body and so are associated with an increased incidence of chest, abdomen and pelvis damage. Although not immediately life-threatening, they present a potential threat to life and prejudice the integrity and survival of the limb. Crush injuries can lead to compartment syndrome and myoglobin release with the risk of renal failure. These injuries must therefore be addressed as soon as the resuscitation priorities have been addressed.

Recognition The casualty must be fully exposed, log-rolled and examined from head to toe in all planes. The limbs are examined visually for:

- colour and perfusion
- wounds
- deformity (angulation and shortening)
- swelling
- discoloration and bruising.

The extremities are then palpated to detect any tenderness, swelling or deformity, indicating potential underlying fractures and dislocations. Crepitus may be felt but should not be specifically elicited. Peripheral circulation is assessed with palpation of pulses and capillary refill. Doppler ultrasound examination may be needed to confirm the presence of pulses – however, the presence of a pulse does not exclude compartment syndrome; it is a very late sign. X-rays should be obtained as indicated as soon as the patient is stable, and early CT scanning or CT angiography should be performed if a vascular injury is suspected. Peripheral perfusion must be restored within two hours from injury to avoid limb-threatening, ischaemic damage.

Management The immediate management is to ensure that the airway and ventilation are optimized, and then control limb haemorrhage with direct pressure, tourniquets, wound packing or haemostatic dressings as described previously. Large tissue deficits may need ongoing fluid and blood replacement as immediate haemorrhage control can be difficult.

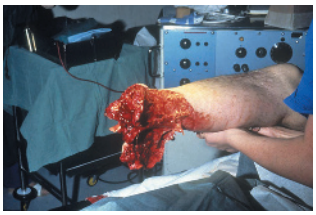
Fractures and dislocations should be reduced and splinted in the anatomical position where possible, to minimize neurovascular compromise and significant analgesia or procedural sedation may be required to facilitate this (e.g. Entonox, morphine or ketamine 0.5–1.0 mg/kg intravenously). The anatomical position should not be forced if significant resistance is felt (e.g. posterior hip dislocation).

Traumatic amputations, de-gloving injuries and blast injuries can be initially managed with specialist blast dressings (Figure 22.30).

Open fractures should be managed as described in Chapter 23 and the patient referred urgently to an orthopaedic surgeon for definitive management. Significant fractures, open fractures and dislocations may need operative intervention whilst life-saving abdominal or neurological surgery is taking place. Vascular and plastic surgery may be required at an early stage.

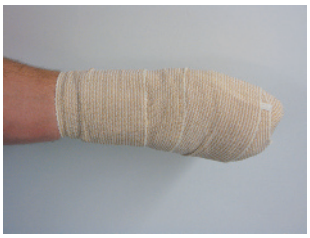
TAKE-HOME MESSAGE

- Limb injuries are not immediately life-threatening in the absence of catastrophic haemorrhage.
- They should be recognized and initially managed in the secondary survey.



(a)

(b)



(c)

Figure 22.30 Musculoskeletal injuries (a) Traumatic amputation; (b) blast dressing and (c) blast dressing *in situ.*

- Ischaemic limbs should be identified and managed early.
- Fracture reduction, splinting and immobilization are instituted before prompt surgical consultation.
- Antibiotics should be given early in open fractures.

Burns (thermal, chemical, electrical, cold injury)

A burn is a broad term that encompasses not only thermal injury to tissues from heat but injury from electric shock, chemicals and cold. In the UK, around 250 000 burn victims attend hospital each year, of whom 16 000 are admitted; in the USA, about 1.25 million burns occur annually, with 51 000 patients hospitalized. The risk is highest in the 18–35 year age group, with a male to female ratio of 2:1 for both injury and death, and serious burns occur most frequently in children under 5 years of age. There are around 4500 burns deaths each year in the USA, and the death rate is much higher in those over the age of 65. The last two decades have seen much improvement in burns care, and the mortality rate is now 4% in those treated in specialist burns centres.

THERMAL BURNS – AWARENESS

Major burns can present a threat to life through compromise of the airway, breathing and circulation. In addition, those burned may suffer other traumatic harm due to explosions, etc., and can present with any of the systemic injuries described previously. Circumferential burns around the neck can cause tissue swelling and airway obstruction, and burns around the chest may cause restrictive respiratory failure. Large burns cause in significant fluid shifts, and resultant shock. In combination with coma from toxin inhalation, burns present a potent mix of assaults on a casualty's life.

Cell damage occurs at a temperature greater than 45 °C (113 °F) owing to denaturation of cellular protein; a burn's size and depth are functions of the burning agent, its temperature and the duration of exposure. Thermal injury to the skin damages the skin's ability to function as a semipermeable barrier to evaporative water loss, resulting in free water loss in moderate to large burns. Other functions such as protection from the environment, control of body temperature, sensation and excretion can also be harmed. Systemic effects include hormonal alterations, changes in tissue acid–base balance, haemodynamic changes and haematological derangement. Massive thermal injury results in an increase in haematocrit with increased blood viscosity during the early phase, followed by anaemia from erythrocyte extravasation and destruction. Vasoactive substances are released and a systemic inflammatory reaction can result.

Inhalational burns Inhalation of super-heated gases and inhalation of toxic smoke in entrapment result in inhalational burns and smoke inhalation. Inhalational injury is now the main cause of mortality in the burns patient, and half of all fire-related deaths are due to smoke inhalation. Direct thermal injury is usually limited to the upper airway above the vocal cords and it can result in rapid development of airway obstruction due to mucosal oedema. Smoke has two noxious components: particulate matter and toxic inhalants. The particles are due to incomplete combustion, are usually less than 0.5 µm in size and can reach the terminal bronchioles, where they initiate an inflammatory reaction, leading to bronchospasm, oedema and respiratory failure.

Toxic inhalants are divided into three main groups: (1) tissue asphyxiants; (2) pulmonary irritants; (3) systemic toxins. The two major tissue asphyxiants are carbon monoxide and hydrogen cyanide. Carbon monoxide poisoning is a well-known consequence of smoke inhalation injury. Severe carbon monoxide poisoning will produce brain hypoxia and coma, with loss of airway protective mechanisms, resulting in aspiration that exacerbates the pulmonary injury from smoke inhalation (Table 22.4). The tight binding of the carbon monoxide to the haemoglobin, forming carboxyhaemoglobin, is resistant to displacement by oxygen, and so hypoxia is persistent. Hydrogen cyanide is formed when nitrogen-containing polymers such as wool, silk, polyurethane or vinyl are burned. Cyanide binds to and disrupts mitochondrial oxidative phosphorylation, leading to profound tissue hypoxia.

Table 22.4 Diagnosis of carbon monoxide poisoning

Carbon monoxide level	Physical symptoms
<20%	No physical symptoms
20–30%	Headache and nausea
30–40%	Confusion
40–60%	Coma
>60%	Death

Depth of burns The depth of a burn is classified according to the degree and extent of tissue damage:

1 *Epidermal burns (first-degree)* involve only the epidermis and cause reddening and pain without blistering. They heal within 7 days and require only symptomatic treatment.
2 *Partial-thickness burns (second-degree)* extend into the dermis. They can be subdivided into superficial partial-thickness and deep partial-thickness burns.
   - In *superficial partial-thickness* burns, the epidermis and the superficial dermis are injured. The deeper layers of the dermis, hair follicles, and sweat and sebaceous glands are spared. A common cause is hot water scalding. There is blistering of the skin and the exposed dermis is red and moist at the blister's base. These burns are very painful to touch. There is good perfusion of the dermis with intact capillary refill. Superficial partial-thickness burns heal in 14–21 days, scarring is usually minimal, and there is full return of function.
   - *Deep partial-thickness* burns extend into the deep dermis. There is damage to hair follicles as well as sweat and sebaceous glands, but their deeper portions usually survive. Hot liquids, steam, grease or flames usually cause deep partial-thickness burns. The skin may be blistered and the exposed dermis is pale white to yellow. The burned area does not blanch, has no capillary refill and has no pain sensation. Deep partial-thickness burns may be difficult to distinguish from full-thickness burns. Healing takes 3 weeks to 2 months. Scarring is common and is related to the depth of the injury. Surgical debridement and skin grafting may be necessary to obtain maximum function.
3 *Full-thickness burns (third-degree)* involve the entire thickness of the skin, and all epidermal and dermal structures are destroyed. They are usually caused by flame, hot oil, steam, or contact with hot objects. The skin is charred, pale, painless and leathery. These injuries will not heal spontaneously, as all dermal elements are destroyed. Surgical repair and skin grafting are necessary, and there will be significant scarring.
4 *Full-thickness burns + (fourth-degree)* are those that extend through the skin to the subcutaneous fat, muscle and even the bone. These are devastating, life-threatening injuries. Amputation or extensive reconstruction is sometimes required.

THERMAL BURNS – RECOGNITION

The initial assessment of burns takes place during the primary survey, and it is designed to recognize immediately life-threatening injuries compromising the airway, breathing and circulation and conscious level. The likelihood of coincidental traumatic injuries should be remembered.

The patient is examined following the *look, listen, feel* format. Diagnosis of an inhalational burn is made from the history of a fire in an enclosed space and physical signs that include facial burns, singed nasal hair, soot in the mouth or nose, hoarseness, carbonaceous sputum and expiratory wheezing. There is no single method capable of demonstrating the extent of inhalation injury. Stridor is a particularly sinister finding, as it indicates an imminent loss of the airway.

Carboxyhaemoglobin levels for carbon monoxide poisoning are useful to document prolonged exposure within an enclosed space with incomplete combustion, as the classically described cherry red skin colour is rare.

The chest X-ray may be normal initially; bronchoscopy and radionuclide scanning are useful in determining the full extent of injury. Arterial blood gas analysis will track hypoxia, ventilatory failure and the development of metabolic acidosis. Signs of shock are looked for, as detailed previously, and the GCS and pupillary response assessed. The patient is fully exposed to allow evaluation of the whole-body surface area.

The burnt areas are assessed for depth of burn. This is a subjective clinical assessment. The extent of the burn is assessed and expressed as a percentage of body surface area (BSA). This can be done using the 'rule of nines' (Figure 22.31), or with aids such as the Lund and Browder charts. The rule of nines is an approximate tool and tends to overestimate the extent of a burn.

For irregular burns, the palmar surface of the patient's hand, including the fingers, represents approximately 1% of the patient's body surface area.

Body surface areas are different in infants; they have a disproportionately larger head surface area and smaller lower limb surface area (Figure 22.32).

THERMAL BURNS – MANAGEMENT

The airway is secured as described previously. Inhalational burns can cause pharyngeal oedema and swelling, which can make tracheal intubation difficult if not impossible. This may leave a surgical airway as the only recourse. The airway may need fibre-optic assessment, and warning signs such as stridor and respiratory distress indicate the need for early intubation. This should be performed under general anaesthesia by an experienced anaesthetist, with a range of equipment for difficult intubation available (e.g. McCoy laryngoscope – Figure 22.33). Needle cricothyroidotomy and surgical airway sets should be immediately accessible.

Breathing should be supported with high-flow oxygen administered via a non-rebreathing, reservoir mask that delivers 85% at a flow rate of 15 L/min.

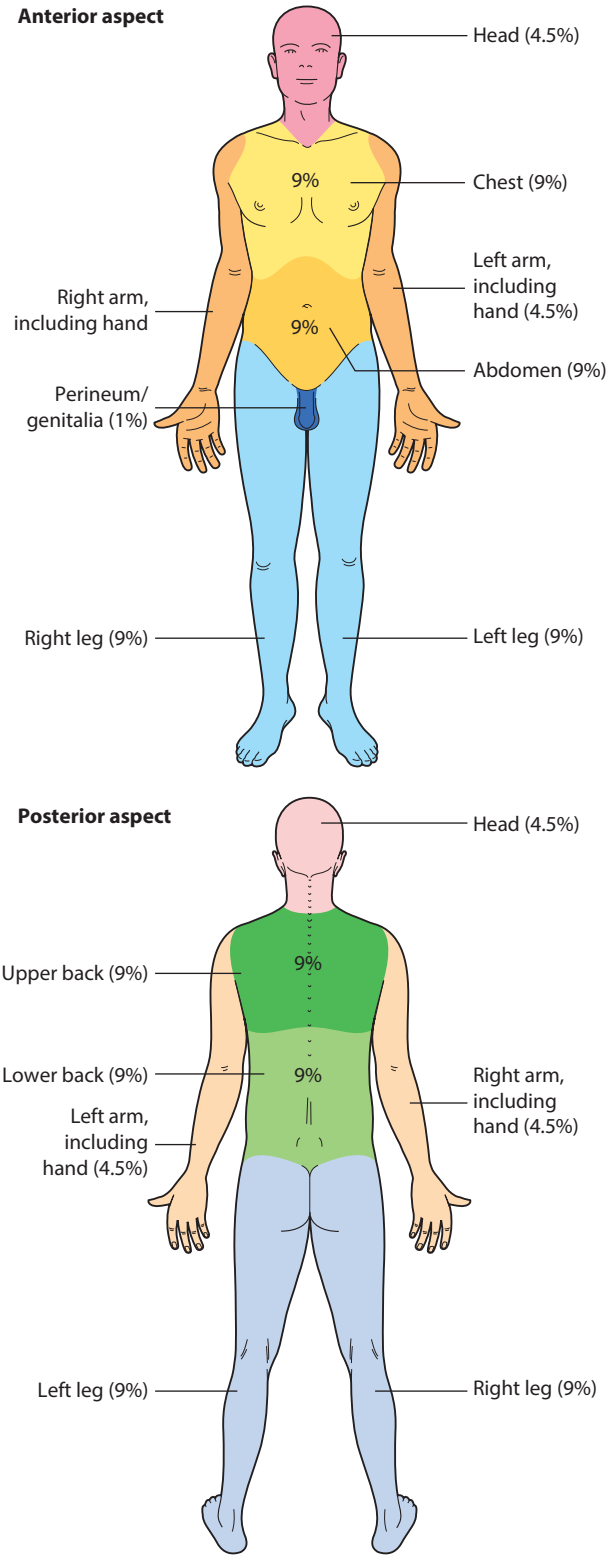


Figure 22.31 Burns 'Rule of nines' for assessment of extent of burns in adults.

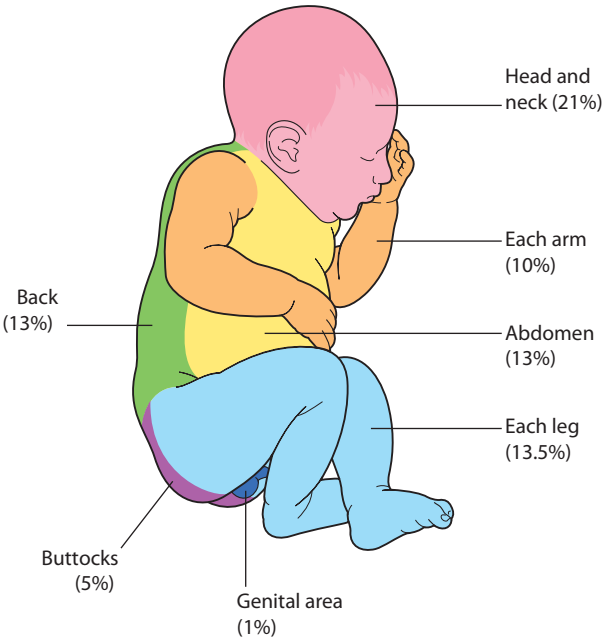


Figure 22.32 Burns in infants Surface areas differ markedly from those in adults.

Figure 22.33 McCoy flexing tip laryngoscope

The ventilation may need support using a BVM assembly with a reservoir and high-flow oxygen. Stridor can be eased, as a holding measure pending airway securement, by administering high-flow helium and oxygen, as this gas mixture has a low density that increases flow through the obstructing airway. However, heliox is only 21% oxygen and will not address hypoxia and carbon monoxide poisoning. Once the airway has been secured by tracheal intubation, the inspired oxygen concentration and ventilation should be adjusted to give optimum SaO_2 levels (>95%) and low normal $EtCO_2$ (4.5 kPa).

The presence of an inhalational burn and pulmonary oedema may hinder oxygenation and ventilation, and a critical-care physician should be involved early. Significant carbon monoxide levels may indicate the need for ventilation with 100% oxygen and hyperbaric therapy, and an early referral should be made to a hyperbaric unit; these are often found located

in diving and naval centres. Circumferential neck and chest burns may need to be incised to allow effective breathing and ventilation.

The circulation should be supported in any burn patient with signs of shock or a burn greater than 20% BSA. Two large-bore intravenous cannulae are sited, preferably although not necessarily, through unburned skin. If intravenous cannulation or central venous cannulation is not possible, intraosseus or intravenous cut-down techniques should be used, as shock will develop rapidly in patients with large and deep burns.

Warmed Hartmann's or Ringer's lactate is the fluid of choice; large volumes of normal saline 0.9% can cause a hyperchloraemic acidosis. Colloids and hypertonic saline have no proven beneficial role. If haemorrhagic shock is excluded, the volume and rate of fluid administration is calculated according to the Parkland formula, as given in Table 22.5. This regimen applies to partial- and full-thickness burns only; superficial burns do not require intravenous fluids. The administration time is calculated from the time of the burn, not from the time of admission or time of assessment. Deeper burns are likely to cause more tissue damage and consequent fluid shifts. The Parkland formula is a guide only and fluid administration should be titrated against response. Blood pressure, central venous pressure, pulse, peripheral perfusion and urine outputs are used, but more sophisticated techniques such as oesophageal Doppler and arterial waveform analysis may aid optimization. Fluid overload should be avoided in patients with inhalational burns and systemic inflammatory reactions. Documented anaemia may indicate the need for blood transfusion.

Wound care starts in the pre-hospital environment with the removal of burnt clothing and the cooling and dressing of wounds. Rings, jewellery, watches and belts are removed as they retain heat and can cause compression as tissues swell. Wounds can initially be dressed with loose, clean, dry dressings. Alternatives are plastic sandwich wrap (known as cling film in the UK, plastic wrap in USA and cling wrap in Australia), specialized gel burns dressings or saline-moistened dressings. Cooling eases pain, but hypothermia should be avoided.

Patients with circumferential deep burns of the limbs may develop eschars (thick, black, dry and necrotic tissue that constricts) with compromise of the distal circulation. Distal pulses need to be monitored closely, with a Doppler probe if not easily palpable. If there is compromise to the circulation, surgical escharotomy will be needed. The eschar should be incised on the midlateral side of the limb, allowing the fat to bulge through. This may be extended to the hand and fingers. Escharotomy may cause substantial soft-tissue bleeding.

Analgesia will be required for partial-thickness burns, which are most painful. Cooling and dressing will help, but opioids may be required. These should be administered intravenously, and can be given by infusion or patient-controlled analgesia (PCA) systems.

Consultation is important. A burns specialist should be involved from the outset for all patients with severe or unusual burns. Transfer will be required for these patients as outcomes are improved in specialist centres. *Indications for transfer* are:

- partial-thickness burns > 20% BSA
- partial-thickness burns > 10% BSA in ages 10–50 years
- full-thickness burns > 5% any age
- partial- and full-thickness burns involving: face, eyes, ears, hands, feet, genitals, perineum, skin over major joints
- significant electrical burns (and lightning)
- significant chemical burns
- inhalational burns
- burns in patients with complicating illness, trauma and long-term rehabilitation needs
- children.

CHEMICAL BURNS

Awareness Most chemical burns result from exposure of the skin to strong alkalis and acids, and phosphorus, phenol and petroleum products can also damage tissue. However, 25 000 products are capable of causing chemical burns, and they account for 5–10% of US burns centre admissions. Full development of chemical burns is slower than thermal injury, so the true extent of the burn can be underestimated on initial evaluation. Alkali burns tend to be more serious and deeper, as the alkalis soften and penetrate tissue, whereas acids tend to form a protective eschar.

Table 22.5 Intravenous fluid requirements in partial- and full-thickness burn patients (Parkland formula)

Adults	Children
Hartmann's or Ringer's lactate: 4 mL × weight (kg) × per cent BSA over initial 24 hours Half over first 8 hours from the time of burn (other half over subsequent 16 hours)	*Hartmann's or Ringer's lactate:* 3 mL × weight (kg) × per cent BSA over initial 24 hours plus maintenance Half over first 8 hours from the time of burn (other half over subsequent 16 hours)
Example: An adult weighing 70 kg with 40% second- and third-degree burns would require 4 mL × 70 kg × 40 = 11 200 mL over 24 hours.	

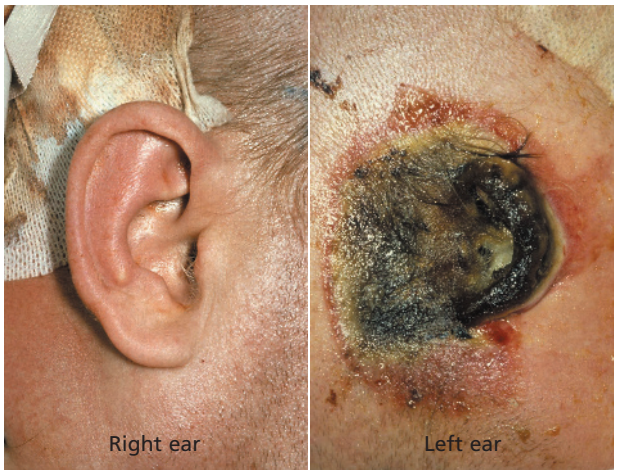


Figure 22.34 Chemical burns Sulphuric acid burn to left ear from car battery acid in roll-over traffic accident.

Recognition Definitive diagnosis depends on the history, and both the chemical involved and its concentration should be determined if possible. Alkali burns are frequently full-thickness injuries, appear pale, and feel leathery and slippery or soapy. Acid burns are often partial-thickness injuries and are accompanied by erythema and erosion. Skin is stained black by hydrochloric acid, yellow by nitric acid, and brown by sulphuric acid (Figure 22.34).

Management The goal of treatment is to minimize any area of irreversible damage, and maximize salvage in the zone of reversible damage. If dry powder is present, it should be brushed off before irrigation with water, which is the mainstay of treatment. Irrigation should be commenced immediately when the injury is recognized, with copious amounts of tap water. Neutralizing agents (e.g. an acid to treat an alkali burn) should not be used, as there is a risk that heat generated by the neutralizing reaction will cause further thermal injury.

After copious water irrigation, some specific treatments are possible, such as calcium gluconate for hydrofluoric acid burns and polyethylene glycol for phenol. An urgent referral to a burns surgeon should be made; eschar formation may make irrigation ineffective and require emergency surgical excision.

ELECTRICAL BURNS

Awareness Electrical burns are caused when an individual makes contact between an electrical source and the earth, and severe, non-lethal electrical injuries constitute 3–5% of admissions to burns units in the USA. Current flows through the skin and variably through different tissues from the point of electrical contact to the ground contact, causing burns and necrosis along its path. The physiological effects of an electric shock are related to the amount, duration, type (AC or DC), and path of current flow. Severe electrical skin burns are associated with high-voltage shocks, whereas most domestic, low-voltage shocks are not associated with skin burns even though they may cause death from ventricular fibrillation. Alternating current (AC) shocks produce tetanic muscle spasm, which can cause the victim's hand to clutch onto the electrical source, and the respiratory muscles can be paralyzed, resulting in respiratory arrest. Electrical muscle damage can result in rhabdomyolysis and renal failure, with hugely raised serum, creatine kinase levels.

Recognition The assessment of an electrical shock victim should follow the ABC principles of ATLS®. The airway may be obstructed if the victim is unconscious, and prolonged apnoea may follow paralysis of the respiratory muscles. The heart may be arrested in ventricular fibrillation or asystole depending on the nature of the shock. Of high voltage electrical shock victims, 50% will have a neurological injury with coma, and spinal injuries can result from violent muscle spasms. The entry and exit points should be examined for burns that may be full thickness, and the true extent of underlying muscle damage may not be apparent. There may be musculoskeletal injuries from associated trauma or muscle spasm and all long bones should be examined and X-rayed when indicated.

Management The immediate priority is to avoid rescuer injury if the casualty is in contact with or even adjacent to a high-voltage electrical source. Power sources must be disconnected; be aware that electrified rail lines retain an electrical charge after disconnection and a rail casualty should not be approached until a rail engineer has earthed the circuit and confirmed it is safe.

Initial management is to secure the airway, protect the cervical spine and oxygenate and ventilate the casualty. Intravenous access is secured, and fluids administered if the casualty is shocked. If in cardiac arrest, advanced life support should be instituted, following the appropriate Advanced Life Support algorithms for VF/VT and non-shockable arrests as indicated.

The heart should be monitored for arrhythmias, which can occur in 30% of high-voltage shock victims. Tissue damage may need surgical debridement, and compartment syndrome may develop, requiring fasciotomies. A urinary catheter is sited, and the urine observed for the brown discoloration indicative of development of myoglobinuria; this is treated by giving intravenous fluids to promote a diuresis, and administration of mannitol. Myoglobinuria should be considered present if a urine dipstick test registers positive for haemoglobin, but the freshly spun urine sediment shows no red blood cells.

As ongoing treatment will be complex in severe electrical injuries and burns, early consultation should be made with a burns surgeon and critical care specialist. Management on a critical care unit will be required.

COLD INJURY BURNS

Awareness Cold injury can be systemic, leading to hypothermia, or localized, leading to localized tissue damage to varying degrees dependent on the degree of freezing.

Hypothermia is defined as a core body temperature of below 35 °C (95 °F). The systemic effects depend on the severity of the drop in core temperature:

Mild	35–32 °C (95–89.6 °F)
Moderate	32–30 °C (89.6–86 °F)
Severe	<30 °C (<86 °F)

As the core temperature drops, the conscious level deteriorates, and the airway can obstruct as coma develops. Respiratory and cardiac functions deteriorate until respiratory and cardiac arrest result.

Localized cold injury is seen in three forms:

1 *Frostnip* – the mildest form, which is reversible on warming.
2 *Frostbite* – due to freezing of tissue and resultant damage from intracellular ice crystals and microvascular occlusion. This can be classified as two types and four degrees:
   - *Superficial frostbite:*
     - First-degree – hyperaemia and oedema without skin necrosis.
     - Second-degree – vesicle formation with partial-thickness skin necrosis.
   - *Deep frostbite:*
     - Third-degree – full-thickness and subcutaneous tissue necrosis, with haemorrhagic vesicle formation.
     - Fourth-degree – full-thickness necrosis, including muscle and bone gangrene.
3 *Non-freezing injury* – trench foot or immersion foot, with microvascular endothelial damage, stasis and vascular occlusion.

Recognition Systemic cold injury is recognized in the primary survey as the airway, breathing and circulation and neurological function are assessed. The patient is cold to the touch, and looks grey and peripherally cyanosed. Strikingly, the expired breath can feel deathly cold on the hand. A low-reading rectal or oesophageal temperature probe will be needed to accurately gauge the degree of hypothermia.

Local injuries are assessed during the secondary survey and the musculoskeletal survey. The affected part of the body initially appears hard, cold, white and anaesthetic, but the appearance changes frequently during treatment.

Management *Hypothermia* is treated by securing the airway, oxygenating and ventilating the patient to normal parameters, gaining intravenous access and treating shock with warmed intravenous fluids. In addition, the patient is rewarmed depending on the degree of hypothermia.

Mild and moderate hypothermia are treated by active external rewarming:

- Heated blankets, warm baths, forced hot air – it is easier to monitor and perform diagnostic and therapeutic procedures using heated blankets.
- Warm bath rewarming is best done in a bath of 40–42 °C moving water (rewarming rate: ~1–2 °C/hour). The warming gradient should not be greater than this to avoid thermal injury. Rewarming should be slow to minimize peripheral dilation, which can cause hypovolaemic shock.

Severe hypothermia and hypothermic cardiac arrest require active internal (core) rewarming:

- Extracorporeal blood rewarming (cardiopulmonary, venovenous, or arteriovenous femorofemoral bypass) is the treatment of choice, especially with cardiac arrest.
- Without equipment for extracorporeal rewarming, left-sided thoracotomy followed by pleural cavity irrigation with warmed saline and cardiac massage is effective in systemic hypothermia <28 °C.
- Thoracic lavage or haemodialysis is also effective.
- Carry out repeated peritoneal dialysis with 2 L of warm (43 °C) potassium-free dialysate solution exchanged every 10–12 minutes until core temperature is raised to ~35 °C.
- Parenteral fluids should be warmed to 43°C.
- Administer humidified air heated to 42 °C through a face mask or tracheal tube.
- *NOTE:* warm colonic and gastrointestinal (GI) irrigations are of less value.

Localized cold injury is initially managed in the field. The hypothermia and dehydration associated with frostbite should be addressed. Wet and constrictive clothing should be removed, the involved extremities should be elevated and wrapped carefully in dry sterile gauze, and affected fingers and toes should be separated. Further cold injury should be avoided. Rapid rewarming is the single most effective therapy for frostbite. As soon as possible, the injured extremity should be placed in gently circulating water at a temperature of 40–42 °C (104–107.6 °F) for approximately 10–30 minutes, until the distal extremity is pliable and erythematous. The current consensus is that clear blisters are aspirated or debrided and dressed. Early surgical intervention in the form of tissue debridement and amputation is not indicated; full demarcation of dead tissue can take 3–4 weeks to occur, and debridement at this point will avoid unnecessary tissue loss.

BURNS – TAKE-HOME MESSAGE

- *Thermal burns* are assessed by depth and extent, and managed by addressing the airway, breathing and circulation.
- Huge volumes of intravenous fluids may be required to maintain homeostasis.
- *Chemical burns* are treated primarily by copious irrigation with water.
- *Electrical burns* may be associated with severe tissue damage and systemic disturbance, and treatment is needed for local burns and systemic cardiac, respiratory and renal complications.
- *Cold injury* can be systemic hypothermia, which is treated by active external and internal rewarming, depending on severity, or localized tissue damage. Localized tissue damage is treated by rapid rewarming and delayed surgical debridement.

INITIAL RESPONSE TO TRAUMA

The physiological effects of trauma are both widespread and predictable, provoking a range of hormonal and cellular mechanisms that have evolved to maximize survival following serious injury. These physiological adaptations are considered as a whole-body strategy for fluid conservation and repair.

Following injury the first survival offensive is a plan to limit blood loss. Direct injury to blood vessels should induce an arterial vasospasm to reduce blood loss followed by the formation of a 'vascular patch' consisting of a fibrin-reinforced aggregation of platelets.

If significant blood loss still occurs, intravascular volume is supported by fluid redistribution between the vascular, cellular and interstitial fluid compartments. The resulting change in compartmental volumes stimulates an endocrine response with the release of a number of renal, adrenal and pituitary hormones (renin, aldosterone, cortisol and antidiuretic hormone [ADH]). This hormonal response constitutes a secondary fluid conservation project and another survival strategy.

Serious injury, which in evolutionary terms would have limited the ability to hunt and feed, produces a metabolic reconditioning. Under endocrine guidance, cellular metabolic priorities and metabolic substrates associated change with a falling basal metabolic rate. These metabolic changes represent an approach to energy conservation, allowing rationing of substrates to allow damage control and repair while still keeping cerebral metabolism as the priority.

Ultimately, a successful outcome following trauma (or major surgery) depends on the integration of these strategies and the maintenance of whole-body physiology. The integrity of the cardiorespiratory system is pivotal. Failure to maintain cellular (organ) perfusion, oxygenation and ATP regeneration will lead to cellular bioenergetics failure, cell apoptosis and death. Any comorbidities present such as pre-existing lung disease or cardiac failure will increase complications and the chance of dying.

The normal physiological response to the increased metabolic demands of trauma, illness and surgery is to increase oxygen delivery in response to an increase in tissue oxygen consumption (Figure 22.35). Failure to respond to this demand will generate an oxygen debt with metabolic consequences. This limitation of oxygen availability will favour anaerobic metabolism over aerobic, reducing metabolic efficiency and generating a lactic acidosis as a consequence. This is clearly unsustainable and clinical studies show that an inability to mount a sustained cardiovascular response is directly proportional to an increase in morbidity and mortality. Survival and outcome relies on the speed of repayment of this oxygen debt. The slower the payback, the greater the ensuing complications.

As a synopsis, trauma and major surgery can be considered to be like running a marathon. To survive, cardiorespiratory function and cellular physiology have to remain intact. Systemic failure, for whatever reason, to maintain tissue perfusion leads to shock, which is one of the most frequently misused and misunderstood terms in medicine and the media. Correctly used it implies tissue hypoperfusion leading to cellular hypoxia and describes a medical emergency with a high mortality rate from multiple organ failure.

From an intensive care perspective, the recognition and appreciation of the type of shock is essential as other reasons for hypoperfusion may coexist (Figure 22.36).

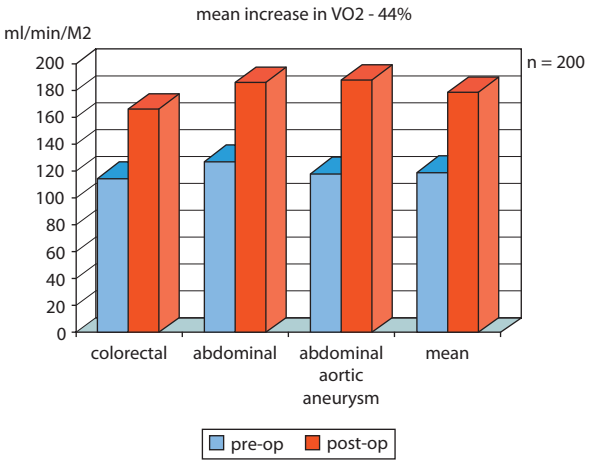


Figure 22.35 Oxygen consumption before and after surgery (Older and Smith. 1988).

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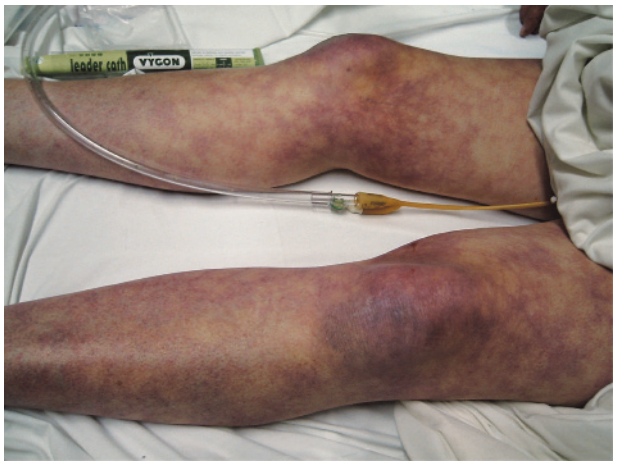


Figure 22.36 Hypoperfusion This 70-year-old man with severe sepsis developed hypoperfusion of the lower limbs. Note the typical marbling of the skin.

SHOCK

In health, cardiac output and the delivery of oxygen (global arterial blood flow multiplied by the blood oxygen content) and local tissue perfusion are closely matched to metabolic requirements. Shock follows a mismatch of metabolic demand to oxygen delivery at tissue level, leading to cellular hypoxia and (if uncorrected) to tissue and organ failure. The causes of circulatory shock can be classified as abnormalities of cardiac output, of systemic vascular resistance, or a combination of both (Box 22.5).

Reduced cardiac output

Impaired performance *Cardiogenic shock* is an intrinsic failure of cardiac function despite adequate circulating volume and venous return, most commonly as a result of acute myocardial infarction. Cardiogenic shock may occur following an apparently minor insult to a heart with any pre-existing functional impairment.

Impaired venous return *Hypovolaemic shock* exists when a fall in circulating volume of sufficient magnitude occurs such that compensatory physiological mechanisms are unable to maintain adequate tissue flow, leading to critical hypoperfusion.

Obstructive shock '*Obstruction*' arises when venous return is compromised by raised intrathoracic or pericardial pressure (pneumothorax and cardiac tamponade), or if right ventricular ejection is blocked by a massive pulmonary embolus, resulting in right ventricular overload and impaired left heart filling. Plain X-rays may not show changes and CT angiography is the initial investigation of choice.

Reduced systemic vascular resistance

Neurogenic shock This occurs when spinal cord injury – usually at a cervical or high thoracic level – leads to loss of sympathetic tone and hence peripheral vasodilatation, venous pooling and reduced venous return. This is aggravated by the absence of direct sympathetic nervous system connection into the heart, and hence impaired compensatory responses.

Anaphylactic shock A drug or parenteral fluid may be the trigger that provokes an immunological response with histamine release, resulting in cardiovascular instability and (potentially) respiratory distress.

Septic shock This condition is defined as severe sepsis with associated hypotension, evidence of tissue hypoperfusion that is unresponsive to fluid resuscitation. Various mechanisms are responsible for the vasodilatatory response and catecholamine resistance, which are characteristic of septic shock. It is becoming clearer that this host response does not appear to be determined by the infecting organism and there is a suggestion of genetic susceptibility being a contributory factor in dictating the severity of subsequent illness.

BOX 22.5 AETIOLOGY OF CIRCULATORY SHOCK

Reduction in cardiac output

- HYPOVOLAEMIC SHOCK – reduced circulating volume causing a reduction in venous return and cardiac output, e.g. haemorrhage
- OBSTRUCTIVE SHOCK – mechanical obstruction to normal venous return or cardiac output, e.g. tension pneumothorax, cardiac tamponade or massive pulmonary embolism
- CARDIOGENIC SHOCK – failure of cardiac pump to maintain cardiac output, e.g. post myocardial infarction

Reduction in peripheral resistance

- DISTRIBUTIVE SHOCK – a drop in peripheral resistance due to vasodilatation, which is often associated with an increase in cardiac output but not sufficient to maintain blood pressure, e.g. anaphylaxis, neurogenic shock, SIRS, septic shock
- ENDOCRINE SHOCK – in the intensive care setting hypothyroidism, hyperthyroidism and adrenal insufficiency can all lead to reduced tissue perfusion

Diagnosis of shock

Early recognition, immediate resuscitation and treatment of the underlying cause are the cornerstones of successful therapy.

There may be an easily identifiable cause of shock, but often the aetiology is difficult to establish. Following massive trauma, shock may be hypovolaemic (blood loss), obstructive (tamponade or tension pneumothorax), cardiogenic (cardiac contusion), neurogenic (spinal cord injury) or anaphylactic (drug reaction).

Careful examination should clarify the aetiology in most cases and will aid in determining severity by identifying end-organ effects. Examination should be thorough and structured to avoid missing useful signs (Box 22.6).

BOX 22.6 CLINICAL EXAMINATION IN SHOCK

Cardiovascular system

- Pulse (rate/rhythm)
- Blood pressure
- JVP (or CVP if central line *in situ*)
- Heart sounds (muffling/murmurs)
- Peripheral perfusion (capillary refill time/skin colour)

Respiratory system

- Respiratory rate
- Work of breathing
- Tracheal deviation
- Air entry
- Added sounds
- Oxygen saturations (relative to inspired oxygen)

Abdomen

- Pain
- Distension
- Peritonitis
- Localizing signs
- Urine output

Central nervous system

- Level of consciousness
- Peripheral neurological signs (e.g. power, reflexes)

Other systems

- Temperature
- Skin signs (e.g. rashes)
- Limbs (bony integrity/perfusion)

Tests should include a full blood count and estimation of electrolytes as well as assessment of renal function, liver function, clotting and blood group/cross-match, serum glucose, blood cultures and inflammatory markers (e.g. C-reactive protein, procalcitonin). Arterial blood gas analysis provides rapid results, and the newer analyzers often measure a serum lactate level. This is a non-specific marker but it may indicate hypoperfusion if elevated.

X-ray examination, ultrasound scanning (e.g. eFAST scan) or CT may identify sources of blood loss and identify likely foci in the case of severe sepsis. An ECG and urgent echocardiography are obligatory if a cardiogenic cause of shock is suspected.

Careful and regularly repeated recording of vital signs (heart rate, respiratory rate, blood pressure, oxygen saturation) and indicators of end-organ perfusion (consciousness level, urine output) are crucial. The initial severity of illness at assessment, and subsequent response to initial resuscitative and treatment measures will dictate the need for more advanced and invasive monitoring tools. Continuous invasive blood pressure and central venous pressure monitoring are generally required and are essential if vasoactive drugs are required, both to enable safe drug delivery and to allow titration of dosing.

Advanced monitoring systems

Figure 22.37 summarizes the investigation and monitoring of shock.

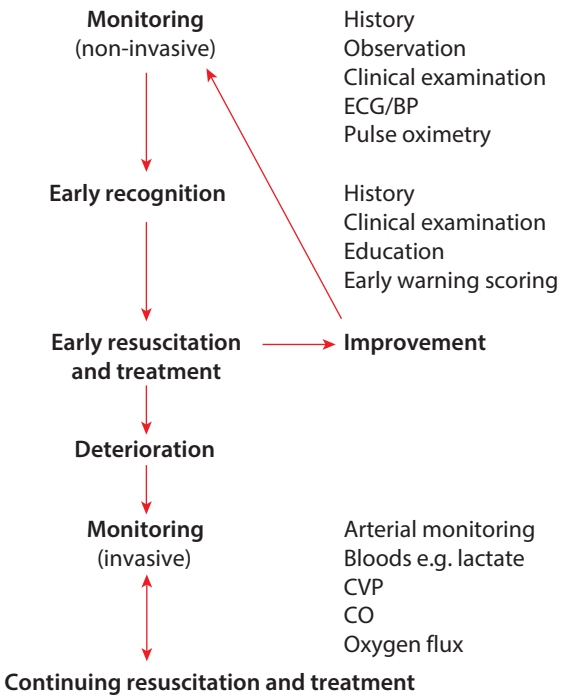


Figure 22.37 Investigation and monitoring of shock

Invasive techniques that allow an estimation of cardiac output – and thereby tissue oxygen delivery – are used in the sickest patients, both as an aid to diagnosis and as a guide to therapy.

PULMONARY ARTERY FLOTATION CATHETERIZATION

In pulmonary artery flotation catheterization (PAFC), a catheter is passed via a central vein through the right heart to rest within a branch of the pulmonary artery. Inflation of the distal balloon permits measurement of the pulmonary artery occlusion pressure (PAOP), which allows an estimate of left atrial pressure and hence (it is assumed) left ventricular preload. Many errors may, however, confound this measurement. The PAFC also allows measurement of cardiac output by way of thermodilution (either by cold injectate or by proximal heating coil, allowing semi-continuous data to be recorded). This is calculated from the area under a curve of distal temperature (recorded by a thermistor at the catheter tip) plotted against time. Cardiac output is inversely proportional to this area.

PAFC use has declined in popularity recently due to concern regarding the complications of what is a highly invasive modality, failure to show outcome benefit in studies of patients monitored by PAFC, and the increasing availability of alternative, less invasive monitors that generate similar data.

CARDIAC OUTPUT FROM ANALYSIS OF ARTERIAL WAVEFORM

Pulse contour analysis The PiCCO® cardiac output monitor employs a mathematical analysis of the shape of the arterial waveform using a dedicated femoral arterial cannula to derive cardiac output data. It is calibrated by a transpulmonary thermodilution technique, following injection of cold saline into a central line.

Pulse power analysis The Lithium Dilution Cardiac Output (LiDCO®) monitor also employs the arterial waveform to derive haemodynamic data but using a power algorithm that can be used in any artery, and it thus does not require insertion of a proprietary arterial line. The monitor is calibrated using either the lithium dilution technique (LiDCO plus) or a nomogram of patient demographics with the LiDCORapid® monitor. As with pulse contour analysis, peripheral resistance and data indicating likely fluid responsiveness are calculated beat-to-beat. It does also have, unlike many other devices, positive outcome data in high-risk patients.

Management of shock

Initial approach Initially attention should be focused on rapid assessment, with airway, breathing and circulation (ABC) addressed in the first instance. High-flow oxygen (FiO_2 0.6 or greater) should be administered via a patent airway and intravenous access should be obtained. Definitive treatment of the underlying cause of shock should be commenced alongside resuscitative measures (Box 22.7). The aim should be to support the circulation to allow adequate tissue oxygen delivery while mitigating or reversing the effects of the initial insult. This may be rapidly successful, for example in decompression of a tension pneumothorax; in other cases it may prove impossible to correct the underlying pathology (e.g. cardiogenic shock due to extensive myocardial infarction).

BOX 22.7 TREATMENT OF THE UNDERLYING CAUSE OF SHOCK

Hypovolaemic
- Control of haemorrhage (may require surgery)
- Restoration of circulating volume (fluid and blood products)

Obstructive
- Needle decompression of tension pneumothorax
- Pericardiocentesis (tamponade)
- Thrombolysis or surgical removal of pulmonary embolus

Cardiogenic
- Inotropes
- Anti-arrhythmics
- Revascularization
- Aortic balloon counterpulsation
- Surgical repair of valve lesions

Distributive
- Early treatment of infection (source control, e.g. drainage, early antibiotic administration)

Fluid therapy Often large volumes are needed, guided by clinical response and monitored indicators of filling (e.g. central venous pressure). The response of these variables to a fluid challenge, and trends, are considerably more useful than 'snapshot' values. Indeed, targeting a particular value of CVP or MAP is physiologically unsound and may be to the patient's detriment. It is always preferable to use fluid boluses or 'challenge techniques' to interpret volaemic status.

In ventilated patients, changes in intrathoracic pressure generate cyclical changes in systolic pressure and using the LiDCO or PiCCO monitors generates a stroke volume variation that is related to volaemic

status under certain conditions. These variations in stroke volume may be more useful indicators of likely fluid responsiveness than other methods.

The choice of fluid is dictated by the underlying cause of the shock and local policies. There is an optimum amount of fluid to target resuscitation and it should be recognized that overenthusiastic transfusion, as with fluid restriction, is also associated with increased complications.

Inotropes/vasopressors This treatment should be instituted if the patient remains hypotensive despite adequate fluid resuscitation. Again, choice is determined by aetiology: vasopressor (e.g. norepinephrine) for distributive shock and inotrope (e.g. dobutamine) for cardiogenic shock. Combinations may be required, guided by haemodynamic data from monitoring equipment and clinical response. Significant doses of either inotropes or vasopressors should be mandatory. Cardiac output monitoring is much better than making decisions based on the arterial blood pressure.

Endocrine support There is recent evidence that treatment with 'physiological' doses of corticosteroid in cases where adrenal response is inadequate may not improve outcomes as had previously been hoped. There is considered to be some benefit from the use of steroids with septic shock with an improvement in haemodynamic response, but this is still the subject of considerable debate and there is a lack of cogent outcome data. The use of vasopressin has traditionally been reserved for patients with catecholamine-resistant septic shock but new evidence suggests that there may be some benefit for those requiring lower doses of noradrenaline.

Tight control of blood glucose levels has also been shown to lead to improved outcomes in the sickest patients in intensive care.

Systemic support Shock leads to multiple organ impairment or failure. Support of other organ systems may well be required during treatment.

Outcome Mortality is determined both by aetiology of circulatory shock and the response to treatment. Early recognition and prompt therapy are the most important factors.

MULTIPLE ORGAN FAILURE

Multiple organ failure or dysfunction syndrome (MODS) is the clinical appearance of a seemingly poorly controlled severe systemic inflammatory reaction, following a triggering event such as infection, inflammation or trauma. It represents the net result of altered host defence and deregulation of the inflammatory response and the immune system. The condition has emerged with medical advances as a result of increasing availability of intensive care facilities. Recognized as a syndrome in the early 1970s, progress in the management of critically ill patients has unmasked this frequently lethal cocktail of sequential pulmonary, hepatic and renal failure.

This pattern of progressive organ impairment and failure complicates illnesses with diverse aetiologies and, despite progress in understanding the underlying mechanisms involved, it carries a mortality rate that remains depressingly high. MODS has now become the commonest cause of stays in surgical ITUs of more than 5 days and (among these patients) the most frequent cause of death.

It is essential to differentiate MODS from postoperative or traumatic, isolated organ dysfunction, which has a different pathogenesis and markedly different survival outcomes.

Epidemiology

Definitions of organ failure use two types of criteria based either on measures of physiological derangement (e.g. hypotension, acidosis, serum creatinine concentration) or on the treatment methods (e.g. dialysis, ventilation, etc.).

The degrees of organ dysfunction, from covert physiological impairment to overt failure, coupled with the difficulties of monitoring the function of all the organs involved has led to controversies about the definition of organ failure and the clinical entities involved. This has hampered epidemiological surveys and the assessment of treatment outcomes. Confusion over the exact incidence of MODS stems from an absence of universal diagnostic criteria; many of the published studies have used differing clinical and temporal definitions of organ failure.

Review of the published studies suggests that MODS develops in 5–15% of patients requiring ICU admission, depending on the diagnostic criteria used and the case-mix of the population of ICU patients studied. The outcome data is remarkably consistent between the studies, with mortality linked to the number of organs failed.

The appearance of MODS broadly follows two clinical courses, differing in onset relative to the initial event, time course and sequence of organ failure. The first pattern usually follows a direct pulmonary insult, such as trauma or aspiration. In this form the overall course of the disease may be relatively short and MODS occurs as a pre-terminal event, becoming evident just prior to death. The second type is the more classical form, as found in severe sepsis, with pulmonary manifestations of acute respiratory distress syndrome (ARDS). MODS is present early in the course of the illness but does not become progressive until

BOX 22.8 INITIATING EVENTS FOR MODS

Severe sepsis
- Peritonitis

Trauma
- Chest injury
- Burns

Shock
- Cardiogenic
- Haemorrhagic

Surgery
- Vascular
- Abdominal

Medical
- Pancreatitis
- Aspiration

Other
- Massive transfusion

after a 7–10-day delay, with manifestations of hepatic and subsequently renal failure becoming apparent.

The initiating events for MODS are many and diverse but by far the most common association is with severe sepsis and ARDS (Box 22.8). The likelihood of occurrence and the progression of disease are related not only to the severity of the initiating event but also to the premorbid physiological reserve of the patient, i.e. old age and pre-existing disease such as cardiac failure, cirrhosis, drug abuse, etc.

Pathogenesis

MODS is now recognized as a systemic disorder resulting in widespread microvascular injury. Most of the initiating events can be characterized as infective, traumatic or ischaemic, and mechanistically it is unravelling as a disorder of the host defence system, with an unregulated and exaggerated immune response, resulting in an excessive release of inflammatory mediators. It is these mediators that produce the widespread microvascular damage leading to organ failure.

As a syndrome, the classical form of MODS appears to progress through four clinical phases:

1 Shock (hypoperfusion)
2 Period of active resuscitation
3 Stable hypermetabolism (systemic inflammatory response)
4 Organ failure.

Shock Common to all the initiating events associated with MODS are periods of relative or total ischaemia relating to regional or global perfusion deficits, which may go clinically unrecognized, i.e. cellular hypoperfusion as discussed earlier. The severity of these deficits, the passage of time to adequate resuscitation and the reserve functional capacity of the organs concerned, appear to provide the key to the path of organ dysfunction and eventual failure.

BOX 22.9 CLINICAL FEATURES OF SIRS

Fever
Tachycardia
Hyperdynamic circulation
Tachypnoea
Oliguria

Active resuscitation If resuscitation is rapid and effective, the sequence of events precipitating MODS may be aborted. However, in many cases, despite apparently adequate management, the syndrome progresses, suggesting a genetic component.

Systemic inflammatory response If resuscitation fails to prevent further progression of the disease, the presence of widespread cellular damage manifests after several days with a picture of panendothelial dysfunction. This endothelial damage is manifest by increased microvascular permeability with the formation of protein-rich oedema fluid. This period of hypermetabolism has characteristic features that are a consequence of the host response. This has been referred to as the systemic inflammatory response (SIRS) (Box 22.9) in the absence of proven sepsis and the sepsis syndrome when associated with an identifiable invading pathogen. Once this phase is entered the mortality rises to the 25–40% range.

Organ failure Failure to adequately control the inciting event and the inexorable progression of the disease is marked in this final stage by increasing organ dysfunction, failure and death. The appearance of clinically overt organ failure is a significant prognostic event signalling another leap in the mortality rate from the 25–40% range to 40–60% in the early stages and 90–100% as the disease progresses with increasing hepatic and renal dysfunction.

MEDIATORS OF THE SIRS/SEPSIS RESPONSE AND MODS

The metabolic and physiological alterations found in the hyperdynamic-hypermetabolic phase and the subsequent cellular damage are caused by complex interactions of endogenous and exogenous mediators. These substances are mainly released from the host endothelial and reticuloendothelial cells, principally macrophages, in response to provocation by a variety of stimuli including ischaemia, sepsis and cytokines. Experimental administration of endogenously produced mediators such as tumour necrosis factor (TNF), interleukins IL1, IL2 and IL6 and platelet-activating factor and exogenously produced mediators such as bacterial endotoxin produce not only similar physiological effects to those

found in the SIRS/sepsis syndrome, but also organ dysfunction similar to that found in patients with MODS.

The wide variety of substances with vastly differing molecular structures implicated in the pathogenesis of the SIRS/sepsis syndrome, all producing the same characteristic physiological response, suggests a 'preprogrammed' or stereotyped host reaction. The effector systems involved in the translation of triggering injury to pathogenesis of MODS are additive and synergistic, and they involve not only the endocrine and central nervous systems but also the cellular and humoral components of the inflammatory responses. Following injury a local inflammatory response occurs resulting from the products of the damaged endothelium and platelets. Leucocytes and macrophages are presumably attracted to the area as a result of these products and secondary activation of complement, coagulation and other components of the inflammatory system occurs. If the injury is severe or persistent enough, this localized reaction may spill over into the systemic circulation, producing the systemic inflammatory response or, if identified with infection, the sepsis syndrome. MODS may subsequently develop.

In health, cytokine production is strongly repressed since cytokines are produced by immune cells following activation by foreign particles such as bacteria. Cytokine induction and production is then closely regulated so as to benefit the host by localizing and destroying the foreign organisms. However, in certain situations, this control system appears inadequate and cytokine production becomes both inappropriate and excessive, leading to destruction of normal cells with a generalized inflammatory response.

A decade of studies has underlined the importance of the immune system and these mediators in the sequence of events ultimately producing MODS. Interleukin-1 is the most extensively investigated cytokine; produced by macrophages, this polypeptide (as well as interleukin-6) can induce fever, hypermetabolism, muscle breakdown and hepatic acute phase protein synthesis. The interleukins, however, appear relatively late in the sequence of events compared to TNF.

TNF appears early in the systemic circulation during critical infective illness, mediating directly or indirectly many of the major features of sepsis. It is probably one of the pivotal mediators with multiple effects, producing endothelial membrane permeability changes and cell death. Many of these effects appear to be secondarily mediated by prostaglandins and TNF-induced release of other cytokines; the full extent of its actions is poorly understood.

SPECIFIC ORGAN INVOLVEMENT IN MODS

The physiological effects of MODS are summarized in Figure 22.39.

Respiratory system

In the majority of critically ill patients who develop MODS the lungs are the first organs to fail, the other organs following in a sequential fashion. The lung appears to be a pivotal organ in the development of MODS, appearing either to generate inflammatory mediators that aggravate peripheral endothelial dysfunction or to allow the persistence of mediators in the circulation following its decreased capacity to clear and metabolize inflammatory substances.

As with other organs, a spectrum of dysfunction exists ranging from minor demonstrable pathology, designated acute lung injury (ALI), to massive alterations in pulmonary pathophysiology – the so-called acute respiratory distress syndrome (ARDS).

ARDS has been defined as a condition characterized by severe hypoxia despite high concentrations of supplemental oxygen, with a radiographic appearance demonstrating diffuse infiltrates in the absence of infection or any other explanation for the respiratory distress (Figure 22.38). Included in this definition are clinical values reflecting the derangement of respiratory function.

ARDS is considered to be a more severe form of ALI, in which the same criteria apply except that the hypoxia is more severe (PaO_2/FiO_2 < 200 mmHg regardless of positive end-expiratory pressure – PEEP). The pathogenesis of this lung injury has in part been suggested to be endothelial damage initiated by complement activation with subsequent leucocyte aggregation and oxygen free radical formation. Platelet clumping and intravascular coagulation have also been implicated. Pathologically in ARDS pulmonary capillary endothelial damage causes fluid leakage and surfactant abnormalities resulting in alveolar and interstitial oedema and fibrosis. This damage to pulmonary architecture causes a reduction in functional residual capacity, increased ventilation/perfusion mismatching and a predilection for secondary infection. The net result

BOX 22.10 FEATURES DEFINING *ARDS*

Hypoxia (PaO_2/FiO_2 < 300 mmHg)
Bilateral infiltrates on chest X-ray
Pulmonary capillary wedge pressure
<18 mmHg or no clinical evidence of increased left atrial pressure

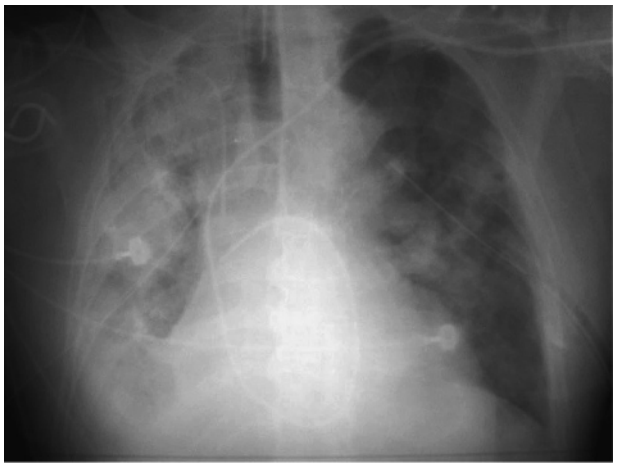


Figure 22.38 ARDS – X-ray Chest radiograph of a patient with ARDS following pulmonary contusion. Infiltrates and patchy consolidation are typical features. Note the pulmonary artery catheter *in situ*.

is failure of gaseous exchange with hypoxia, hypercarbia and therefore an aggravation of the peripheral tissue hypoxia.

Cardiovascular system

Under normal physiological conditions, tissue oxygen utilization is closely matched by its delivery to the tissues. Oxygen uptake by cells is normally dictated by need. Cardiac output, minute ventilation and regional blood flow in the microcirculation are regulated to prevent cellular ischaemia. If stressed in this situation, cells cope with increasing metabolic demands by increasing oxygen extraction. However, under the pathological conditions found in patients with SIRS who are developing MODS, the tissues appear unable to extract oxygen efficiently from the blood, thus resulting in cellular oxygenation having to rely on increased oxygen delivery rather than extraction – the so-called pathological oxygen, supply or flow, dependency.

There may be a number of reasons for this. Microvascular inflammatory injury with endothelial and interstitial oedema hinders the diffusion of oxygen, and furthermore altered membrane characteristics of the erythrocytes render them less deformable and therefore less accessible to transit within the microcirculation.

In the hypermetabolic SIRS phase, the response to increased metabolic demands coupled with less effective utilization of oxygen must be met by an increased cardiac output. This increase, in conjunction with mediator-induced systemic vasodilation, gives rise to the hyperdynamic state characteristic of the SIRS/sepsis syndrome. Failure to meet this increased oxygen demand heralds a diminished likelihood of survival.

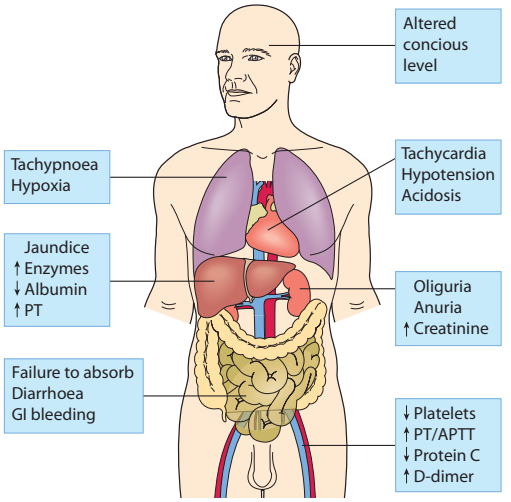


Figure 22.39 Physiological effects of MODS

Poor cardiac performance may also contribute to the oxygen supply–utilization disequilibrium. It is well documented in sepsis that certain circulating factors adversely affect ventricular compliance and contractility. Furthermore, if pre-existing coronary artery disease coexists with this hyperdynamic state, myocardial ischaemia and failure may progressively ensue. The effects of this may not only cause a decrease in organ perfusion but also aggravate existing pulmonary dysfunction with raised left atrial pressures and the generation of pulmonary oedema, further aggravating oxygen delivery.

Gastrointestinal tract

The gastrointestinal tract is particularly vulnerable to the processes occurring in MODS. There is a growing body of evidence to suggest that the persistence of the SIRS/sepsis syndrome may be driven by abnormal colonization of the normally sterile upper gastrointestinal tract with pathogenic enteric bacteria. Some investigators believe that the development of MODS in the absence of a recognized focus of infection is caused by gut failure with translocation of bacteria and toxins from the gut eventually into the systemic circulation. This abnormal colonization of the gut, coupled with potentially toxic gut luminal contents, forms a deadly reservoir of pathogenic substances.

The body relies on the epithelial integrity of the gut wall to prevent seepage of these contents into the circulation. This epithelial barrier is, however, also involved in the systemic disease process, especially as preferential redistribution of the blood from the splanchnic circulation to muscle predisposes the gut mucosa to ischaemia and membrane reperfusion injury. The epithelial barrier is then likely to fail, allowing translocation of pathogenic bacteria, or endotoxins into the portal circulation. Under normal

circumstances, overspill of gut luminal toxic products into the portal circulation would be cleared by the hepatic reticuloendothelial system. In the presence of MODS the hepatic clearance of these substances is greatly reduced and spillage of toxins will be washed into the pulmonary microcapillary network. The appearance of endotoxin and bacteria in the lung will activate pulmonary alveolar macrophages with local damage occurring from macrophage-derived mediator release, adding to the destruction of pulmonary architecture already occurring in ARDS.

Kidneys

The involvement of renal dysfunction and failure as part of classical MODS heralds a large increase in mortality. The explanation for this excess mortality is unknown; perhaps the failing kidneys act as a further source of inflammatory mediators 'fuelling' the systemic disease process further. The loss of intravascular volume control may exacerbate ARDS and heart failure with the potential for volume overload. In addition, institution of methods of renal support will have the potential for further activation of the reticuloendothelial cells caused by bioincompatibility problems of the extracorporeal circuit and haemofilter/dialyser.

Haematological system

Coagulopathy is common after major trauma. Initially this may just reflect massive fluid replacement and transfusion. Massive transfusion – the replacement of greater than one circulating blood volume (approximately 10 units of blood) in less than 24 hours – may result in diffuse microvascular bleeding from surgical wounds, intravenous catheter sites and areas of minor trauma. The source of the coagulopathy, ignoring the presumed continuing consumption, is the dilution of coagulation factors through the infusion of products deficient in these factors (e.g. packed red blood cells, crystalloids and colloids). Laboratory tests demonstrate thrombocytopaenia, hypofibrinogenemia and prolongation of the prothrombin times.

An insidious complication of severe injury and blood loss is a widespread disorder of coagulation and haemostasis. This is due, at least in part, to the release of tissue thromboplastins into the circulation, endothelial damage and platelet activation. The result is a complex mixture of intravascular coagulation, depletion of clotting factors, fibrinolysis and thrombocytopaenia. Microvascular occlusion causes haemorrhagic infarctions and tissue necrosis, while deficient haemostasis leads to abnormal bleeding. This resulting coagulopathy is termed disseminated intravascular coagulation (DIC). The pathophysiology results from the generation of excessive amounts of thrombin. Thrombin generation in florid DIC is sufficiently intense that anticoagulant mechanisms such as antithrombin and activated protein C systems become ineffective. Fibrin deposition in the microvasculature undergoes fibrinolysis and promotes the consumption of clotting factors (especially fibrinogen, and platelet factors V and VIII). This in turn leads to a consumptive coagulopathy characterized by thrombocytopaenia, hypofibrinogenaemia and ongoing thrombolysis.

The consequences of DIC are variable but include excessive bleeding due to consumption of haemostatic factors and secondary fibrinolysis, organ dysfunction, skin infarction, haemolysis and disseminated thrombosis. The clinical features are those of diffuse microvascular thrombosis: restlessness, confusion, neurological dysfunction, skin infarcts, oliguria and renal failure. Abnormal haemostasis causes excessive bleeding at operation, oozing drip sites and wounds, spontaneous bruising, gastrointestinal bleeding and haematuria. The diagnosis is confirmed by finding a low haemoglobin concentration, prolonged prothrombin and thrombin times, thrombocytopaenia, hypofibrinogenaemia and raised levels of fibrinogen degradation products.

Management of MODS

Once the clinical syndrome of MODS is established, despite major advances in ITU technology and management strategies, the chances of survival are much reduced. The best treatment for MODS remains prevention. This entails early aggressive resuscitation following the original insult, avoidance of hypotensive episodes and removal of risk factors, for example by early excision of necrotic tissue, early fracture stabilization and ambulation, and appropriate antibiotic usage following drainage of sources of sepsis.

Early circulatory resuscitation is of paramount importance and this should be guided by invasive monitoring. Oxygen delivery should be maximized to a point where oxygen consumption no longer rises or to the level where markers of anaerobic metabolism such as serum lactate fall. It appears that the use of less invasive clinical markers for the adequacy of the circulation, such as mean arterial pressure, temperature gradients and urine output, may not entirely reflect the success of microcirculatory resuscitation. Once the sequence of MODS is established, early appropriate institution of organ support (e.g. endotracheal intubation and ventilation) is essential.

The treatment of ALI/ARDS remains mainly supportive and includes the management of precipitating causes. A large prospective study, supported by the National Heart Lung and Blood Institute in the USA, has shown that the use of low tidal volume ventilatory strategies (6 mL/kg) and limited plateau pressure (<30 cm H_2O) was effective in reducing the mortality rate from 40% to 31%. Other measures to improve

oxygenation – (e.g. prone positioning, high-frequency ventilation, nitrous oxide inhalation and extracorporeal life support) have limited success in improving overall outcome.

Renal and haematological management strategies are also largely supportive with renal replacement therapy and blood products frequently requiring expert involvement.

Malnutrition is a common and major contributing factor to MODS. Nutritional starvation combined with hypermetabolism leads to structural catabolism. Unlike starvation the substrates metabolized are mixed, with a significant increase in amino-acid oxidation. With the temporal progression of MODS, direct amino-acid oxidation increasingly becomes prevalent with rapid dissolution of skeletal muscle. Metabolic support in terms of providing adequate calories and maintaining nitrogen balance is essential if lean body mass is to be preserved and 'autocannabilism' slowed. This has led to recommendations for early parenteral feeding (although this is still controversial). Providing a calorie source for these patients requires care and a balance of substrates has to be given to prevent adding iatrogenic problems to the metabolic mayhem already occurring. While it is known that glucose has a protein-sparing effect, excessive amounts confer no additional advantages and may cause complications such as fatty liver, hyperosmolarity, hyperglycaemia, and increased CO_2 production, increasing the excretory load of the lungs and further exacerbating respiratory failure. The glucose load should not therefore exceed 4–5 mg/kg/minute, with a non-protein calorific load of 25–30 kcal/kg/day and 0.5–1.0 g/kg/day of lipids. Protein requirements run at 1–2 g/kg/day with modified amino acid preparations as these appear to be the most efficient protein source, producing less urea and better nitrogen retention.

Rigorous attention to these details has brought improvements in prevention and outcome in MODS. Other newer treatment strategies are still largely unproven in terms of outcome. Selective digestive tract decontamination (SDD) by administration of non-absorbable antimicrobial agents may reduce the incidence of nosocomial pneumonia by resterilizing the upper gastrointestinal tract. Trials of SDD have shown some benefit but large-scale effects on antibiotic resistance from widespread use of antibiotics are awaited. The use of aggressive early enteral feeding in patients without an ileus may not only reduce the effects of catabolism but also prevent upper gut colonization by bacteria and hence nosocomial pneumonia by stimulation of bactericidal gastric acid secretion. Recent studies appear to suggest that this may have a positive effect on outcome.

Probably the most recent advances in treatment of MODS have been in relation to modulation of the hypermetabolic inflammatory response by use of specific agents. These include monoclonal antibodies against endotoxin and TNF inhibitors of nitric oxide synthase and receptor antagonists for interleukin-1. Unfortunately, interim reports of the therapeutic effectiveness are conflicting and it would appear as yet that the 'magic bullet' remains elusive.

Again it must be emphasized that prevention is better than attempting cure for MODS, the major killer of critically ill patients in intensive care.

TETANUS

The pathogenic Gram-positive tetanus bacterium, *Clostridium tetani*, grows only in necrotic tissue under anaerobic conditions. The exotoxin released passes to the central nervous system via the blood and the perineural lymphatics from the infected region. The toxin is fixed in the anterior horn cells and therefore cannot be neutralized by antitoxin.

Established tetanus is characterized by tonic, and later clonic, contractions, especially of the muscles of the jaw and face (trismus, risus sardonicus), those near the wound itself, and later of the neck and trunk. Ultimately, the diaphragm and intercostal muscles may be 'locked' by spasm resulting in asphyxia.

Treatment

With established tetanus, intravenous antitoxin (human for choice) is advisable. Heavy sedation and muscle-relaxant drugs may help; tracheal intubation and ventilation are the only options to treat respiratory muscle involvement.

Prophylaxis against tetanus by active immunization with tetanus toxoid vaccine is a valuable goal. If the patient has been immunized, booster doses of toxoid are given after all but trivial skin wounds. In the non-immunized patient prompt and thorough wound toilet together with antibiotics may be adequate but, if the wound is contaminated, and particularly with a delay before operation, antitoxin is advisable.

FAT EMBOLISM SYNDROME

Fat embolism is a common phenomenon following limb fractures. Circulating fat globules larger than 10 μm in diameter occur in most adults after closed fractures of long bones and histological traces of fat can be found in the lungs and other internal organs. A small percentage of these patients develop clinical features similar to those of ARDS; this was recognized as the fat embolism syndrome long before ARDS entered the medical literature. Whether the fat embolism syndrome is an expression of the same condition or whether it is an entirely separate entity is still uncertain.

The source of the fat emboli is probably the bone marrow, and the condition is more common in patients with multiple fractures.

Clinical features

Early warning signs of fat embolism (usually within 72 hours of injury) are a slight rise of temperature and pulse rate. In more pronounced cases there is breathlessness and mild mental confusion or restlessness. Pathognomonic signs are petechiae on the trunk and axillae and in the conjunctival folds and retinae. In more severe cases there may be respiratory distress and coma, due both to brain emboli and hypoxia from involvement of the lungs. The features at this stage are essentially those of ARDS.

There is no infallible test for fat embolism; however, urinalysis may show fat globules in the urine and the blood PO_2 should always be monitored; values below 8 kPa (60 mmHg or less) within the first 72 hours of any major injury must be regarded as suspicious. A chest X-ray may show classical changes in the lungs.

Management

Management of severe fat embolism is supportive. Symptoms of the syndrome can be reduced with the use of supplemental high inspired oxygen concentrations immediately after injury and the incidence appears to be reduced by the prompt stabilization of long-bone fractures. Intramedullary nailing is not thought to increase the risk of developing the syndrome. Fixation of fractures also allows the patient to be nursed in the sitting position, which optimizes the ventilation–perfusion match in the lungs.

CRUSH SYNDROME

This is seen when a limb is compressed for extended periods (e.g. following entrapment in a vehicle or rubble) but also after prolonged use of a pneumatic antishock garment.

The crushed limb is underperfused and myonecrosis follows, leading to the release of toxic metabolites when the limb is freed and so generating a reperfusion injury. Reactive oxygen metabolites create further tissue injury. Membrane damage and capillary fluid reabsorption failure result in swelling that may lead to a compartment syndrome, thus creating more tissue damage from escalating ischaemia. Tissue necrosis also causes systemic problems such as renal failure from free myoglobin, which is precipitated in the renal glomeruli. Myonecrosis may cause a metabolic acidosis with hyperkalaemia and hypocalcaemia.

Clinical features and treatment

The compromised limb is pulseless and becomes red, swollen and blistered; sensation and muscle power may be lost. The most important measure is prevention. From an intensive care perspective a high urine flow is encouraged with alkalization of the urine with sodium bicarbonate, which prevents myoglobin precipitating in the renal tubules. If oliguria or renal failure occurs, renal haemofiltration will be needed.

If a compartment syndrome develops, and is confirmed by pressure measurements, then a fasciotomy is indicated. Excision of dead muscle must be radical to avoid sepsis. Similarly, any open wound should be managed aggressively. If there is no open wound and the compartment pressures are not high, then the risk of infection is probably lower if early surgery is avoided.

INTENSIVE CARE UNIT SCORING SYSTEMS

The role of scoring systems in medicine has expanded since the 1950s. The concepts of 'severity of illness' scoring systems were developed to evaluate care delivery and provide outcome predictions in groups of critically ill patients. Scoring systems usually consist of two parts: a numerical severity score (the higher the number the more severe the condition) and a probability of mortality calculated from analysis of the data.

There are now many scoring systems catering for most organ dysfunction, disease states, trauma and critical illnesses. New scoring systems are regularly being developed and older systems refined. Their widespread use relates to a pivotal role in communication, audit and research as well as clinical management.

Scoring systems can theoretically be created from many types of variable. However, to be useful, scoring systems must have predictive properties, and the information has to be unambiguous, reliable and easy to determine and collect. Ideally, the variables should be frequently recorded or measured. Variables can be selected using clinical judgement and recognized physiological associations, or by statistical analysis of prospectively recorded variables to determine which are associated with the outcome of interest. The variables are then assigned a weighting in relation to their importance to the predictive power of the scoring system, again either by clinical relevance or from statistical models.

Logistic regression analysis, a multivariate statistical procedure, is used to convert a score to a predicted probability of the outcome measured, usually morbidity or mortality, using a large patient database suitable to the scoring system being developed. Finally, the scoring system has to be validated on a population of

patients independent from those used to develop the scoring system.

Patients form a heterogeneous population and differ in many respects including age, previous health status, reason for admission and severity of illness. When comparing patients on intensive care for the purpose of research or audit, it is often difficult to standardize for all physiological variables due to the diversity of patients and their conditions. Scoring systems are therefore used to standardize for the physiological variables, age and reason for admission, allowing comparisons to be made between patients with different severity of illness.

In the majority of scoring systems a high score reflects a patient who is more sick than one with a lower score (with the notable exception of the Glasgow Coma Score), but the score does not always follow a linear scale. Therefore a patient with a score of 20 is neither necessarily twice as sick nor has double the chance of dying as a patient with a score of 10. However, using logistic regression, it is possible to derive from the score a probability of morbidity, or mortality in hospital.

Audit

The most common use for scoring systems is for audit. This allows ICUs to assess their performance in comparison to other units and also their own performance from year to year. If an ICU admitted patients who were not very sick, then their actual mortality on that unit would be lower than on a unit that admitted extremely sick patients and therefore it would be difficult to compare the performance between those units. This has led to the comparisons of actual mortality to a predicted mortality. The ratio of the actual to predicted mortality gives a figure for the standardized mortality ratio (SMR). Therefore an ICU with an SMR of less than 1 is theoretically performing better than expected and a unit with an SMR of more than 1 is performing worse than expected. The SMR can then be used to compare performance between units. Also, if the severity of illness of patients varies, or if different types of patient are admitted from year to year, the SMR can be used to assess the performance of a unit over time. Statistical significance of different SMRs can be evaluated using confidence intervals.

Research

The diversity of patients and different pathologies on the ICU makes comparisons between treatments or procedures difficult. Scoring systems can be used to adjust for the differences in case-mix in patients recruited for trials, so if an intervention is used on all patients, the scoring systems can standardize for any heterogenicity between the groups prior to the intervention being initiated. Stratification of the risk of death can also be inferred from the scoring systems, allowing for investigation in different subgroups of patients in the ITU, and allowing researchers to assess response to interventions in patients at different risk of mortality.

Clinical management

As well as quantifying the degree of physiological derangement or clinical intervention, and promoting better communication between clinicians, scoring systems can also be used to guide patient management. Some scoring systems lend themselves to sequential reassessment and thus can be used to monitor a patient's progress over time. Also, as most research conducted in ICUs uses scoring systems, the recommendations from research can sometimes be applied to subsets of patients with a severity of illness score within a certain range. This allows therapies to be directed sensibly at patients with an appropriate severity of illness. As most ITU scoring systems are an assessment of risk of mortality they have also been used to trigger admission to high-dependency or intensive care.

Scoring systems on the ICU

Scoring systems are often classified into three subsets: (1) anatomical (e.g. the injury severity score); (2) physiological (e.g. the GCS) and therapeutic (e.g. therapeutic intervention scoring systems). Most intensive care scoring systems are based on physiological variables; however, other data are also included in the score, making simple classification very difficult.

An ideal scoring system would be simple to use and be applicable to all intensive care patients irrespective of age, diagnosis and urgency of admission. It should also not be dependent upon treatment given prior to or on admission to ICU. The outcome prediction modelling should have a high sensitivity and specificity. The intensive care scoring systems are developed from large databases incorporating data from many ICUs. The data include physiological variables, comorbidities, age, diagnoses, urgency of admission, and outcome at discharge from hospital.

Acute physiology and chronic health evaluation

Knaus and colleagues introduced the acute physiology and chronic health evaluation (APACHE) model in 1981 and revised it to APACHE II in 1985. APACHE III was presented in 1991 but as the regression analysis modelling used is not in the public domain its uptake has been slow.

APACHE II is made up of four basic components: (1) acute physiology score; (2) chronic health evaluation; (3) age; (4) urgency of admission to critical care. The acute physiology score is composed of 12 variables, with the most deranged measurement during the first 24 hours of admission to critical care being used to calculate the score. The original data collection for APACHE II occurred between 1979 and 1982 from ICUs in North America, and the population studied included relatively few surgical and trauma patients. Also, there have been many advances in patient care since the 1980s, which have made APACHE II dated, despite its continued popularity. The latest iteration is the APACHE IV published in 2006. It is the most complex of all the APACHE models with 146 variables and was developed in the USA for the US ICU model.

Simplified acute physiology score

The simplified acute physiology score (SAPS) initially used 14 variables and did not provide any probability of survival. In 1993 it was revised to SAPS II with the data originating from European and North American ICUs. The score includes 12 physiological variables (the worst value within the first 24 hours), age, type of admission and three underlying disease variables (acquired immune deficiency syndrome (AIDS), metastatic cancer and haematological malignancy). Using logistic regression, SAPS II can also be used to estimate the probability of survival. It is a simpler scoring system than APACHE and is also in the public domain, resulting in its widespread use, particularly in Europe. It suffers similar disadvantages when compared to APACHE with regards to the timing of data collection, but it is based on more recent and international data. SAPS III was introduced in 2005 and calculates scores based on data collected within the first hour after admission. The rationale is that this allows a prediction of outcome before ICU intervention, and is a better evaluation of an individual patient rather than an ICU. There are limitations including the timing of data collection which may lead to data omissions

Mortality prediction model

The original mortality prediction model (MPM) was derived in the late 1980s with data from a single hospital and differed from many of the scoring systems by being dependent not on physiological data but on the presence or absence of pathology. Therefore there was less of an impact by treatment of the physiology prior to and on admission to intensive care. In 1993 the MPM was revised to MPM II based on the same data set as SAPS II but with the inclusion of six extra ICUs. Initially the model was constructed of two time points: within 1 hour (MPM II_0) and the first 24 hours (MPM II_{24}) of admission. Now it can be used for 48- and 72-hour points as well, giving a prediction of mortality at those time points. Its variables include physiological parameters, age, acute diagnoses, chronic diseases, type of admission, as well as others. The MPM II_0 is useful as it is minimally affected by the treatment given in an ICU. It is not applicable for paediatric patients under 14 years, or to burns or cardiac patients.

Therapeutic intervention scoring system

The original therapeutic intervention scoring system (TISS) was devised in 1976, consisting of 76 therapeutic activities and was used initially to stratify the severity of illness, i.e. measuring sickness based on the type and amount of treatment received. Its use for this purpose has largely been superseded by the newer scoring systems, but it is still commonly used to assess nursing workload and in resource management, for which it was not designed. A simplified TISS was developed in 1996, which included only 28 therapeutic activities.

Sequential organ failure assessment

The sequential organ failure assessment (SOFA) system was developed in the early 1990's and was known originally as the sepsis-related organ failure assessment score, describing the severity of organ dysfunction associated with sepsis in a mixed ICU population. It has been recalibrated to represent various groups of ICU patients with organ dysfunction which may not be sepsis-related. The SOFA scoring system utilizes assessment of six organ systems (respiratory, cardiovascular, renal, hepatic, central nervous system and coagulation). The assessment of function is scored from 0 (normal) to 4 for all six systems, giving a score range of 0 to 24. Mortality increases with increasing organ dysfunction scores and the cardiovascular component is calculated from vasopressor and inotropic dosage. The highest total (maximal) SOFA score is the sum of the highest scores per individual during the patient's ICU stay and is a sensitive predictor of mortality. A score of 15+ predicts a mortality of over 90%.

Limitations

Overall there is very little to choose between the third-generation scoring systems (APACHE III, SAPS II and MPM II) in terms of their predictive power. Despite this, APACHE II continues to dominate the literature and continues to be the most widely used score to date.

The APACHE II/III and SAPS I/II scoring systems measure physiological variables during the first 24 hours of ITU admission and there has been concern that this can lead to bias. If a patient is treated prior to admission to ITU, their physiological variables will normally have been improved and the patients will have lower scores. Similarly, if a patient is admitted to the ITU and receives inappropriate treatment over the first 24 hours, their scores will suggest that the ITU is dealing with sicker patients. Lastly, if a patient dies within 24 hours, their scores before death will be very high and will therefore skew the SMR of a unit to suggest that it is admitting very sick patients. MPM II measures variables during the first hour and within the first 24 hours, thereby reducing the bias that may occur in the score when measured over 24 hours.

Limitations and errors associated with the use of the scoring systems include missing data, observer error and inter-observer variability. Even the method of data collection (manual data entry versus data collected automatically from monitoring systems) leads to wide variations in scores. Although the above scoring systems are useful to assess and compare outcomes in patient populations, such scores may not be appropriate to provide individual risk assessment in critically ill patients.

FURTHER READING

American College of Surgeons Committee on Trauma. *Advanced Trauma Life Support® Program for Doctors*, 8th edn. Chicago: American College of Surgeons, 2010.

Earlam R. *Trauma Care*. London: Helicopter Emergency Medical Service (HEMS), 1997.

Findlay G, Martin IC, Carter S, *et al.* (compilers). *Trauma: Who cares? A report of the National Confidential Enquiry into Patient Outcome and Death (2007)*. NCEPOD, 2007.

Knaus WA, Zimmerman JE, Wagner DP. APACHE: Acute Physiology and Chronic Health Evaluation, a physiologically based classification system. *Crit Care Med* 1981; **16**: 470–8.

Mock C, Lormand JD, Goosen J, *et al.* *Guidelines for Essential Trauma Care*. Geneva: World Health Organization, 2004.

National Institute for Clinical Excellence. Pre-hospital initiation of fluid replacement therapy in trauma. *Technology Appraisal* **74**, January 2004.

National Institute for Health and Clinical Excellence. *Head injury. Triage, assessment, investigation and early management of head injury in infants, children and adults.* NICE clinical guideline 56, London, September 2007.

Peden M, Scurfield R, Sleet D, *et al.* *The World Report on Road Traffic Injury Prevention*. Geneva: World Health Organization, 2004.

Principles of fractures

23

Boyko Gueorguiev, Fintan T. Moriarty, Martin Stoddart, Yves P. Acklin, R. Geoff Richards & Michael Whitehouse

INTRODUCTION

A fracture is a break in the structural continuity of bone. It may be no more than a crack, a crumpling or a splintering of the cortex; more often the break is complete. The resulting bone fragments may be displaced or undisplaced. If the overlying skin remains intact, it is a *closed fracture*; if the skin or one of the body cavities is breached, it is an *open fracture (also known as a compound fracture)*, liable to contamination and infection.

HOW FRACTURES OCCUR

Fractures are caused by (1) injury; (2) repetitive stress; or (3) abnormal weakening of the bone (a 'pathological' fracture).

Fractures due to injury

Most fractures are caused by sudden and excessive force (overloading), which may be direct or indirect.

With a *direct force (direct injury)* the bone breaks at the point of impact; the soft tissues are also damaged. A direct blow usually splits the bone transversely or may bend it over a fulcrum so as to create a break with a 'butterfly' fragment. Damage to the overlying skin is common; if crushing occurs or in high-energy injuries, the fracture pattern will be comminuted with extensive soft-tissue damage.

With an *indirect force (indirect injury)* the bone breaks at a distance from where the force is applied; soft-tissue damage at the fracture site is not inevitable. Although most fractures are due to a combination of forces (twisting, bending, compression or tension – see Figure 23.1), the dominant mechanism is revealed by X-rays:

- *Twisting* causes a spiral fracture.
- *Compression* causes a short oblique fracture.
- *Bending* results in a fracture with a triangular 'butterfly' fragment.
- *Tension* tends to break the bone transversely; in some cases it may simply avulse a small fragment of bone at the points of ligament or tendon insertion.

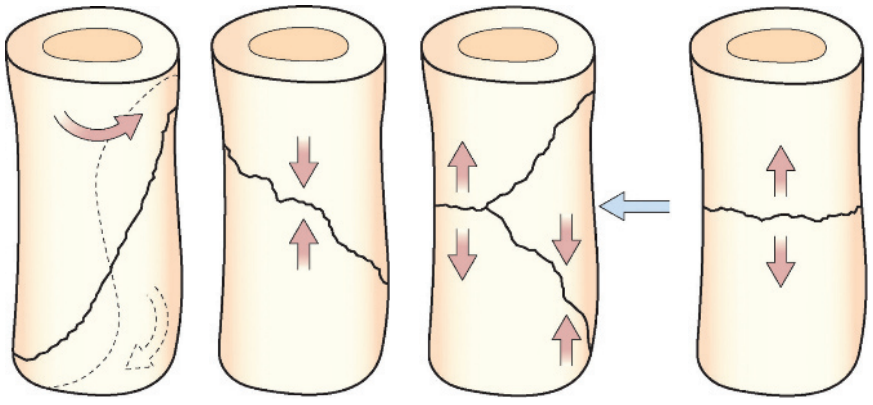


Figure 23.1 Mechanism of injury Some fracture patterns suggest the causal mechanism: (a) spiral pattern (twisting); (b) short oblique pattern (compression); (c) triangular 'butterfly' fragment (bending, the fragment shows the direction of the force) and (d) transverse pattern (tension). Spiral and some (long) oblique patterns are usually due to low-energy indirect injuries; bending and transverse patterns are caused by high-energy direct trauma.

NOTE: The above description applies mainly to the long bones. A cancellous bone, such as a vertebra or a calcaneum, when subjected to sufficient force, will split or be crushed into an abnormal shape.

Fatigue or stress fractures

These fractures occur in normal bone which is subjected to repeated heavy loading, typically in athletes, dancers or military personnel who have gruelling exercise programmes or when the intensity of exercise is significantly increased from baseline. The heavy loading creates minute deformations that initiate the normal process of remodelling – a combination of bone resorption and new bone formation in accordance with Wolff's law. When exposure to stress and deformation is repeated and prolonged, bone resorption occurs faster than replacement (new bone formation) and leaves the area liable to fracture. A similar problem exists in individuals who are on medication that alters the normal balance of bone resorption and replacement; stress fractures are increasingly seen in patients with chronic inflammatory diseases who are on treatment with steroids or methotrexate.

Pathological fractures

Fractures may occur even with normal stresses if the bone has been weakened by a change in its structure (e.g. in patients with osteoporosis, osteogenesis imperfecta or Paget's disease, bisphosphonate therapy) or through a lytic lesion (e.g. a bone cyst or a metastasis).

TYPES OF FRACTURE

Fractures are variable in appearance but for practical reasons they are divided into a few well-defined groups (Figure 23.2).

Complete fractures

The bone is split into two or more fragments. The fracture pattern on X-ray can help predict the behaviour after successful reduction: in a *transverse fracture* the fragments usually remain in place after reduction; in an *oblique* or *spiral*, they tend to shorten and redisplace even if the bone is splinted. In an *impacted fracture* the fragments are jammed tightly together and the fracture line is indistinct. A *comminuted fracture* is one with more than two fragments with interlocking of the fracture surfaces; it is often unstable.

Incomplete fractures

The bone is incompletely divided and the periosteum remains in continuity. In a *greenstick fracture* the bone is buckled or bent (like snapping a green twig); this is seen in children, whose bones are less brittle than those of adults. Children can also sustain injuries where the bone is plastically deformed (misshapen) without any crack visible on the X-ray. In contrast, *compression fractures* occur when cancellous bone is crumpled, typically in adults where this type of bone structure is present, such as in vertebral bodies, calcaneum and the tibial plateau.

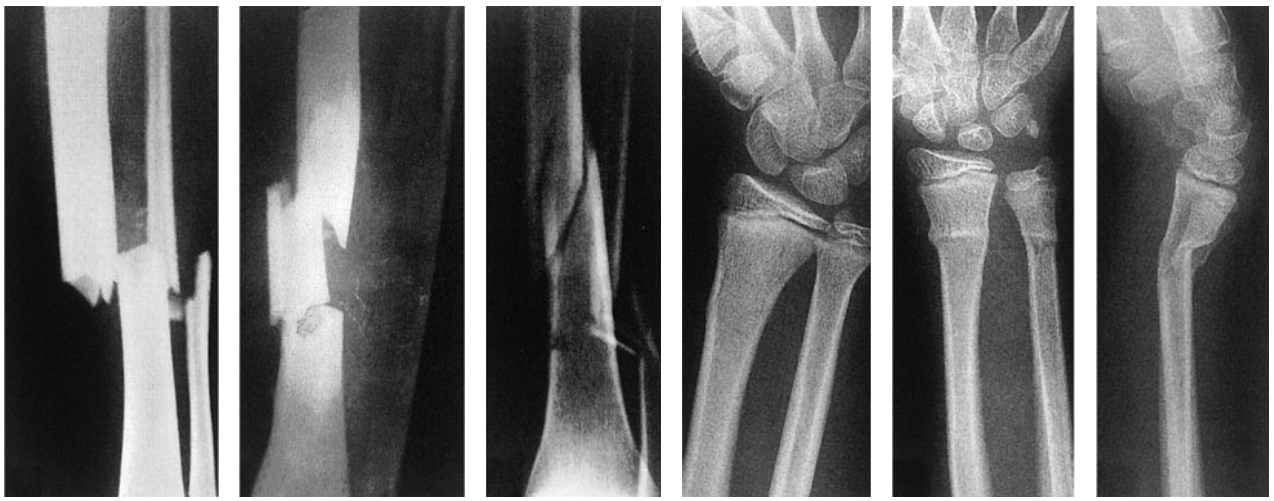


Figure 23.2 Varieties of fracture Complete fractures: (a) transverse; (b) segmental, (c) spiral. Incomplete fractures: (d) buckle or torus, (e,f) greenstick.

CLASSIFICATION OF FRACTURES

Sorting fractures into groups with similar features brings advantages when done well: it allows any information about a fracture to be applied to others in the group (i.e. generalizability whether related to treatment or prognosis) and facilitates a common dialogue between surgeons and other personnel involved in the care of such injuries.

Traditional classifications, which often bear the originator's name, are hampered by being applicable to a specific type of injury only; even then the term is often inaccurately applied, famously in the case of Pott's fracture, which is often applied to any fracture around the ankle though that is not what Sir Percival Pott implied when he described the injury in 1765.

A universal, anatomically based system facilitates communication and exchange of data from a variety of countries and populations, thus contributing to advances in research and treatment. An alphanumeric classification developed by Müller and colleagues, known as AO/OTA classification and fulfilling the objective of being comprehensive, has now been adapted and revised (Figure 23.3). In this system, the first digit specifies the bone (1 = humerus, 2 = radius/ulna, 3 = femur, 4 = tibia/fibula, 5 = spine, 6 = pelvis/acetabulum, 7 = hand, 8 = foot, 9 = craniomaxillofacial bones) and the second digit specifies the segment (1 = proximal, 2 = diaphyseal, 3 = distal, 4 = malleolar). A letter specifies the fracture pattern (for diaphysis: A = simple, B = wedge, C = complex; for metaphysis: A = extra-articular, B = partial articular, C = complete articular). Two further numbers specify the detailed morphology of the fracture.

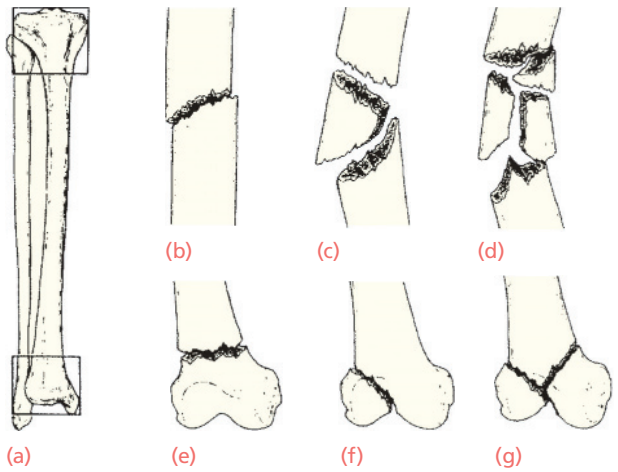


Figure 23.3 Müller's classification (a) Each long bone has three segments – proximal, diaphyseal and distal; the proximal and distal segments are defined by a square based on the widest part of the bone. (b,c,d) Diaphyseal fractures may be simple, wedge or complex. (e,f,g) Proximal and distal fractures may be extra-articular, partial articular or complete articular.

HOW FRACTURES ARE DISPLACED

After a complete fracture the fragments usually become displaced, partly by the force of injury, partly by gravity and partly by the pull of muscles attached to them. Displacement is usually described in terms of translation, alignment (angulation), rotation and altered length:

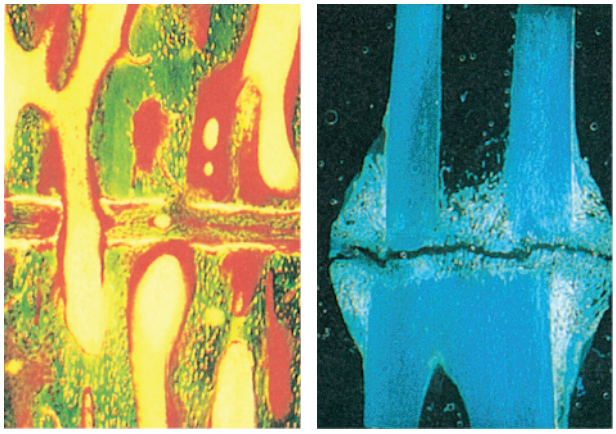
- *Translation (shift)* – The fragments may be shifted sideways, backward or forward in relation to each other, such that the fracture surfaces lose part or all of their contact. The fracture will usually unite as long as sufficient contact between surfaces remains or can be achieved by reduction; this may occur even if reduction is imperfect, or indeed even if the fracture ends are off-ended but the bone segments come to lie side by side.
- *Angulation (tilt)* – The fragments may be tilted or angulated in relation to each other. Malalignment, if uncorrected, may lead to deformity of the limb.
- *Rotation (twist)* – One of the fragments may be twisted around its longitudinal axis; the bone often looks aligned on X-ray, but the limb ends up with a rotational deformity which is best observed on examination of the patient.
- *Length* – The fragments may be distracted and separated, or they may overlap, due to muscle spasm, causing shortening of the bone.

HOW FRACTURES HEAL

Fracture healing is characterized by a process of new bone formation with fusion of the bone fragments. The bone either heals by primary (without callus formation) or secondary (with callus formation) fracture healing. The process of fracture repair varies according to the type of bone involved and the amount of movement at the fracture site. The mechanical strain applied across the fracture gap plays a major role in directing the healing response. Absolute stability and compression leads to direct healing (primary bone healing), while relative stability leads to indirect healing (secondary bone healing). However, excessive motion may lead to delayed or non-union. Clinical and experimental studies have shown that callus formation occurs in response to movement at the fracture site. It serves to stabilize the fragments as rapidly as possible – a necessary precondition for bridging by new bone formation. Therefore, most fractures are splinted in order to: (1) alleviate pain; (2) ensure that union takes place in good position; and (3) permit early movement of the limb and a return of function.

Healing by direct union (primary bone healing)

If the fracture site is absolutely stable – for example, an impacted fracture in cancellous bone, or a fracture held by a metal plate with absolute stability – there is no stimulus for callus. Instead, osteoblastic new bone formation occurs directly between the fragments. Where the exposed fracture surfaces are in intimate contact and held from the outset with absolute stability, internal bridging may occasionally occur without any intermediate stages (*contact healing*). Gaps between the fracture surfaces are invaded by new capillaries and osteoprogenitor cells growing in from the edges, and new bone is laid down on the exposed surface (*gap healing*). Where the crevices are very narrow (less than 200 µm), osteogenesis produces lamellar bone; wider gaps are filled first by woven bone, which is then remodelled to lamellar bone. By 3–4 weeks the fracture is solid enough to allow penetration and bridging of the area by bone remodelling units, i.e. osteoclastic 'cutting cones' followed by osteoblasts (Figure 23.4). With rigid metal fixation, however, the absence of callus means that there is a long period during which the bone depends entirely upon the metal implant for its integrity, increasing the risk of implant failure. Moreover, the implant diverts stress away from the bone, which may become osteoporotic and may not recover fully until the metal is removed.



(a) (b)

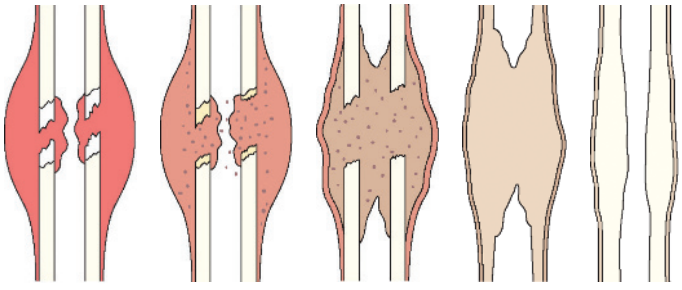
Figure 23.4 Fracture healing – histology Experimental fracture healing: (a) by direct penetration of the fracture gap by a cutting cone; (b) by bridging callus.

Healing by callus (secondary bone healing)

Healing by callus, though less direct (indirect healing) has distinct advantages: it ensures mechanical strength while the bone ends heal, and with increasing stress the callus grows stronger and stronger (according to Wolff's law) (see Figure 23.6).

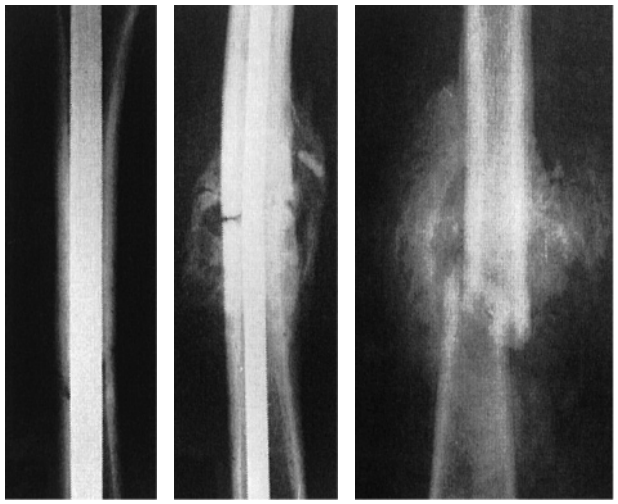
At least for the secondary bone healing, surgical stabilization is not always necessary but it can prevent malunion. Secondary bone healing is the most common form of healing in tubular bones; in the absence of rigid fixation, it proceeds in five stages (Figure 23.5):

1 *Haematoma formation* – At the time of injury, bleeding occurs from the bone and soft tissues.
2 *Inflammation* – The inflammatory process starts rapidly when the fracture haematoma forms and cytokines are released, and lasts until fibrous tissue, cartilage, or bone formation begins (1–7 days postfracture). Osteoclasts are formed to remove the necrotic ends of bony fragments.
3 *Soft callus formation* – After 2–3 weeks, the first soft callus is formed. This is about the time when the fragments can no longer move freely. The strain applied to the cells in the fracture gap modifies their growth factor expression and progenitor cells are stimulated to become osteoblasts. The cells form a cuff of woven bone periosteally. The fracture can now still angulate but is stable in length.
4 *Hard callus formation* – When the fracture ends are linked together, the hard callus starts and lasts until the fragments are firmly united (3–4 months). Bone callus forms at the periphery of the fracture and progressively moves centrally.



(a) (b) (c) (d) (e)

Figure 23.5 Fracture healing Five stages of healing: (a) *Haematoma:* there is tissue damage and bleeding at the fracture site; (b) *Inflammation:* inflammatory cells (cytokines) appear in the haematoma; the bone ends die back for a few millimetres. (c) *Soft callus formation:* the cell population changes to osteoblasts; over time, the callus calcifies and woven bone appears in the fracture callus. (d) *Hard callus formation:* the fracture is solidly united. (e) *Remodelling:* the newly formed bone is remodelled to resemble the normal structure. Woven bone is replaced by laminar bone.



(a) (b) (c)

Figure 23.6 Callus and movement Three clinical cases with femoral shaft fractures. (a) and (b) are both 6 weeks post fixation: in (a) the Kuntscher nail fitted tightly, preventing movement, and there is no callus; in (b) the nail fitted loosely, permitting some movement, so there is callus. (c) This patient had cerebral irritation and thrashed around wildly; at 3 weeks callus is excessive.

5 *Remodelling* – The woven bone is slowly replaced by lamellar bone. This process can last from a few months to several years.

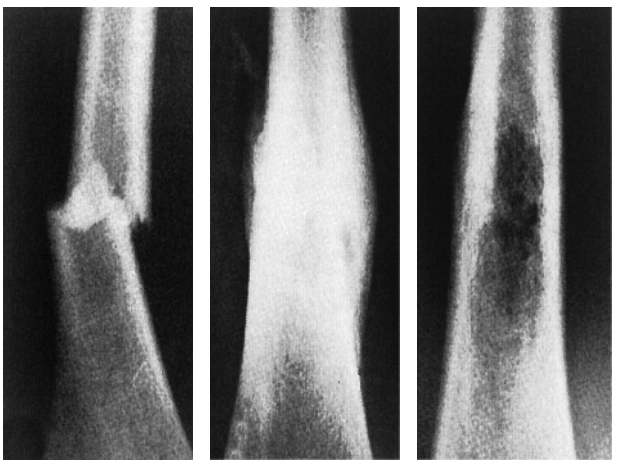
The periosteum is an excellent source of local mesenchymal stem cells that can enhance bone repair. For this reason it is imperative that, as much as it is possible, the periosteum is left in place and remains viable. This should be taken into account when considering direct contact plates that press against the periosteum, potentially damaging the vascular supply and leading to periosteal necrosis.

Union, delayed union and non-union

Repair of a fracture is a continuous process (Figure 23.7): any stages into which it is divided are necessarily arbitrary. In this chapter the terms 'union', 'delayed union' and 'non-union' are used, and they are defined as follows:

- *Union* – Union is incomplete repair; the ensheathing callus is calcified. Clinically, the fracture site is painless on palpation and weight-bearing. X-rays show bridging callus. The fracture line is completely or almost obliterated and crossed by bone trabeculae. Repair is complete and further protection is unnecessary.
- *Timetable* – How long does a fracture take to unite? No precise answer is possible because age, constitution, blood supply, type of fracture and other factors all influence the time taken. A spiral fracture in the upper limb takes 6–8 weeks to unite; in the lower limb it needs twice as long. Add 25% if the fracture is not spiral or if it involves the femur. Children's fractures, of course, join more quickly. These figures are only a rough guide; there must be clinical and radiological evidence of consolidation before full stress is permitted without splintage.
- *Delayed union* – Delayed union means that fracture healing is not taking place at the expected rate and time but healing is still possible. Additional effort should be aimed at achieving fracture healing as fast as possible. Clinically, the fractured limb has local swelling and movement or partial weight-bearing is painful.
- *Non-union* – Sometimes the normal process of fracture repair is thwarted and the bone fails to unite. Unless there is bone loss, non-union is usually defined as fracture that has not healed 9 months post operation and there is no visible progress of healing during the last 3 months. Causes of aseptic non-union are: (1) mechanical instability or (2) impaired vascularity. Septic non-unions occur with infected osteosynthesis.

  Aseptic non-unions can be either stiff or mobile as judged by clinical examination. The mobile ones can be as free and painless as to give the impression of a false joint (pseudoarthrosis). On X-rays, non-unions are typified by a lucent line still present between the bone fragments; sometimes there is exuberant callus trying – but failing – to bridge the gap (*hypertrophic non-union*) or at times none at all (*atrophic non-union*) with a withered appearance to the fracture ends (Figure 23.8).



(a) (b) (c)

Figure 23.7 Fracture repair (a) Fracture; (b) union; (c) bone remodelling. The fracture must be protected until consolidated.

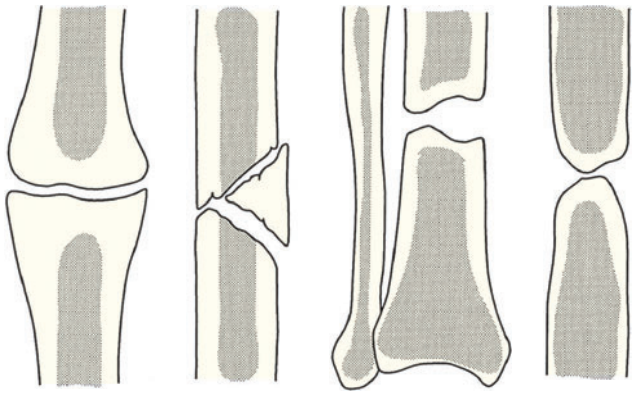


Figure 23.8 Non-unions Aseptic non-unions are generally divided into hypertrophic and atrophic types. Hypertrophic non-unions often have florid streams of callus around the fracture gap – the result of insufficient stability. They are sometimes given colourful names, such as (a) elephant's foot. In contrast, atrophic non-unions usually arise from an impaired repair process; they are classified according to the X-ray appearance as (b) necrotic; (c) gap and (d) atrophic.

CLINICAL FEATURES

History

There is usually a history of injury, followed by inability to use the injured limb – but beware! The fracture is not always at the site of the injury: a blow to the knee may fracture the patella, femoral condyles, shaft of the femur or even acetabulum depending upon the size and direction of the applied load. The patient's age and mechanism of injury are important. If a fracture occurs with trivial trauma, suspect a pathological lesion. Pain, bruising and swelling are common symptoms but they do not distinguish a fracture from a soft-tissue injury. *Deformity* is much more suggestive of a fracture or dislocation.

Always enquire about symptoms of associated injuries: pain and swelling elsewhere (it is a common mistake to get distracted by the main injury, particularly if it is severe), numbness or loss of movement, skin pallor or cyanosis, blood in the urine, abdominal pain, difficulty with breathing or transient loss of consciousness.

Once the acute emergency has been dealt with, ask about previous injuries, or any other musculoskeletal abnormality that might cause confusion when the X-ray is seen. Finally, a general medical history is important, in preparation for anaesthesia or operation.

General management

Unless it is obvious from the history that the patient has sustained a localized and fairly modest injury, priority must be given to dealing with the general effects of trauma (see Chapter 22). Follow the ABCs: look for, and if necessary attend to, **A**irway obstruction, **B**reathing problems, **C**irculatory problems and **C**ervical spine injury/immobilization. During the secondary survey it will also be necessary to exclude other previously unsuspected injuries and to be alert to any possible predisposing cause (such as Paget's disease or a metastasis).

Local signs

Injured tissues must be handled gently. Eliciting crepitus or abnormal movement is unnecessarily painful; X-ray diagnosis is more reliable. Nevertheless, the familiar headings of clinical examination should always be considered, or damage to arteries, nerves and ligaments may be overlooked. A systematic approach is always helpful:

1 Examine the most obviously injured part.
2 Test for artery and nerve damage.
3 Look for associated injuries in the region.
4 Look for associated injuries in distant parts.

Look

Swelling, bruising and deformity may be obvious, but the important point is whether the skin is intact; if the skin is breached and the wound communicates with the fracture, the injury is 'open' ('compound'). Note also the posture of the distal extremity and the colour of the skin (for telltale signs of nerve or vessel damage).

Feel

The injured part is gently palpated for localized tenderness. Some fractures would be missed if not specifically looked for; for example, the classical sign of a fractured scaphoid is tenderness on pressure precisely in the anatomical snuffbox. The common and characteristic associated injuries should also be felt for, even if the patient does not complain of them. For example, an isolated fracture of the proximal fibula should always alert to the likelihood of an associated fracture or ligament injury of the ankle, and in high-energy injuries, always examine the spine and pelvis. Vascular and peripheral nerve abnormalities should be tested for, both before and after treatment.

Move

Crepitus and abnormal movement may be present, but why inflict pain when X-rays are available? It is more important to ask if the patient can move the joints distal to the injury.

X-rays

X-ray examination is mandatory. Remember the '*rule of twos*' (Figure 23.9):

- *Two views* – A fracture or a dislocation may not be seen on a single X-ray view, and at least two views (anteroposterior and lateral) must be taken.
- *Two joints* – In the forearm or leg, one bone may be fractured and angulated. Angulation, however, is impossible unless the other bone is also broken, or a joint dislocated. The joints above and below the fracture must both be included on the X-rays.
- *Two limbs* – In children, the appearance of immature epiphyses may confuse the diagnosis of a fracture; X-rays of the uninjured limb can be used for comparison.
- *Two injuries* – Severe force often causes injuries at more than one level. Thus, with fractures of the calcaneum or femur it is important also to X-ray the pelvis and spine.
- *Two occasions* – Some fractures are notoriously difficult to detect soon after injury, but another X-ray examination a week or two later may show the lesion. Common examples are undisplaced fractures of the distal end of the clavicle, scaphoid, femoral neck and lateral malleolus, and also stress fractures and physeal injuries wherever they occur.

Special imaging

Sometimes the fracture – or the full extent of the fracture – is not apparent on a plain X-ray. *Computed tomography (CT)* may be helpful in lesions of the spine or for complex joint fractures; indeed, these crosssectional images are essential for accurate visualization of fractures in 'difficult' sites such as the calcaneum or acetabulum. *Magnetic resonance imaging (MRI)* may be the only way of showing whether a fractured vertebra is threatening to compress the spinal cord. *Radioisotope scanning* may be helpful in diagnosing

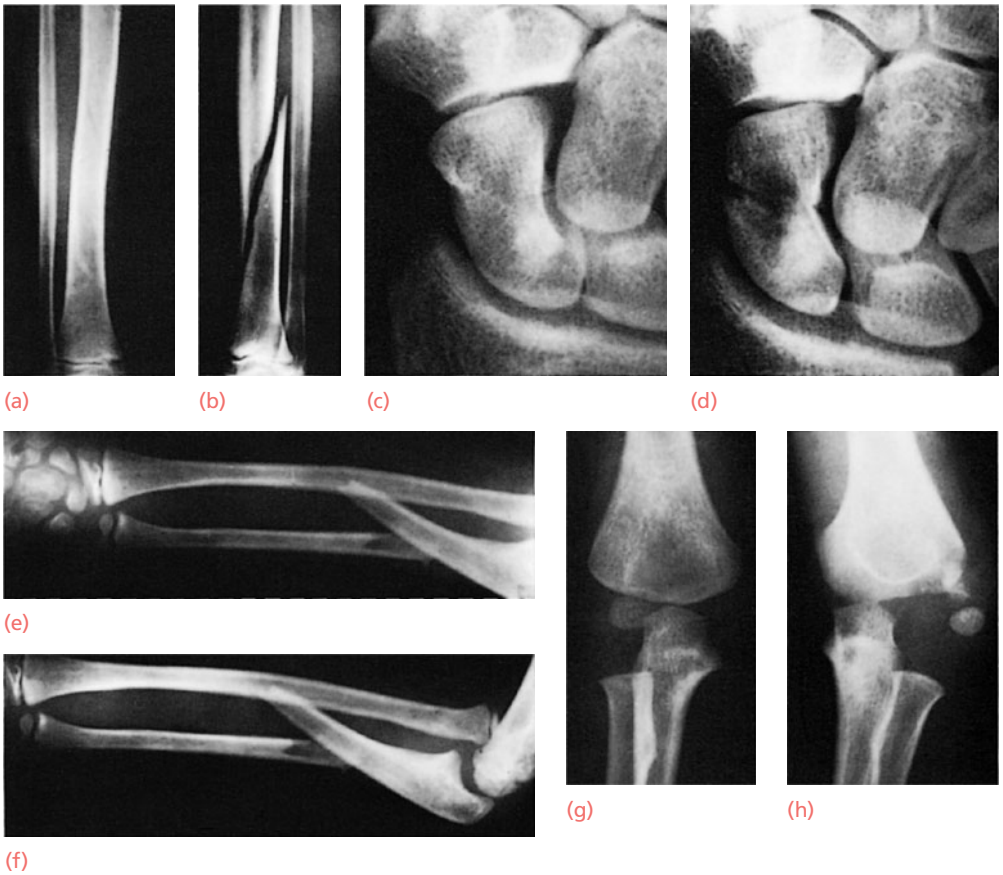


Figure 23.9 X-ray examination must be 'adequate' (a,b) Two views of the same tibia: the fracture may be 'invisible' in one view and perfectly plain in a view at right angles to that. (c,d) More than one occasion: A fractured scaphoid may not be obvious on the day of injury, but clearly seen 2 weeks later. (e,f) Two joints: the first X-ray (e) did not include the elbow; this was, in fact, a Monteggia fracture – the head of the radius is dislocated; (f) shows the dislocated radiohumeral joint. (g,h) Two limbs: sometimes the abnormality can be appreciated only by comparison with the normal side; in this case there is a fracture of the lateral condyle on the left side (h).

a suspected stress (fatigue) fracture or other undisplaced fractures. *Ultrasound* can be used in children to diagnose fracture (imaging fracture line or haematoma is possible).

Description

Diagnosing a fracture is not enough; the surgeon should picture (and describe) it with its properties. Patient characteristics and comorbidities, secondary injuries and last but not least the fracture pattern and displacement are important features that will help the surgeon begin to formulate a management plan. For the description of the fracture, the AO/OTA classification may be used. This classification system offers an easy-to-use and globally accepted system for all bones (see description above).

Shape of the fracture

A *transverse fracture* is slow to join because the area of contact is small; if the broken surfaces are accurately apposed, however, the fracture is stable under compression. A *spiral fracture* joins more rapidly (because of the large contact area) but is not stable under compression. *Comminuted fractures* are often slow to join because they are: (1) associated with more severe soft-tissue damage, and (2) likely to be unstable.

Displacement

For each fracture, four components must be assessed:

- *translation (shift)* – backwards, forwards, sideways
- *angulation (tilt)* – in any direction
- *rotation (twist)* – in any direction along the longitudinal bone axis
- *length* – impaction or distraction.

A problem often arises in the description of angulation. 'Anterior angulation' could mean that the apex of the angle points anteriorly or that the distal fragment is tilted anteriorly: in this chapter it is always the latter meaning that is intended ('anterior tilt of the distal fragment' is probably clearer).

SECONDARY INJURIES

Certain fractures are apt to cause secondary injuries and these should always be assumed to have occurred until proved otherwise.

- *Thoracic injuries* – Fractured ribs or sternum may be associated with injury to the lungs or heart. It is essential to check cardiorespiratory function.
- *Spinal cord injury* – With any fracture of the spine, neurological examination is essential to: (1) establish whether the spinal cord or nerve roots have been damaged; and (2) obtain a baseline for later comparison if neurological signs should change.
- *Pelvic and abdominal injuries* – Fractures of the pelvis may be associated with visceral injury. It is especially important to enquire about urinary function; if a urethral or bladder injury is suspected, diagnostic urethrograms or cystograms may be necessary.
- *Pectoral girdle injuries* – Fractures and dislocations around the pectoral girdle may damage the brachial plexus or the large vessels at the base of the neck. Neurological and vascular examination is essential.

TREATMENT OF CLOSED FRACTURES

General treatment is the first consideration: *treat the whole patient, not only the fracture*. The principles are discussed in Chapter 22.

Treatment of the fracture consists of *manipulation* to improve the position of the fragments, followed by *splintage* to hold them together until they unite; meanwhile joint *movement* and function must be preserved. Fracture healing is promoted by physiological loading of the bone, so muscle activity and early *partial or full weight-bearing* are encouraged. These objectives are covered by three simple injunctions:

- reduce
- hold
- exercise.

Two existential conflicts have to be overcome. The first is how to hold a fracture adequately and yet permit the patient to use the limb sufficiently; this is a conflict that the surgeon seeks to resolve as rapidly as possible (e.g. by internal fixation). However, the surgeon also wants to avoid unnecessary risks – here is a second conflict. This dual conflict epitomizes the four factors that dominate fracture management (the term 'fracture quartet' seems appropriate).

The fact that the fracture is closed (not open) is no cause for complacency. The most important factor in determining the natural tendency to heal is the state of the surrounding soft tissues and the local blood supply. Low-energy (or low-velocity) fractures cause only moderate soft-tissue damage; high-energy (or high-velocity) fractures cause severe soft-tissue damage, no matter whether the fracture is open or closed.

Tscherne has devised a helpful classification of closed injuries:

- *Grade 0* – a simple fracture with little or no soft-tissue injury
- *Grade 1* – a fracture with superficial abrasion or bruising of the skin and subcutaneous tissue

- *Grade 2* – a more severe fracture with deep soft-tissue contusion and swelling
- *Grade 3* – a severe injury with marked soft-tissue damage and a threatened compartment syndrome.

The more severe grades of injury are more likely to require some form of mechanical fixation; good skeletal stability aids soft-tissue recovery.

REDUCTION

Although general treatment and resuscitation must always take precedence, there should not be undue delay in attending to the fracture; swelling of the soft tissues during the first 12 hours makes reduction increasingly difficult. However, there are some situations in which reduction is unnecessary: (1) when there is little or no displacement; (2) when displacement does not matter initially (e.g. in fractures of the clavicle); and (3) when reduction is unlikely to succeed (e.g. with compression fractures of the vertebrae).

Reduction should aim for *adequate apposition* and *normal alignment* of the bone fragments. The greater the contact surface area between fragments, the more likely healing is to occur. A gap between the fragment ends is a common cause of delayed union or non-union. On the other hand, as long as there is contact and the fragments are properly aligned, some overlap at the fracture surfaces is permissible. The exception is a fracture involving an articular surface; this should be reduced as near to perfection as possible because any irregularity will cause abnormal load distribution between the surfaces and predispose to degenerative changes in the articular cartilage.

There are two existing methods of reduction: closed and open.

Closed reduction

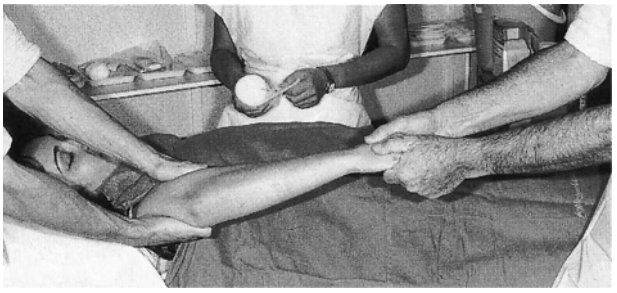
Under appropriate anaesthesia and muscle relaxation, the fracture is reduced by a three-fold manoeuvre: (1) the distal part of the limb is pulled in the line of the bone; (2) as the fragments disengage, they are repositioned (by reversing the original direction of force if this can be deduced); and (3) alignment is adjusted in each plane. This is most effective when the periosteum and muscles on one side of the fracture remain intact; the soft-tissue strap prevents over-reduction and stabilizes the fracture after it has been reduced.

Some fractures are difficult to reduce by manipulation because of powerful muscle pull and these may need prolonged traction.

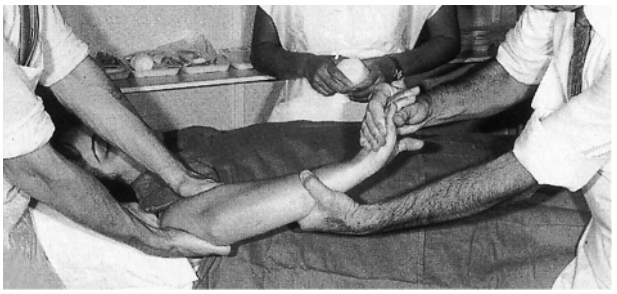
In general, closed reduction can be used for all minimally displaced fractures, for most fractures in children and for fractures that are not unstable after reduction and can be held in some form of splint or cast (Figures 23.10 and 23.11). Unstable fractures can also be reduced using closed methods prior to stabilization with internal or external fixation. Surgeons resort to reduction manoeuvres that avoid fracture-site exposure, even when the aim is some form of internal or external fixation. Traction, which reduces fracture fragments through *ligamentotaxis* (ligament pull), can usually be applied by using a fracture table or bone distractor.

Open reduction

Operative reduction of the fracture under direct vision is indicated: (1) when closed reduction fails, either because of difficulty in controlling the fragments or because soft tissues are interposed between them; (2) when there is a large articular fragment that needs accurate positioning; or (3) for traction (avulsion)



(a)



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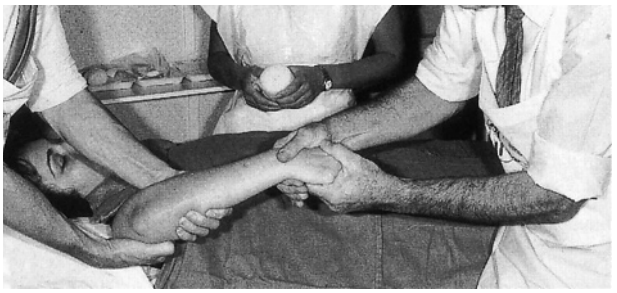


(c)

Figure 23.10 Closed reduction (a) Traction in the line of the bone. (b) Disimpaction. (c) Pressing fragment into reduced position.

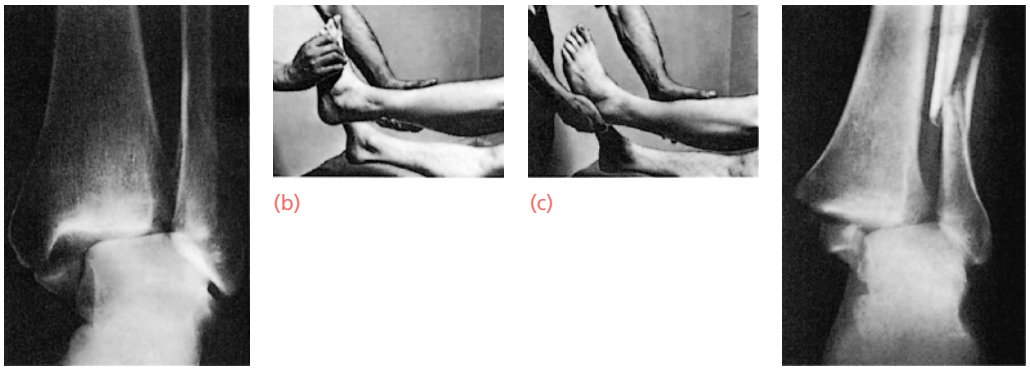


Figure 23.11 Closed reduction These two ankle fractures look somewhat similar but are caused by different forces. The causal force must be reversed to achieve reduction: (a) requires internal rotation (b); an adduction force (c) is needed for (d).

fractures in which the fragments are held apart. As a rule, however, open reduction is merely the first step to fracture fixation.

RETAINING (HOLDING) REDUCTION

The word 'immobilization' has been deliberately avoided because the objective is seldom complete immobility; usually it is the prevention of displacement. Nevertheless, some restriction of movement is needed to promote soft-tissue healing and to allow free movement of the unaffected parts.

The available methods of holding reduction are:

- continuous traction
- cast splintage
- functional bracing
- internal fixation
- external fixation.

In the modern technological age, 'closed' methods are often scorned – an attitude arising from ignorance rather than experience. The muscles surrounding a fracture, if they are intact, act as a fluid compartment; traction or compression creates a hydraulic effect that is capable of splinting the fracture. Therefore, closed methods are most suitable for fractures with intact soft tissues and are liable to fail if they are used as the primary method of treatment for fractures with severe soft-tissue damage. Other contraindications to non-operative methods are inherently unstable fractures, multiple fractures and fractures in confused or uncooperative patients. If these constraints are borne in mind, closed reduction can be sensibly considered in choosing the most suitable method of fracture splintage. Remember, too, that the objective is to splint the fracture, not the entire limb!

CONTINUOUS TRACTION

Traction is applied to the limb distal to the fracture, so as to exert a continuous pull in the long axis of the bone, with a counterforce in the opposite direction (to prevent the patient being merely dragged along the bed). This is particularly useful for shaft fractures that are oblique or spiral and easily displaced by muscle contraction. It can also be used for acetabular fractures with femoral head subluxation or dislocation.

Traction cannot *hold* a fracture still (Figure 23.12); it can pull a long bone straight and hold it out to length but to maintain accurate reduction is sometimes difficult. Meanwhile the patient can *move* the joints and exercise the muscles.

Traction is safe enough, provided that it is not excessive and care is taken during insertion of the traction pin. In contemporary orthopaedics, traction is generally used as a temporizing measure to bridge the time from fracture to definitive treatment. Traction methods include the following (Figure 23.13):

- *Traction by gravity* – This applies only to upper limb injuries. Thus, with a wrist sling the weight of the arm provides continuous traction to the humerus. For comfort and stability, especially with a transverse fracture, a U-slab of plaster may be bandaged on or, better, a removable plastic sleeve from the axilla to just above the elbow is held on with Velcro.
- *Skin traction* – Skin traction will sustain a pull of no more than 4–5 kg. Holland strapping or one-way-stretch Elastoplast is stuck to the shaved skin and held on with a bandage. The malleoli are protected by Gamgee tissue, and cords or tapes are used for traction.

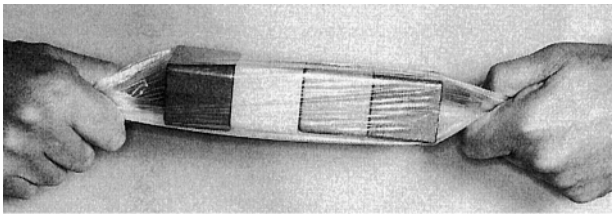


Figure 23.12 Hold reduction Showing how traction will align the bony fragments if the soft tissues around a fracture are intact.

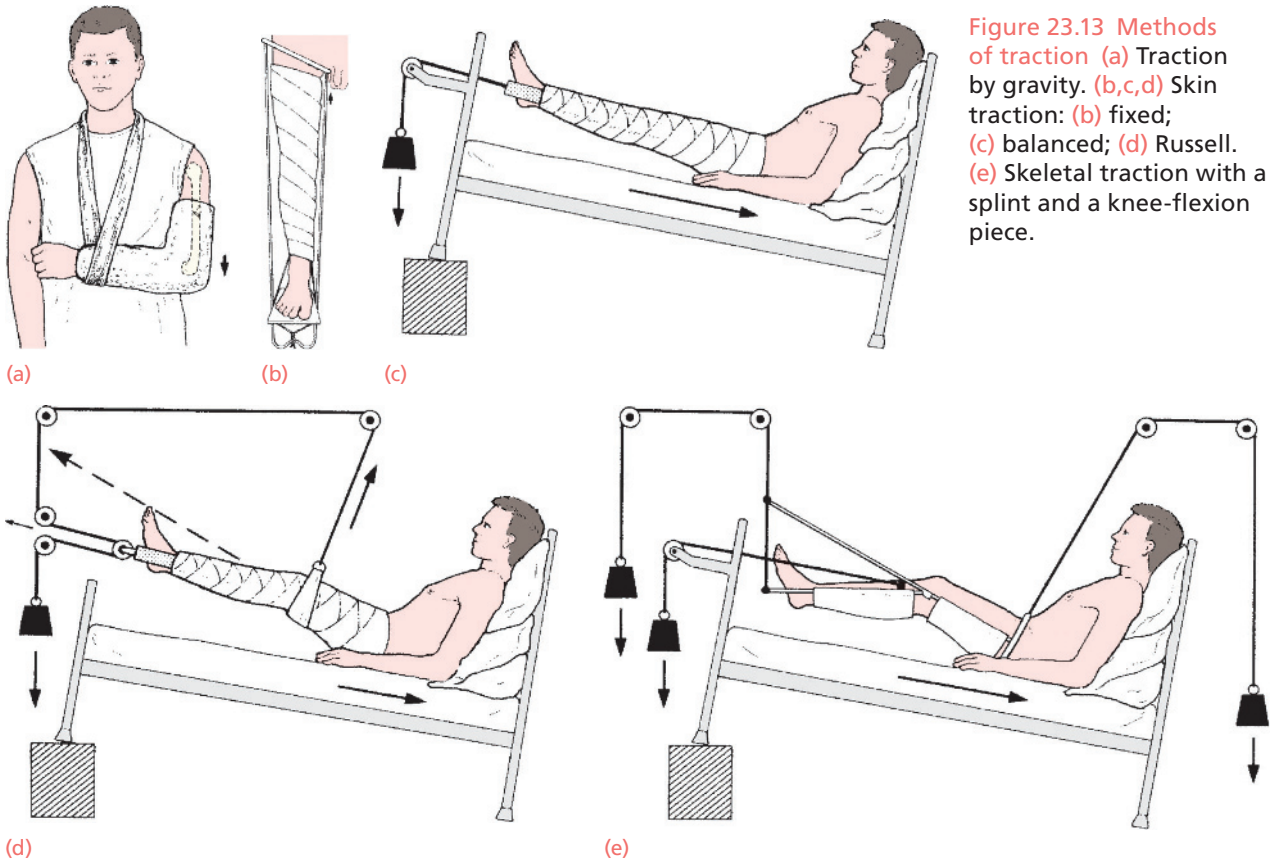


Figure 23.13 Methods of traction (a) Traction by gravity. (b,c,d) Skin traction: (b) fixed; (c) balanced; (d) Russell. (e) Skeletal traction with a splint and a knee-flexion piece.

- *Skeletal traction* – A stiff wire or pin is inserted – usually behind the tibial tubercle for hip, thigh and knee injuries, or through the calcaneum for tibial fractures – and cords tied to them for applying traction. Whether by skin or skeletal traction, the fracture is reduced and held in one of three ways: fixed traction, balanced traction or their combination.

Complications of traction

Circulatory embarrassment Especially in children, traction tapes and circular bandages may constrict the circulation; for this reason 'gallows traction', in which the baby's legs are suspended from an overhead beam, should never be used for children over 12 kg in weight.

Nerve injury In older people, leg traction may predispose to peroneal nerve injury and cause a drop foot; the limb should be checked repeatedly to see that it does not roll into external rotation during traction.

Pin site infection Pin sites must be kept clean and should be checked daily.

Cast splintage

Plaster is still widely used as a splint, especially for distal limb fractures and for most children's fractures. It is safe enough, as long as the practitioner is alert to the danger of a tight cast and provided that pressure sores are prevented. Holding reduction is usually no problem. However, joints encased in plaster cannot move and are liable to stiffen; stiffness, which has earned the sobriquet 'fracture disease', is the problem with conventional casts. While the swelling and haematoma resolve, adhesions may form bind muscle fibres to each other and to the bone; with articular fractures, plaster perpetuates surface irregularities (closed reduction is seldom perfect) and lack of movement inhibits the healing of cartilage defects. Newer substitutes have some advantages over plaster – they are impervious to water, and also lighter – but as long as they are used as full casts the basic drawback is the same.

Technique

After the fracture has been reduced, stockinette is threaded over the limb and the bony prominences are protected with wool. Plaster is then applied (Figure 23.14). While it is setting, the surgeon moulds it away from bony prominences; with shaft fractures three-point pressure can be applied to keep the intact periosteal hinge under tension and thereby maintain reduction.

If the fracture is recent, further swelling is likely to occur; the plaster and stockinette are therefore split

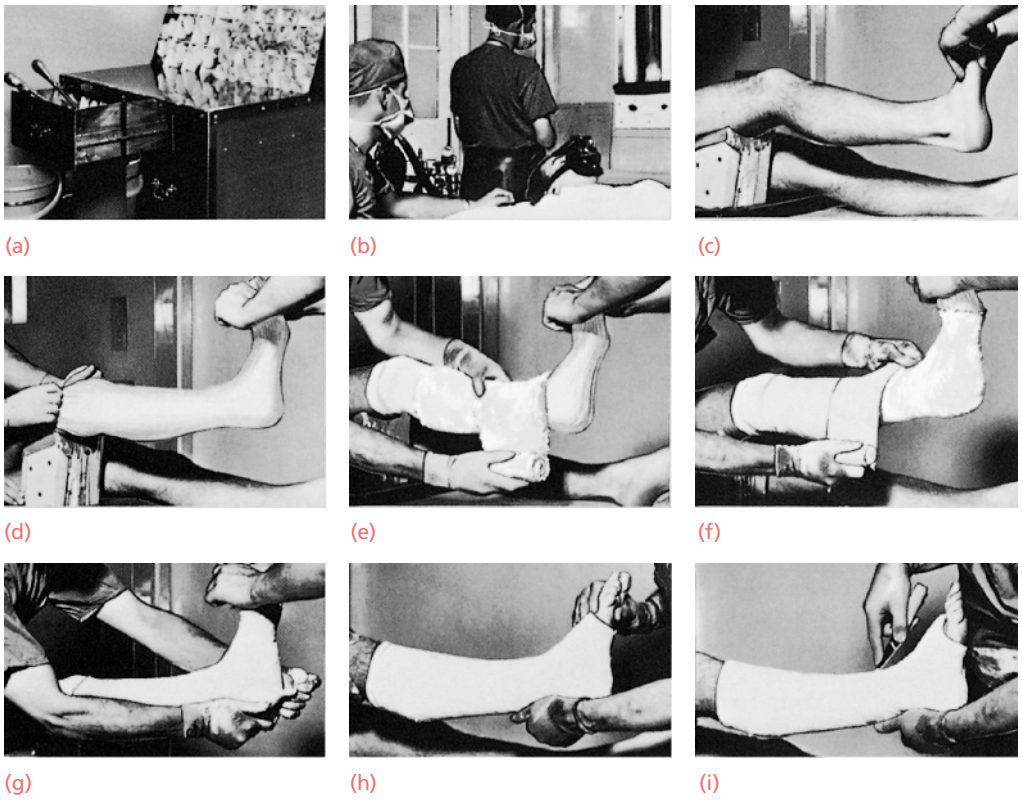


Figure 23.14 Plaster technique Applying a well-fitting and effective plaster needs experience and attention to detail. (a) A well-equipped plaster trolley is invaluable. (b) Careful study of the X-rays is indispensable. (c) For a below-knee plaster the thigh is best supported on a padded block. (d) Stockinette is threaded smoothly onto the leg. (e) For a padded plaster the wool is rolled on and it must be even. (f) Plaster is next applied smoothly, taking a tuck with each turn, and (g) smoothing each layer firmly onto the one beneath. (h) While still wet the cast is moulded away from the point points. (i) With a recent injury the plaster is then split.

from top to bottom, exposing the skin. Check X-rays are essential and the plaster can be wedged if further correction of angulation is necessary.

With shaft fractures of long bones, rotation is controlled only if the plaster includes the joints above and below the fracture. In the lower limb, the knee is usually held slightly flexed, the ankle at a right angle and the tarsus and forefoot neutral (this 'plantigrade' position is essential for normal walking). In the upper limb the position of the splinted joints varies with the fracture. Splintage must not be discontinued (though a functional brace may be substituted) until the fracture is consolidated; if plaster changes are needed, check X-rays are essential.

Complications

Plaster immobilization is safe, but only if care is taken to prevent certain complications. These are tight cast, pressure sores, abrasion or laceration of the skin and loose cast.

Tight cast The cast may be put on too tightly, or it may become tight if the limb swells. The patient complains of diffuse pain; only later – sometimes much later – do the signs of vascular compression appear. The only safe course is to split the cast and ease it open: (1) throughout its length, and (2) through all the padding down to skin. Whenever swelling is anticipated, the cast should be applied over thick padding and the plaster should be split before it sets, so as to provide a firm but not absolutely rigid splint.

Pressure sores Even a well-fitting cast may press upon the skin over a bony prominence (e.g. the patella, heel, elbow or head of the ulna). The patient complains of localized pain precisely over the pressure spot. Such localized pain demands immediate inspection through a window in the cast.

Skin abrasion or laceration This is really a complication of removing plasters, especially if an electric saw is used. Complaints of nipping or pinching during plaster removal should never be ignored; a ripped forearm is a good reason for litigation.

Loose cast Once the swelling has subsided, the cast may no longer hold the fracture securely. If it is loose, the cast should be replaced.

Functional bracing

Functional bracing, using either plaster or one of the lighter thermoplastic materials, is one way of preventing joint stiffness while still permitting fracture splintage and loading. Functional bracing is commonly used in fracture-dislocations. Segments of a cast are applied only over the shafts of the bones, leaving the joints free; the cast segments are connected by metal or plastic hinges that allow movement of the joint in one plane. The splints are 'functional' in that joint movements are much less restricted than with conventional casts.

Functional bracing is used most widely for fractures of the femur or tibia, but since the brace is not very rigid, it is usually applied only when the fracture is beginning to unite, i.e. after 3–6 weeks of traction or conventional plaster.

Technique

Considerable skill is needed to apply an effective brace. First the fracture is 'stabilized' during a few days under traction or in a conventional plaster. Then a hinged cast or splint is applied, which holds the fracture snugly but permits joint movement; functional activity is encouraged. Unlike internal fixation, functional bracing holds the fracture through compression of the soft tissues; the small amount of movement that occurs at the fracture site through using the limb encourages vascular proliferation and callus formation.

For details of the rationale, technique and applications of functional bracing, see Sarmiento and Latta (1999, 2006) in Further reading.

Internal fixation

Bone fragments may be fixed with screws, a metal plate held by screws, a long intramedullary rod or nail (with or without interlocking screws), circumferential bands or a combination of these methods.

Properly applied, internal fixation holds a fracture securely so that movement can begin immediately; with early movement 'fracture disease' (stiffness and oedema) is abolished.

The greatest danger, however, is infection; if infection supervenes, all the manifest advantages of internal fixation (reduction, immediate stability and early mobilization) may be lost. The risk of infection depends upon: (1) the patient – devitalized tissues, a dirty wound and an unfit patient are all risk factors for infection; (2) the surgeon – thorough training, a high degree of surgical dexterity and adequate assistance are all essential; and (3) the facilities – a guaranteed aseptic routine, a full range of implants and staff familiar with their use are all indispensable.

Indications

Internal fixation is often the most desirable form of treatment (Figure 23.15). The most important indications (see Figure 23.16) are:

- fractures that cannot be reduced except by operation
- fractures that are inherently unstable and prone to redisplace after reduction (e.g. midshaft fractures of the forearm and some displaced ankle fractures) plus those fractures liable to be pulled apart by muscle action (e.g. transverse fracture of the patella or olecranon)
- pathological fractures in which bone disease may inhibit healing
- multiple fractures where early fixation (by either internal or external fixation) reduces the risk of general complications and late multisystem organ failure
- fractures in patients who present nursing difficulties (e.g. paraplegics, those with multiple injuries and the very elderly).

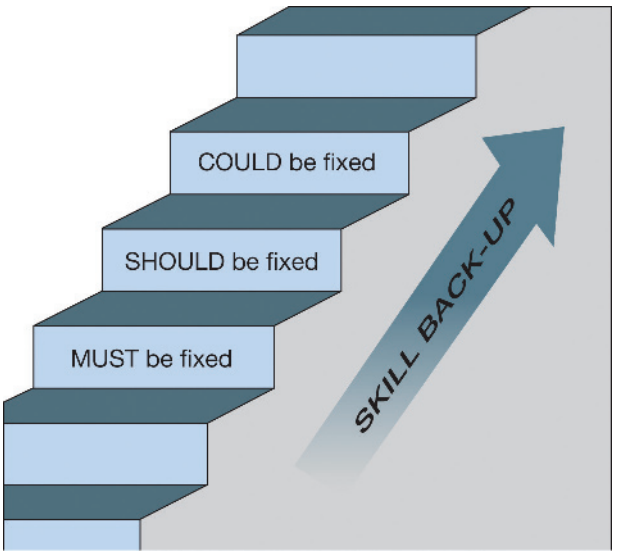


Figure 23.15 Indications staircase The indications for internal fixation are not immutable; thus, if the surgical skill or back-up facilities (staff, sterility and equipment) are of a low order, internal fixation is indicated only when the alternative is unacceptable (e.g. with femoral neck fractures). With average skill and facilities, fixation is indicated when alternative methods are possible but very difficult or unwise (e.g. multiple injuries). With the highest levels of skill and facilities, internal fixation is reasonable if it saves time, money or beds.

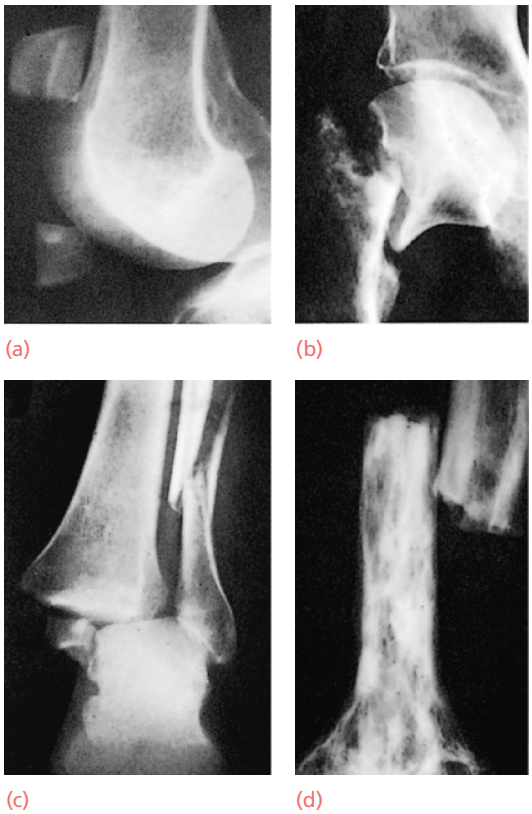


Figure 23.16 Indications for internal fixation (a) This patella has been pulled apart and can be held together only with internal fixation. (b) This patient was considered to be too ill for operation; her femoral neck fracture has failed to unite without rigid fixation. (c) Fracture dislocation of the ankle is often unstable after reduction and usually requires fixation. (d) Pathological fracture in Paget bone; without fixation, union may not occur.

Types of internal fixation

Interfragmentary lag screws Screws that are partially threaded exert a compression or 'lag' effect when inserted across two fragments. A similar effect is achieved by overdrilling the 'near' bone cortex. This technique is useful for reducing single fragments onto the main shaft of a tubular bone or fitting together fragments of a metaphyseal fracture.

Cerclage and tension-band wires are essentially loops of wire passed around two bone fragments and then tightened to compress the fragments together. When using cerclage wires, make sure that the wires hug the bone and do not embrace any of the nerves or vessels in the vicinity.

Plates and screws This form of fixation is useful for articular, metaphyseal and diaphyseal fractures. Plates have five different functions:

- *Neutralization (protection)* – Plates provide protection when used to bridge a fracture and supplement the effect of interfragmentary lag screws; the plate is applied to resist torque and shortening.
- *Compression* – Plates are often used in simple metaphyseal and diaphyseal fractures to achieve primary bone healing (no callus). This technique is less appropriate for multifragmentary fractures. To preserve vascularity, a minimally invasive technique may be used.
- *Buttressing* – Here the plate resists axial load by applying force against the axis of deformity (e.g. in treating fractures of the proximal tibial plateau).
- *Tension-band* – Using a plate in this manner on the tensile surface of the bone allows compression to be applied to the biomechanically more advantageous side of the fracture preventing its opening.
- *Bridging* – The plate bridges simple or multifragmentary fractures to restore correct length, axis and rotation with minimal stripping of soft tissues.

Intramedullary nails They are suitable for long bones. A nail (or long rod) is inserted into the medullary canal to splint the fracture; rotational forces are resisted by introducing transverse *interlocking screws* that transfix the bone cortices and the nail proximal and distal to the fracture. Nails are used with or without prior reaming of the medullary canal; reamed nails achieve an interference fit in addition to the added stability from interlocking screws. Published data suggest that reamed nails have a higher union rate.

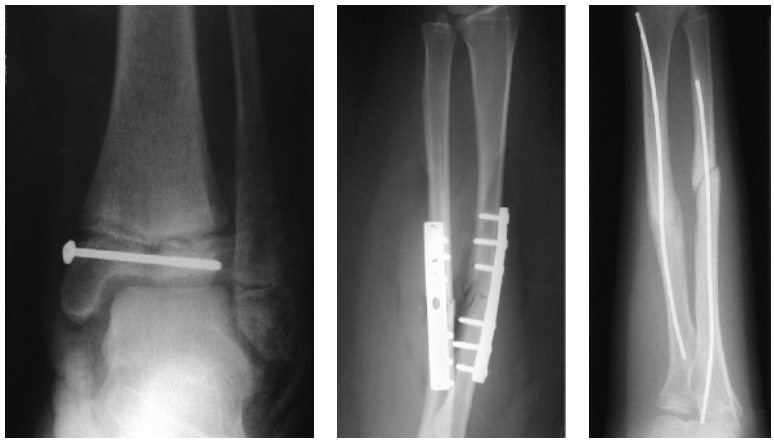
The type of fixation used must be appropriate to the situation (Figure 23.17).

Complications of internal fixation

Most of the complications of internal fixation are due to poor technique, poor equipment or poor operating conditions (Figure 23.18).

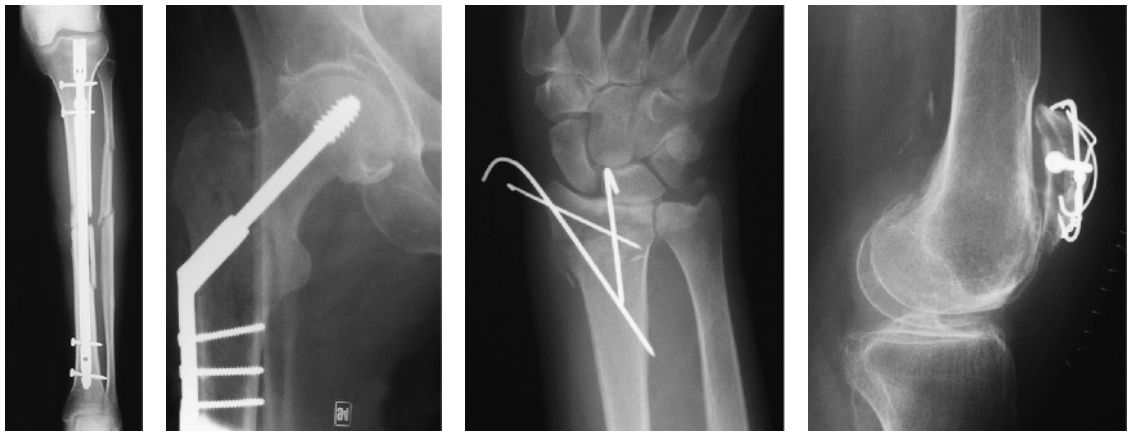
Infection Iatrogenic infection is now the most common cause of chronic osteomyelitis, which can necessitate multiple revision surgeries and delay healing; in experimental studies, the presence of an implanted medical device has been shown to lower the number of bacteria required to cause an infection and so, in all cases in which orthopaedic hardware is introduced, there should be an awareness of this risk. The operation and quality of the patient's tissues, i.e. tissue handling, can influence the risk of infection.

Non-union If the bones have been fixed rigidly with a gap between the ends, the fracture may fail to unite. This is more likely in the leg or the forearm if one bone is fractured and the other remains intact. Other causes of non-union are stripping of the soft

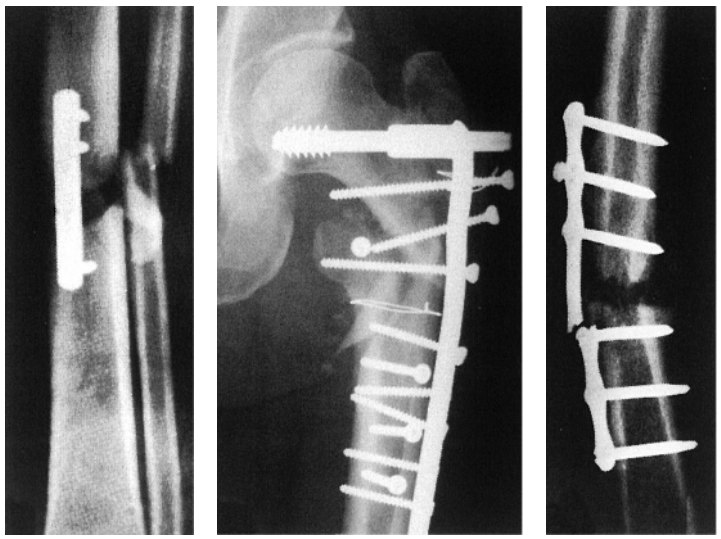


(a) (b) (c)

Figure 23.17 Internal fixation The method used must be appropriate to the situation: (a) lag screws – interfragmentary compression; (b) plate and screws – here at the forearm diaphysis; (c) flexible intramedullary nails – for long bones in children, particularly forearm bones and femur; (d) interlocking nail and screws – ideal for long bone shaft fractures in adults; (e) dynamic compression screw and plate – ideal for the proximal and distal ends of the femur; (f) simple K-wires – for fractures around the elbow and wrist and (g) tension-band wiring – for olecranon fractures or fractures of the patella.



(d) (e) (f) (g)



(a) (b) (c)

Figure 23.18 Poor fixation (how not to do it) (a) Too little. (b) Too much. (c) Too weak.

tissues and damage to the blood supply in the course of operative fixation.

Implant failure Metal is subject to fatigue and can fail unless some bone union of the fracture has occurred. If the device used to fix the fracture is not capable of supporting the full load transferred through the limb on normal activity, a period of protected (or partial) weight-bearing may be required until callus or other radiological sign of fracture healing is seen on X-ray. Pain at the fracture site is a danger signal and must be investigated.

Refracture It is important not to remove metal implants too soon, or the bone may refracture. A year is the minimum and 18–24 months safer; several weeks after removal the bone is still weak, and care or protection is needed.

External fixation

A fracture may be held by transfixing screws that pass through the bone above and below the fracture and are attached to an external frame. This is especially applicable to the tibia and pelvis, but the method is also used for fractures of the femur, humerus, distal radius and even bones of the hand.

Indications

External fixation is particularly useful for the following:

- fractures associated with severe soft-tissue damage (including open fractures) or those that are contaminated, where internal fixation is risky and repeated access is needed for wound inspection, dressing or plastic surgery
- fractures around joints that are potentially suitable for internal fixation but the soft tissues are too swollen to allow safe surgery – a spanning external fixator provides stability until soft-tissue conditions improve
- patients with severe multiple injuries, especially if there are bilateral femoral fractures, pelvic fractures with severe bleeding, and those with limb and associated chest or head injuries
- ununited fractures, which can be excised and compressed; sometimes this is combined with bone lengthening to replace the excised segment
- infected fractures, for which internal fixation might not be suitable.

Technique

The principle of external fixation is simple: the bone is transfixed above and below the fracture with Schanz screws or tensioned wires and these are then connected to each other by rigid bars. There are numerous types of external fixation devices; they vary in the technique of application and each type can be constructed to provide varying degrees of rigidity and stability. Most of them permit adjustment of length and alignment after application on the limb (Figure 23.19).

The fractured bone can be thought of as broken into segments – a simple fracture has two segments whereas a two-level (segmental) fracture has three and so on. Each segment should be held securely, ideally with the half-pins or tensioned wires straddling the length of that segment.

The wires and half-pins must be inserted with care. Knowledge of 'safe corridors' is essential so as to avoid injuring nerves or vessels; in addition, the entry sites should be irrigated during drilling to prevent burning of the bone (a temperature of only 50 °C can cause bone death – bone necrosis). The fracture

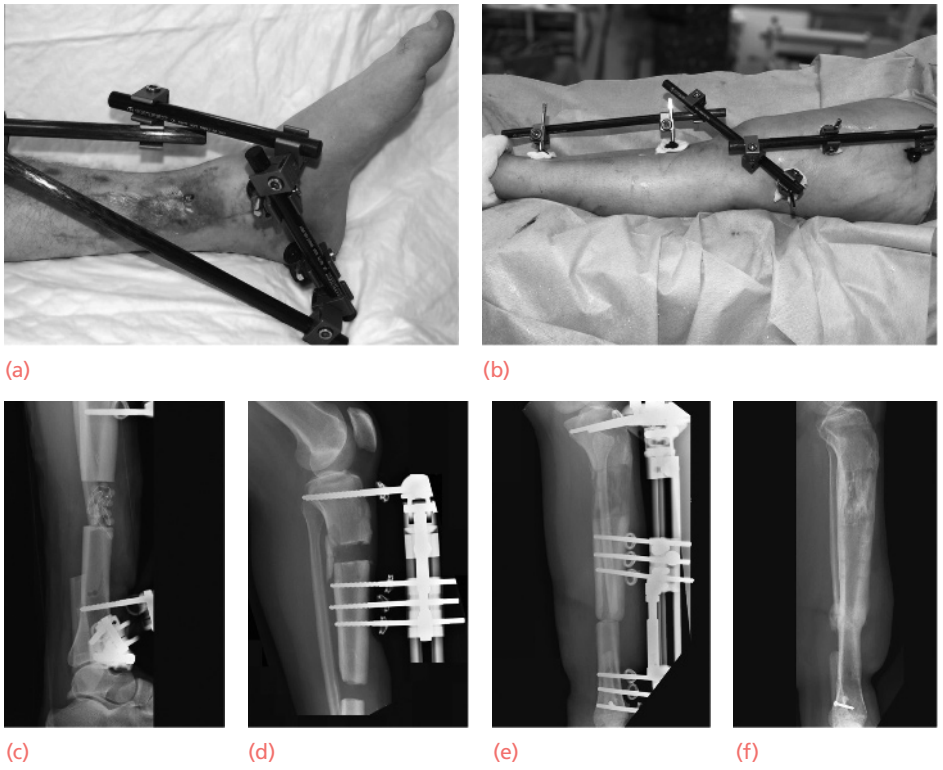


Figure 23.19 External fixation of fractures External fixation is widely used for 'damage control' (a,b) temporary stabilization of fractures in order to allow the patient's general condition or the state of soft tissues to improve prior to definitive surgery or (c–f) reconstruction of limbs using distraction osteogenesis. (c) A bone defect after surgical resection with gentamicin beads used to fill the space temporarily. (d) Bone transport from a more proximal osteotomy. (e) 'Docking' of the transported segment and (f) final union and restoration of structural integrity.

is then reduced by connecting the various groups of pins by rods.

Depending on the stability of fixation and the underlying fracture pattern, partial weight-bearing can be started as early as possible to 'stimulate' fracture healing. Some fixators incorporate a telescopic unit that allows 'dynamization'; this will convert the forces of weight-bearing into axial micromovement at the fracture site, thus promoting callus formation and accelerating bone union.

Complications of external fixation

Damage to soft-tissue structures Transfixing pins or wires may injure nerves or vessels, or may tether ligaments and inhibit joint movement. The surgeon must be thoroughly familiar with the cross-sectional anatomy before operating.

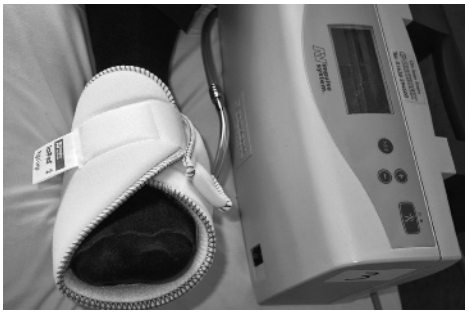
Overdistraction If there is no contact between the fragments, bone union is unlikely to occur.

Pin-track infection This is less likely with good operative technique. Nevertheless, meticulous pin-site care is essential to avoid infection.

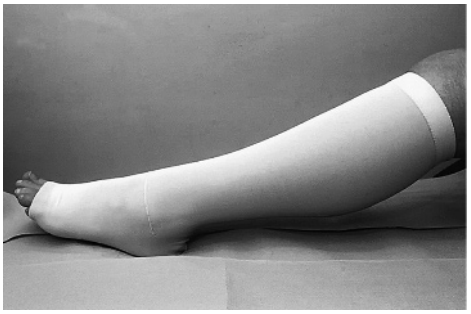
EXERCISE

More correctly, restore function – not only to the injured parts but also to the patient as a whole. The objectives are to reduce oedema, preserve joint movement, restore muscle power and guide the patient back to normal activity (Figure 23.20).

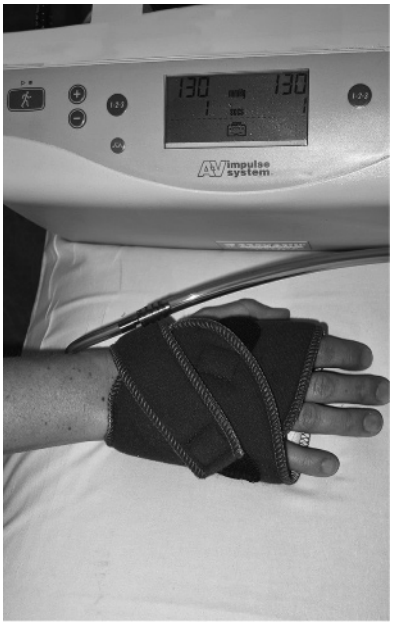
Prevention of oedema Swelling is almost inevitable after a fracture and may cause skin stretching and blisters. Persistent oedema is an important cause of joint stiffness, especially in the hand; it should be prevented if possible, and treated energetically if it is already present, by a combination of elevation and exercise. Not every patient needs admission to hospital, and less severe injuries of the upper limb are successfully managed by placing the arm in a sling; however, it is then essential to insist on active use, with movement of all the joints that are free. As with most closed fractures, in all open fractures and all fractures treated by internal fixation it must be assumed that swelling will occur; the limb should be elevated and active exercises should start as soon as the patient will tolerate this. The essence of soft-tissue care may be summed up like this: elevate and exercise; never dangle, never force.



(a)



(b)



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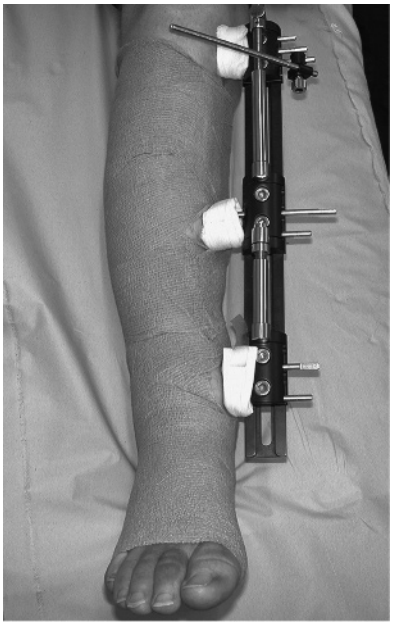


(d)

Figure 23.20 Some aspects of soft-tissue treatment Swelling is minimized by improving venous drainage. This can be accomplished by (1) elevation and (2) firm support. Stiffness is minimized by exercise. (a,c) Intermittent venous plexus pumps for use on the foot or palm to help reduce swelling. (b) A made-to-measure pressure garment that helps reduce swelling and scarring after treatment. (d) Coban wrap around a limb to control swelling during treatment.

Elevation An injured limb usually needs to be elevated; after reduction of a leg fracture the foot is raised off the bed and exercises start. If the leg is in plaster, the limb must, at first, be dependent for only short periods; between these periods, the leg is elevated on a chair. In order to reduce swelling, the limb should be elevated to the same level as the heart or above. The patient is allowed and encouraged to exercise the limb actively, but not to let it dangle. When the plaster is finally removed, a similar routine of activity, punctuated by elevation, is practised until circulatory control is fully restored.

Injuries of the upper limb also need elevation. A sling must not be a permanent passive arm-holder; the limb must be elevated intermittently or, if needed, continuously.

Active exercise Active movement helps to pump away oedema fluid, stimulates the circulation, prevents soft-tissue adhesion and promotes fracture healing. A limb encased in plaster is still capable of static muscle contraction and the patient should be taught how to do this. When splintage is removed the joints are mobilized and muscle-building exercises are steadily increased. Remember that the unaffected joints need exercising too; it is all too easy to neglect a stiffening shoulder while caring for an injured wrist or hand.

Assisted movement It has long been taught that passive movement can be deleterious, especially with injuries around the elbow, where there is a high risk of developing myositis ossificans. Certainly, forced movements should never be permitted, but gentle assistance during active exercises may help to retain function or regain movement after fractures involving the articular surfaces. Nowadays this is done with machines that can be set to provide a specified range and rate of movement ('continuous passive motion' – Figure 23.21).

Functional activity As the patient's mobility improves, an increasing amount of directed activity is included in the programme. The patient may need to be taught again how to perform everyday tasks such as walking, getting in and out of bed, bathing, dressing or handling eating utensils. Experience is the best teacher and the patient is encouraged to use the injured limb as much as possible. Those with very severe or extensive injuries may benefit from spending time in a special rehabilitation unit, but the best incentive to full recovery is the promise of re-entry into family life, recreational pursuits and meaningful work.

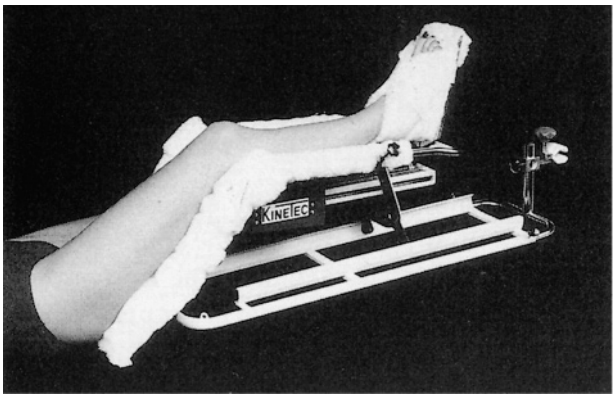


Figure 23.21 Continuous passive motion The motorized frame provides continuous flexion and extension to pre-set limits.

TREATMENT OF OPEN FRACTURES

Initial management

Patients with open fractures may have multiple injuries; a rapid general assessment is the first step and any life-threatening conditions are addressed (see Chapter 22).

The open fracture may draw attention away from other more important conditions and it is essential that the step-by-step approach in advanced trauma life support is not forgotten.

When the treatment of the patient allows the fracture to be dealt with, the wound is first carefully inspected; any gross contamination is removed, the wound is photographed to record the injury and the area is then covered with a saline-soaked dressing under an impervious seal to prevent desiccation. This is left undisturbed until the patient is in the operating theatre. The patient is given antibiotics, usually co-amoxiclav or cefuroxime, but clindamycin if the patient is allergic to penicillin. Tetanus prophylaxis is administered: toxoid for those previously immunized, human antiserum if not. The limb is then splinted until surgery is undertaken.

The limb circulation and distal neurological status will need to be checked repeatedly, particularly after any fracture reduction manoeuvres. Compartment syndrome is not prevented by an existing open fracture; vigilance for this complication is essential.

Classifying the injury

Treatment is determined by the type of fracture, the nature of the soft-tissue injury (including the wound size) and the degree of contamination. *Gustilo's classification of open fractures* into three types is widely used (Gustilo *et al.*, 1984) – note that final grading can only be done at the time of surgery.

- *Type 1* – The wound is usually a small, clean puncture through which a bone spike has protruded. There is little soft-tissue damage with no crushing and the fracture is not comminuted (i.e. a low-energy fracture).

- *Type II* – The wound is more than 1 cm long, but there is no skin flap. There is not much soft-tissue damage and no more than moderate crushing or comminution of the fracture (also a low- to moderate-energy fracture).
- *Type III* – There is a large laceration, extensive damage to skin and underlying soft tissue and, in the most severe examples, vascular compromise. The injury is caused by high-energy transfer to the bone and soft tissues. Contamination can be significant.

There are three grades of type III severity. In *type III A* the fractured bone can be adequately covered by soft-tissue despite the laceration, fracture pattern (e.g. multifragmentary, segmental) or bone loss irrespective of the size of skin wound. In *type III B* there is extensive periosteal stripping and fracture cover is not possible without use of local or distant flaps. The fracture is classified as *type III C* if there is an arterial injury that needs to be repaired, regardless of the amount of other soft-tissue damage.

The incidence of wound infection correlates directly with the extent of soft-tissue damage, rising from less than 2% in type I to more than 20% in type III fractures.

PRINCIPLES OF TREATMENT

All open fractures, no matter how trivial they may seem, must be assumed to be contaminated; it is important to try to prevent them from becoming infected. The four essentials are:

- antibiotic prophylaxis
- urgent wound and fracture debridement
- early definitive wound cover
- stabilization of the fracture.

Sterility and antibiotic cover

The wound should be kept covered until the patient reaches the operating theatre. Antibiotic prophylaxis in open fractures is an adjunct to meticulous wound debridement and should not be expected to overcome failings in aseptic technique or debridement. According to the combined recommendations of the British Orthopaedic Association (BOA) and British Association of Plastic, Reconstructive and Aesthetic Surgeons (BAPRAS), co-amoxiclav or cefuroxime (or clindamycin if penicillin allergy is an issue) is given as soon as possible in most cases. This is often in the Accident and Emergency Department but some services can provide antibiotics pre-hospital. The antibiotics provide prophylaxis against the majority of Gram-positive and Gram-negative bacteria that may have entered the wound at the time of injury. The co-amoxiclav or cefuroxime (or clindamycin) is continued until wound debridement.

Wounds of Gustilo type I and II fractures can be closed primarily at the time of debridement. With Gustilo type IIIA fractures, some surgeons prefer to delay closure until after a 'second look' procedure. Delayed cover is also usually practised in most cases of type IIIB and IIIC injuries. As the wounds have now been present in a hospital environment for some time, and there are data to indicate infections after such open fractures are caused mostly by hospital-acquired bacteria and not seeded at the time of injury, further antibiotics are given at the time of debridement (the BOA/BAPRAS recommendations are co-amoxiclav and gentamicin; vancomycin or teicoplanin are alternatives) and continued for 72 hours or until definitive wound closure, whichever is sooner. These antibiotics are effective against methicillin-resistant *Staphylococcus aureus* and *Pseudomonas*, both of which are near the top of the league table of responsible bacteria (Table 23.1).

Table 23.1 Antibiotics for open fractures[1]

	Type I	Type II	Type IIIA	Type IIIB/IIIC
As soon as possible (within 3 hours of injury)	Co-amoxiclav[2]	Co-amoxiclav[2]	Co-amoxiclav[2]	Co-amoxiclav[2]
At debridement	Co-amoxiclav[2] and gentamicin	Co-amoxiclav[2] and gentamicin	Co-amoxiclav[2] and gentamicin	Co-amoxiclav[2] and gentamicin
At definitive fracture cover	Wound cover is usually possible at debridement Delayed closure unnecessary	Wound cover is usually possible at debridement If delayed, gentamicin and vancomycin (or teicoplanin) at the time of cover	Wound cover is usually possible at debridement If delayed, gentamicin and vancomycin (or teicoplanin) at the time of cover	Gentamicin and vancomycin (or teicoplanin)
Continued prophylaxis	Only co-amoxiclav[2] continued after surgery	Only co-amoxiclav[2] continued between procedures and after final surgery	Only co-amoxiclav[2] continued between procedures and after final surgery	Only co-amoxiclav[2] continued between procedures and after final surgery
Maximum period	24 hours	72 hours	72 hours	72 hours

[1] Based on the Standards for the Management of Open Fractures of the Lower Limb, BOA/BAPRAS, 2009.
[2] Or cefuroxime (clindamycin for those with penicillin allergy).

Debridement

The operation aims to render the wound free from foreign material and of dead tissue (for example, avascular bone fragments), leaving a clean surgical field and tissues with a good blood supply throughout. Under general anaesthesia the patient's clothing is removed, while an assistant maintains traction on the injured limb and holds it still. The dressing previously applied to the wound is replaced by a sterile pad and the surrounding skin is cleaned. The pad is then taken off and the wound is irrigated thoroughly with copious amounts of physiological saline. The wound is covered again and the patient's limb then prepped and draped for surgery.

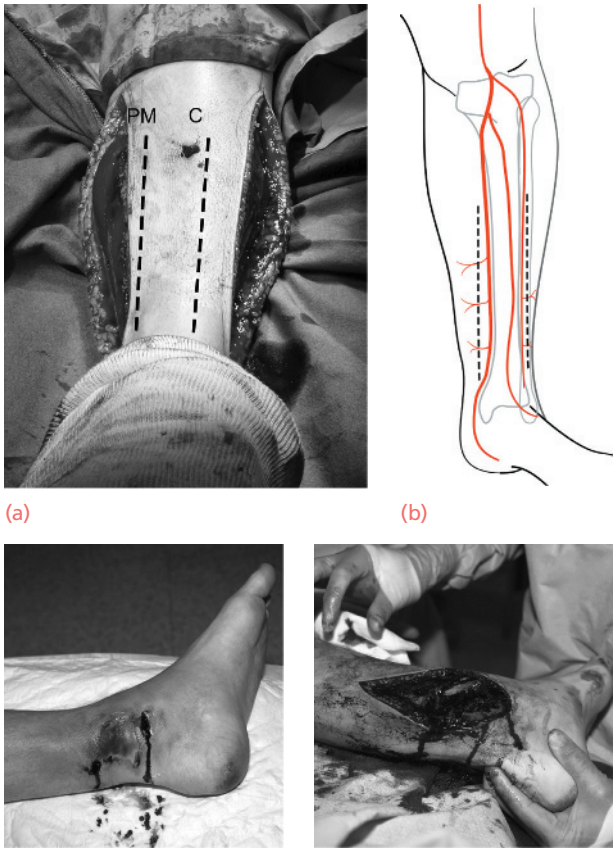


Figure 23.22 Wound extensions for access in open fractures of the tibia Wound incisions (extensions) for adequate access to an open tibial fracture are made along standard fasciotomy incisions: 1 cm behind the posteromedial border of the tibia and 2–3 cm lateral to the crest of the tibia as shown in this example of a two-incision fasciotomy. The dotted lines mark out the crest (C) and posteromedial corner (PM) of the tibia (a). These incisions avoid injury to the perforating branches supplying areas of skin that can be used as flaps to cover the exposed fracture (b). This clinical example shows how local skin necrosis around an open fracture is excised and the wound extended proximally along a fasciotomy incision (c,d).

Many surgeons prefer to use a tourniquet as this provides a bloodless field. However, this induces ischaemia in an already badly injured leg and can make it difficult to recognize which structures are devitalized. A compromise is to apply the tourniquet but not to inflate it during the debridement unless absolutely necessary.

Because open fractures are often high-energy injuries with severe tissue damage, the operation should be performed by someone skilled in dealing with both skeletal and soft tissues; ideally, this will be a joint effort by orthopaedic and plastic surgeons. The following principles must be observed.

Wound excision The wound margins are excised, but only enough to leave healthy skin edges.

Wound extension Thorough cleansing necessitates adequate exposure; poking around in a small wound to remove debris can be dangerous. If extensions are needed, they should not jeopardize the creation of skin flaps for wound cover if this should be needed. The safest extensions are to follow the line of fasciotomy incisions; these avoid damaging important perforator vessels that can be used to raise skin flaps for eventual fracture cover (Figure 23.22).

Delivery of the fracture Examination of the fracture surfaces cannot be adequately performed without extracting the bone from within the wound (Figure 23.23). The simplest (and gentlest) method is to bend the limb in the manner in which it was forced at the moment of injury; the fracture surfaces will be exposed through the wound without any additional damage to the soft tissues. Large bone levers and retractors should not be used.

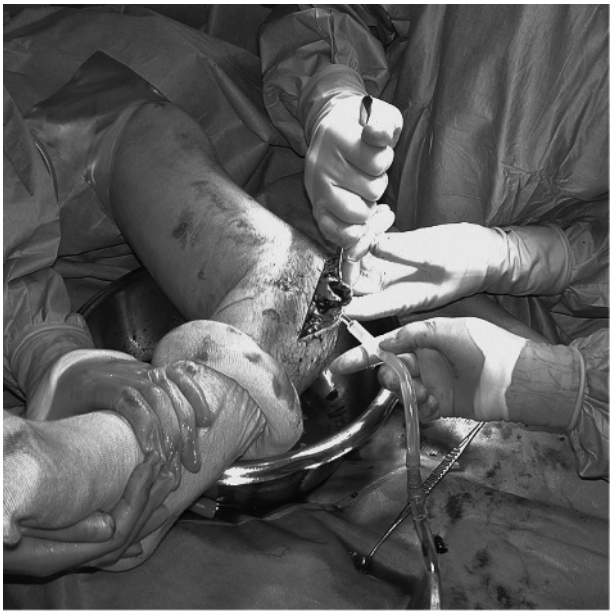


Figure 23.23 Delivering the fracture Debridement is only possible if the fracture is adequately seen; for this, the fracture ends have to be delivered from within.

Removal of devitalized tissue Devitalized tissue provides a nutrient medium for bacteria. Dead muscle can be recognized by its purplish colour, its mushy consistency, its failure to contract when stimulated and its failure to bleed when cut. All doubtfully viable tissue, whether soft or bony, should be removed. The fracture ends can be curetted or nibbled away until seen to bleed.

Wound cleansing All foreign material and tissue debris is removed by excision or through a wash with copious quantities of saline. A common mistake is to inject fluid through a small aperture using a syringe – this only serves to push contaminants further in; up to 12 L of saline may be needed to irrigate and clean an open fracture of a long bone. Adding antibiotics or antiseptics to the solution has no added benefit.

Nerves and tendons As a general rule it is best to leave cut nerves and tendons alone, though if the wound is absolutely clean and no dissection is required – and provided the necessary expertise is available – they can be repaired.

Wound closure

A small, uncontaminated wound in a type I or II fracture may be sutured (after debridement), provided this can be done without tension. In more severe injuries, immediate fracture stabilization and wound cover using split-skin grafts, local or distant flaps are ideal, provided that both orthopaedic and plastic surgeons are satisfied with a clean, viable wound achieved after debridement. In the absence of this combined approach at the time of debridement, the fracture is temporarily stabilized and the wound left open and dressed with an impervious dressing (Figure 23.24). Adding vacuum dressings has been shown to help.

Return to surgery for a 'second look' should have definitive fracture fixation and wound coverage as an objective. It should be done within 48–72 hours, and not later than 5 days. Open fractures do not fare well if left exposed for long periods, and multiple returns to theatre for repeated debridement can be self-defeating.

Stabilizing the fracture

Stabilizing the fracture is important in reducing the likelihood of infection and assisting recovery of the soft tissues. The method of fixation selected depends on the degree of contamination, time from injury to operation and amount of soft-tissue damage. If there is no obvious contamination and definitive wound cover can be achieved at the time of debridement, open fractures of all types can be treated as for a closed injury; internal or external fixation may be appropriate depending on the individual characteristics of the fracture and wound (Figure 23.25). This ideal scenario of judicious soft-tissue and bone debridement, wound cleansing, immediate stabilization and cover is only possible if surgeons with orthopaedic and plastic surgical expertise are present at the time of initial surgery.

If wound cover is delayed, external fixation can be used as a temporary measure; however, the surgeon must take care to insert the fixator pins away from potential flaps needed by the plastic surgeon.

The external fixator may be exchanged for internal fixation at the time of definitive wound cover as long as: (1) the delay to wound cover is less than 7 days; (2) wound contamination is not visible; and (3) internal fixation can control the fracture as well as the external fixator. This approach is less risky than introducing internal fixation at the time of initial surgery

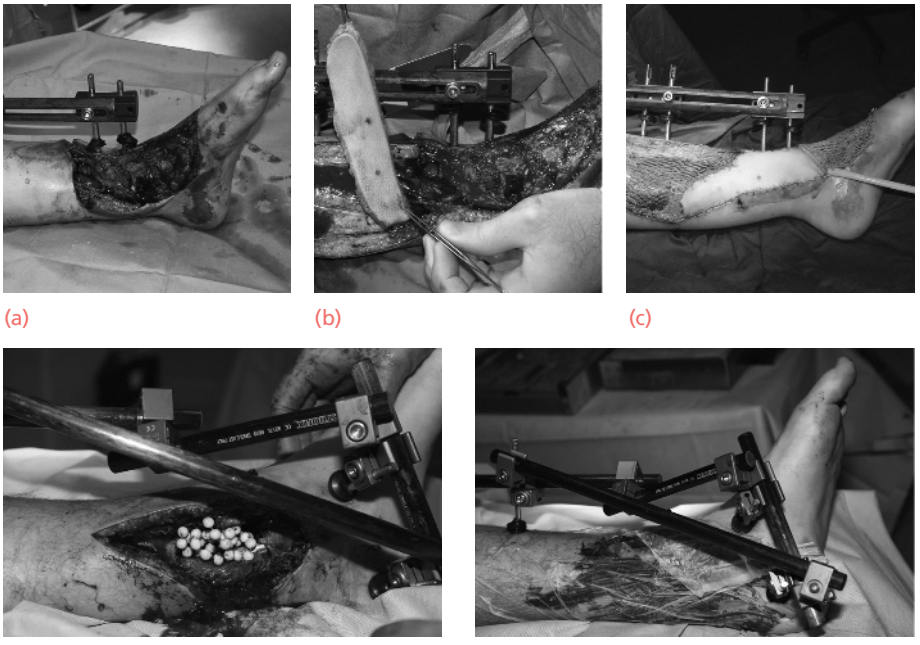
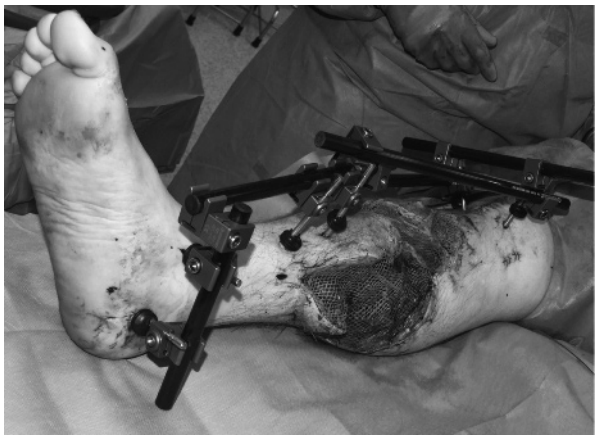


Figure 23.24 Covering the fracture The best fracture cover is skin or muscle – with the help of a plastic surgeon (a–c). If none is available, gentamicin beads can be inserted and sealed with an impervious dressing until the second operation, where a further debridement and, ideally, definitive fracture cover is obtained (d,e).



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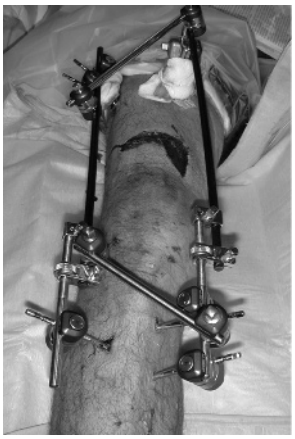


(b)

Figure 23.25 Stabilizing the limb in open fractures Spanning external fixation is a useful method of holding the fracture in the first instance (a,b). When definitive fracture cover is carried out, this can be substituted with internal fixation, provided the wound is clean and the interval between the two procedures is less than 7 days.

and leaving both metalwork and bone exposed until definitive cover several days later.

Aftercare

In the ward, the limb is elevated and its circulation carefully watched. Wound cultures are seldom helpful as osteomyelitis, if it were to ensue, is often caused by hospital-derived organisms; this emphasizes the need for meticulous debridement and early appropriate wound closure or coverage.

SEQUELS TO OPEN FRACTURES

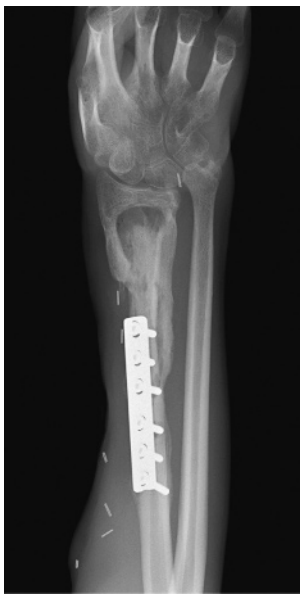
Skin

If split-thickness skin grafts are used inappropriately (e.g. to cover tendons or bone), particularly where flap cover is more suited, there can be areas of contracture or friable skin that breaks down intermittently. Reparative or reconstructive surgery by a plastic surgeon is desirable.

Bone

Infection involves the bone and any implants that may have been used. Early infection may present as wound inflammation with or without discharge. Identifying the causal organism without tissue samples is difficult but, at best guess, it is likely to be *S. aureus* (including methicillin-resistant varieties) or *Pseudomonas*. Suppression by appropriate antibiotics, as long as the fixation remains stable, may allow the fracture to proceed to union, but further surgery is likely to be needed later, when the antibiotics are stopped.

Late occurrence of infection may be with a sinus and X-ray evidence of sequestra (Figure 23.26). The implants and all avascular pieces of bone should



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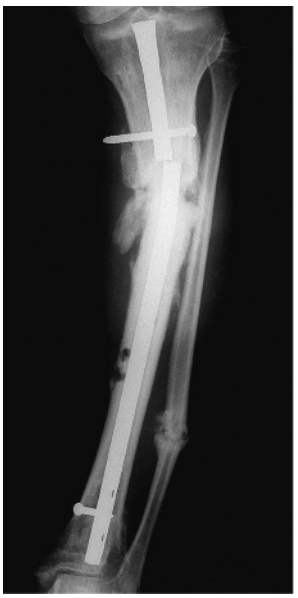

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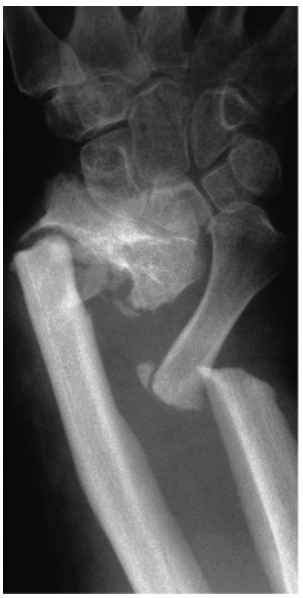

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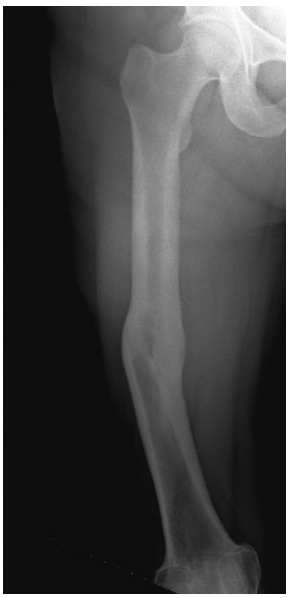


(d)

Figure 23.26 Complications of fractures Fractures can become infected (a,b), fail to unite (delayed union, non-union) (c) or unite in poor alignment (malunion) (d).

be removed; robust soft-tissue cover (ideally a flap) is needed. An external fixator can be used to bridge the fracture. If the resulting defect is too large for bone grafting at a later stage, the patient should be referred to a centre with the necessary experience and facilities for bone transport/limb reconstruction.

Joints

When an infected fracture communicates with a joint, the principles of treatment are the same as with bone infection, namely debridement and drainage, drugs and splintage. On resolution of the infection, attention can be given to stabilizing the fracture so that joint movement can recommence. Permanent stiffness is a real threat; where fracture stabilization cannot be achieved to allow movement, the joint should be splinted in the optimum position for ankylosis.

GUNSHOT INJURIES

Missile wounds are looked upon as a special type of open injury. Tissue damage is produced by: (1) direct injury in the immediate path of the missile; (2) contusion of muscles around the missile track; and (3) bruising and congestion of soft tissues at a greater distance from the primary track. The exit wound (if any) is usually larger than the entry one.

With high-velocity missiles (bullets, usually from rifles, travelling at speeds above 600 m/s) there is marked cavitation and tissue destruction over a wide area. The splintering of bone, resulting from the transfer of large quantities of energy, creates secondary missiles, causing greater damage. With low-velocity missiles (bullets from civilian hand-guns travelling at speeds of 300–600 m/s) cavitation is much less, and with smaller weapons tissue damage may be virtually confined to the bullet track. However, with all gunshot injuries debris is sucked into the wound, which is therefore contaminated from the outset.

Emergency treatment

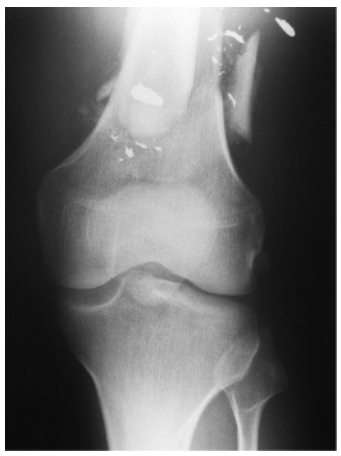
As always, the arrest of bleeding and general resuscitation take priority. Each wound should be covered with a sterile dressing and the area examined for artery or nerve damage. Antibiotics should be given immediately, following the recommendations for open fractures (see Table 23.1).

Definitive treatment

Traditionally, all missile injuries were treated as severe open injuries, by exploration of the missile track and formal debridement. However, it has been shown that low-velocity wounds with relatively clean entry and exit wounds can be treated as Gustilo type I injuries, by superficial debridement, splintage of the limb and antibiotic cover; the fracture is then treated in the same way as similar open fractures. If the injury involves soft tissues only, with minimal bone splinters, the wound may be safely treated without surgery but with local wound care and antibiotics.

High-velocity injuries demand thorough cleansing of the wound and debridement, with excision of deep damaged tissues and, if necessary, splitting of fascial compartments to prevent ischaemia; the fracture is stabilized and the wound is treated as for a Gustilo type III fracture. If there are comminuted fractures, these are best managed by external fixation. The method of wound closure will depend on the state of tissues after several days; in some cases delayed primary suture is possible but, as with other open injuries, close collaboration between plastic and orthopaedic surgeons is needed.

Close-range shotgun injuries, although the missiles may be technically low velocity, are treated as high-velocity wounds because the mass of shot transfers large quantities of energy to the tissues (Figure 23.27).



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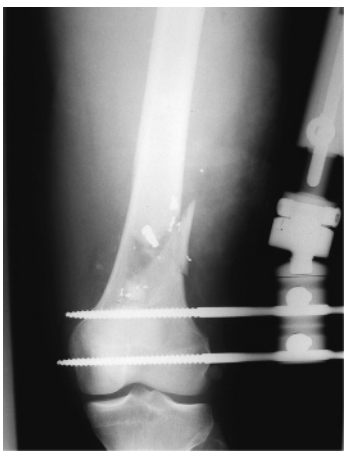

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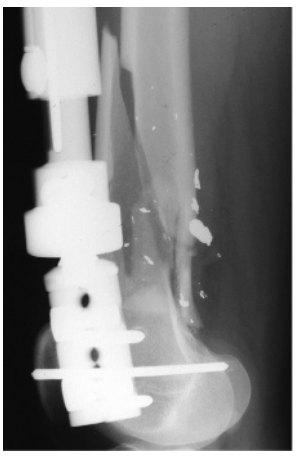


(c)

Figure 23.27 Gunshot injuries (a) Close range shotgun blasts, although technically low velocity, transfer large quantities of destructive force to the tissues due to the mass of shot. They should be treated like high-energy open fractures (b,c).

COMPLICATIONS OF FRACTURES

The general complications of fractures (blood loss, shock, fat embolism, cardiorespiratory failure, etc.) are dealt with in Chapter 22.

Local complications can be divided into *early* (arising during the first few weeks following injury) and *late* (Table 23.2).

EARLY COMPLICATIONS

Early complications may present as part of the primary injury or may appear only after a few days or weeks.

VISCERAL INJURY

Fractures around the trunk are often complicated by injuries to underlying viscera, the most important being penetration of the lung with life-threatening pneumothorax following rib fractures and rupture of the bladder or urethra in pelvic fractures. These injuries require early recognition and emergency treatment.

Table 23.2 Local complications of fractures

Early		Late
Urgent	**Less urgent**	
Visceral injury	Fracture blisters	Delayed union
Vascular injury	Plaster and pressure sores	Non-union
Nerve injury	Heterotopic ossification	Malunion
Compartment syndrome		Avascular necrosis
Haemarthrosis	Ligament injury	Growth disturbance
Infection	Tendon lesions	Bed sores
Gas gangrene	Nerve compression	Muscle contracture
	Joint stiffness	Joint instability
	Complex regional pain syndrome (algodystrophy)	Osteoarthritis

VASCULAR INJURY

The fractures most often associated with damage to a major artery are those around the knee and elbow, and those of the humeral and femoral shafts (Table 23.3). The artery may be cut, torn, compressed or contused, either by the initial injury or subsequently by swelling, bone fragments, reduction manoeuvres or surgery. Even if its outward appearance is normal, the intima may be detached and the vessel blocked by thrombus, or a segment of artery may be in spasm. The effects vary from transient diminution of blood flow to profound ischaemia, tissue death and peripheral gangrene (Figure 23.28).

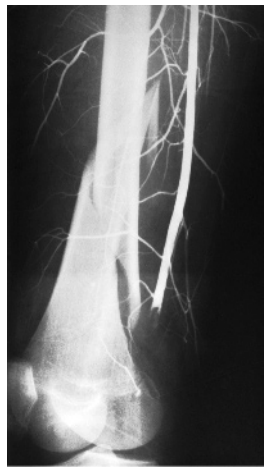
Clinical features

The patient may complain of paraesthesia or numbness in the toes or the fingers. The injured limb is cold and pale, or slightly cyanosed, and the pulse is weak or absent. X-rays will probably show one of the 'high-risk' fractures listed in Table 23.3. If a vascular injury is suspected, an angiogram or duplex should be performed immediately; if it is positive, emergency treatment must be started without further delay.

Table 23.3 Common vascular injuries

Injury	Vessel
First rib fracture	Subclavian
Shoulder dislocation	Axillary
Humeral supracondylar fracture	Brachial
Elbow dislocation	Brachial
Pelvic fracture	Presacral and internal iliac
Femoral supracondylar fracture	Femoral
Knee dislocation	Popliteal
Proximal tibial	Popliteal or its branches

(a)



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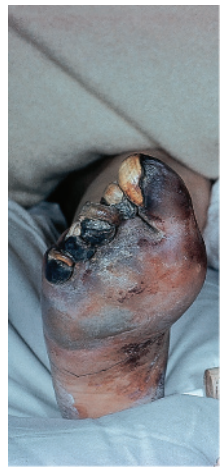


(c)

Figure 23.28 Vascular injury This patient was brought into hospital with a fractured femur and early signs of vascular insufficiency. The plain X-ray (a) looked as if the proximal bone fragment might have speared the popliteal artery. The angiogram (b) confirmed these fears. Despite vein grafting the patient developed peripheral gangrene (c).

Treatment

All bandages and splints should be removed. The fracture is re-X-rayed and, if the position of the bones suggests that the artery is being compressed or kinked, prompt reduction is necessary. The circulation is then reassessed repeatedly over the next half hour. If there is no improvement, the vessels must be explored by operation – preferably with the benefit of preoperative or perioperative angiography. A cut vessel can be sutured, or a segment may be replaced by a vein graft; if it is thrombosed, endarterectomy may restore the blood flow. If vessel repair is undertaken, stable fixation is a must and where it is practicable, the fracture should be fixed internally.

Nerve injury

Nerve injury is particularly common with fractures of the humeral shaft or injuries around the elbow or the knee (Table 23.4; see also Chapter 11). The telltale signs should be looked for (*and documented*) during the initial examination and again after reduction of the fracture.

Closed nerve injuries

In closed injuries the nerve is seldom severed, and spontaneous recovery should be awaited – it occurs in 90% of cases within 4 months. If recovery has not occurred within the expected time, and if nerve conduction studies and EMG fail to show evidence of recovery, the nerve should be explored.

Open nerve injuries

With open fractures the nerve injury is more likely to be complete. In these cases the nerve should be explored at the time of debridement and repaired at the time of debridement or at wound closure.

Acute nerve compression

Nerve compression, as distinct from a direct injury, sometimes occurs with fractures or dislocations around the wrist. Complaints of numbness or paraesthesia in the distribution of the median or ulnar nerves should be taken seriously and the patient monitored closely; if there is no improvement within 48 hours of fracture reduction or splitting of bandages around the splint, the nerve should be explored and decompressed.

Table 23.4 Common nerve injuries

Injury	Nerve
Shoulder dislocation	Axillary
Humeral shaft fracture	Radial
Humeral supracondylar fracture	Radial or median
Elbow medial condyle	Ulnar
Monteggia fracture-dislocation	Posterior interosseous
Hip dislocation	Sciatic
Knee dislocation	Peroneal

BOX 23.1 INDICATIONS FOR EARLY EXPLORATION

- Nerve injury associated with open fracture
- Nerve injury with fractures that need internal fixation
- Presence of a concomitant vascular injury
- Nerve damage diagnosed after manipulation of the fracture

Compartment syndrome

Fractures of the arm or leg can give rise to severe ischaemia, even if there is no damage to a major vessel. Bleeding, oedema or inflammation (infection) may increase the pressure within one of the osseofascial compartments; there is reduced capillary flow, which results in muscle ischaemia, further oedema, greater pressure and yet more profound ischaemia – a vicious circle that ends, after 6 hours or less, in necrosis of nerve and muscle within the compartment. Nerve is capable of regeneration but muscle, once infarcted, can never recover and is replaced by inelastic fibrous tissue (*Volkmann's ischaemic contracture*). A similar cascade of events may be caused by swelling of a limb inside a tight plaster cast (Figure 23.29).

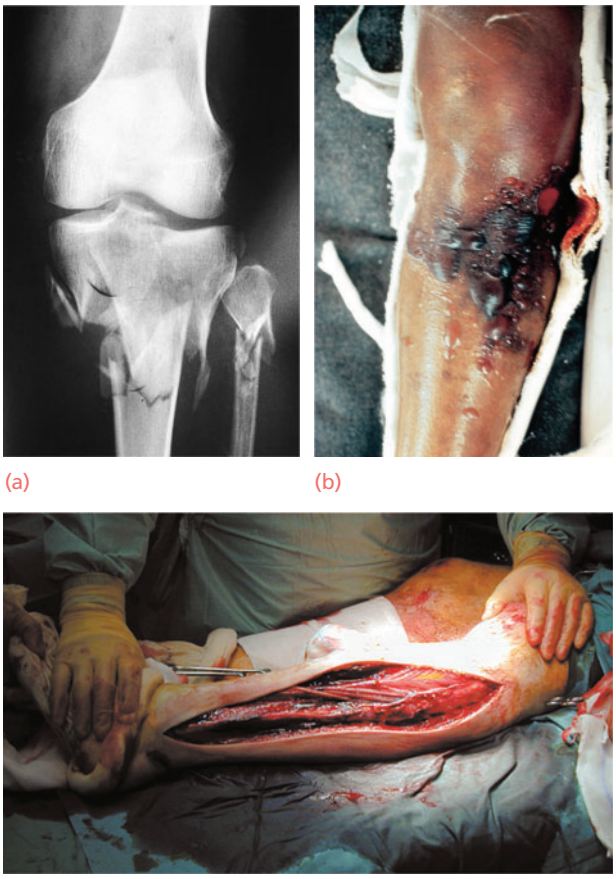
Clinical features

High-risk injuries are fractures of the elbow, forearm bones, proximal third of the tibia, and also multiple fractures of the hand or foot, crush injuries and circumferential burns. Other precipitating factors are operation (usually for internal fixation) or infection.

The classic features of ischaemia are the five Ps:

- Pain
- Paraesthesia
- Pallor
- Paralysis
- Pulselessness.

However, in compartment syndrome the ischaemia occurs at the capillary level, so pulses may still be felt and the skin may not be pale! The earliest of the 'classic' features is severe pain (or a 'bursting' sensation) and this may be the only feature seen. Altered sensibility and paresis (or more usually, weakness in active muscle contraction) may also occur. Skin sensation should be carefully and repeatedly checked.



(a) (b)

(c)

Figure 23.29 Compartment syndrome (a) A fracture at this level is always dangerous. This man was treated in plaster. Pain became intense and when the plaster was split (which should have been done immediately after its application), the leg was swollen and blistered (b). Tibial compartment decompression (c) requires fasciotomies of *all* the compartments in the leg.

Ischaemic muscle is highly sensitive to stretch. If the limb is unduly painful, swollen or tense, the muscles (which may be tender) should be tested by stretching them. When the toes or fingers are passively hyperextended, there is increased pain in the calf or forearm.

Confirmation of the diagnosis can be made by measuring the intracompartmental pressures. The need for early diagnosis is so important that some surgeons advocate the use of continuous compartment pressure monitoring for high-risk injuries (e.g. fractures of the tibia and fibula) and especially for forearm or leg fractures in patients who are unconscious. A split catheter is introduced into the compartment and the pressure is measured close to the level of the fracture. A differential pressure (ΔP) – the difference between diastolic pressure and compartment pressure – of less than 30 mmHg (4.00 kilopascals) is an indication for immediate compartment decompression.

Treatment

The threatened compartment (or compartments) must be promptly decompressed. Casts, bandages and dressings must be completely removed – merely splitting the plaster is utterly useless – and the limb should be nursed flat (elevating the limb causes a further decrease in end capillary pressure and aggravates the muscle ischaemia). The ΔP should be carefully monitored; if it falls below 30 mmHg, immediate open fasciotomy is performed. Compartment syndrome is, however, a clinical diagnosis and, if the surgeon believes there is a compartment syndrome present, fasciotomy is justified even if a predetermined pressure threshold has not been reached or the facility to measure pressure is not immediately available. If the clinical signs are 'soft', the limb should be examined at 30-minute intervals and, if there is no improvement within 2 hours of splitting the dressings, fasciotomy should be performed. Muscle will be dead after 4–6 hours of total ischaemia – there is no time to lose!

In the case of the leg, 'fasciotomy' means opening all four compartments through medial and lateral incisions (see Figure 23.22). The wounds should be left open and inspected 2 days later: if there is muscle necrosis, debridement can be carried out; if the tissues are healthy, the wounds can be sutured (without tension) or skin-grafted.

Haemarthrosis

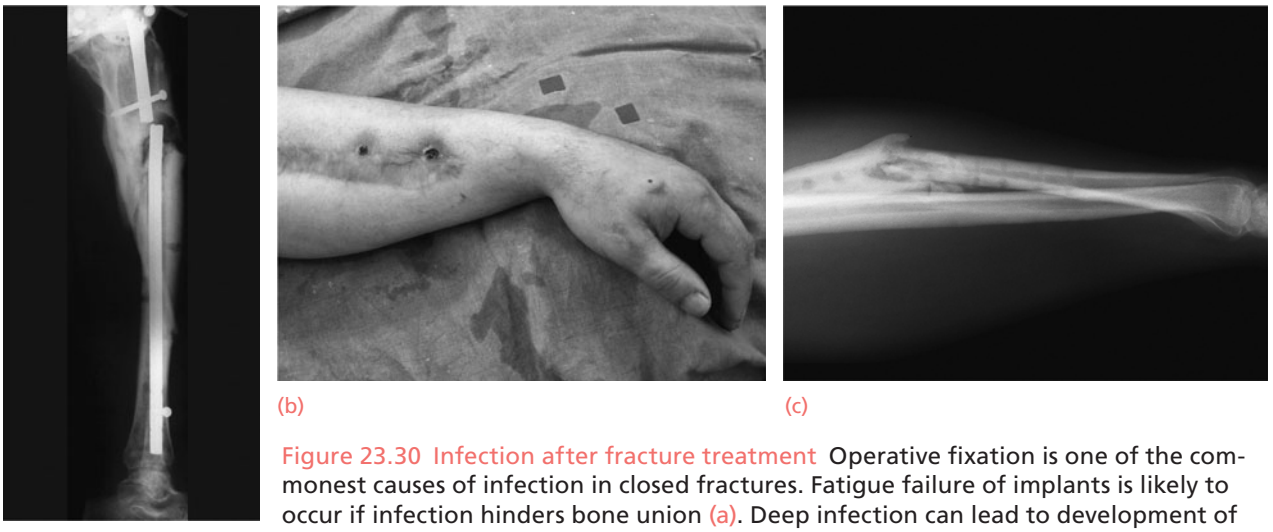
Fractures involving a joint may cause acute haemarthrosis. The joint is swollen and tense and the patient resists any attempt at moving it. The blood should be aspirated before dealing with the fracture.

Infection

Open fractures may become infected; closed fractures hardly ever do unless they are opened by operation. Post-traumatic wound infection is now the most common cause of chronic osteitis (Figure 23.30). The management of early and late infection is summarized in 'Sequels to open fractures' above.

Gas gangrene

This terrifying condition is produced by clostridial infection (especially *Clostridium welchii*). These are anaerobic organisms that can survive and multiply only in tissues with low oxygen tension; the prime site for infection, therefore, is a dirty wound with dead muscle that has been closed without adequate debridement. Toxins produced by the organisms destroy the cell wall and rapidly lead to tissue necrosis, thus promoting the spread of the disease.

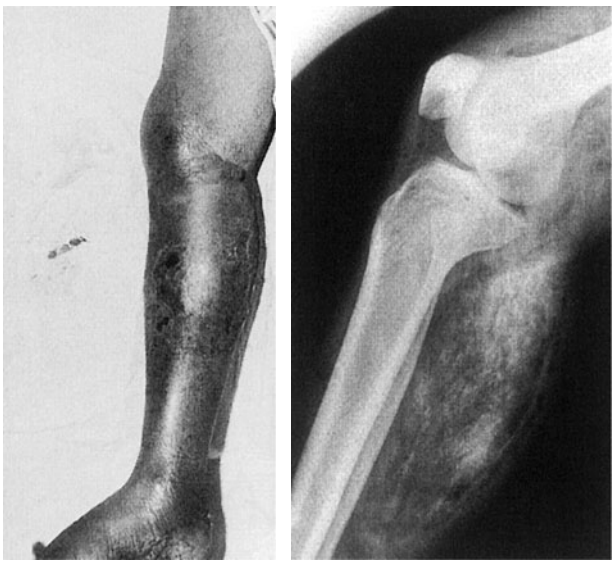


(a) (b) (c)

Figure 23.30 Infection after fracture treatment Operative fixation is one of the commonest causes of infection in closed fractures. Fatigue failure of implants is likely to occur if infection hinders bone union (a). Deep infection can lead to development of discharging sinuses (b,c).

Clinical features appear within 24 hours of the injury: the patient complains of intense pain and swelling around the wound (Figure 23.31), and a brownish discharge may be seen; gas formation is usually not very marked. There is little or no pyrexia but the pulse rate is increased and a characteristic smell becomes evident (once experienced this is never forgotten). Rapidly the patient becomes toxaemic and may lapse into coma and death.

It is essential to distinguish gas gangrene, which is characterized by myonecrosis, from anaerobic cellulitis, in which superficial gas formation is abundant but toxaemia usually slight. Failure to recognize the difference may lead to unnecessary amputation for the non-lethal cellulitis.



(a) (b)

Figure 23.31 Gas gangrene (a) Clinical picture of gas gangrene. (b) X-rays show diffuse gas in the muscles of the calf.

Prevention

Deep, penetrating wounds in muscular tissue are dangerous; they should be explored, all dead tissue should be completely excised and, if there is the slightest doubt about tissue viability, repetitive debridement should be performed. Unhappily, there is no effective antitoxin against *Clostridium perfringens (C. welchii)*.

Treatment

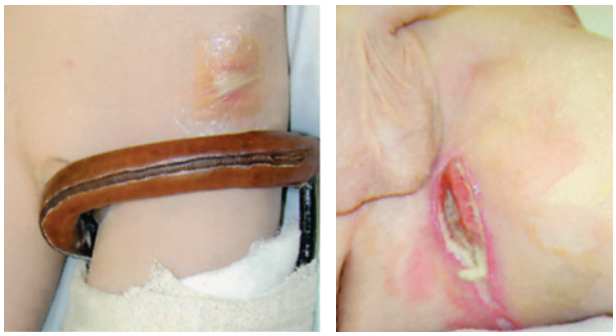
The key to life-saving treatment is early diagnosis. General measures, such as fluid replacement and intravenous antibiotics, are started immediately. Hyperbaric oxygen has been used as a means of limiting the spread of gangrene. However, the mainstay of treatment is prompt decompression of the wound and removal of all dead tissue. In advanced cases, amputation may be essential.

Fracture blisters

Two distinct blister types are sometimes seen after fractures: clear fluid-filled vesicles and blood-stained ones. Both occur during limb swelling and are due to elevation of the epidermal layer of skin from the dermis. There is no advantage to puncturing the blisters (it may even lead to increased local infection) and surgical incisions through blisters, although generally safe, should be undertaken only when limb swelling has decreased.

Plaster and pressure sores

Plaster sores occur where skin presses directly onto bone. They should be prevented by padding the bony points and by moulding the wet plaster so that pressure is distributed to the soft tissues around the bony points. While a plaster sore is developing, the patient



(a) (b)

Figure 23.32 Pressure sores Pressure sores are a sign of carelessness. (a,b) Sores from poorly supervised treatment in a Thomas splint.

feels localized burning pain. A window must immediately be cut in the plaster, or warning pain quickly abates and skin necrosis proceeds unnoticed.

Even traction on a Thomas splint requires skill in nursing care; careless selection of ring size, excessive fixed (as opposed to balanced) traction, and neglect can lead to pressure sores around the groin and iliac crest (Figure 23.32).

LATE COMPLICATIONS

Delayed union

If the time before union is unduly prolonged, the term 'delayed union' is used.

Causes

Factors causing delayed union can be summarized as: *biological, biomechanical* or *patient-related*.

BIOLOGICAL

Inadequate blood supply A poorly reduced fracture of a long bone will cause tearing of both the periosteum and interruption of the intramedullary blood supply. The fracture edges will become necrotic and dependent on the formation of an ensheathing callus mass to bridge the break. If the zone of necrosis is extensive, as might occur in highly comminuted fractures, union may be hampered.

Severe soft tissue damage Severe damage to the soft tissues affects fracture healing by: (1) reducing the effectiveness of muscle splintage; (2) damaging the local blood supply; and (3) diminishing or eliminating the osteogenic input from mesenchymal stem cells within muscle.

Periosteal stripping Overenthusiastic stripping of periosteum during internal fixation is an avoidable cause of delayed union.

BIOMECHANICAL

Imperfect splintage Excessive traction (creating a fracture gap) or excessive movement at the fracture site will delay ossification in the callus. In the forearm and leg a single-bone fracture may be held apart by an intact fellow bone.

Over-rigid fixation Contrary to popular belief, completely rigid fixation delays rather than promotes fracture union. It is only because the fixation device holds the fragments so securely that the fracture seems to be 'uniting'. Union by primary bone healing is slow, but, provided stability is maintained throughout, it does eventually occur.

Infection Both biology and stability are hampered by active infection: not only is there bone lysis, necrosis and pus formation, but implants which are used to hold the fracture tend to loosen.

PATIENT-RELATED

In a less than ideal world, there are patients who are:

- Immense
- Immoderate
- Immovable
- Impossible!

These factors must be accommodated in an appropriate fashion.

Clinical features

Fracture tenderness persists and, if the bone is subjected to stress, pain may be acute.

On X-ray, the fracture line remains visible and there is very little or incomplete callus formation or periosteal reaction. However, the bone ends are not sclerosed or atrophic. The appearances suggest that, although the fracture has not united, it eventually will.

Treatment

CONSERVATIVE TREATMENT

The two important principles are: (1) to eliminate any possible cause of delayed union; and (2) to promote healing by providing the most appropriate environment. Immobilization should be sufficient to prevent shear at the fracture site, but fracture loading is an important stimulus to union and can be enhanced by: (1) encouraging muscular exercise; and (2) at least partial weight-bearing in the cast or brace. The watchword is patience; however, at a certain time point prolonged immobilization outweighs the advantages of non-operative treatment.

OPERATIVE TREATMENT

Each case should be treated on its merits; however, if union is delayed for more than 6 months and there

is no sign of callus formation, internal fixation and bone grafting are indicated. The operation should be planned in such a way as to cause the least possible damage to the soft tissues, therefore optimizing the biology.

Non-union

In a minority of cases delayed union gradually turns into 'non-union' – it becomes apparent that the fracture will never unite without intervention. Movement can be elicited at the fracture site and pain diminishes.

X-rays The fracture is clearly visible but the bone on either side of it may show either exuberant callus that is failing to bridge or atrophy. This contrasting appearance has led to non-union being divided into hypertrophic and atrophic types (Figure 23.33). In *hypertrophic non-union* the bone ends are enlarged, suggesting that osteogenesis is still active but not quite capable of bridging the gap. In *atrophic non-union*, osteogenesis seems to have ceased. The bone ends are tapered or rounded with no indication of new bone formation.

Causes

When dealing with the problem of non-union, four questions must be addressed. They have given rise to the acronym *CASS*:

1 *Contact* – Was there sufficient contact between the bone fragments?
2 *Alignment* – Was the fracture adequately aligned to reduce shear?
3 *Stability* – Was the fracture held with sufficient stability?
4 *Stimulation* – Was the fracture sufficiently 'stimulated'?

There are, of course, also biological and patient-related reasons that may lead to non-union: (1) poor soft tissues (from either the injury or surgery); (2) local infection; (3) associated drug abuse, anti-inflammatory or cytotoxic immunosuppressant medication; and (4) non-compliance on the part of the patient.

Treatment

CONSERVATIVE TREATMENT

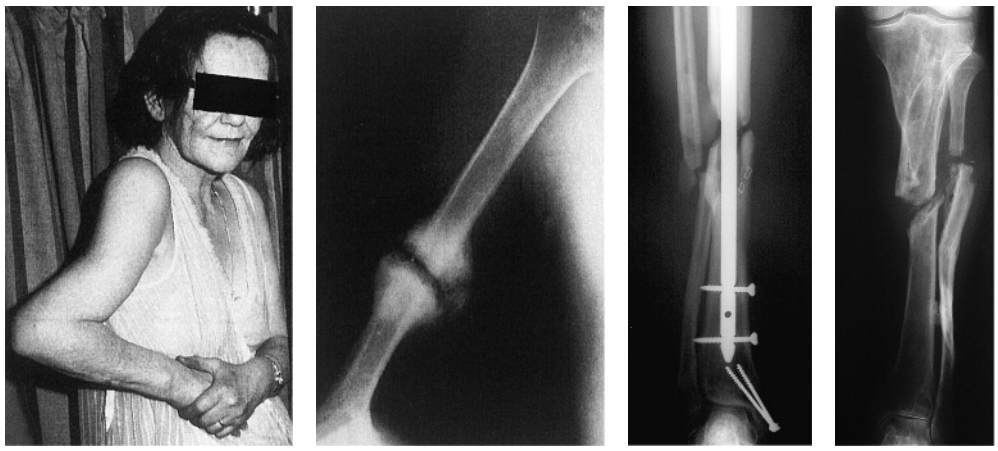
Non-union is occasionally symptomless, needing no treatment or, at most, a removable splint. Even if symptoms are present, operation is not the only answer; with hypertrophic non-union, functional bracing may be sufficient to induce union, but splintage often needs to be prolonged. Pulsed electromagnetic fields and low-frequency, pulsed ultrasound can also be used to stimulate bone union.

OPERATIVE TREATMENT

With hypertrophic non-union and in the absence of deformity, very rigid fixation alone (internal or external) may lead to union. With atrophic non-union, fixation alone is not enough (Figure 23.34). Fibrous tissue in the fracture gap as well as the hard, sclerotic bone ends are excised and bone grafts are packed around the fracture. If there is significant 'die-back', this will require more extensive excision and the gap is then dealt with by bone advancement methods such as the Ilizarov technique (Figure 23.35).

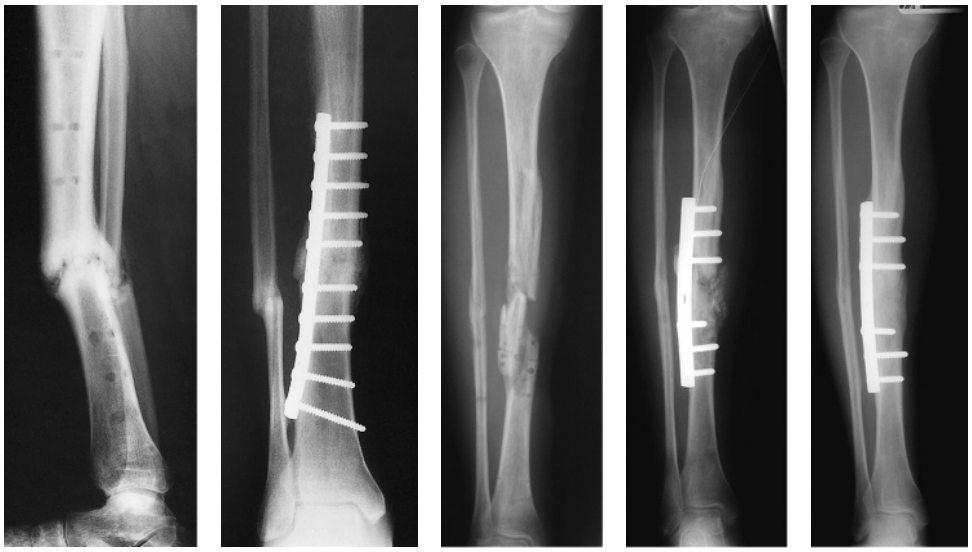
Malunion

When the fragments join in an unsatisfactory position (unacceptable angulation, rotation or shortening) the fracture is defined as 'malunited'. Causes are failure to reduce a fracture adequately, failure to hold reduction while healing proceeds, or gradual collapse of comminuted or osteoporotic bone.



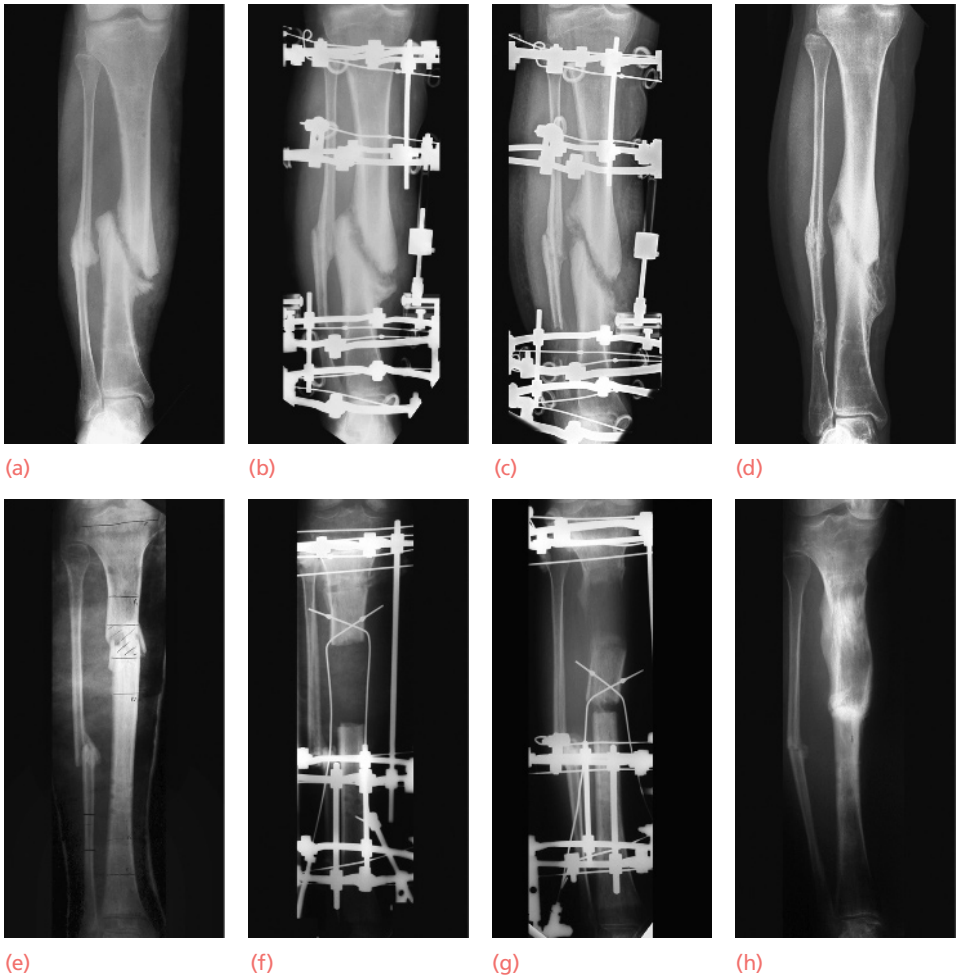
(a) (b) (c) (d)

Figure 23.33 Non-union (a) This patient has an obvious pseudarthrosis of the humerus. The X-ray (b) shows a typical hypertrophic non-union. (c,d) Examples of atrophic non-union.



(a) (b) (c) (d) (e)

Figure 23.34 Non-union – treatment (a) This patient with fractures of the tibia and fibula was initially treated by internal fixation with a plate and screws. The fracture failed to heal, and developed the typical features of hypertrophic non-union. (b) After a further operation, using more rigid fixation (and no bone grafts), the fractures healed solidly. (c,d) This patient with atrophic non-union needed both internal fixation and bone grafts to stimulate bone formation and union (e).



(a) (b) (c) (d)

(e) (f) (g) (h)

Figure 23.35 Non-union – treatment by the Ilizarov technique Hypertrophic non-unions can be treated by gradual distraction and realignment in an external fixator (a–d). Atrophic non-unions will need extended surgery; the necrotic tissue is excised (e,f) and replaced through bone transport (g,h).

Clinical features

The deformity is usually obvious, but sometimes the true extent of malunion is apparent only on X-ray. Rotational deformity of the femur, tibia, humerus or forearm may be missed unless the limb is clinically compared with the contralateral side. Rotational deformity of a metacarpal fracture is detected by asking the patient to flatten the fingers onto the palm and seeing whether the normal regular fan-shaped appearance is reproduced (Chapter 26).

X-rays are essential to check the position of the fracture while it is uniting. This is particularly important during the first 2–3 weeks when the situation may change without warning. At this stage it is sometimes difficult to decide what constitutes 'malunion'; acceptable norms differ and these are discussed under the individual fractures.

Treatment

Incipient malunion may call for treatment even before the fracture has fully united; the decision on the need for re-manipulation or correction may be extremely difficult. A few guidelines are offered:

1 In adults, fractures should be reduced as near to the anatomical position as possible. Angulation of more than 10–15 degrees in a long bone or a noticeable rotational deformity may need correction by remanipulation, or by osteotomy and fixation (Figure 23.36). Deformity in the plane of motion of a nearby joint may be better tolerated than deformity in a plane in which the nearby joint does not move.
2 In children, angular deformities near the bone ends (and especially if the deformity is in the same plane as that of movement of the nearby joint) will usually remodel with time; rotational deformities will not.
3 In the lower limb, shortening of more than 2 cm is seldom acceptable to the patient and a limb length equalizing procedure may be indicated.
4 The patient's expectations (often prompted by cosmesis) may be quite different from the surgeon's; they are not to be ignored.
5 Early discussion with the patient, a guided view of the X-rays will help in deciding the need for treatment and may prevent later misunderstanding of what degree of deformity is considered acceptable.
6 Very little is known about the long-term effects of small angular deformities on joint function. However, it seems likely that malalignment of more than 15 degrees in any plane may cause asymmetrical loading of the joint above or below and late development of secondary osteoarthritis; this applies particularly to the large weight-bearing joints.

Avascular necrosis

Certain regions are notorious for their propensity to develop ischaemia and bone necrosis after injury (see also Chapter 6). They are: (1) the head of the femur (after fracture of the femoral neck or dislocation of the hip) (Figure 23.37); (2) the proximal part of the scaphoid (after fracture through its waist);

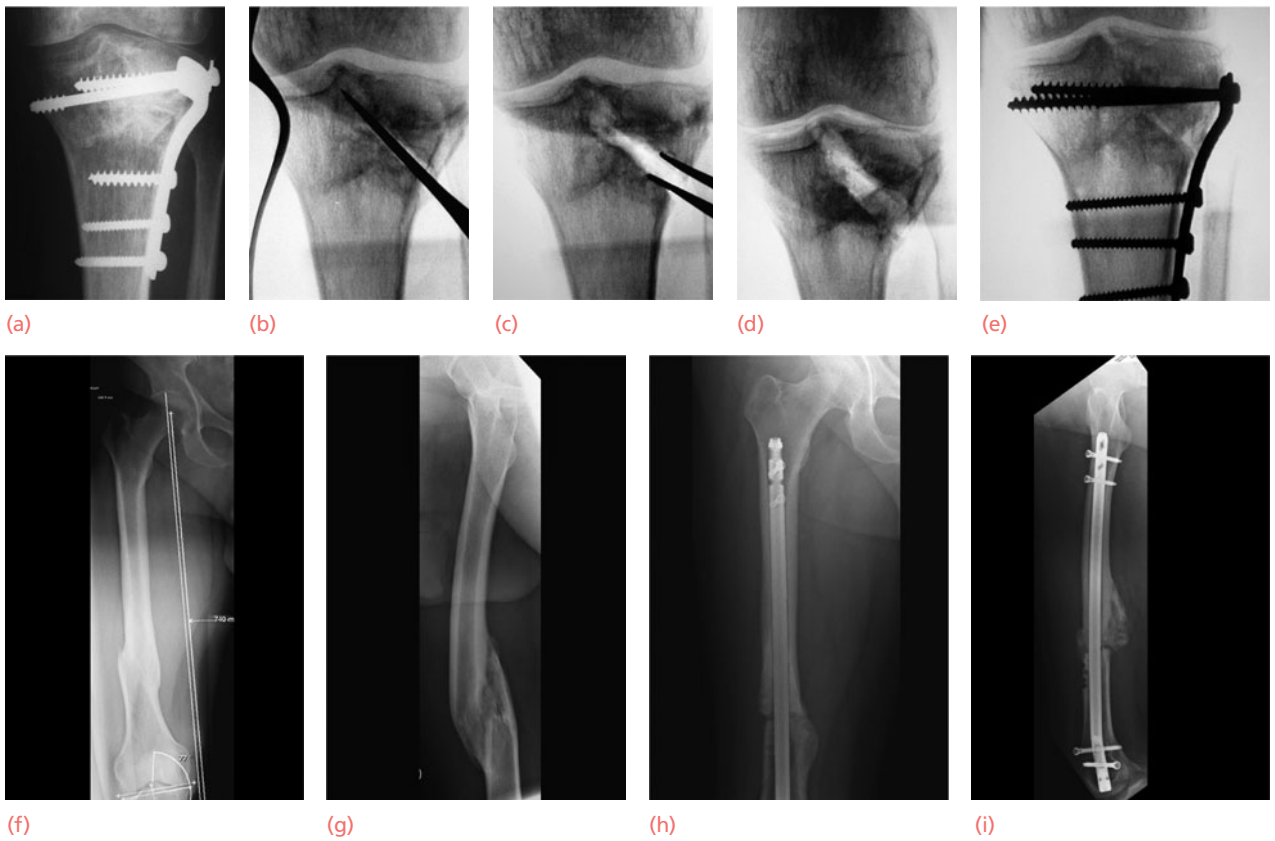
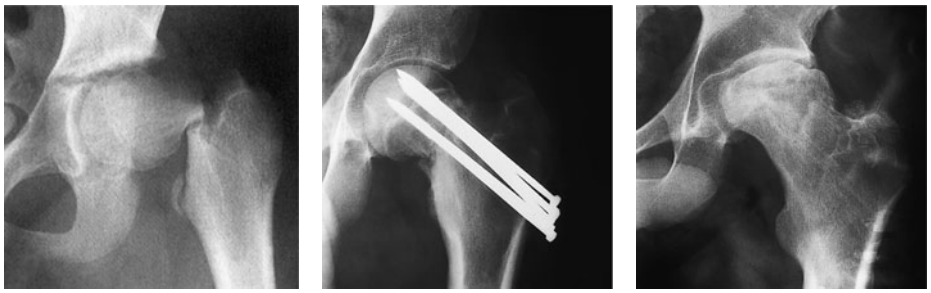


Figure 23.36 Malunion – treatment by internal fixation An osteotomy, correction of deformity and internal fixation can be used to treat both intra-articular deformities (a–e) and those in the shaft of a long bone (f–i).



(a) (b) (c)

Figure 23.37 Avascular necrosis (a) Displaced fractures of the femoral neck are at considerable risk of developing avascular necrosis. Despite internal fixation within a few hours of the injury (b), the head-fragment developed avascular necrosis. (c) X-ray after removal of the fixation screws.

(3) the lunate (following dislocation); and (4) the body of the talus (after fracture of its neck).

Accurately speaking, this is an early complication of bone injury, because ischaemia occurs during the first few hours following fracture or dislocation. However, the clinical and radiological effects are not seen until weeks or even months later.

Clinical features

There are no symptoms associated with avascular necrosis but, if the fracture fails to unite or if the bone collapses, the patient may complain of pain. X-rays show the characteristic increase in X-ray density, which occurs as a consequence of two factors: disuse osteoporosis in the surrounding parts gives the impression of 'increased density' in the necrotic segment, and collapse of trabeculae compacts the bone and increases its density. Where normal bone meets the necrotic segment, a zone of increased radiographic density may be produced by new bone formation.

Treatment

Treatment usually becomes necessary when joint function is threatened. In elderly people with necrosis of the femoral head, an arthroplasty is the obvious choice; in younger people, realignment osteotomy (or, in some cases, arthrodesis) may be wiser. Avascular necrosis in the scaphoid or talus may need no more than symptomatic treatment, but arthrodesis of the wrist or ankle is sometimes needed.

Growth disturbance

In children, damage to the physis may lead to abnormal or arrested growth. A transverse fracture through the growth plate is not always disastrous; the fracture runs through the hypertrophic and calcified layers and not through the germinal zone, so provided it is accurately reduced, there may not be any disturbance of growth. However, fractures that split the epiphysis inevitably traverse the growing portion of the physis, and so further growth may be asymmetrical and the bone end characteristically angulated; if the entire physis is damaged, there may be slowing or complete cessation of growth. The subject is dealt with in more detail in the section on 'Injuries of the physis' below.

Bed sores

Bed sores occur in elderly or paralysed patients (Figure 23.38). The skin over the sacrum and heels is especially vulnerable. Careful nursing and early activity can usually prevent bed sores; once they have developed, treatment is difficult – it may be necessary to excise the necrotic tissue and apply skin grafts. Vacuum-assisted closure (a form of negative pressure dressing) may be used for sacral bed sores.

Heterotopic ossification

Heterotopic ossification in the muscles sometimes occurs after an injury, particularly dislocation of the elbow or a blow to the brachialis, deltoid or quadriceps, and is accentuated in craniocerebral injury. It is thought

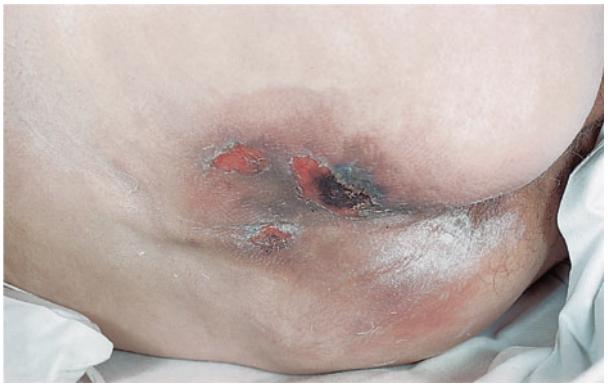


Figure 23.38 Bed sores Bed sores in an elderly patient that kept her in hospital for months.

to be due to muscle damage, but it may also occur without a local injury in unconscious or paraplegic patients.

Clinical features

Soon after the injury, the patient (usually a fit young man) complains of pain; there is local swelling and soft-tissue tenderness. X-rays are normal but a bone scan may show increased activity. Over the next 2–3 weeks the pain gradually subsides, but joint movement is limited; X-rays may show fluffy calcification in the soft tissues. By 8 weeks the bony mass is easily palpable and is clearly defined in the X-rays (Figure 23.39).

Treatment

The worst treatment is to attack an injured and slightly stiff elbow with vigorous muscle-stretching exercises; this is liable to precipitate or aggravate the condition. The joint should be rested in the position of function until pain subsides; gentle active movements are then begun.

Months later, when the condition has stabilized, it may be helpful to excise the bony mass. Indomethacin or radiotherapy should be given to help prevent a recurrence.

Tendon lesions

Tendinitis may affect the tibialis posterior tendon following medial malleolar fractures. The risk can be reduced by accurate reduction, if necessary at surgery. Rupture of the extensor pollicis longus tendon may occur 6–12 weeks after a fracture of the lower radius.

Direct suturing is seldom possible and the resulting disability is treated by transferring the extensor indicis proprius tendon to the distal stump of the ruptured thumb tendon. Late rupture of the long head of biceps after a fractured neck of humerus usually requires no treatment.

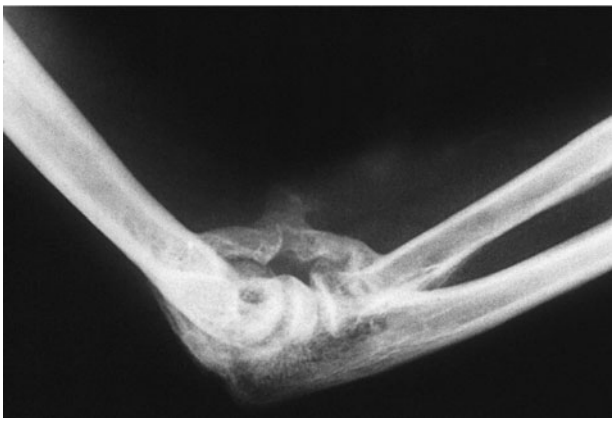


Figure 23.39 Heterotopic ossification This followed a fractured head of the radius.

Nerve compression

Nerve compression may damage the lateral popliteal nerve if an elderly or emaciated patient lies with the leg in full external rotation. Radial palsy may follow the faulty use of crutches. Both conditions are due to lack of supervision.

Bone or joint deformity may result in local nerve entrapment with typical features such as numbness or paraesthesia, loss of power and muscle wasting in the distribution of the affected nerve. Common sites are: (1) the ulnar nerve, due to a valgus elbow following a malunited lateral condyle or supracondylar fracture; (2) the median nerve, following injuries around the wrist; and (3) the posterior tibial nerve, following fractures around the ankle.

Treatment is by early decompression of the nerve; in the case of the ulnar nerve this may require anterior transposition.

Muscle contracture

Following arterial injury or compartment syndrome, the patient may develop ischaemic contractures of the affected muscles (*Volkmann's ischaemic contracture*). Nerves injured by ischaemia sometimes recover, at least partially; thus the patient presents with deformity and stiffness, but numbness is inconstant. The most commonly affected sites are the forearm and hand, leg and foot.

In a severe case affecting the forearm, there will be wasting of the forearm and hand, and clawing of the fingers (Figure 23.40). If the wrist is passively flexed, the patient can extend the fingers, showing that the deformity is largely due to contracture of the forearm muscles. Detachment of the flexors at their origin and along the interosseous membrane in the forearm may improve the deformity, but function is no better if sensation and active movement are not restored. A pedicle nerve graft, using the proximal segments of the median and ulnar nerves may restore protective sensation in the hand, and tendon transfers (wrist extensors to finger and thumb flexors) will allow active grasp. In less severe cases, median nerve sensibility may be quite good and, with appropriate tendon releases and transfers, the patient regains a considerable degree of function.

Ischaemia of the hand may follow forearm injuries, or swelling of the fingers associated with a tight forearm bandage or plaster. The intrinsic hand muscles fibrose and shorten, pulling the fingers into flexion at the metacarpophalangeal joints, but the interphalangeal joints remain straight. The thumb is adducted across the palm (Bunnell's 'intrinsic-plus' position).

Ischaemia of the calf muscles may follow injuries or operations involving the popliteal artery or its divisions. This is more common than

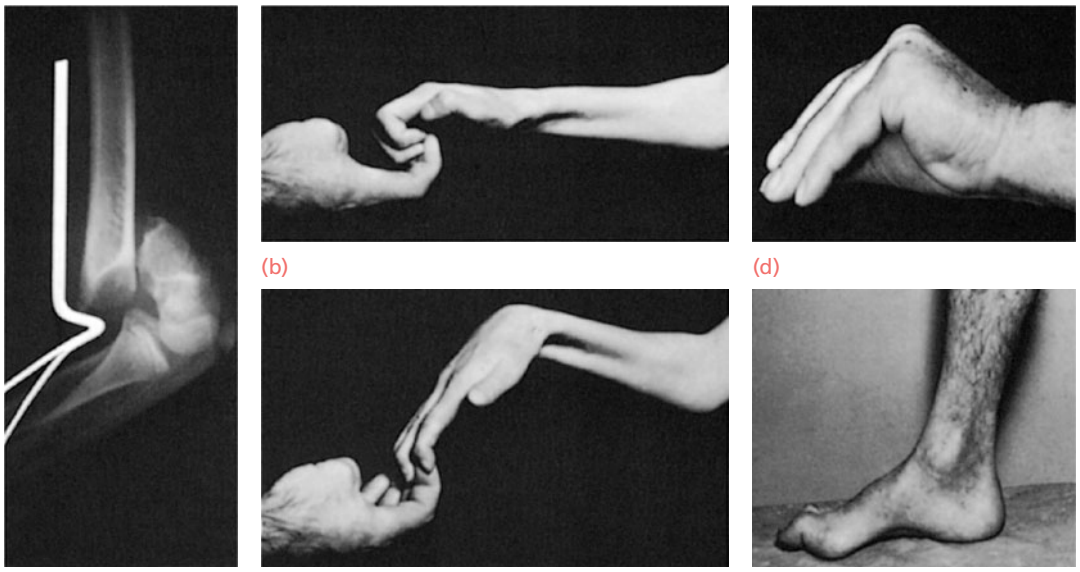


Figure 23.40 Volkmann's ischaemia (a) Kinking of the main artery is an important cause, but intimal tears may also lead to blockage from thrombosis. A delayed diagnosis of compartment syndrome carries the same sorry fate. (b,c) Volkmann's contracture of the forearm. The fingers can be straightened only when the wrist is flexed (the constant length phenomenon). (d) Ischaemic contracture of the small muscles of the hand. (e) Ischaemic contracture of the calf muscles with clawing of the toes.

usually supposed. The symptoms, signs and subsequent contracture are similar to those following ischaemia of the forearm. One of the causes of late claw-toe deformity is an undiagnosed compartment syndrome.

Joint instability

Following injury, a joint may give way. Causes include the following:

- *ligamentous laxity* – especially at the knee, ankle and metacarpophalangeal joint of the thumb
- *muscle weakness* – especially if splintage has been excessive or prolonged, and exercises have been inadequate – again the knee and ankle are most often affected
- *bone loss* – especially after a gunshot fracture or severe compound injury, or from crushing of metaphyseal bone in joint depression fractures.

Injury may also lead to *recurrent dislocation*. The commonest sites are: (1) the shoulder – if the glenoid labrum has been detached or the glenoid fractured (a Bankart lesion); and (2) the patella – if, after traumatic dislocation, the restraining patellofemoral ligament heals poorly.

A more subtle form of instability is seen after fractures around the wrist. Patients complaining of persistent discomfort or weakness after wrist injury should be fully investigated for *chronic carpal instability* (see Chapters 15 and 25).

Joint stiffness

Joint stiffness after a fracture commonly occurs in the knee, elbow, shoulder and (worst of all) small joints of the hand. Sometimes the joint itself has been injured; a haemarthrosis forms and leads to synovial adhesions. More often the stiffness is due to oedema and fibrosis of the capsule, ligaments and muscles around the joint, or adhesions of the soft tissues to each other or to the underlying bone. All these conditions are made worse by prolonged immobilization; moreover, if the joint has been held in a position where the ligaments are at their shortest, no amount of exercise will afterwards succeed in stretching these tissues and restoring the lost movement completely.

In a small percentage of patients with fractures of the forearm or leg, early post-traumatic swelling is accompanied by tenderness and progressive stiffness of the distal joints. These patients are at great risk of developing a *complex regional pain syndrome*; whether this is an entirely separate entity or merely an extension of the 'normal' post-traumatic soft-tissue reaction is uncertain. It is important to recognize this type of 'stiffness' when it occurs and to insist on skilled physiotherapy until normal function is restored.

Treatment

The best treatment is prevention – by exercises that keep the joints mobile from the outset. If a joint has to be splinted, make sure that it is held in the 'position of safe immobilization'. (see Chapter 16).

Joints that are already stiff take time to mobilize, but prolonged and patient physiotherapy can work wonders. If the situation is due to intra-articular adhesions, arthroscopic-guided releases may free the joint sufficiently to permit a more pliant response to further exercise. Occasionally, adherent or contracted tissues need to be released by operation (e.g. when knee flexion is prevented by adhesions in and around the quadriceps).

Complex regional pain syndrome

In 1900 Sudeck described a condition characterized by painful osteoporosis of the hand. The same condition sometimes occurs after fractures of the extremities and for many years it was called *Sudeck's atrophy*. It is now recognized that this advanced atrophic disorder is the late stage of a post-traumatic *reflex sympathetic dystrophy* (also known as *algodystrophy*), which is much more common than originally believed and that it may follow relatively trivial injury. Because of continuing uncertainty about its nature, the term *complex regional pain syndrome (CRPS)* has been introduced (see Chapter 10).

Two types of CRPS are recognized:

- *Type 1* – a reflex sympathetic dystrophy that develops after an injurious or noxious event
- *Type 2* – causalgia that develops after a nerve injury.

The patient complains of continuous pain, often described as 'burning' in character. At first there is local swelling, redness and warmth, as well as tenderness and moderate stiffness of the nearby joints. As the weeks go by, the skin becomes pale and atrophic, movements are increasingly restricted and the patient may develop fixed deformities. X-rays characteristically show patchy rarefaction of the bone (Figure 23.41).

The earlier the condition is recognized and treatment begun, the better the prognosis. Elevation and active exercises are important after all injuries, but in CRPS they are essential. In the early stage of the condition anti-inflammatory drugs and adequate analgesia are helpful. Involvement of a pain specialist who is familiar with desensitization methods, regional anaesthesia, and use of drugs such as amitriptyline, carbamazepine and gabapentin may help; this, combined with prolonged and dedicated physiotherapy, is the mainstay of treatment.

Osteoarthritis

A fracture involving a joint may severely damage the articular cartilage and give rise to post-traumatic osteoarthritis within a period of months. Even if the cartilage heals, irregularity of the joint surface may cause localized stress and so predispose to secondary osteoarthritis years later. If the step-off in the

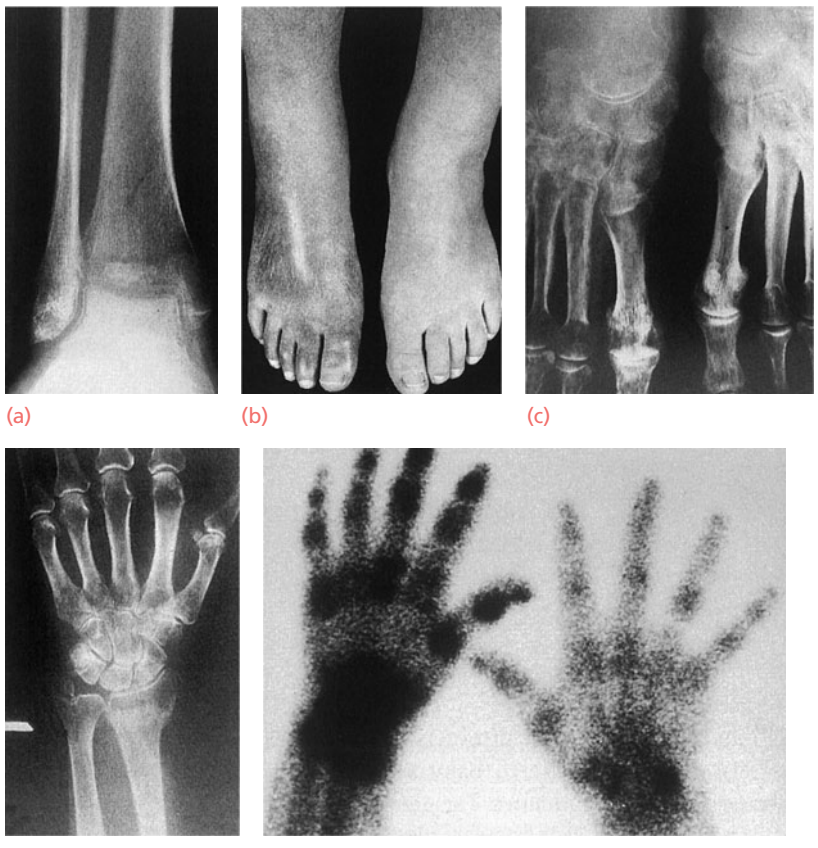


Figure 23.41 Complex regional pain syndrome (a) Regional osteoporosis is common after fractures of the extremities. (b) In CRPS the picture is exaggerated and the soft tissues are also involved: here the right foot is somewhat swollen and the skin has become dusky, smooth and shiny. (c) In the full-blown case, X-rays show a typical patchy osteoporosis. (d) Similar changes may occur in the wrist and hand; they are always accompanied by (e) increased activity in the radionuclide scan.

articular surface involves a large fragment in a joint that is readily accessible to surgery, intra-articular osteotomies and repositioning of the fragment may help. Often, although the problem arises from areas that were previously comminuted and depressed, little can be done once the fracture has united.

Malunion of a metaphyseal fracture may radically alter the mechanics of a nearby joint and this can give rise to secondary osteoarthritis too. It is often asserted that malunion in the shaft of a long bone (e.g. the tibia) may act in a similar manner; however, there is little evidence to show that residual angulation of less than 15 degrees can cause proximal or distal osteoarthritis.

STRESS (FATIGUE) FRACTURES

A stress or fatigue fracture is one occurring in the normal bone of a healthy patient, due not to any specific traumatic incident but to small repetitive stresses of two main types: bending and compression.

Bending stress causes deformation, and bone responds by changing the pattern of remodelling. With repeated stress, osteoclastic resorption exceeds osteoblastic formation and a zone of relative weakness develops, ultimately leading to a breach in the cortex. This process affects young adults undertaking strenuous physical routines and is probably due to muscular forces acting on bone. Athletes in training, dancers and military recruits build up muscle power quickly but bone strength only slowly; this accounts for the high incidence of stress fractures in these groups.

Compressive stresses act on soft cancellous bone; with frequent repetition an impacted fracture may result.

A combination of compression and shearing stresses may account for the osteochondral fractures that characterize some of the so-called osteochondritides.

'*Spontaneous fractures*' occur with even greater ease in people with osteoporosis or osteomalacia and in patients treated with drugs that affect bone remodelling in a similar way (e.g. corticosteroids and methotrexate). These are often referred to as *insufficiency fractures*.

Sites affected

Least rare are the following: shaft of humerus (adolescent cricketers); pars interarticularis of fifth lumbar vertebra (causing spondylolysis); pubic rami (inferior in children, both rami in adults); femoral neck (at any age); femoral shaft (mainly lower third); patella (children and young adults); tibial shaft (proximal third in children, middle third in athletes and trainee paratroopers, distal third in the elderly); distal shaft of the fibula (the 'runner's fracture'); calcaneum (adults); navicular (athletes) and metatarsals (especially the second).

Clinical features

There may be a history of unaccustomed and repetitive activity or one of a strenuous physical exercise programme. A common sequence of events is: *pain after exercise – pain during exercise – pain without exercise*. Occasionally the patient presents only after the fracture has healed and may then complain of a lump (the callus).

The patient is usually healthy. The affected site may be swollen or red. It is sometimes warm and usually tender; the callus may be palpable. 'Springing' the bone (attempting to bend it) is often painful.

Imaging

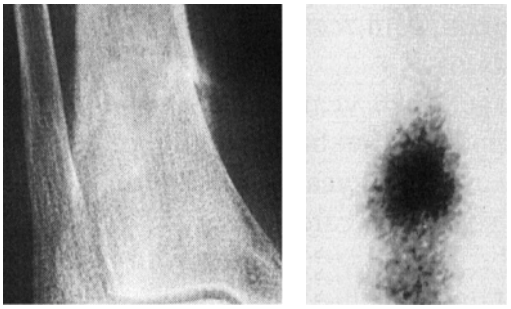
X-RAYS

Early on, the fracture is difficult to detect, but radioscintigraphy will show increased activity at the painful spot (Figure 23.42). Plain X-rays taken a few weeks later may show a small transverse defect in the cortex and/or localized periosteal new-bone formation. These appearances have, at times, been mistaken for those of an osteosarcoma, a horrifying trap for the unwary (Figure 23.43). Compression stress fractures (especially of the femoral neck and upper tibia) may show as a hazy transverse band of sclerosis with peripheral callus (in the tibia).

Another typical picture is that of a small osteoarticular fracture – most commonly of the dome of the medial femoral condyle at the knee or the upper surface of the talus at the ankle. Later, ischaemic necrosis of the detached fragment may render the lesion even more obvious.

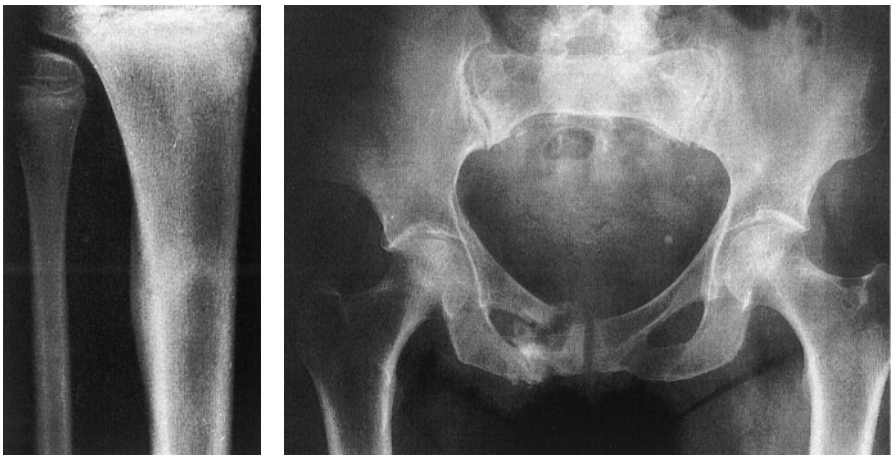
MRI

The earliest changes, particularly in 'spontaneous' undisplaced osteoarticular fractures, are revealed by MRI. This investigation should be requested in older



(a) (b)

Figure 23.42 Stress fracture (a) The stress fracture in this tibia is only just visible on X-ray, but it had already been suspected 2 weeks earlier when the patient first complained of pain and a radioisotope scan revealed a 'hot' area just above the ankle (b).



(a) (b)

Figure 23.43 Stress fractures
Stress fractures are often missed or wrongly diagnosed. (a) This tibial fracture was at first thought to be an osteosarcoma. (b) Stress fractures of the pubic rami in elderly women can be mistaken for metastases.

patients (possibly with osteoporosis) complaining of sudden onset of pain over the anteromedial part of the knee.

Diagnosis

Many disorders, including osteomyelitis, scurvy and battered baby syndrome, may be confused with stress fractures. The great danger, however, is a mistaken diagnosis of osteosarcoma; scanning shows increased uptake in both conditions and even biopsy may be misleading.

Treatment

Most stress fractures need no treatment other than an elastic bandage and avoidance of the painful activity until the lesion heals; surprisingly, this can take many months and the forced inactivity is not easily accepted by the hard-driving athlete or dancer.

An important exception is stress fracture of the femoral neck. This should be suspected in all elderly people who complain of pain in the hip for which no obvious cause can be found. If the diagnosis is confirmed by bone scan, the femoral neck should be internally fixed with screws as a prophylactic measure.

PATHOLOGICAL FRACTURES

When abnormal bone gives way, this is referred to as a pathological fracture. The causes are numerous and varied; often the diagnosis is not made until a biopsy is examined (Table 23.5 and Figure 23.44).

History

Bone that fractures spontaneously, or after trivial injury, must be regarded as abnormal until proved otherwise. Older patients should always be asked about previous illnesses or operations. A malignant tumour, no matter how long ago it occurred, may be the source of a late metastatic lesion; a history of gastrectomy, intestinal malabsorption, chronic alcoholism or prolonged drug therapy should suggest a metabolic bone disorder.

Symptoms such as loss of weight, pain, a lump, cough or haematuria suggest that the fracture may be through a secondary deposit.

In younger patients, a history of several previous fractures may suggest a diagnosis of osteogenesis imperfecta, even if the patient does not show the classic features of the disorder.

Examination

Local signs of bone disease (an infected sinus, an old scar, swelling or deformity) should not be missed. The site of the fracture may suggest the diagnosis: patients with involutional osteoporosis develop fractures of the vertebral bodies and corticocancellous junctions of long bones; a fracture through the shaft of the

Table 23.5 Causes of pathological fracture

Generalized bone disease	**Primary malignant tumours**
Osteogenesis imperfecta Postmenopausal osteoporosis Metabolic bone disease Myelomatosis Polyostotic fibrous dysplasia Paget's disease	Chondrosarcoma Osteosarcoma Ewing's tumour
Local benign conditions	**Metastatic tumours**
Chronic infection Solitary bone cyst Fibrous cortical defect Chondromyxoid fibroma Aneurysmal bone cyst Chondroma Monostotic fibrous dysplasia	Carcinoma in breast, lung, kidney, thyroid, colon and prostate

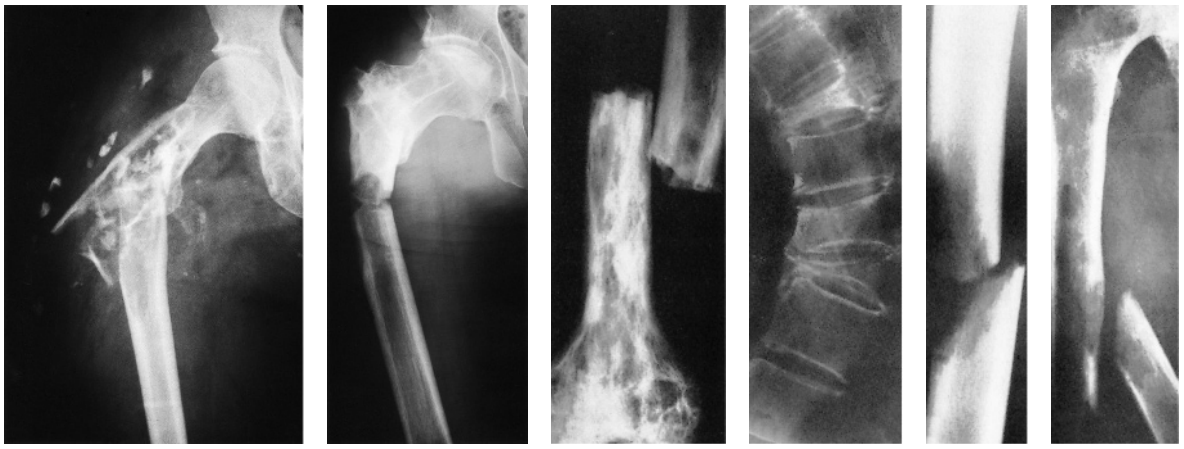


Figure 23.44 Pathological fractures Six examples of pathological fractures, due to: (a) primary chondrosarcoma; (b) postoperative bone infection at a screw-hole following plating of an intertrochanteric fracture; (c) Paget's disease; (d) vertebral metastases; (e) metastasis from carcinoma of the breast; (f) myelomatosis.

bone in an elderly patient, especially in the subtrochanteric region, should be regarded as a pathological fracture until proved otherwise.

General examination may be informative. Congenital dysplasias, fibrous dysplasia, Cushing's syndrome and Paget's disease all produce characteristic appearances. The patient may be wasted (possibly due to malignant disease). The lymph nodes or liver may be enlarged. It should be noted whether there is a mass in the abdomen or pelvis. Old scars should not be overlooked, and rectal and vaginal examinations are mandatory.

Under the age of 20 the common causes of pathological fracture are benign bone tumours and cysts. Over the age of 40 the common causes are multiple myeloma, metastatic lesions and Paget's disease.

X-rays

Understandably, the fracture itself attracts most attention but the surrounding bone must also be examined, and features such as cyst formation, cortical erosion, abnormal trabeculation and periosteal thickening should be sought. The type of fracture is important too: vertebral compression fractures may be due to severe osteoporosis or osteomalacia, but they can also be caused by skeletal metastases or myeloma. Middle-aged men, unlike women, do not normally become osteoporotic: X-ray signs of bone loss and vertebral compression in a male younger than 75 years should be regarded as 'pathological' until proven otherwise.

Additional investigations

Local radionuclide imaging may help elucidate the diagnosis, and whole-body scanning is important in revealing or excluding other deposits.

X-rays of other bones, the lungs and the urogenital tract may be necessary to exclude malignant disease. Investigations should always include a full blood count, ESR, protein electrophoresis, and tests for syphilis and metabolic bone disorders.

Urine examination may reveal blood from a tumour or Bence–Jones protein in myelomatosis.

Biopsy

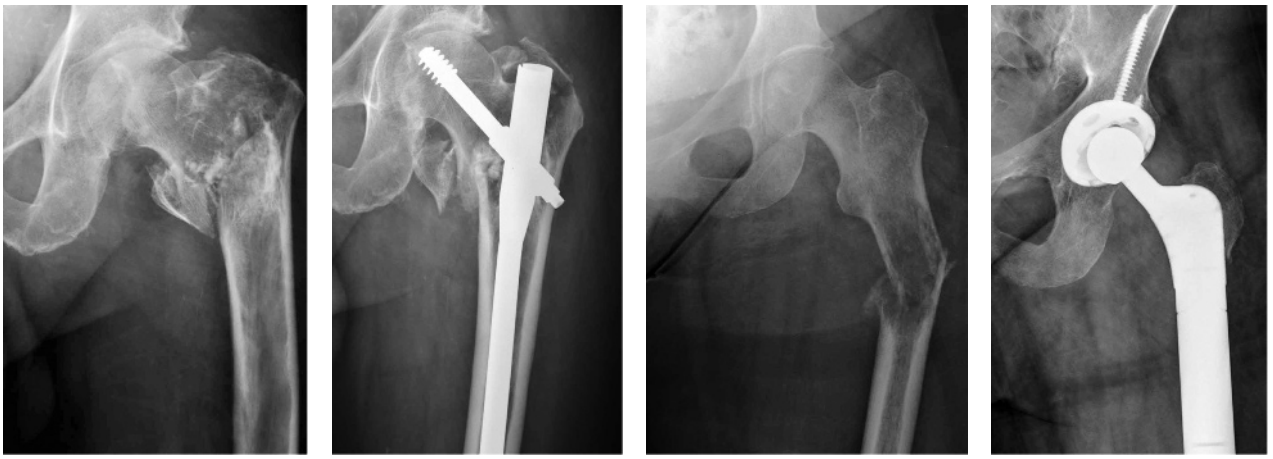
Some lesions are so typical that a biopsy is unnecessary (solitary cyst, fibrous cortical defect, Paget's disease). Others are more obscure and a biopsy is essential for diagnosis. If open reduction of the fracture is indicated, the biopsy can be carried out at the same time; otherwise a definitive procedure should be arranged.

Treatment

The principles of fracture treatment remain the same: *reduce, hold, exercise.* However, the choice of method is influenced by the condition of the bone (Figure 23.45); the underlying pathological disorder may need treatment in its own right (see Chapter 9).

Generalized bone disease In most of these conditions (including Paget's disease) the bones fracture more easily, but they heal quite well provided that the fracture is properly immobilized. Internal fixation is therefore advisable (and for Paget's disease almost essential). Patients with osteomalacia, hyperparathyroidism, renal osteodystrophy and Paget's disease will need systemic treatment as well.

Local benign conditions Fractures through benign cyst-like lesions usually heal quite well and



(a) (b) (c) (d)

Figure 23.45 Pathological fractures – treatment (a,b) Paget's disease of the femur increases the brittleness of bone, making it more likely to fracture. Intramedullary fixation allows the entire femur to be supported. (c,d) A fracture through a solitary metastasis from a previously excised renal cell carcinoma can be resected in order to achieve cure. In this case replacement of the proximal femur with an endoprosthesis is needed.

they should be allowed to do so before tackling the local lesion. Treatment is therefore the same as for simple fractures in the same area, although in some cases it will be necessary to take a biopsy before immobilizing the fracture. When the bone has healed, the tumour can be dealt with by curettage or local excision.

Primary malignant tumour The fracture may need splinting but this is merely a prelude to definitive treatment of the tumour, which will have spread by now to the surrounding soft tissues. The prognosis is almost always very poor.

Metastatic tumours Metastasis is a frequent cause of pathological fracture in older people. Breast cancer is the commonest source and the femur the commonest site. Nowadays cancer patients (even those with metastases) often live for many years and effective treatment of the fracture will vastly improve their quality of life.

Fracture of a long-bone shaft should be treated by internal fixation; if necessary, the site is also packed with acrylic cement. Bear in mind that the implant will function as a load-*bearing* and not a load-*sharing* device and will need to be strong enough to not break due to fatigue failure during the lifespan of the patient; intramedullary nails are more suitable than plates and screws.

Fracture near a bone end can often be treated by excision and total joint or endoprosthetic replacement; this is especially true of hip fractures.

Preoperatively, imaging studies should be performed to detect other bone lesions; these may be amenable to prophylactic fixation as part of the same procedure. Once the wound has healed, local irradiation can be considered to reduce the risk of progressive osteolysis.

Pathological compression fractures of the spine cause severe pain. This is due largely to spinal instability and treatment options should include operative stabilization. If there are either clinical or imaging features of actual or threatened spinal cord or cauda equina compression, the segment should also be decompressed. Postoperative irradiation is given as usual.

With all types of metastatic lesion, the primary tumour should be sought if not known, investigated and treated as well.

INJURIES OF THE PHYSIS

In children, over 10% of fractures involve injury to the growth plate (or physis). Because the physis is a relatively weak part of the bone, joint strains that might cause ligament injuries in adults are liable to result in separation of the physis in children. The fracture most commonly runs transversely through the hypertrophic or the calcified layer of the growth plate, often veering off into the metaphysis at one of the edges to include a triangular lip of bone. This has little effect on longitudinal growth, which takes place in the germinal and proliferating layers of the physis. However, if the fracture traverses the cellular 'reproductive' layers of the physis, it may result in premature ossification of the injured part and serious disturbances of bone growth.

Classification

The most widely used classification of physeal injuries is that of Salter and Harris, which distinguishes five basic types of injury (Figure 23.46).

- *Type 1* – This is a transverse fracture through the hypertrophic or calcified zone of the plate. Even if the fracture is quite alarmingly displaced, the growing zone of the physis is usually not injured and growth disturbance is uncommon.
- *Type 2* – This is essentially similar to type 1, but towards the edge the fracture deviates away from the physis and splits off a triangular metaphyseal fragment of bone (sometimes referred to as the Thurston–Holland fragment).
- *Type 3* – This is a fracture that splits the epiphysis and then veers off transversely to one or the other side, through the hypertrophic layer of the physis. Inevitably it damages the 'reproductive' layers of the physis (as these layers are closer to the epiphysis than the metaphysis) and may result in growth disturbance.
- *Type 4* – As with type 3, the fracture splits the epiphysis, but it extends into the metaphysis. These fractures are liable to displacement and a consequent misfit between the separated parts of the physis, resulting in asymmetrical growth.
- *Type 5* – This is a longitudinal compression injury of the physis. There is no visible fracture but the growth plate is crushed and this may result in growth arrest.

Mechanism of injury

Physeal fractures usually result from falls or traction injuries. They occur mostly in road accidents and during sporting activities or playground tumbles. Non-accidental injury should always be considered by the treating doctors and surgeons and investigated appropriately.

Clinical features

These fractures are more common in boys than in girls and are usually seen either in infancy or between the ages of 10 and 12. Deformity is usually minimal, but any injury in a child followed by pain and tenderness near the joint should arouse suspicion, and X-ray examination is essential.

X-rays

The physis itself is radiolucent and the epiphysis may be incompletely ossified; this makes it hard to tell whether the bone end is damaged or deformed. The younger the child, the smaller the 'visible' part of the epiphysis and thus the more difficult it is to make the diagnosis; comparison with the normal side is a great help. Telltale features are widening of the physeal 'gap', incongruity of the joint or tilting of the epiphyseal axis. If there is marked displacement the diagnosis is obvious, but even a type 4 fracture may at first be so little displaced that the fracture line is hard to see; if there is the faintest suspicion of a physeal fracture, a repeat X-ray after 4 or 5 days is essential. Type 5 injuries are usually diagnosed only in retrospect.

Treatment

Undisplaced fractures may be treated by splinting the part in a cast or a close-fitting plaster slab for 2–4 weeks (depending on the site of injury and the age of the child). However, with undisplaced type 3 and type 4 fractures, a check X-ray after 4 days and again at about 10 days is mandatory in order not to miss late displacement.

Displaced fractures should be reduced as soon as possible (Figure 23.47). With types 1 and 2 this can usually be done closed; the part is then splinted securely for 3–6 weeks. Type 3 and type 4 fractures demand perfect anatomical reduction. An attempt can be made to achieve this by gentle manipulation under

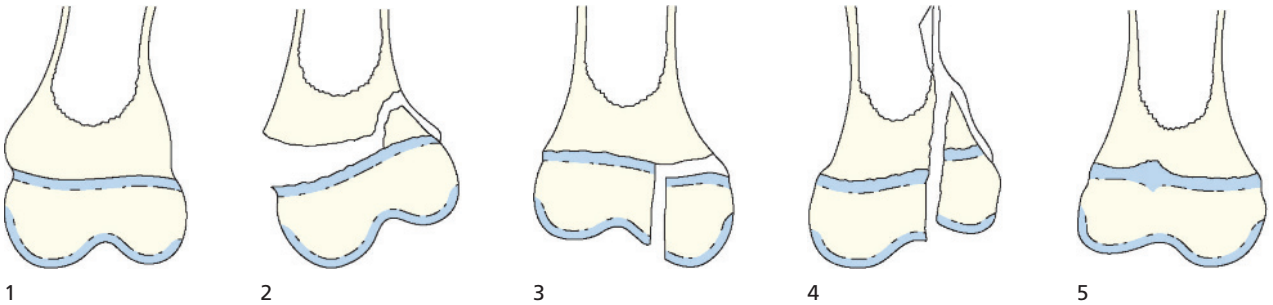


Figure 23.46 Physeal injuries *Type 1* – separation of the epiphysis – which usually occurs in infants but is also seen at puberty as a slipped femoral epiphysis. *Type 2* – fracture through the physis and metaphysis – is the commonest; it occurs in older children and seldom results in abnormal growth. *Type 3* – an intra-articular fracture of the epiphysis – needs accurate reduction to restore the joint surface. *Type 4* – splitting of the physis and epiphysis – damages the articular surface and may also cause abnormal growth; if it is displaced it needs open reduction. *Type 5* – crushing of the physis – may look benign but ends in arrested growth.

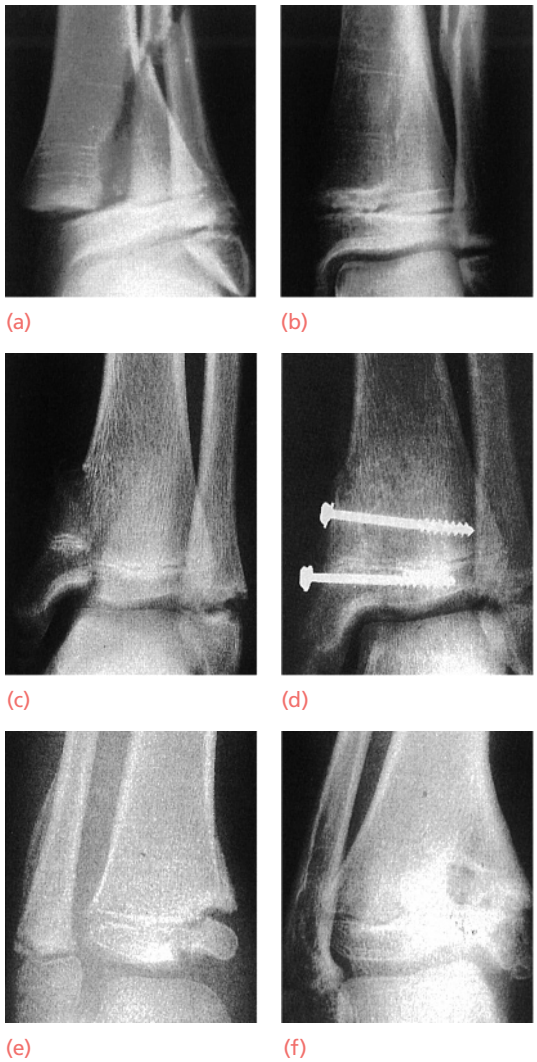


Figure 23.47 Physeal injuries (a) Type 2 injury. The fracture does not traverse the width of the physis; after reduction (b) bone growth is not distorted. (c,d) This type 4 fracture of the tibial physis was treated immediately by open reduction and internal fixation and a good result was obtained. (e,f) In this case accurate reduction was not achieved and the physeal fragment remained displaced; the end result was partial fusion of the physis and severe deformity of the ankle.

general anaesthesia; if this is successful, the limb is held in a cast for 4–8 weeks (the longer periods for type 4 injuries). If a type 3 or 4 fracture cannot be reduced accurately by closed manipulation, immediate open reduction and internal fixation with smooth K-wires is essential. The limb is then splinted for 4–6 weeks, but it takes that long again before the child is ready to resume unrestricted activities.

Complications

Type 1 and type 2 injuries, if properly reduced, have an excellent prognosis and bone growth is not adversely affected. Exceptions to this rule are injuries around the knee involving the distal femoral or proximal tibial physis; both growth plates are undulating in shape, so a transverse fracture plane may actually pass through more than just the hypertrophic zone and also damage the proliferative zone. Complications such as malunion or non-union may also occur if the diagnosis is missed and the fracture remains unreduced (e.g. fracture separation of the medial humeral epicondyle).

Type 3 and type 4 injuries may result in premature fusion of part of the growth plate or asymmetrical growth of the bone end. Type 5 fractures cause premature fusion and retardation of growth. The size and position of the bony bridge across the physis can be assessed by CT or MRI. If the bridge is relatively small (less than one-third the width of the physis), it can be excised and replaced by a fat graft, with some prospect of preventing or diminishing the growth disturbance. If the bone bridge is more extensive, however, the operation is contraindicated as it can end up doing more harm than good.

Established deformity, whether from asymmetrical growth or from malunion of a displaced fracture (e.g. a valgus elbow due to proximal displacement of a lateral humeral condylar fracture) should be treated by corrective osteotomy. If further growth is abnormal, the osteotomy may have to be repeated.

INJURIES TO JOINTS

Joints are usually injured by twisting or tilting forces that stretch the ligaments and capsule. If the force is great enough, the ligaments may tear, or the bone to which they are attached may be pulled apart. The articular cartilage may be damaged too if the joint surfaces are compressed or if there is a fracture into the joint (Figure 23.48).

As a general principle, forceful angulation will tear the ligaments rather than crush the bone, but in older people with porotic bone the ligaments may hold and the bone on the opposite side of the joint is crushed instead, while in children there may be a fracture separation of the physis.

Sprains, strains and ruptures

There is much confusion about the use of the terms ‘sprain’, ‘strain’ and ‘rupture’. Strictly speaking, a *sprain* is any painful wrenching (twisting or pulling) movement of a joint, but the term is generally reserved for joint injuries less severe than actual complete tearing of the capsule or ligaments. *Strain* is a physical effect of stress, in this case tensile stress associated with some stretching of the ligaments; in colloquial usage, ‘strained ligament’ is often meant to denote an injury somewhat more severe than a ‘sprain’, which

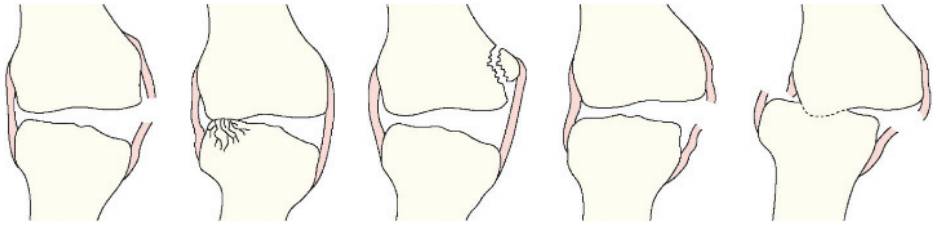


Figure 23.48 Joint injuries Severe stress may cause various types of injury. (a) A ligament may rupture, leaving the bone intact. If the soft tissues hold, the bone on the opposite side may be crushed (b), or a fragment may be pulled off by the taut ligament (c). Subluxation (d) means the articular surfaces are partially displaced; dislocation (e) refers to complete displacement of the joint.

possibly involves tearing of some fibres. If the stretching or twisting force is severe enough, the ligament may be strained to the point of complete *rupture*.

Strained ligament

Only some of the fibres in the ligament are torn and the joint remains stable. The injury is one in which the joint is momentarily twisted or bent into an abnormal position. The joint is painful and swollen and the tissues may be bruised. Tenderness is localized to the injured ligament and tensing the tissues on that side causes a sharp increase in pain.

Treatment

The joint should be firmly strapped or braced to support the injured ligament but range of motion permitted. Symptoms are improved by rest until the acute pain subsides. Thereafter, active movements are encouraged, and exercises should be practised to strengthen the muscles.

Ruptured ligament

The ligament is completely torn and the joint is unstable. Sometimes the ligament holds and the bone to which it is attached is avulsed; this is effectively the same lesion but easier to deal with because the bone fragment can be securely reattached with good chance of healing.

As with a strain, the joint is suddenly forced into an abnormal position; sometimes the patient actually hears or feels a snap. The joints most likely to be affected are the ones that are insecure by virtue of their shape or least well protected by surrounding muscles: the knee, ankle and finger joints.

Pain is severe and there may be considerable bleeding under the skin; if the joint is swollen, this is probably due to a haemarthrosis and typically comes on quickly. The patient is unlikely to permit a searching examination in the acute phase, but under general anaesthesia the instability can be demonstrated; this distinguishes the lesion from a strain. X-rays may show a detached flake of bone where the ligament is inserted.

Treatment

Torn ligaments heal by fibrous scarring. Previously this was thought inevitable and the surgeon's task was to ensure that the torn ends were securely sutured so as to restore the ligament to its normal length. In some injuries, such as rupture of the ulnar collateral ligament of the metacarpophalangeal joint of the thumb, this approach is still valid. In others, however, it has changed; thus, solitary medial collateral ligament ruptures of the knee, even complete ruptures, are often treated non-operatively in the first instance. The joint is supported in a splint or locked brace and local measures are taken to reduce swelling. After 1–2 weeks, the splint is exchanged for a functional brace (or the locked brace is unlocked) thus allowing joint movement. At the same time, the brace support prevents repeat injury to the ligament, especially if some instability is also present. Physiotherapy is applied to maintain muscle strength and later proprioceptive exercises are added. This non-operative approach has shown better results not only in the strength of the healed ligament but also in the nature of healing – there is less fibrosis.

An exception to this non-operative approach is when the ligament is avulsed with an attached fragment of bone; reattachment of the fragment is indicated if the piece is large enough. Occasionally, non-operative treatment may result in some residual instability that is clinically detectable; often this is not symptomatic but, if it is, surgical reconstruction should be considered.

Dislocation and subluxation

'Dislocation' means that the joint surfaces are completely displaced and are no longer in contact;

'subluxation' implies a lesser degree of displacement, such that the articular surfaces are still partly apposed.

Clinical features

Following an injury, the joint is painful and the patient tries at all costs to avoid moving it. The shape of the joint is abnormal and the bony landmarks may be displaced. The limb is often held in a characteristic position; movement is painful and restricted. X-rays will usually clinch the diagnosis but appropriate views must be taken; they will also show whether there is an associated bony injury affecting joint stability, i.e. a fracture-dislocation.

Apprehension test If the dislocation is reduced by the time the patient is seen, the joint can be tested by stressing it as if almost to reproduce the suspected dislocation: the patient develops a sense of impending disaster and violently resists further manipulation.

Recurrent dislocation If the ligaments and joint margins are damaged, repeated dislocation may occur. This is seen especially in the shoulder and patellofemoral joint.

Habitual (voluntary) dislocation Some patients acquire the knack of dislocating (or subluxating) the joint by voluntary muscle contraction. Ligamentous laxity may make this easier, but the habit often betrays a manipulative and neurotic personality. It is important to recognize this because such patients are seldom helped by operation which can lead to worsening of their symptoms.

Treatment

The dislocation must be reduced as soon as possible; usually sedation or general anaesthetic is required, and sometimes a muscle relaxant as well. The joint is then rested or immobilized until soft-tissue swelling reduces – usually after 2 weeks. Controlled movements then begin in a functional brace; progress with physiotherapy is monitored. Occasionally surgical reconstruction for residual instability is called for.

Complications

Many of the complications of fractures are seen also after dislocations: vascular injury, nerve injury, avascular necrosis of bone, heterotopic ossification, joint stiffness and secondary osteoarthritis. The principles of diagnosis and management of these conditions have been discussed earlier.

The authors would like to thank Selvadurai Nayagam for his contribution to this chapter in the previous edition.

FURTHER READING

Atkins RM. Complex regional pain syndrome. *J Bone Joint Surg* 2003; **85B**: 1100–6.

Charnley J. *The Closed Treatment of Common Fractures.* Edinburgh: Churchill Livingstone, 1961.

Dicpinigaitis PA, Koval KJ, Tejwani NC, Egol KA. Gunshot wounds to the extremities. *Bull NYU Hosp Jt Dis* 2006; **64**: 139–55.

Giordano CP, Koval KJ, Zuckerman JD, Desai P. Fracture blisters. *Clin Orthop* 1994; **307**: 214–21.

Gustilo RB, Mendoza RM, Williams DN. Problems in the management of type III (severe) open fractures: a new classification of type III open fractures. *J Trauma* 1984; **24**: 742–6.

Kenwright J, Richardson JB, Cunningham JL, et al. Axial movement and tibial fractures. A controlled randomised trial of treatment. *J Bone Joint Surg* 1991; **73B**: 654–9.

Langenskiold A. An operation for partial closure of an epiphyseal plate in children, and its experimental basis. *J Bone Joint Surg* 1975; **57B**: 325–30.

Langenskiold A. Surgical treatment of partial closure of the growth plate. *J Pediatr Orthop* 1981; **1**: 3–11.

Marsh JL, Slongo TF, Agel J, et al. Fracture and dislocation classification compendium – 2007: Orthopaedic Trauma Association classification, database and outcomes committee. *J Orthop Trauma* 2007; **21**(Suppl): S1–133.

McKibbin B. The biology of fracture healing in long bone. *J Bone Joint Surg* 1978; **60B**: 150–62.

McQueen MM, Court-Brown CM. Compartment monitoring in tibial fractures. The pressure threshold for decompression. *J Bone Joint Surg Br* 1996; **78**: 99–104.

Müller M, Nazarian S, Koch P, Schatzker J. *The Comprehensive Classification of Fractures of Long Bones.* New York: Springer Verlag, 1990.

Oestern H, Tscherne H. Pathophysiology and classification of soft tissue injuries associated with fractures. In: H. Tscherne H, Gotzen L (eds), *Fractures with Soft Tissue Injuries.* Berlin: Springer Verlag, 1984.

Pape HC, Giannoudis PV, Kretteck C, Trentz O. Timing of fixation of major fractures in blunt polytrauma: role of conventional indicators in clinical decision making. *J Orthop Trauma* 2005; **19**: 551–62.

Rang M. *The Growth Plate and its Disorders.* Edinburgh: Churchill Livingstone, 1969.

Roberts CS, Pape HC, Jones AL, et al. Damage control orthopaedics. Evolving concepts in the treatment of patients who have sustained orthopaedic trauma. *J Bone Joint Surg* 2005; **87A**: 434–49.

Rüedi TP, Buckley RE, Moran CG (eds). *AO Principles of Fracture Management.* Stuttgart: Thieme, 2007.

Salter RB, Harris WR. Injuries involving the epiphyseal plate. *J Bone Joint Surg* 1963; **45A**: 587–622.

Sarmiento A, Latta L. Functional fracture bracing. *J Am Acad Orthop Surg* 1999; **7**: 66–75.

Sarmiento A, Latta L. The evolution of functional bracing of fractures. *J Bone Joint Surg* 2006; **88B**: 141–8.

Sarmiento A, Mullis DL, Latta L, et al. A quantitative comparative analysis of fracture healing under the influence of compression plating vs. closed weight-bearing treatment. *Clin Orthop* 1980; **149**: 232–9.

Slongo TF, Audige L. Fracture and dislocation classification compendium for children: the AO pediatric comprehensive classification of long bone fractures (PCCF). *J Orthop Trauma* 2007; **21**(Suppl): S135–60.

Woo SL, Vogrin TM, Abramowitch SD. Healing and repair of ligament injuries in the knee. *J Am Acad Orthop Surg* 2000; **8**: 364–72.

Injuries of the shoulder and upper arm

24

Andrew Cole

The great bugbear of upper-limb injuries is stiffness – particularly of the shoulder but sometimes of the elbow and hand as well. Two points should be constantly borne in mind:

- Whatever the injury, and however it is treated, all the joints that are not actually immobilized – and especially the finger joints – should be exercised from the start.
- In elderly patients it is sometimes best to disregard the fracture and concentrate on regaining movement.

FRACTURES OF THE CLAVICLE

In children the clavicle fractures easily, but it almost invariably unites rapidly and without complications. In adults this can be a much more troublesome injury. In adults clavicle fractures are common, accounting for 2.6–4% of fractures and approximately 35% of all shoulder girdle injuries. Fractures of the midshaft account for 69–82%, lateral fractures for 21–28% and medial fractures for 2–3%.

Mechanism of injury

A fall on the shoulder or the outstretched hand may break the clavicle. In the common midshaft fracture, the lateral fragment is pulled down by the weight of the arm and the inner, medial half is held up by the sternomastoid muscle. In fractures of the lateral end, if the ligaments are intact, there is little displacement; but if the coracoclavicular ligaments are torn, or if the fracture is just medial to these ligaments, displacement may be more severe and closed reduction impossible. The clavicle is also a reasonably common site for pathological fractures.

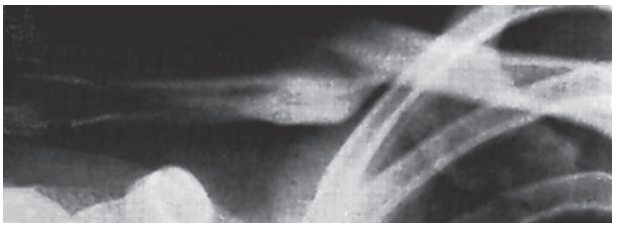
Clinical features

The arm is clasped to the chest to prevent movement. A subcutaneous lump may be obvious and occasionally a sharp fragment threatens the skin. Although vascular complications are rare, it is prudent to feel the pulse and gently to palpate the root of the neck. Outer third fractures are easily missed or mistaken for acromioclavicular joint injuries.

Imaging

Radiographic analysis requires at least an anteroposterior view and another taken with a 30 degree cephalic tilt. The fracture is usually in the middle third of the bone, and the outer fragment usually lies below the inner (Figure 24.1). Fractures of the outer third may be missed, or the degree of displacement underestimated, unless additional views of the shoulder are obtained. With medial-third fractures it is also wise to obtain *X-rays* of the sternoclavicular joint. In assessing clinical progress, remember that 'clinical' union usually precedes 'radiological' union by several weeks.

(a)



(b)

Figure 24.1 Fracture of the clavicle (a) Displaced fracture of the middle third of the clavicle – the most common injury. (b) The fracture usually unites in this position, leaving a barely noticeable 'bump'.

CT scanning with three-dimensional reconstructions may be needed to determine accurately the degree of shortening or for diagnosing a sternoclavicular fracture-dislocation, and also to establish whether a fracture has united.

Classification

Clavicle fractures are usually classified on the basis of their location:

- *Group I* – middle-third fractures
- *Group II* – lateral-third fractures
- *Group III* – medial-third fractures.

While it is helpful to describe the position of the fracture, this does not describe any of the prognostic indicators such as comminution, shortening or displacement. Lateral-third fractures can be further sub-classified into *Neer type I*, those with the coracoclavicular ligaments intact, *Neer type II*, those where the coracoclavicular ligaments are torn or detached from the medial segment but the trapezoid ligament remains intact to the distal segment, and *Neer type III* factures, which are intra-articular. An even more detailed classification of midshaft fractures, proposed by Robinson, is useful for managing data and comparing clinical outcomes.

Treatment

MIDDLE-THIRD FRACTURES

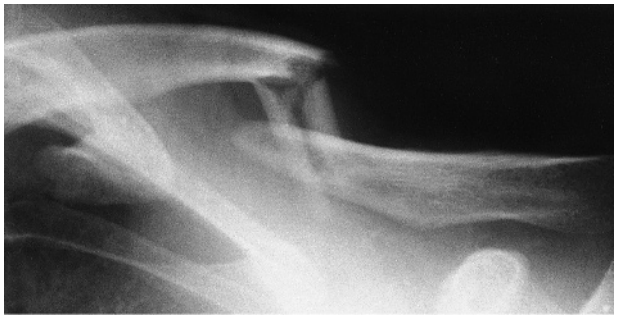
There is general agreement that undisplaced fractures should be treated non-operatively. Most will go on to unite uneventfully with a non-union rate below 5% and a return to normal function.

Non-operative management consists of applying a simple sling for comfort. It is discarded once the pain subsides (after 1–3 weeks) and the patient is then encouraged to mobilize the limb as pain allows. There is no evidence that the traditional figure-of-eight bandage confers any advantage and it carries the risk of increasing the incidence of pressures sores over the fracture site and causing harm to neurological structures; it may even increase the risk of non-union.

There is less agreement about the management of displaced middle-third fractures. Treating those with shortening of more than 2 cm by simple splintage is now believed to incur a considerable risk of symptomatic malunion – mainly pain and lack of power during shoulder movements – and an increased incidence of non-union. There is, therefore, a growing trend towards internal fixation of acute clavicular fractures associated with severe displacement, fragmentation or shortening (Figure 24.2). Methods include plating (specific contoured locking plates are available) and intramedullary fixation.

LATERAL-THIRD FRACTURES

Most lateral clavicle fractures are minimally displaced and extra-articular. The fact that the coracoclavicular ligaments are intact prevents further displacement and non-operative management is usually appropriate. Treatment consists of a sling for 2–3 weeks until



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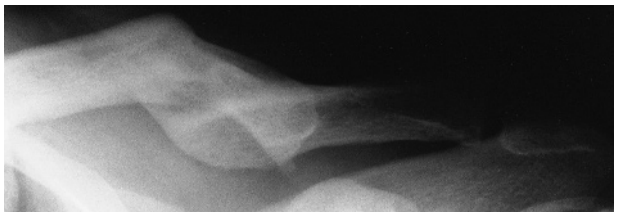

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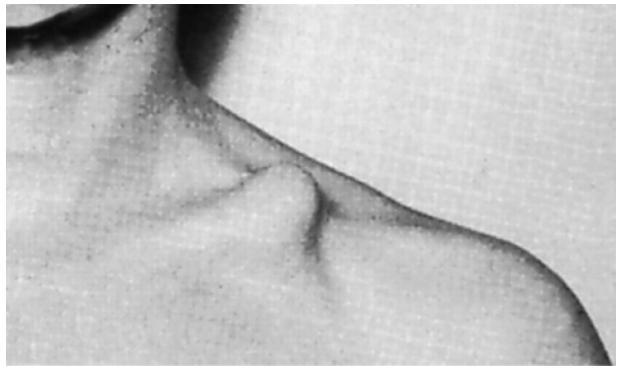

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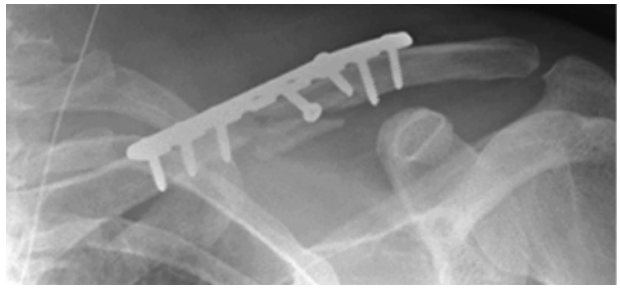
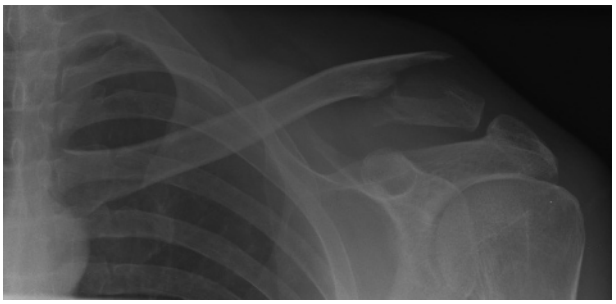


(d)

Figure 24.2 Severely displaced fracture (a) A comminuted fracture which united in this position (b) leaving an unsightly deformity (c). This fracture may have been better managed by (d) open reduction and internal fixation.



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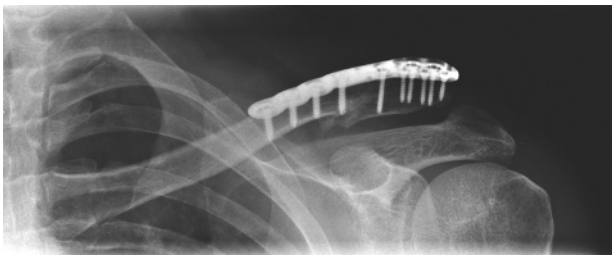


(b)

Figure 24.3 Fracture of the outer (lateral) third (a) The shaft of the clavicle is elevated, suggesting that the medial part of the coracoclavicular ligament is ruptured. (b) This was treated by open reduction and internal fixation, using a lateral clavicular locking plate.

the pain subsides, followed by mobilization within the limits of pain.

Displaced lateral-third fractures are associated with disruption of the coracoclavicular ligaments and are therefore unstable injuries (Figure 24.3). A number of studies have shown that these particular fractures have a higher than usual rate of non-union if treated non-operatively. Surgery to stabilize the fracture is often recommended. However, the converse argument is that many of the fractures that develop non-union do not cause any symptoms and surgery can therefore be reserved for patients with symptomatic non-union. Operations for these fractures have higher complication rates and no single procedure has been shown to be better than the others. Techniques include the use of a coracoclavicular screw and plate, hook plate fixation, suture and sling techniques with Dacron graft ligaments and the more recent lateral clavicle locking plates.

MEDIAL-THIRD FRACTURES

Most of these rare fractures are extra-articular. They are mainly managed non-operatively unless the fracture displacement threatens the mediastinal structures. Initial fixation is associated with significant complications, including migration of the implants into the mediastinum, particularly when K-wires are used. Other methods of stabilization include suture and graft techniques and the newer locking plates.

Complications

EARLY

Despite the close proximity of the clavicle to vital structures, a pneumothorax, damage to the subclavian vessels and brachial plexus injuries are all very rare.

LATE

Non-union In displaced fractures of the shaft non-union occurs in 1–15%. Risk factors include increasing age, displacement, comminution and female sex, but accurate prediction of those fractures most likely to go on to non-union remains difficult.

Symptomatic non-unions are generally treated with plate fixation and bone grafting if necessary. This procedure usually produces a high rate of union and satisfaction.

Lateral clavicle fractures have a higher rate of non-union (11.5–40%). Treatment options for symptomatic non-unions are excision of the lateral part of the clavicle (if the fragment is small and the coracoclavicular ligaments are intact) or open reduction, internal fixation and bone grafting if the fragment is large. Locking plates and hooked plates are used.

Malunion All displaced fractures heal in a non-anatomical position with some shortening and angulation, although most do not produce symptoms. Some may go on to develop periscapular pain and this is more likely with shortening of more than 1.5 cm. In these circumstances the difficult operation of corrective osteotomy and plating can be considered.

Stiffness of the shoulder This is common but usually temporary.

FRACTURES OF THE SCAPULA

Mechanisms of injury

Fractures of the scapula are often as a consequence of high-energy trauma and considerable force due to its protected position. Common causes include direct blunt trauma, crushing injuries, falls and seizures. Other associated injuries such as rib fractures and other intra-thoracic injuries are common. The neck of the scapula may be fractured by a blow or by a fall on the shoulder; the attached long head of triceps may drag the glenoid downwards and laterally. The coracoid process may fracture across its base or be avulsed at the tip. Fracture of the acromion is due to direct force. Fracture of the glenoid fossa usually suggests a medially directed force (impaction of the joint) but may occur with dislocation of the shoulder.

Clinical features

The arm is held immobile and there may be severe bruising over the scapula or the chest wall. Because of the energy required to damage the scapula, fractures of the body of the scapula are often associated with severe injuries to the chest, brachial plexus, spine, abdomen and head. Careful neurological and vascular examinations are essential.

Imaging

Scapular fractures can be difficult to define on plain *X-rays* because of the surrounding soft tissues. A true AP of the scapula, axillary view and scapular Y views are the most helpful. *CT and 3D reconstruction* are useful for demonstrating glenoid fractures or body fractures.

BOX 24.1 FRACTURES OF THE SCAPULA

Fractures of the scapular body

Fractures of the glenoid neck

Intra-articular glenoid fossa fractures (Ideberg modified by Goss) (Figure 24.4)

- Type I Fractures of the glenoid rim
- Type II Fractures through the glenoid fossa, inferior fragment displaced with subluxed humeral head
- Type III Oblique fracture through glenoid exiting superiorly (may be associated with acromioclavicular dislocation or fracture)
- Type IV Horizontal fracture exiting through the medial border of the scapula
- Type V Combination of type IV and a fracture separating the inferior half of the glenoid
- Type VI Severe comminution of the glenoid surface

Fractures of the acromion

- Type I Minimally displaced
- Type II Displaced but not reducing subacromial space
- Type III Inferior displacement and reduced subacromial space

Fractures of the coracoid process

- Type I Proximal to attachment of the coracoclavicular ligaments and usually associated with acromioclavicular separation
- Type II Distal to the coracoacromial ligaments

Classification

Fractures of the scapula are divided anatomically into scapular body, glenoid neck, glenoid fossa, acromion and coracoid processes. Scapular neck fractures are the most common. Further subdivisions are shown in Box 24.1.

Treatment

Body fractures Surgery is not usually necessary. The patient wears a sling for comfort, and from the start practises active exercises to the shoulder, elbow and fingers.

Isolated glenoid neck fractures This is the second most common fracture of the scapula. A CT scan is usually required to confirm that it is extra-articular. The fractures are often displaced, but further displacement is uncommon as long as the clavicle is not fractured. The fracture is frequently impacted and the glenoid surface is intact. A sling is worn for comfort and early exercises are begun.

Intra-articular fractures See Figures 24.4 and 24.5. *Type I* glenoid fractures, if displaced, may result in

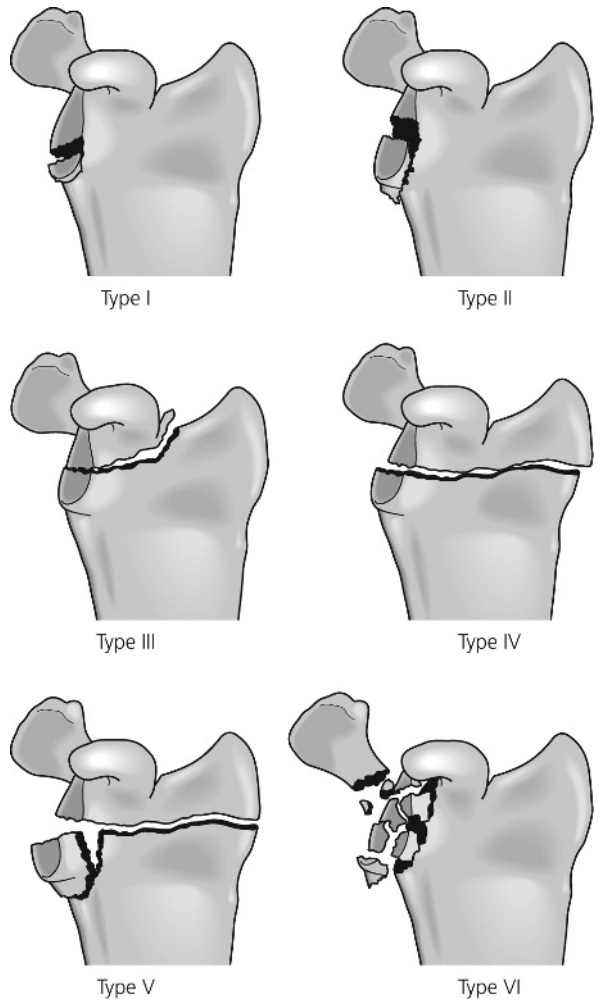
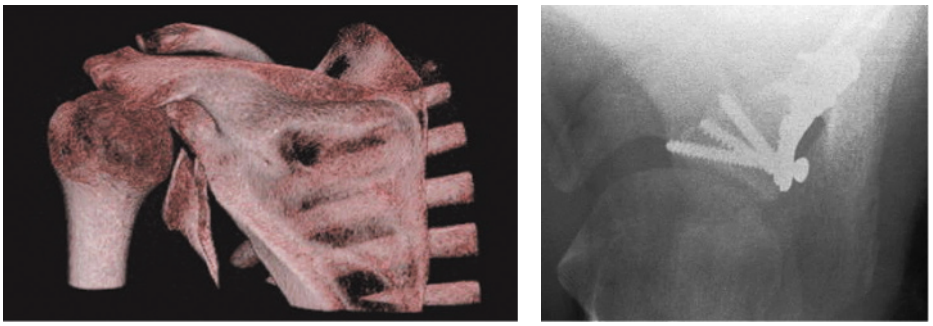


Figure 24.4 Fractures of the glenoid – classification
Diagrams showing the main types of glenoid fracture.



(a) (b)

Figure 24.5 Glenoid fracture –imaging
(a) Three-dimensional CT of a type II glenoid fracture. (b) X-ray after open reduction and internal fixation.

instability of the shoulder. If the fragment involves more than a quarter of the glenoid surface and is displaced by more than 5 mm, surgical fixation should be considered. Anterior rim fractures are approached through a deltopectoral incision and posterior rim fractures through the posterior approach. *Type II* fractures are associated with inferior subluxation of the head of the humerus and require open reduction and internal fixation. *Type III, IV, V and VI* fractures have poorly defined indications for surgery, but indications include excessive medialization of the glenoid or intra-articular steps of more than 5 mm. Generally speaking, if the head is centred on the major portion of the glenoid and the shoulder is stable, a non-operative approach is adopted. Comminuted fractures of the glenoid fossa are likely to lead to osteoarthritis in the longer term.

Fractures of the acromion Undisplaced fractures are treated non-operatively. Only type III acromial fractures, in which the subacromial space is reduced, require operative intervention to restore the anatomy.

Fractures of the coracoid process Fractures distal to the coracoacromial ligaments do not result in serious anatomical displacement; those proximal to the ligaments are usually associated with acromioclavicular separations and may need operative treatment.

Combined fractures Whereas an isolated fracture of the glenoid neck is stable, if there is an associated fracture of the clavicle or disruption of the acromioclavicular ligament, the glenoid mass may become markedly displaced, giving rise to a 'floating shoulder'. Diagnosis can be difficult and may require advanced imaging and 3D reconstructions. At least one of the injuries (and sometimes both) will need operative fixation before the fragments are stabilized.

SCAPULOTHORACIC DISSOCIATION

This is a high-energy injury. The scapula and arm are wrenched away from the chest, rupturing the subclavian vessels and brachial plexus.

Clinical features

The limb is flail and ischaemic. The diagnosis is usually made on the chest X-ray. There is swelling above the clavicle from an expanding haematoma. A distraction of more than 1 cm of a fractured clavicle should give rise to suspicion of this injury. Diagnosis can be made on a chest X-ray, showing lateral displacement of the scapula. There is a high mortality rate associated with this injury.

Treatment

The patient is resuscitated. The outcome for the upper limb is very poor. Functional outcome is dependent on the neurological injury but in many cases early amputation may be the outcome.

ACROMIOCLAVICULAR JOINT INJURIES

Acute injury of the acromioclavicular joint is common and usually follows direct trauma. It represents around 12% of all shoulder injuries. Chronic sprains, often associated with degenerative changes, are seen in people engaged in athletic activities like weightlifting or occupations such as working with jack-hammers and other heavy vibrating tools.

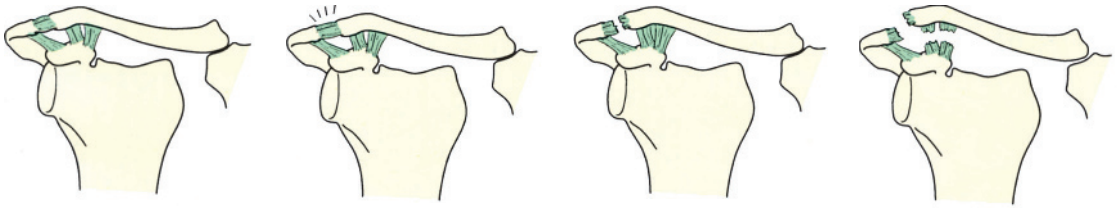
Mechanism of injury

A fall on the shoulder with the arm adducted may strain or tear the acromioclavicular ligaments and upward subluxation of the clavicle may occur; if the force is severe enough, the coracoclavicular ligaments will also be torn, resulting in complete dislocation of the joint.

Pathological anatomy and classification

The injury is graded according to the type of ligament injury and the amount of displacement of the joint (Figure 24.6).

- *Type I* – This is an acute sprain of the acromioclavicular ligaments; the joint is undisplaced.



(a) (b) (c) (d)

Figure 24.6 Acromioclavicular joint injuries (a) Normal joint. (b) Sprained acromioclavicular joint; no displacement. (c) Torn capsule and subluxation but coracoclavicular ligaments intact. (d) Dislocation with torn coracoclavicular ligaments.

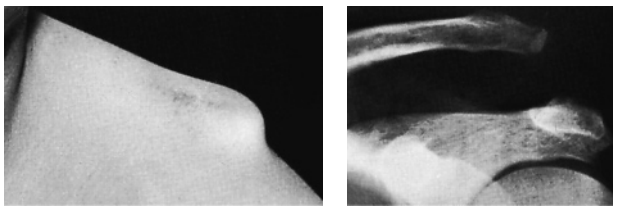
- *Type II* – The acromioclavicular ligaments are torn and the joint is subluxated with slight elevation of the clavicle.
- *Type III* – The acromioclavicular and coracoclavicular ligaments are torn and the joint is dislocated; the clavicle is elevated (or the acromion depressed) creating a visible and palpable 'step'. Other types of displacement are less common, but occasionally the clavicle is displaced posteriorly (type IV), very markedly upwards (type V) or inferiorly beneath the coracoid process (type VI).

Clinical features

The patient can usually point to the site of injury and the area may be bruised. If there is tenderness but no deformity, the injury is probably a sprain or a subluxation. With dislocation the patient may be in severe pain and a prominent 'step' can be seen and felt (Figure 24.7). Shoulder movements may also be limited.

X-rays

The acromioclavicular joint is not always easily visualized; anteroposterior, cephalic tilt and axillary views are advisable. In addition, a stress view is sometimes helpful in distinguishing between a type II and type III injury: this is an anteroposterior X-ray including both shoulders with the patient standing upright, arms by the side and holding a 5 kg weight in each hand. The distance between the coracoid process and the inferior border of the clavicle is measured on each side; a difference of more than 50% is diagnostic of acromioclavicular dislocation.



(a) (b)

Figure 24.7 Acromioclavicular dislocation (a) Clinically one sees a definite 'step' in the contour at the lateral end of the clavicle. (b) The X-ray shows complete separation of the acromioclavicular joint.

Treatment

Sprains and subluxations often do not affect function and do not require any special treatment; the arm is rested in a sling until pain subsides (usually no more than a week) and shoulder exercises are then begun.

Dislocations are poorly controlled by padding and bandaging, yet the role of surgery in type III injuries remains controversial. The large number of operations described suggests that none is ideal. There is no convincing evidence that surgery provides a better functional result than conservative treatment for a straightforward type III injury. Operative repair should be considered only for patients with extreme prominence of the clavicle, those with posterior or inferior dislocation of the clavicle and those who aim to resume strenuous overarm or overhead activities.

While there is no consensus regarding the best surgical solution, there are a number of underlying principles to consider if surgery is contemplated. Accurate reduction should be the goal. The ligamentous stability can be recreated either by transferring existing ligaments (the coracoacromial or conjoined tendons), or by using a free graft (e.g. autogenous semitendinosis or a synthetic ligament). This reconstruction must have sufficient stability to prevent re-dislocation during recovery. Any rigid implants which cross the joint will need to be removed at a later date to prevent loosening or fracture.

In the modified Weaver–Dunn procedure the lateral end of the clavicle is excised and the coracoacromial ligament is transferred to the outer end of the clavicle and attached by transosseous sutures. Tension on the repair can be reduced either by anchoring the clavicle to the coracoid with various techniques such as anchors or slings around the coracoid and clavicle. Great care is needed to avoid entrapment or damage to a nerve or vessel. Elbow and forearm exercises are begun on the day after operation and active-assisted shoulder movements 2 weeks later, increasing gradually to active movements at 4–6 weeks. Strenuous lifting movements

are avoided for 4–6 months. An alternative procedure is to use a synthetic graft which wraps around the coracoid and is secured around the clavicle by various techniques.

Recent advances in instrumentation have made it feasible to perform this type of reconstructive surgery arthroscopically.

Complications

Rotator cuff problems An acute strain of the acromioclavicular joint is sometimes followed by supraspinatus tendinitis. Whether this is directly due to the primary injury or whether it results from post-traumatic oedema or inflammation of the overlying acromioclavicular joint is unclear. Treatment with anti-inflammatory preparations may help.

Unreduced dislocation An unreduced dislocation can be disfiguring and sometimes affects function. Simple excision of the distal clavicle will only make matters worse. An attempt should be made to reconstruct the coracoclavicular ligament. The Weaver–Dunn type procedure may be suitable (Figure 24.8).

Ossification of the ligaments The more severe injuries are quite often followed by ossification of the coracoclavicular ligaments. Bony spurs may predispose to later rotator cuff dysfunction, which may require operative treatment.

Secondary osteoarthritis A late complication of type I and II injuries is osteoarthritis of the acromioclavicular joint. This can usually be managed conservatively, but if pain is marked the outer 2 cm of the clavicle can be excised. The patient will be aware of some weakness during strenuous overarm activities and pain is often not completely abolished.

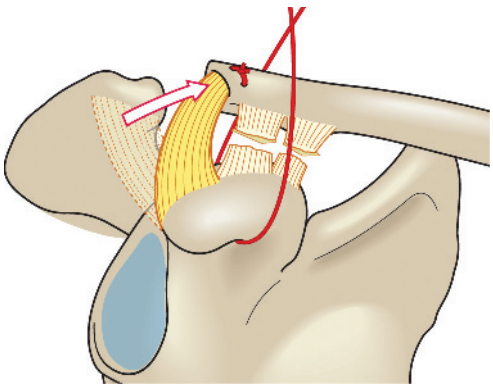


Figure 24.8 Modified Weaver–Dunn operation The lateral end of the clavicle is excised; the acromial end of the coracoacromial ligament is detached and fastened to the lateral end of the clavicle. Tension on the ligament is lessened by placing a 'sling' around the clavicle and the coracoid process. (Dotted lines show former position of coracoacromial ligament.)

STERNOCLAVICULAR DISLOCATION

Mechanism of injury

This uncommon injury is usually caused by lateral compression of the shoulders, such as when someone is pinned to the ground following an RTC or other crushing injuries. Rarely, it follows a direct blow to the front of the joint. Anterior dislocation is much more common than posterior. The joint can be sprained, subluxed or dislocated.

Clinical features

Anterior dislocation is easily diagnosed; the dislocated medial end of the clavicle forms a prominent bump over the sternoclavicular joint (Figure 24.9). The condition is painful but there are usually no cardiothoracic complications.

Posterior dislocation, though rare, is much more serious. Discomfort is marked; there may be pressure on the trachea or large vessels, causing venous congestion of the neck and arm and circulation to the arm may be decreased.

Imaging

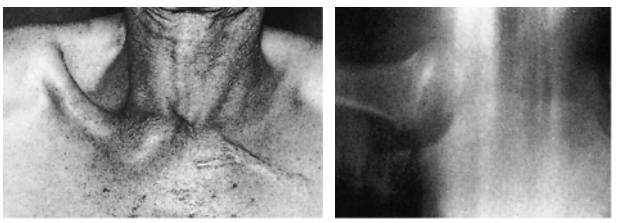
Because of overlapping shadows, plain *X-rays* are difficult to interpret. Special oblique views are helpful and *CT* is the ideal method.

Treatment

Sprains and subluxations often do not require specific treatment.

Anterior dislocation can be reduced by exerting pressure over the clavicle and pulling on the arm with the shoulder abducted. However, the joint usually redislocates. Not that this matters much; full function is often regained, though this may take several months.

Internal fixation is usually unnecessary and potentially hazardous (because of the large vessels behind the sternum). Posterior dislocation should be reduced



(a) (b)

Figure 24.9 Sternoclavicular dislocation (a) The bump over the sternoclavicular joint may be obvious, though this is difficult to demonstrate on plain X-rays. (b) Tomography (or, better still, CT) will show the lesion.

as soon as possible. This can usually be done closed (if necessary under general anaesthesia) by lying the patient supine with a sandbag between the scapulae and then pulling on the arm with the shoulder abducted and extended. The joint reduces with a snap and stays reduced. If this manoeuvre fails, the medial end of the clavicle is grasped with bone forceps and pulled forwards. If this also fails (a very rare occurrence), open reduction is justified, but great care must be taken not to damage the mediastinal structures. After reduction, the shoulders are braced back with a figure-of-eight bandage, which is worn for 3 weeks.

DISLOCATION OF THE SHOULDER

Of the large joints, the shoulder is the one that most commonly dislocates. This is due to a number of factors: (1) the shallowness of the glenoid socket; (2) the extraordinary range of movement; (3) underlying conditions such as ligamentous laxity or glenoid dysplasia; and (4) the sheer vulnerability of the joint during stressful activities of the upper limb.

In this chapter, acute anterior and posterior dislocations are described. Chronic instability is described in Chapter 13.

ANTERIOR DISLOCATION OF THE SHOULDER

Mechanism of injury

Dislocation is usually caused by a fall on the hand. The head of the humerus is driven forward, tearing the capsule and producing avulsion of the glenoid labrum (the Bankart lesion). Occasionally the posterolateral part of the head is crushed, giving rise to a Hill–Sachs lesion. Rarely, the acromion process levers the head downwards and the joint dislocates with the arm pointing upwards (*luxation erecta*); nearly always the arm then drops, bringing the head to its subcoracoid position. In patients with constitutionally lax shoulders, minimal trauma may be involved.

Clinical features

Pain is often severe. The patient supports the arm with the opposite hand and is loathe to permit any kind of examination. The lateral outline of the shoulder may be flattened and, if the patient is not too muscular, a bulge may be felt just below the clavicle (Figure 24.10). The arm must always be examined for nerve and vessel injury before reduction is attempted.

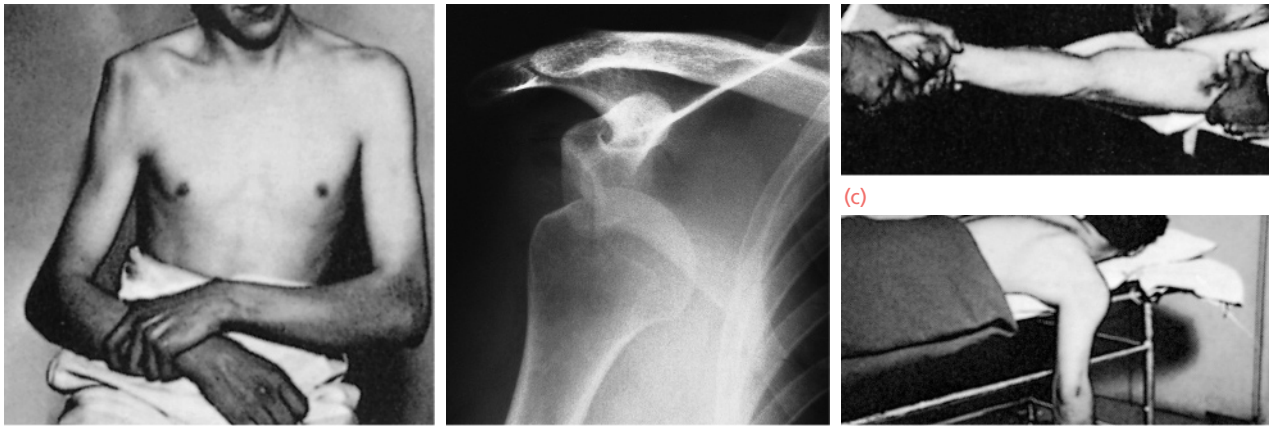
X-rays

The anteroposterior X-rays will show the overlapping shadows of the humeral head and glenoid fossa, with the head usually lying below and medial to the socket. A lateral view aimed along the blade of the scapula will show the humeral head out of line with the socket.

If the joint has dislocated before, special views may show flattening or an excavation of the posterolateral contour of the humeral head, where it has been indented by the anterior edge of the glenoid socket, the Hill–Sachs lesion.

Treatment

Various methods of reduction have been described, some of them now of no more than historical interest. In a patient who has had previous dislocations, simple traction on the arm may be successful.



(a) (b) (c) (d)

Figure 24.10 Anterior dislocation of the shoulder (a) The prominent acromion process and flattening of the contour over the deltoid are typical signs. (b) X-rays confirm the diagnosis of anterior dislocation. (c,d) Two methods of reduction.

Usually, sedation and occasionally general anaesthesia is required.

- *Stimson's technique* – The patient is left prone with the arm hanging over the side of the bed. After 15 or 20 minutes the shoulder may reduce.
- *Hippocratic method* – Gently increasing traction is applied to the arm with the shoulder in slight abduction, while an assistant applies firm countertraction to the body (a towel slung around the patient's chest, under the axilla, is helpful).
- *Kocher's method* – The elbow is bent to 90 degrees and held close to the body; no traction should be applied. The arm is slowly rotated 75 degrees laterally, then the point of the elbow is lifted forwards, and finally the arm is rotated medially. *This technique carries the risk of nerve, vessel and bone injury and is not recommended.*

Another technique has the patient sitting on a reduction chair and with gentle traction of the arm over the back of the padded chair the dislocation is reduced.

An X-ray is taken to confirm reduction and exclude a fracture. When the patient is fully awake, active abduction is gently tested to exclude an axillary nerve injury and rotator cuff tear. The median, radial, ulnar and musculocutaneous nerves are also tested and the pulse is felt.

The arm is rested in a sling for about 3 weeks in those under 30 years of age (who are most prone to recurrence) and for only 1 week in those over 30 (who are most prone to stiffness). Then movements are begun, but combined abduction and lateral rotation must be avoided for at least 3 weeks. Throughout this period, elbow and finger movements are practised every day.

There has been some interest in the use of external rotation splints, based on the theory that this would reduce the Bankart lesion into a better position for healing. However, a recent Cochrane review has concluded that there is insufficient evidence to inform on the choices for conservative treatment and that further trials are needed to compare different types and duration of immobilization.

Young athletes who dislocate their shoulder traumatically and who continue to pursue their sports (particularly contact sports) are at a much higher risk of re-dislocation in the future. The rates of recurrent instability depend on the size of the Hill–Sachs lesion, the presence of other bony injuries such as glenoid rim fractures and the state of the soft tissues. The rates of recurrent instability vary in particular with the age of the patient. After traumatic dislocation these rates can vary from 17% to 96% with increased rates in younger patients. This knowledge, combined with better arthroscopic instrumentation, has led to a recent increase in surgical stabilization after a single anterior dislocation in young sporting patients. Currently the evidence supports this course, but there is little evidence for the role of surgery after a single dislocation in older age groups.

Complications

EARLY

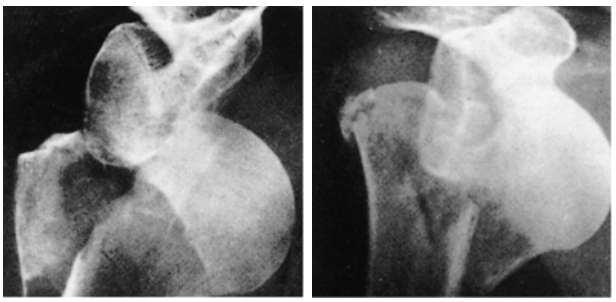
Rotator cuff tear This commonly accompanies anterior dislocation, particularly in older people. The patient may have difficulty abducting the arm after reduction; palpable contraction of the deltoid muscle excludes an axillary nerve palsy. Some do not require surgical attention, but young active individuals with large tears will benefit from early repair.

Nerve injury The axillary nerve is most commonly injured; the patient is unable to contract the deltoid muscle and there may be a small patch of anaesthesia over the muscle. The inability to abduct must be distinguished from a rotator cuff tear. The nerve lesion is usually a neuropraxia, which recovers spontaneously after a few weeks; if it does not, then surgery should be considered as the results of repair are less satisfactory if the delay is more than a few months.

Occasionally the radial nerve, musculocutaneous nerve, median nerve or ulnar nerve can be injured. Rarely there is a complete infraclavicular brachial plexus palsy. This is somewhat alarming, but fortunately it usually recovers with time.

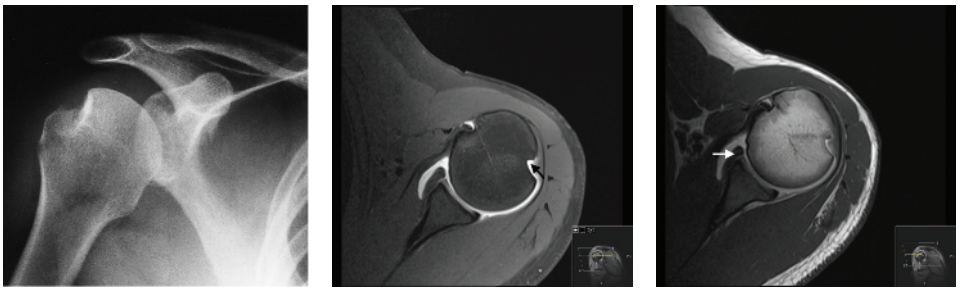
Vascular injury The axillary artery may be damaged, particularly in older patients with fragile vessels. This can occur either at the time of injury or during overzealous reduction. The limb should always be examined for signs of ischaemia both before and after reduction.

Fracture-dislocation If there is an associated fracture of the proximal humerus, open reduction and internal fixation may be necessary (Figure 24.11). The greater tuberosity may be sheared off during dislocation. It usually falls into place during reduction,



(a) (b)

Figure 24.11 Anterior fracture–dislocation Anterior dislocation of the shoulder may be complicated by fracture of (a) the greater tuberosity or (b) the neck of the humerus – this often needs open reduction and internal fixation.



(a) (b) (c)

Figure 24.12 Recurrent dislocation of the shoulder (a) The classic X-ray sign is a depression in the posterosuperior part of the humeral head (the Hill–Sachs lesion). (b,c) MRI scans showing both the Hill–Sachs lesion and a Bankart lesion of the glenoid rim (arrows).

and no special treatment is then required. If it remains displaced, surgical reattachment is recommended to avoid later subacromial impingement.

LATE

Shoulder stiffness Prolonged immobilization may lead to stiffness of the shoulder, especially in patients over the age of 40. There is loss of lateral rotation, which automatically limits abduction. Active exercises will usually loosen the joint. They are practised vigorously, bearing in mind that full abduction is not possible until lateral rotation has been regained. Manipulation under anaesthesia or arthroscopic capsular release is advised only if progress has halted and at least 6 months have elapsed since injury.

Unreduced dislocation Surprisingly, a dislocation of the shoulder sometimes remains undiagnosed. This is more likely if the patient is either unconscious or very elderly. Closed reduction is worth attempting up to 6 weeks after injury; manipulation later may fracture the bone or tear vessels or nerves. Operative reduction is indicated after 6 weeks only in the young, because it is difficult, dangerous and followed by prolonged stiffness. An anterior approach is used, and the vessels and nerves are carefully identified before the dislocation is reduced. 'Active neglect' summarizes the treatment of unreduced dislocation in the elderly. The dislocation is disregarded and gentle active movements are encouraged. Moderately good function is often regained.

Recurrent dislocation If an anterior dislocation tears the shoulder capsule, repair occurs spontaneously following reduction and the dislocation may not recur; but if the glenoid labrum is detached, or the capsule is stripped off the front of the neck of the glenoid, repair is less likely and recurrence is more common. Detachment of the labrum occurs particularly in young patients, and, if at injury a bony defect has been gouged out of the posterolateral aspect of the humeral head, recurrence is even more likely. In older patients, especially if there is a rotator cuff tear or greater tuberosity fracture, recurrent dislocation is unlikely. The period of postoperative immobilization makes no difference.

The history is diagnostic. The patient complains that the shoulder dislocates with relatively trivial everyday actions. Often he can reduce the dislocation himself. Any doubt as to diagnosis is quickly resolved by the *apprehension test*: if the patient's arm is passively placed behind the coronal plane in a position of abduction and lateral rotation, his immediate resistance and apprehension are pathognomonic. An AP X-ray with the shoulder medially rotated may show an indentation in the back of the humeral head (the Hill–Sachs lesion) (Figure 24.12).

Even more common, but less readily diagnosed, is recurrent subluxation. The management of both types of instability is dealt with in Chapter 13.

Posterior dislocation of the shoulder

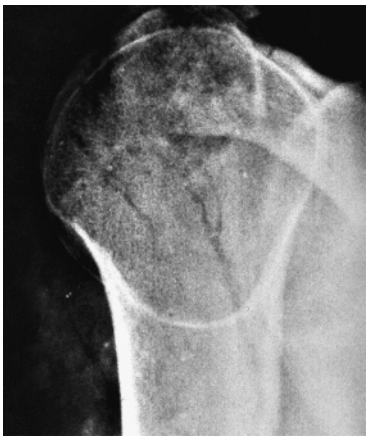
Posterior dislocation is rare, accounting for less than 2% of all dislocations around the shoulder.

Mechanism of injury

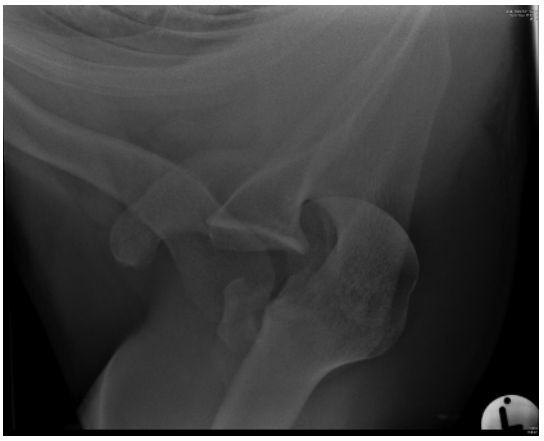
Indirect force producing marked internal rotation and adduction needs be very severe to cause a dislocation. This happens most commonly during a fit or convulsion, or with an electric shock. Posterior dislocation can also follow a fall on the flexed, adducted arm, a direct blow to the front of the shoulder or a fall on the outstretched hand.

Clinical features

The diagnosis is frequently missed – partly because reliance is placed on a single AP X-ray (which may look almost normal) and partly because those attending to the patient fail to think of it. There are, in fact, several well-marked clinical features. The arm is held in internal rotation and is locked in that position. The front of the shoulder looks flat with a prominent coracoid, but swelling may obscure this deformity; seen from above, however, the posterior displacement is usually apparent.



(a)



(b)

Figure 24.13 Posterior dislocation of the shoulder (a) The characteristic X-ray image. Because the head of the humerus is internally rotated, the anteroposterior X-ray shows a head-on projection giving the classic 'electric light-bulb' appearance. (b) Locked (unreduced) posterior dislocation.

Imaging

On AP X-rays the humeral head, because it is medially rotated, looks abnormal in shape (like an electric light bulb) and it stands away somewhat from the glenoid fossa (the 'empty glenoid' sign) (Figure 24.13). A lateral film and axillary view is essential; it shows posterior subluxation or dislocation and sometimes a deep indentation on the anterior aspect of the humeral head. Posterior dislocation is sometimes complicated by fractures of the humeral neck, posterior glenoid rim or lesser tuberosity. Sometimes the patient is too uncomfortable to permit adequate imaging and in these difficult cases *CT* is essential to rule out posterior dislocation of the shoulder.

Treatment

The acute dislocation is reduced (usually under general anaesthesia) by pulling on the arm with the shoulder in adduction; a few minutes are allowed for the head of the humerus to disengage and the arm is then gently rotated laterally while the humeral head is pushed forwards. If reduction feels stable, the arm is immobilized in a sling; otherwise the shoulder is held widely abducted and laterally rotated in an airplane-type splint for 3–6 weeks to allow the posterior capsule to heal in the shortest position. Shoulder movement is regained by active exercises.

Complications

Unreduced dislocation At least half of patients with posterior dislocation have 'unreduced' lesions when first seen. Sometimes weeks or months elapse before the diagnosis is made and up to two-thirds of posterior dislocations are not recognized initially. Typically patients hold the arm internally rotated; they cannot abduct the arm more than 70–80 degrees and, if they lift the extended arm forwards, they cannot then turn the palm upwards. If the patient is young, or is uncomfortable and the dislocation fairly recent, open reduction is indicated. This is a difficult procedure. It is generally done through a deltopectoral approach; the shoulder is reduced and the defect in the humeral head can then be treated by transferring the subscapularis tendon into the defect (McLaughlin procedure). Alternatively, the defect on the humeral head can be bone grafted. A useful technique for treating a defect smaller than 40% of the humeral head is to transfer of the lesser tuberosity together with the subscapularis into the defect. For defects larger than this some form of joint resurfacing may be considered.

Late dislocations, especially in the elderly, are best left, but movement is encouraged.

Recurrent dislocation or subluxation Chronic posterior instability of the shoulder is discussed in Chapter 13.

Inferior dislocation of the shoulder (*luxatio erecta*)

Inferior dislocation is rare but it demands early recognition because the consequences are potentially very serious. Dislocation occurs with the arm in nearly full abduction/elevation. The humeral head is levered out of its socket and lies in the axilla; the arm remains fixed in abduction.

Mechanism of injury and pathology

The injury is caused by a severe hyper-abduction force. With the humerus as the lever and the acromion as the fulcrum, the humeral head is lifted across the inferior rim of the glenoid socket; it remains in the subglenoid position, with the humeral shaft pointing upwards (Figure 24.14). Soft-tissue injury may be severe and includes avulsion of the capsule and surrounding tendons, rupture of muscles, fractures of the glenoid or proximal humerus and damage to the brachial plexus and axillary artery.

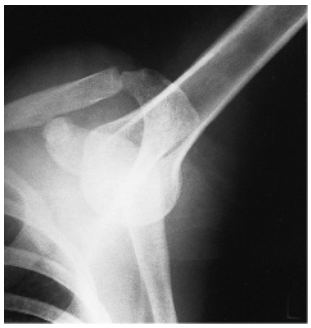


Figure 24.14 Inferior dislocation of the shoulder You can see why the condition is called *luxatio erecta*. The shaft of the humerus points upwards and the humeral head is displaced downwards.

Clinical features

The startling picture of a patient with his arm locked in almost full abduction should make diagnosis quite easy. The head of the humerus may be felt in or below the axilla. Always examine for neurovascular damage.

X-rays

The humeral shaft is shown in the abducted position with the head sitting below the glenoid. It is important to search for associated fractures of the glenoid or proximal humerus.

NOTE: True inferior dislocation must not be confused with postural downward displacement of the humerus, which results quite commonly from weakness and laxity of the muscles around the shoulder, especially after trauma and shoulder splintage; here the shaft of the humerus lies in the normal anatomical position at the side of the chest. The condition is harmless and resolves as muscle tone is regained.

Treatment

Inferior dislocation can usually be reduced by pulling upwards in the line of the abducted arm, with countertraction downwards over the top of the shoulder. If the humeral head is stuck in the soft tissues, open reduction is needed. It is important to examine again, after reduction, for evidence of neurovascular injury.

The arm is rested in a sling until pain subsides and movement is then allowed, but avoiding abduction for 3 weeks to allow the soft tissues to heal.

Shoulder dislocation in children

Traumatic dislocation of the shoulder is exceedingly rare in children. Children who give a history of the shoulder 'slipping out' almost invariably have either voluntary or involuntary (atraumatic) dislocation or subluxation. With voluntary dislocation, the child can demonstrate the instability at will. With involuntary dislocation, the shoulder slips out unexpectedly during everyday activities. Most of these children have generalized joint laxity and some have glenoid dysplasia or muscle patterning disorders (Chapter 13). Examination may show that the shoulder subluxates in almost any direction; X-rays may confirm the diagnosis.

Treatment

Atraumatic dislocation should be viewed with great caution. Some of these children have behavioural or muscle patterning problems and this is where treatment should be directed. A prolonged exercise programme may also help. Only if the child is genuinely distressed by the disorder, and provided psychological factors have been excluded, should one consider reconstructive surgery.

FRACTURES OF THE PROXIMAL HUMERUS

Fractures of the proximal humerus usually occur after middle age and most of the patients are osteoporotic, postmenopausal women. Fracture displacement is usually not marked and treatment presents few problems. However, in about 20% of cases there is considerable displacement of one or more fragments and a significant risk of complications due to bone fragility, damage to the rotator cuff and the prevailing comorbidities. Deciding between operative and non-operative treatment can be very difficult.

Mechanism of injury

Fracture usually follows a fall on the outstretched arm – the type of injury which, in younger people, might cause dislocation of the shoulder. Sometimes, indeed, there is both a fracture and a dislocation.

Classification and pathological anatomy

The most widely accepted classification is that proposed by Neer in 1970 who drew attention to the four major segments involved in these injuries: the head of the humerus, the lesser tuberosity, the greater tuberosity and the shaft (Figure 24.15). Neer's classification distinguishes between the number of displaced fragments, with displacement defined as greater than 45 degrees of angulation or 1 cm of separation. Thus, however many fracture lines there are, if the fragments are undisplaced, it is regarded as a one-part fracture; if one segment is separated from the others, it is a two-part fracture; if two fragments are displaced, that is a three-part fracture; if all the major parts are displaced, it is a four-part fracture. Furthermore, a fracture-dislocation exists when the head is dislocated

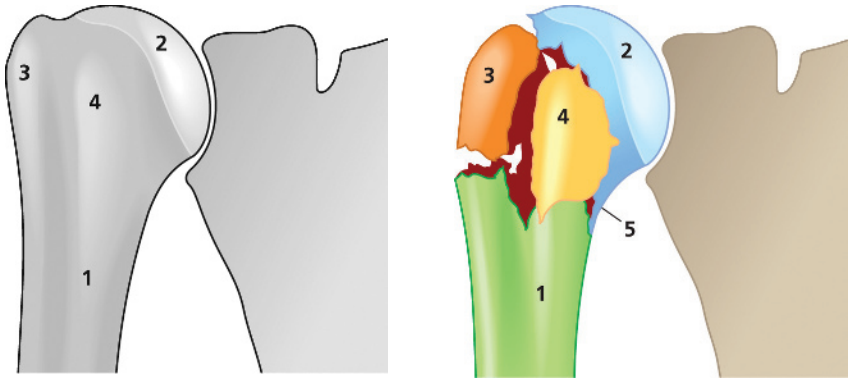


Figure 24.15 Fractures of the proximal humerus Diagram of (a) the normal and (b) a fractured proximal humerus, showing the four main fragments, two or more of which are seen in almost all proximal humeral fractures. 1 shaft of humerus; 2 head of humerus; 3 greater tuberosity; 4 lesser tuberosity. In this figure there is a sizeable medial calcar spike, 5, suggesting a low risk of avascular necrosis.

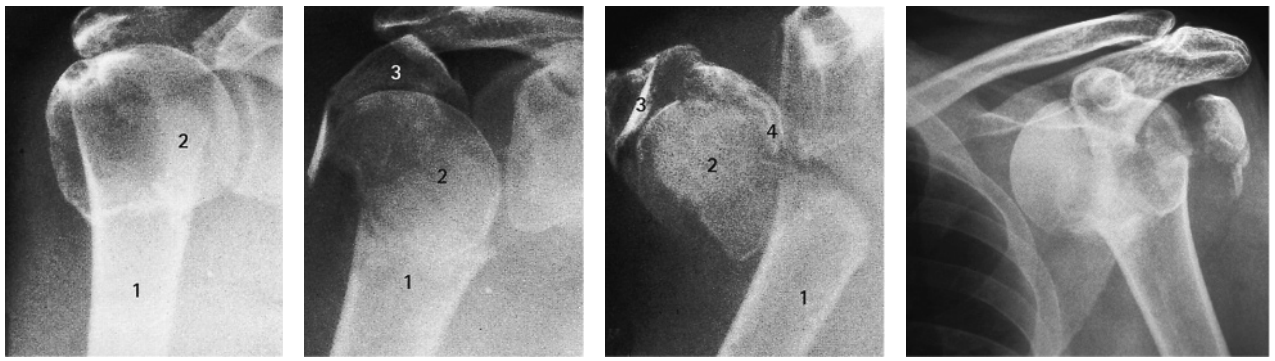


Figure 24.16 X-rays of proximal humeral fractures Classification is all very well, but X-rays are more difficult to interpret than line drawings. (a) Two-part fracture. (b) Three-part fracture involving the neck and the greater tuberosity. (c) Four-part fracture. (1 shaft of humerus; 2 head of humerus; 3 greater tuberosity; 4 lesser tuberosity). (d) X-ray showing fracture dislocation of the shoulder.

and there are two, three or four parts. This grading is based on X-ray appearances, although observers do not always agree with each other on which class a particular fracture falls into (Figure 24.16).

Clinical features

Because the fracture is often firmly impacted, pain may not be severe. However, the appearance of a large bruise on the upper part of the arm is suspicious. Signs of axillary nerve or brachial plexus injury should be sought.

Imaging

In elderly patients there often appears to be a single, impacted fracture extending across the surgical neck. However, with good *X-rays*, several undisplaced fragments may be seen. In younger patients, the fragments are usually more clearly separated. Axillary and scapular-lateral views should always be obtained, to exclude dislocation of the shoulder.

It has always been difficult to apply Neer's classification when based on plain X-rays, and not surprisingly there is a relatively high level of both inter- and intraobserver disagreement. Neer himself later noted that, when this classification was developed, the criteria for displacement (distance >1 cm, angulation >45 degrees) were set arbitrarily. The classification was not intended to dictate treatment, but simply to help clarify the pathoanatomy of the different fracture patterns.

The advent of *3D CT reconstruction* has helped to reduce the degree of inter- and intraobserver error, enabling better planning of treatment than in the past (Figure 24.17).

As the fracture heals, the humeral head is sometimes seen to be subluxated downwards (inferiorly); this is due to muscle atony and it usually recovers once exercises are begun.

Treatment

MINIMALLY DISPLACED FRACTURES

These comprise the vast majority. They need no treatment apart from period of a week or two of rest with the arm in a sling until the pain subsides, and then gentle passive movements of the shoulder. Once the fracture has united (usually after 6 weeks), active exercises are encouraged; the elbow and hand are, of course, actively exercised from the start.

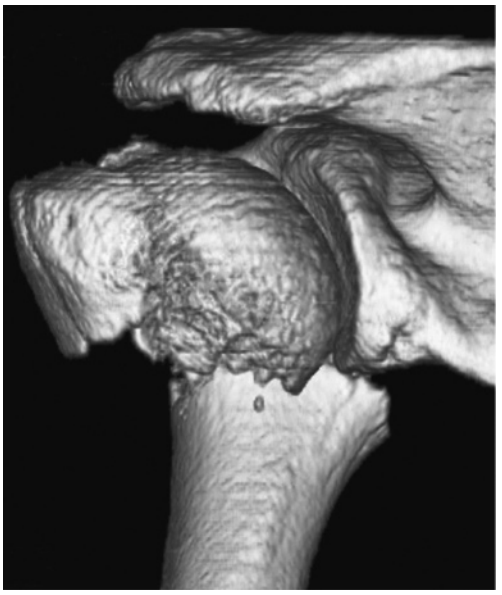


Figure 24.17 CT with three-dimensional reconstruction
Advanced imaging provides a much clearer picture of the injury, allowing better pre-operative planning.

TWO-PART FRACTURES

Surgical neck fractures The fragments are gently manipulated into alignment and the arm is immobilized in a sling for about 4 weeks or until the fracture feels stable and the X-rays show some signs of healing. Elbow and hand exercises are encouraged throughout this period; shoulder exercises are commenced at about 4 weeks. The results of conservative treatment are generally satisfactory, considering that most of these patients are over 65 and do not demand perfect function. If the fracture cannot be reduced closed or if the fracture is very unstable after closed reduction, then fixation is required. Options include percutaneous pins, bone sutures, intramedullary pins with tension band wiring or a locked intramedullary nail. Plate fixation requires a wider exposure and the newer locking plates offer a stable fixation without the need for extensive periosteal stripping.

Greater tuberosity fractures Fracture of the greater tuberosity is often associated with anterior dislocation and it reduces to a good position when the shoulder is relocated. If it does not reduce, the fragment can be reattached through a small incision with interosseous sutures or, in young hard bone, cancellous screws.

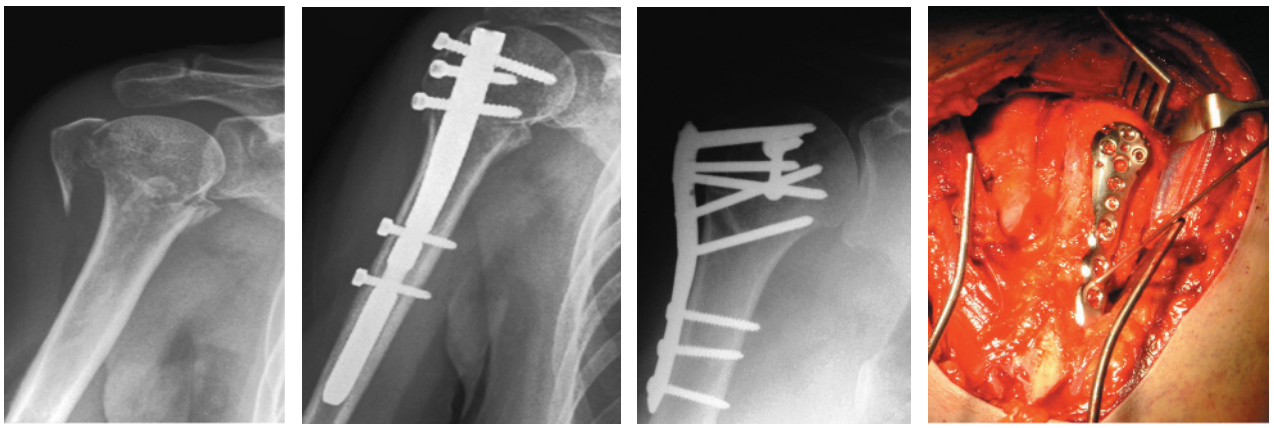
Anatomical neck fractures These are very rare. In young patients the fracture should be fixed. In older patients prosthetic replacement (hemiarthroplasty) is preferable because of the high risk of avascular necrosis of the humeral head.

THREE-PART FRACTURES

These usually involve displacement of the surgical neck and the greater tuberosity; they are extremely difficult to reduce closed. In active individuals this injury is best managed by open reduction and internal fixation (Figure 24.18a,b). There is little evidence that one technique is better than another although the newer implants with locked plating and nailing are biomechanically superior in osteoporotic bone.

FOUR-PART FRACTURES

The surgical neck and both tuberosities are displaced. These are severe injuries with a high risk of complications such as vascular injury, brachial plexus damage, injuries of the chest wall and (later) avascular necrosis of the humeral head. The X-ray diagnosis is difficult (how many fragments are there, and are they displaced?). Often the most one can say is that there are 'multiple displaced fragments', sometimes together with glenohumeral dislocation. In young patients an attempt should be made at reconstruction (Figure 24.18c,d).



(a) (b) (c) (d)

Figure 24.18 Proximal humerus fractures – treatment (a) Three-part fracture, treated by (b) locked nail fixation. (c) Four-part fracture fixed with a locked plate; the intraoperative picture (d) shows how the plate was positioned.

In older patients, closed treatment and attempts at open reduction and fixation can result in continuing pain and stiffness and additional surgical treatment can compromise the blood supply still further. If the fracture pattern is such that the blood supply is likely to be compromised, or that reconstruction and internal fixation will be extremely difficult, then the treatment of choice is prosthetic replacement of the proximal humerus.

The results of hemiarthroplasty are somewhat unpredictable. Anatomical reduction, fixation and healing of the tuberosities are prerequisites for a satisfactory outcome; even then, secondary displacement of the tuberosities may result in a poor functional outcome. In addition the prosthetic implant should be perfectly positioned. Be warned – these are operations for the expert.

More recently the reverse shoulder replacements have been used in these fractures although the long-term outcomes are not yet known. Decision making in all of these fractures remains difficult and current evidence remains unclear whether surgical management actually improves the overall outcome in these serious fractures.

FRACTURE-DISLOCATIONS

Two-part fracture-dislocations (greater tuberosity with anterior dislocation and lesser tuberosity with posterior) can usually be reduced by closed means.

Three-part fracture-dislocations, when the surgical neck is also broken, usually require open reduction and fixation; the brachial plexus is at particular risk during this operation.

Four-part fracture-dislocations have a poor prognosis; prosthetic replacement is recommended in all but young and very active patients.

Complications

Vascular injuries and nerve injuries The patient should always be carefully assessed for signs of vascular and nerve injuries, both at the initial examination and again after any operation. The axillary nerve is at particular risk, both from the injury and from surgery.

Avascular necrosis The reported incidence of avascular necrosis (AVN) of the humeral head is in the range of 10–30% in three-part fractures and 10–50+% in four-part fractures. The ability to predict the likelihood of this outcome is important in making the choice between internal fixation and hemiarthroplasty for complex fractures.

The blood supply of the humeral head is provided mainly by the anterior circumflex artery and its ascending branch (the arcuate artery) which penetrates into the humeral head and arches across subchondrally. Additional blood supply is provided by vessels entering the posteromedial aspect of the proximal humerus, metaphyseal vessels and vessels of the greater and lesser tuberosities that anastomose with the intraosseous arcuate artery. Thus, in three- and four-part fractures with the only supply coming from the posteromedial vessels, there may still be sufficient perfusion of the humeral head if the head fragment includes a sizeable part of the calcar on the medial side of the anatomical neck. Fractures at the anatomical neck with a medial metaphyseal (calcar) spike shorter than 8 mm carry a high risk of developing humeral head avascular necrosis (see Figure 24.15). Disruption of the medial periosteal hinge is another predictor of avascular necrosis and the presence of these two factors combined has a positive predictive value of 98% for avascular necrosis of the humeral head. Contrariwise, fractures with an intact medial hinge and/or a large posteromedial metaphyseal spike carry a much better prognosis. The mere number of fracture parts, their degree of displacement and split-head fractures are rated as poor predictors of avascular necrosis, as is the presence of dislocation.

Stiffness of the shoulder This is a common complication, particularly in elderly patients. Unlike a frozen shoulder, the stiffness is maximal at the outset. It can be prevented, or at least minimized, by starting exercises early.

Malunion Malunion usually causes little disability, but loss of rotation may make it difficult for the patient to reach behind the neck or up the back.

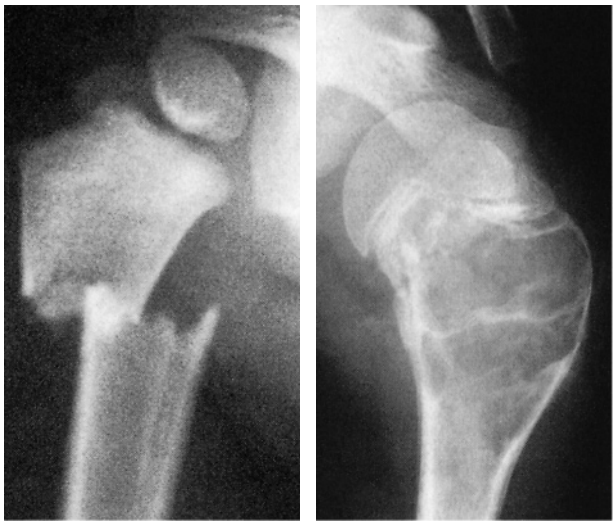
FRACTURES OF THE PROXIMAL HUMERUS IN CHILDREN

At birth, the shoulder is sometimes dislocated or the proximal humerus fractured. Diagnosis is difficult and a clavicular fracture or brachial plexus injury should also be considered.

In infancy, the physis can separate (Salter–Harris I); reduction does not have to be perfect and a good outcome is usual.

In older children, metaphyseal fractures or type II physeal fractures occur (Figure 24.19). Considerable displacement and angulation can be accepted; because of the marked growth and remodelling potential of the proximal humerus, malunion is readily compensated for during the remaining growth period.

Pathological fractures are not unusual, as the proximal humerus is a common site of bone cysts and tumours in children. Fracture through a simple cyst usually unites and the cyst often heals spontaneously;



(a) (b)

Figure 24.19 Fractures of the proximal humerus in children (a) The typical metaphyseal fracture. Reduction need not be perfect as remodelling will compensate for malunion. (b) Fracture through a benign cyst.

all that is needed is to rest the arm in a sling for 4–6 weeks. Other lesions require treatment in their own right (see Chapter 9).

FRACTURED SHAFT OF HUMERUS

Mechanism of injury

A fall on the hand may twist the humerus, causing a spiral fracture. A fall on the elbow with the arm abducted exerts a bending force, resulting in an oblique or transverse fracture. A direct blow to the arm causes a fracture which is either transverse or comminuted. Fracture of the shaft in an elderly patient may be due to a metastasis.

Pathological anatomy

With fractures above the deltoid insertion, the proximal fragment is adducted by pectoralis major. With fractures lower down, the proximal fragment is abducted by the deltoid. Injury to the radial nerve is common, though fortunately recovery is usual.

Clinical features

The arm is painful, bruised and swollen. It is important to test for radial nerve function before and after treatment. This is best done by assessing active extension of the metacarpophalangeal joints; active extension of the wrist can be misleading because extensor carpi radialis longus is sometimes supplied by a branch arising proximal to the injury.

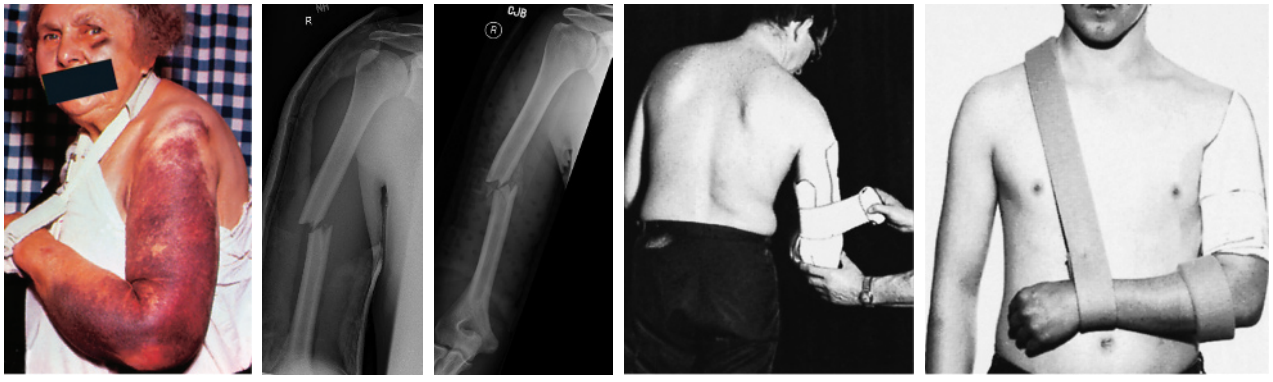
X-rays

The site of the fracture, its line (transverse, spiral or comminuted) and any displacement are readily seen. The possibility that the fracture may be pathological should be remembered.

Treatment

NON-OPERATIVE TREATMENT

Fractures of the humerus heal readily. They require neither perfect reduction nor immobilization; the weight of the arm with an external cast is usually enough to pull the fragments into alignment. A 'hanging cast' is applied from shoulder to wrist with the elbow flexed 90 degrees, and the forearm section is suspended by a sling around the patient's neck. This cast may be replaced after 2–3 weeks by a short (shoulder to elbow) cast or a functional polypropylene brace which is worn for a further 6 weeks (Figure 24.20).



(a) (b) (c) (d) (e)

Figure 24.20 Fractured shaft of humerus (a) Bruising is always extensive. (b,c) Closed transverse fracture with moderate displacement. (d) Applying a U-slab of plaster (after a few days in a shoulder-to-wrist hanging cast) is usually adequate. (e) Ready-made braces are simpler and more comfortable, though not suitable for all cases. These conservative methods demand careful supervision if excessive angulation and malunion are to be prevented.

The wrist and fingers are exercised from the start. Pendulum exercises of the shoulder are begun within a week, but active abduction is postponed until the fracture has united (about 6 weeks for spiral fractures but often twice as long for other types); once united, only a sling is needed until the fracture is consolidated.

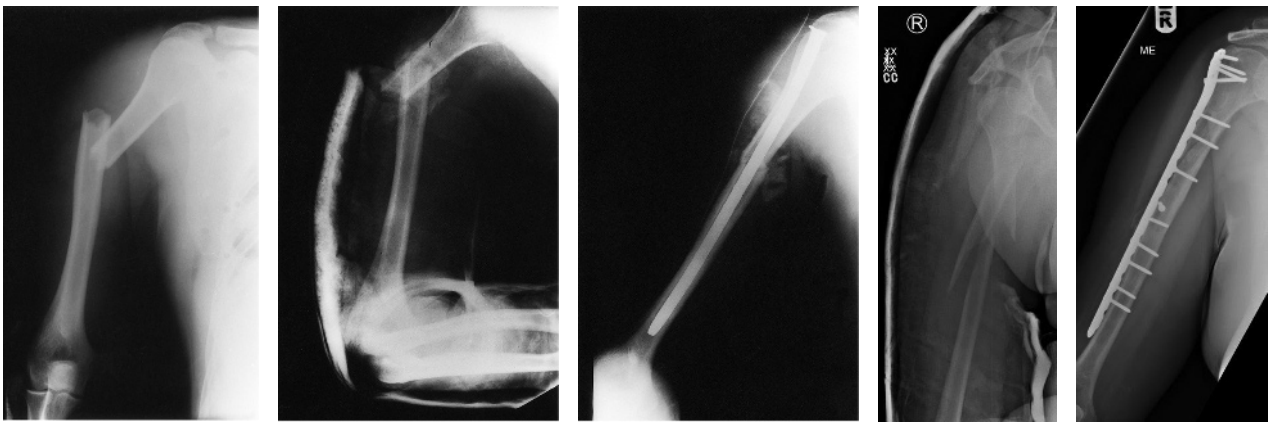
OPERATIVE TREATMENT

Patients often find the hanging cast uncomfortable, tedious and frustrating; they can feel the fragments moving and that is sometimes quite distressing. The temptation is to 'do something', and the 'something' usually means an operation. It is as well to remember (1) that the complication rate after internal fixation of the humerus is high (2) that the great majority of humeral fractures unite with non-operative treatment and (3) there is no good evidence that the union rate is higher with fixation (and the rate may be lower if there is distraction with nailing or periosteal stripping with plating). There are, nevertheless, some well-defined indications for surgery:

- severe multiple injuries
- an open fracture
- segmental fractures
- displaced intra-articular extension of the fracture
- a pathological fracture
- a 'floating elbow' (simultaneous unstable humeral and forearm fractures)
- radial nerve palsy after manipulation
- non-union
- problems with nursing care in a dependent person.

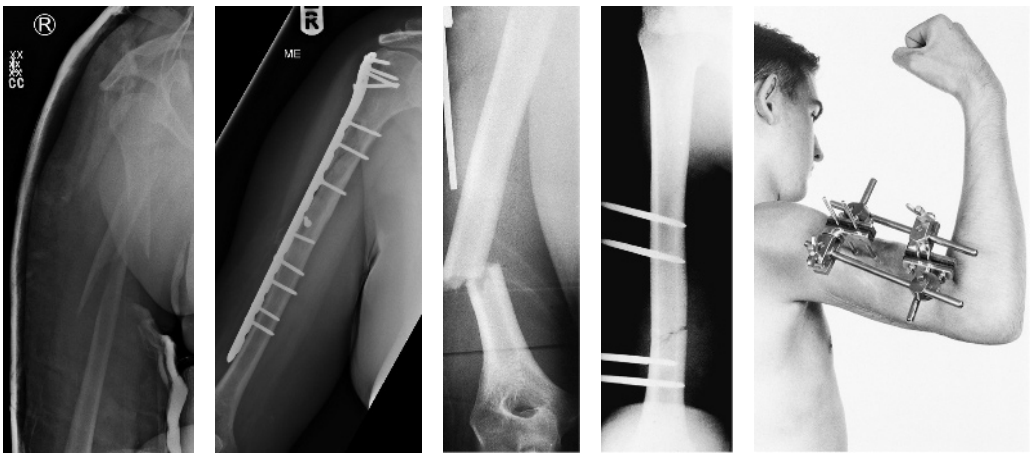
Fixation can be achieved with: (1) a compression plate and screws; (2) an interlocking intramedullary nail or semi-flexible pins or (3) an external fixator (Figures 24.21 and 24.22).

Plating permits excellent reduction and fixation, and it has the added advantage that it does not interfere with shoulder or elbow function. However, it



(a) (b) (c) (d) (e)

Figure 24.21 Fractured shaft of humerus – treatment (a,b) Most shaft fractures can be treated in a hanging cast or functional brace, but beware the upper third fracture which tends to angulate at the proximal border of a short cast. This fracture would have been better managed by (c) intramedullary nailing or open reduction and internal fixation. (d) Segmental fracture of the humerus. (e) Open reduction and internal fixation.



(a) (b) (c) (d) (e)

Figure 24.22 Fractured humerus – other methods of fixation (a,b) Compression plating, and (c,d,e) external fixation.

requires wide dissection and the radial nerve must be protected. Too much periosteal stripping or inadequate fixation will probably increase the risk of non-union.

Antegrade nailing is performed with a rigid interlocking nail inserted through the rotator cuff under fluoroscopic control. It requires minimal dissection but has the disadvantage that it causes rotator cuff problems in a significant proportion of cases (the reported incidence is 5–40%). The nail can also distract the fracture which will inhibit union; if this happens, exchange nailing and bone grafting of the fracture may be needed.

Retrograde nailing with multiple flexible rods is not entirely stable. Retrograde nailing with an interlocking nail is suitable for some fractures of the middle third.

External fixation can be an option for high-energy segmental fractures and open fractures, but great care must be taken in placing the pins as the radial nerve is vulnerable.

Complications

EARLY

Vascular injury If there are signs of vascular insufficiency in the limb, brachial artery damage must be excluded. Angiography will show the level of the injury. This is an emergency, requiring exploration and either direct repair or grafting of the vessel. In these circumstances, internal fixation is advisable.

Nerve injury Radial nerve palsy (wrist drop and paralysis of the metacarpophalangeal extensors) may occur with shaft fractures, particularly oblique fractures at the junction of the middle and distal thirds of the bone (Holstein–Lewis fracture). If nerve function was intact before manipulation but is defective afterwards, it must be assumed that the nerve has been snagged and surgical exploration is necessary. Otherwise, in closed injuries the nerve is very seldom divided, so there is no hurry to operate as it will usually recover. The wrist and hand must be regularly moved through a full passive range of movement to preserve joint motion until the nerve recovers. If there is no sign of recovery by 12 weeks, the nerve should be explored. It may just need a neurolysis but, if there is loss of continuity of normal-looking nerve, a graft is needed. The results are often satisfactory but, if necessary, function can be largely restored by tendon transfers (see Chapter 11).

LATE

Delayed union and non-union Transverse fractures sometimes take months to unite, especially if excessive traction has been used (a hanging cast must not be too heavy). Simple adjustments in technique may solve the problem; as long as there are signs of callus formation it is worth persevering with non-operative treatment, but remember to keep the shoulder moving. The rate of non-union in conservatively treated low-energy fractures is less than 3%. Segmental high-energy fractures and open fractures are more prone to both delayed union and non-union.

Intramedullary nailing may contribute to delayed union but, if rigid fixation can be maintained (if necessary by exchange nailing), the rate of non-union can probably be kept below 10%.

A particularly vicious combination is incomplete union and a stiff joint. If elbow or shoulder movements are forced before consolidation of the fracture, or if an intramedullary nail is removed too soon (e.g. because of shoulder problems), the humerus may refracture and non-union is then more likely.

The treatment of established non-union is operative. The bone ends are freshened, cancellous bone graft is packed around them and the reduction is held with an intramedullary nail or a compression plate.

Joint stiffness Joint stiffness is common. It can be minimized by early activity, but transverse fractures (in which shoulder abduction is ill-advised) may limit shoulder movement for several weeks.

Special features in children

Fractures of the humerus are uncommon; in children under 3 years of age the possibility of child abuse should be considered and tactful examination for other injuries performed.

Taking advantage of the robust periosteum and the power of rapid healing in children, the humeral fracture can usually be treated by applying a collar and cuff bandage for 3–4 weeks. If there is gross shortening, manipulation may be needed. Older children may require a short plaster splint.

Injuries of the elbow and forearm

25

Adam Watts & David Warwick
Children's sections: Mike Uglow & Joanna Thomas

FRACTURES OF THE DISTAL HUMERUS IN ADULTS

In adults, fractures around the elbow, especially those of the distal humerus, are often high-energy injuries but an increasing number of comminuted osteoporotic fractures are now seen. High-energy injuries may be associated with vascular or nerve damage or breech of the soft-tissue envelope. Some can be reduced and stabilized only by complex surgical techniques; and the tendency to stiffness of the elbow is a constant challenge, best met by prevention through early stabilization and mobilization.

The AO-ASIF Group has defined three types of distal humeral fracture:

- *Type A* – extra-articular supracondylar fracture
- *Type B* – intra-articular unicondylar fracture (one condyle sheared off)
- *Type C* – bicondylar fracture with varying degree of comminution.

Type A – supracondylar fractures

These extra-articular fractures are rare in adults. When they do occur, they are usually displaced and unstable – probably because there is no tough periosteum to tether the fragments. In high-energy injuries there may be comminution of the distal humerus.

Treatment

Closed reduction is unlikely to be stable and K-wire fixation is not strong enough to permit early mobilization. Open reduction and internal fixation is therefore the treatment of choice. The distal humerus is approached through a posterior exposure. It is sometimes possible to fix the fracture without recourse to an olecranon osteotomy using a triceps elevating approach. A simple transverse or oblique fracture can usually be reduced and fixed with a medial and lateral contoured plate and screws.

Types B and C – intra-articular fractures

Except in osteoporotic individuals, intra-articular condylar fractures should be regarded as high-energy injuries with soft-tissue damage. A severe blow on the point of the elbow drives the olecranon process upwards, splitting the condyles apart. Swelling is considerable but, if the bony landmarks can be felt, the elbow is found to be distorted. The patient should be carefully examined for evidence of vascular or nerve injury; if there are signs of vascular insufficiency, this must be addressed as a matter of urgency.

Imaging

X-rays show that the fracture extends from the lower humerus into the elbow joint; it may be difficult to tell whether one or both condyles are involved, especially with an undisplaced condylar fracture. There is also often comminution of the bone between the condyles, the extent of which is usually underestimated. Sometimes the fracture extends into the metaphysis as a T- or Y-shaped break, or else there may be multiple fragments (comminution). *CT scans* can be helpful in planning the surgical approach but the surgeon should be prepared for the worst case.

Treatment

These are severe injuries associated with joint damage; prolonged immobilization will certainly result in a stiff elbow. Early movement is therefore a prime objective.

Undisplaced fractures These can be treated by applying a posterior slab with the elbow flexed almost

90 degrees; movements are commenced after 2 weeks. However, great care should be taken to avoid the dual pitfalls of underdiagnosis (displacement and comminution are not always obvious on the initial X-rays) and late displacement (always obtain check X-rays a week after injury).

Displaced type B and C fractures If the appropriate expertise and facilities are available, open reduction and internal fixation is the treatment of choice for displaced fractures and for most undisplaced fractures in adults (Figure 25.1). Minor displacement and comminution may be underappreciated and can lead to displacement. The danger with conservative treatment is the strong tendency to stiffening of the elbow and persistent pain.

Bridging external fixation can be used for the initial management of open fractures with soft-tissue contamination. This allows for eradication of contaminants prior to staged early definitive treatment. If there is substantial soft-tissue loss that is likely to require involvement of a plastic surgeon, they should be consulted prior to application of the external fixator as this may substantially reduce the reconstructive options.

If the articular involvement is minimal, a triceps-preserving approach can be used to access the humerus. For more comminuted fractures a good exposure of the joint is needed. This may require an olecranon osteotomy that can be intra- or extra-articular. The ulnar nerve should be identified, decompressed and protected throughout; some favour transposition in all cases. The fragments are reduced and held temporarily with K-wires. In adults the use of plates and screws is preferred over lag screws or cannulated screws, even for unicondylar fractures. Parallel or orthogonal plates are used depending on the fracture configuration of the lateral column. Pre-contoured locking plates are available that help maintain position in osteoporotic bone. Independent lag screws or headless compression screws may be required for coronal plane fractures but otherwise it is preferable for transverse screws to pass through a plate to engage fragments on the opposite side.

Postoperatively the patient is provided with a sling for comfort but immediate active mobilization is initiated with the patient lying supine and the shoulder flexed to 90 degrees. The use of a splint or cast is not recommended and passive stretch should be avoided. Fracture healing usually occurs by 12 weeks. Despite these measures the patient often does not regain full extension and they should be counselled accordingly before surgery. In some cases movement may be severely restricted. A description of this sort fails to convey the real difficulty of these operations, which can provide a real challenge even in the most skilled hands.

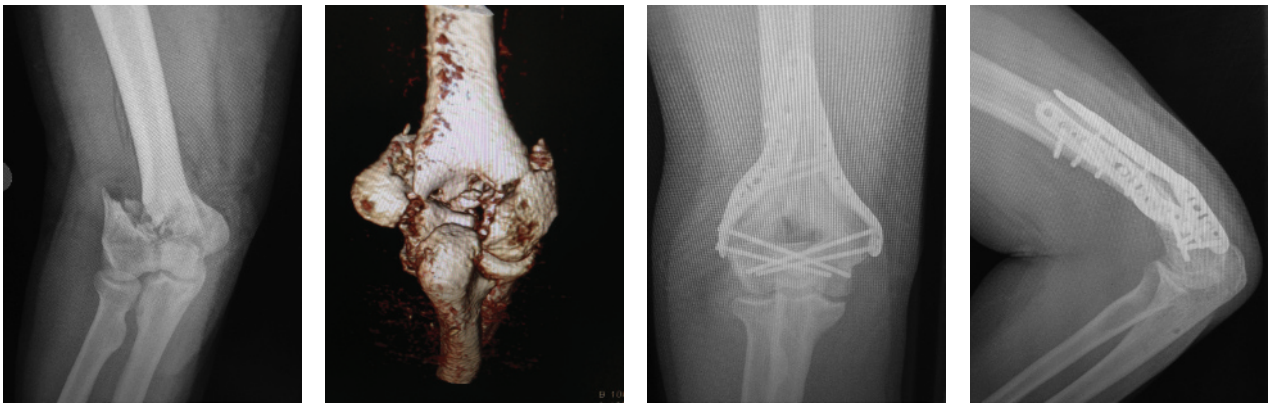
ALTERNATIVE METHODS OF TREATMENT

If it is anticipated that the outcome of osteosynthesis will be poor (because of the degree of comminution, bone quality, soft-tissue damage or patient compliance), other options can be considered.

Elbow hemiarthroplasty Replacement of the distal humerus alone is finding an increasing role for the treatment of very comminuted fractures in elderly osteoporotic patients as it avoids additional surgery to the ulna, the part of a total elbow replacement that tends to fail first.

Total elbow replacement Total joint replacement is an option for unreconstructable distal humerus fractures and in particular those with pre-existing joint disease.

The 'bag of bones technique' This method is reserved for patients in whom the risks of surgery



(a) (b) (c) (d)

Figure 25.1 Bicondylar fractures X-rays taken (a,b) before and (c,d) after open reduction and internal fixation. An excellent reduction was obtained in this case; however, the elbow sometimes ends up with considerable loss of movement even though the general anatomy has been restored.

outweigh the benefits due to pre-existing medical conditions, or those in whom poor compliance would compromise the outcome of surgery (e.g. patients with severe dementia or with poorly controlled alcoholism). The elbow can be placed in a cast at 90 degrees of flexion or collar and cuff if tolerated for 2–3 weeks to allow initial healing and for the pain to settle. Active range of movement exercises are started as soon as tolerated and continued until gains in range of movement have plateaued. A useful range of movement with a pain-free elbow can result.

Complications

EARLY

Vascular injury This should be excluded by thorough examination of pulses and capillary refill distal to the injury. Vigilance is required to make the diagnosis and institute treatment as early as possible.

Nerve injury This is most commonly to the ulnar nerve but the radial nerve may be injured by a long lateral plate and median nerve injury has been reported. It is important to examine the hand and record the findings before any treatment is commenced.

LATE

Stiffness Stiffness is the most common complication of distal humerus fracture. The causes of elbow stiffness may be intrinsic (intra-articular adhesions, capsular contracture, joint incongruity, instability) or extrinsic (heterotopic ossification or nerve entrapment). Prevention is the best management plan with early restoration of skeletal stability and early active mobilization and avoidance of painful passive stretch.

For those patients who develop a very stiff elbow with unacceptable function, elbow arthrolysis may be indicated. The success of this procedure is dependent on establishing the cause of the stiffness and addressing this directly. The average gain in range of movement is approximately 30 degrees.

FRACTURES OF THE RADIAL HEAD

Radial head fractures are the most common fracture of the elbow in adults. In children radial neck fractures are more common because the head is largely cartilaginous. Radial head fractures may be isolated bony injuries or part of a pattern of fracture dislocation of the elbow, which will be dealt with separately later.

Mechanism of injury

With an isolated radial head fracture the patient reports a fall on the elbow or a fall on an outstretched hand with the elbow extended. The injury may be associated with lateral ligament avulsion and/or medial ligament tear.

Clinical features

This fracture is sometimes missed, but tenderness on pressure over the radial head and pain on pronation and supination should suggest the diagnosis. A firm block to forearm rotation should not be ignored.

Imaging

Three types of fracture are identified and classified by Mason (Figure 25.2):

- *Type I* – undisplaced partial articular fracture of the radial head
- *Type II* – displaced (>2 mm) partial articular fracture of the radial head
- *Type III* – comminuted radial head fracture.

An additional type IV has been proposed, for those fractures with a radiograph showing a dislocated elbow, but this is quite arbitrary as the elbow may have been dislocated or subluxated at the time of injury but spontaneously reduced. The most reliable predictor of elbow instability is loss of cortical contact of the radial head fragment(s).

CT or *MRI* may be indicated to better understand the injury; the associated lateral ligament avulsion

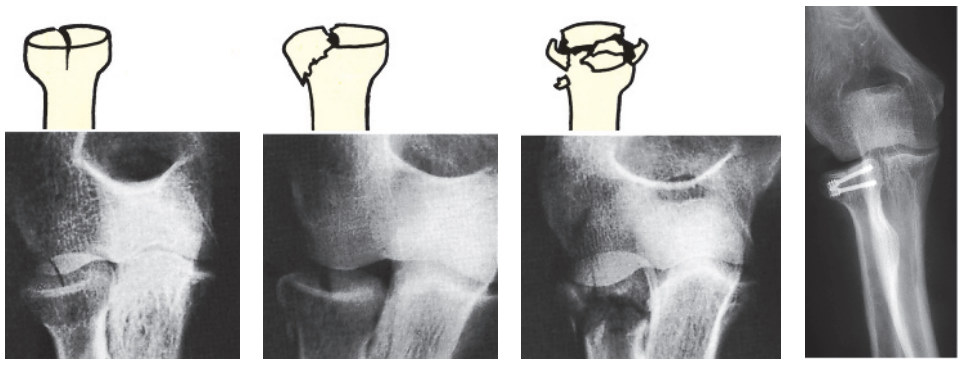


Figure 25.2 Fractured head of radius There are three main types of adult radial head fracture: (a) a chisel-like split of head; (b) a marginal fracture; or (c) a comminuted fracture. Displaced marginal fractures can often be treated by (d) internal fixation.

increases in prevalence with higher grade of injury, from 50% with type I, to 100% for type III.

Treatment

TYPE I

Undisplaced partial articular fractures (or head splits) have a good prognosis with non-surgical management of pain relief and mobilization as comfort allows. Most will regain a pain-free elbow with only a slight loss of extension the most common sequel. Aspiration of the haematoma from the elbow joint and injection of local anaesthetic may provide relief of initial pain but may also increase the risk of infection.

TYPE II

In displaced partial articular fractures, it is important to identify those which are associated with a mechanical block to rotation and those associated with elbow instability. Assessment of those with loss of forearm rotation can be a challenge; is there truly a mechanical block or is there simply too much pain to permit movement? Aspiration and local anaesthetic injection can be used but gentle assessment of passive range of movement or reassessment at 1 week with repeat clinical examination may provide useful information. If there is doubt, a CT or MRI scan will demonstrate the presence of a displaced fragment engaging on the lip of the lesser sigmoid notch that would require open reduction and internal fixation with headless screws. Loss of cortical contact of the radial head fragment or a fragment that is greater than a quarter of the radial head surface can be associated with instability of the elbow. If so, it should be reduced and fixed. In all surgical cases the associated soft-tissue injury (lateral ligament avulsion) should be addressed with suture anchor repair back to the humerus.

TYPE III

A comminuted fracture is challenging and is usually associated with a fracture-dislocation.

FRACTURE-DISLOCATIONS OF THE ELBOW

Successful management of elbow fracture-dislocations depends on correct identification of the pattern of injury, application of a treatment algorithm specific to that pattern and based on anatomical principles, and recognition of the underlying 'hidden' soft-tissue injury that can be inferred from the fracture pattern. Consider that elbow fracture-dislocations occur as a result of forces applied through the forearm that act as a long 'lever arm' to multiply forces across the elbow. These forces can be 'twisting' or rotational forces, bending forces or axial loads.

Injury patterns

ROTATIONAL INJURY

The forearm can twist posterolaterally off the humerus or posteromedially. Each will produce a different pattern of injury. In some cases, despite surgical reconstruction, the elbow is still not stable and an external fixator may have to be applied across the elbow to maintain acceptable alignment.

LATERAL OR EXTERNAL ROTATION INJURY (TERRIBLE TRIAD)

Elbow dislocation with fracture of the radial head, coronoid process and medial collateral ligament rupture is known as a 'terrible triad' injury. The injury is now well understood and no longer earns the 'terrible' title, but as with other elbow injuries stiffness is a common problem. The treatment is usually surgical, with radial head fixation or replacement and lateral ligament repair. Coronoid fixation is required only if the fracture extends to the medial facet, and opinion varies about repair of the medial ligament.

MEDIAL OR INTERNAL ROTATION INJURY (POSTEROMEDIAL FRACTURE-DISLOCATION)

This results in an isolated fracture of the anteromedial facet of the coronoid and lateral ligament complex. Often dismissed as a 'tip of coronoid' fracture, this injury pattern has only been recognized relatively recently. CT will identify the subtle coronoid fracture in a patient who typically reports a fall backwards on the hand. In many cases non-operative management will result in rapid progression to osteoarthritis. Treatment consists of lateral ligament repair with or without fixation of the coronoid fragment, depending on its size.

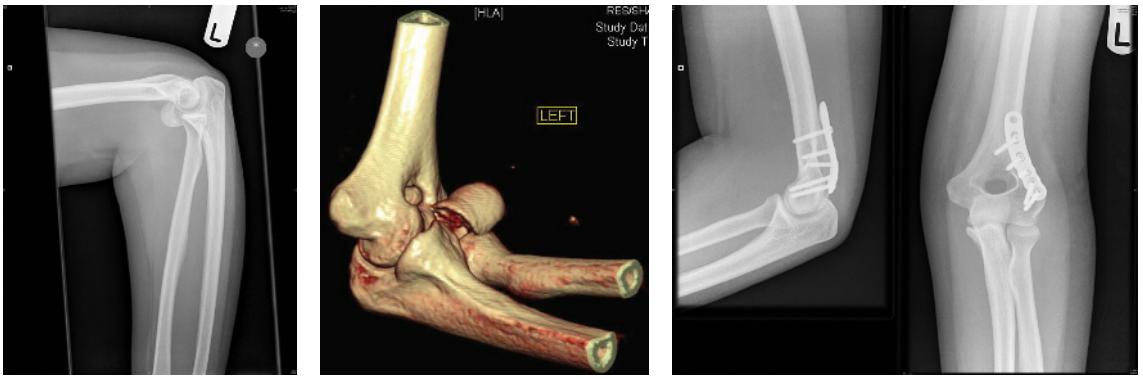
BENDING INJURY

Monteggia fracture-dislocation is a proximal ulna fracture with dislocation of the radial head from the radiocapitellar joint. These can be divided into those with an apex anterior ulna fracture and those with an apex posterior ulna fracture. Apex anterior fractures carry a better prognosis because the radial head is often intact. In apex posterior fractures the radial head is driven into the capitellum, resulting in a comminuted radial head fracture that may be associated with coronoid fracture and ligament injury.

AXIAL LOAD

Axial loads through the forearm from a high-energy fall on the hand result in fracture of the radial head and proximal migration of the radius relative to the ulna. This rare injury is associated with rupture of the central condensation of the interosseous membrane of the forearm, which should be addressed to produce a satisfactory outcome. It is known as the *Essex Lopresti lesion*.

If any of these is present, excision of the radial head is contraindicated; this may lead to intractable instability



(a) (b) (c)

Figure 25.3 Fractured capitellum (a) Lateral X-ray; (b) 3D CT reconstruction; and (c) surgical fixation.

of the elbow or forearm. The head must be meticulously reconstructed with small headless screws or replaced.

Complications

Joint stiffness is common and may involve both the elbow and the radioulnar joints. Even with minimally displaced fractures the elbow can take several months to recover, and stiffness may occur. Heterotopic ossification is an occasional complication that can result in dramatic stiffness. Recurrent instability of the elbow can occur.

FRACTURES OF THE RADIAL NECK

In adults a displaced fracture of the radial neck may need open reduction; if so, a pre-contoured locking plate can be applied, carefully placed to avoid the articular surface during rotation. If undisplaced, non-operative management can produce acceptable results despite a relatively high rate of non-union. Most are asymptomatic.

FRACTURES OF THE CAPITELLUM

This is a rare articular fracture that is usually more extensive than it initially appears, with involvement of the trochlea and posterior humerus common.

Clinical features

The elbow is typically held at around 70 degrees of flexion as this is the most relaxed position of the joint capsule accommodating the haemarthrosis. The lateral side of the elbow is tender. Bruising on the lateral side of the elbow indicates disruption of the superficial fascia and usually indicates a more significant soft-tissue injury.

Imaging

In the lateral *X-ray* view the capitellum is displaced such that the radial head no longer articulates congruently with it, and often it is rotated through 90 degrees to face the shoulder (Figure 25.3). A double arc sign (two crescent shapes on the lateral view) indicates that the fracture extends into the trochlea, which is a more unstable situation than an isolated capitellum fracture. *CT scans* can be helpful in clarifying the diagnosis and extent of the injury. Bryan and Morrey classify these as:

- *Type I* – complete simple fracture of capitellum
- *Type II* – cartilagenous shell
- *Type III* – comminuted fracture.

Treatment

Undisplaced fractures are rare and can be treated with analgesia and a collar and cuff.

Displaced fractures should be reduced and held. While closed reduction is feasible, prolonged immobilization may result in a stiff elbow and therefore open reduction and internal fixation is preferred. The fragment is always larger than expected. If there is no dorsal comminution and good quality bone, one or two headless screws or lag screws can be passed from anterior to posterior to stabilize the fragment. If there is comminution, a dorsal lateral plate can be used with or without a block bone graft, depending on the amount of bone loss. Highly comminuted fractures may have to be excised. Movements are commenced as soon as possible. The long-term outcome may be compromised by stiffness, non-union or instability. Injury to the lateral ligament complex must be addressed acutely.

FRACTURES OF THE OLECRANON

Olecranon fractures can be divided into two broad categories: (1) a simple transverse fracture that may occur as an avulsion due to a fall on an outstretched hand with the triceps contracting; and (2) a comminuted fracture which is due to a direct blow or a fall on the elbow. These can be further sub-classified into

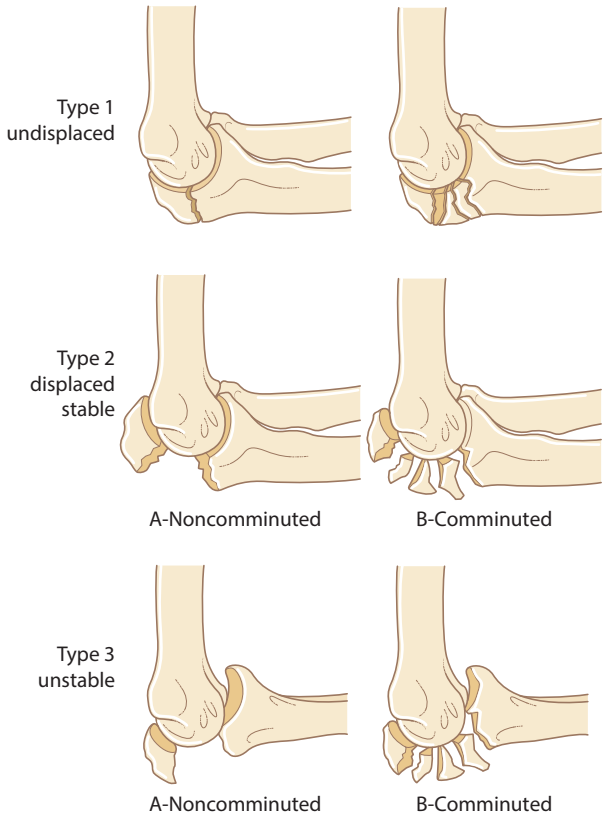


Figure 25.4 Mayo classification of olecranon fractures

(a) displaced or (b) undisplaced, and whether there is subluxation of the ulnohumeral joint, which is largely related to how distal the fracture is on the olecranon (Figure 25.4).

The fracture always enters the elbow joint and therefore may damage the articular cartilage. With transverse fractures the triceps aponeurosis sometimes remains intact, in which case the fracture fragments stay together. Stress fractures are a rare occurrence in young athletic individuals.

Clinical features

Swelling and bruising are usually evident. A breach of the skin indicates a direct blow to the elbow. A gap may be palpable and the patient will be unable to extend the elbow against resistance. The clinician should be alerted to the possibility of a stress fracture by an absence of these clinical signs and a history of prodromal pain.

X-rays

A properly oriented lateral view and AP of the elbow are required. Note should be made of the fracture pattern, the presence of comminution, the degree of displacement and in particular signs of subluxation of the ulnohumeral joint or radiocapitellar joint.

Treatment

In the elderly or infirm many of these fractures can be treated with analgesia and mobilization with a satisfactory outcome. An undisplaced transverse fracture that does not separate when the elbow is X-rayed in flexion can be treated without surgery. Repeat X-rays are needed to exclude displacement. Casting is not recommended but a sling can be used for comfort.

Operative repair is recommended for displaced fractures and those with instability of the ulnohumeral or radiocapitellar joints (Figure 25.5). A plate and screws should be used in all except those with

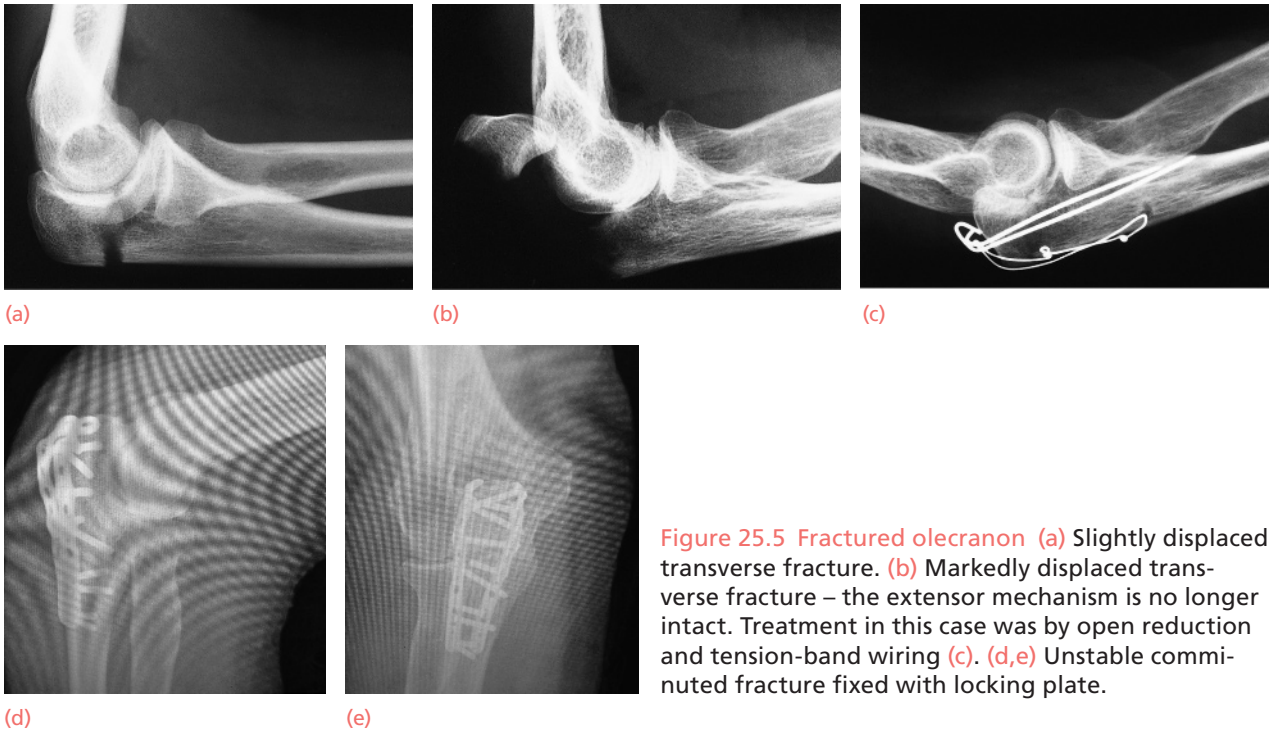


Figure 25.5 Fractured olecranon (a) Slightly displaced transverse fracture. (b) Markedly displaced transverse fracture – the extensor mechanism is no longer intact. Treatment in this case was by open reduction and tension-band wiring (c). (d,e) Unstable comminuted fracture fixed with locking plate.

simple transverse fractures with a stable joint, in whom a suture repair or tension-band wiring can be used. Immediate postoperative mobilization is recommended.

Complications

Stiffness This used to be common but with early mobilization the residual loss of movement should be minimal. Non-union sometimes occurs after inadequate reduction and fixation. If elbow function is good, it can be ignored; if not, rigid internal fixation and bone grafting will be needed.

Ulnar nerve symptoms Although these may occur, they usually settle spontaneously.

Osteoarthritis This can be a late complication, especially if reduction is less than perfect. It can usually be treated with analgesia and activity modification.

SIMPLE DISLOCATION OF THE ELBOW

Dislocation of the ulnohumeral joint is the second most common major joint dislocation after the shoulder. A simple dislocation is one without a fracture (although flake avulsions at the ligament insertions may be seen). Injuries are usually classified according to the direction of displacement. In over 90% of cases the forearm dislocates in a posterior direction relative to the humerus. Approximately 2% dislocate anteriorly and very rarely the radius goes laterally and the ulna medially (divergent dislocation) (Figure 25.6).

Anatomy

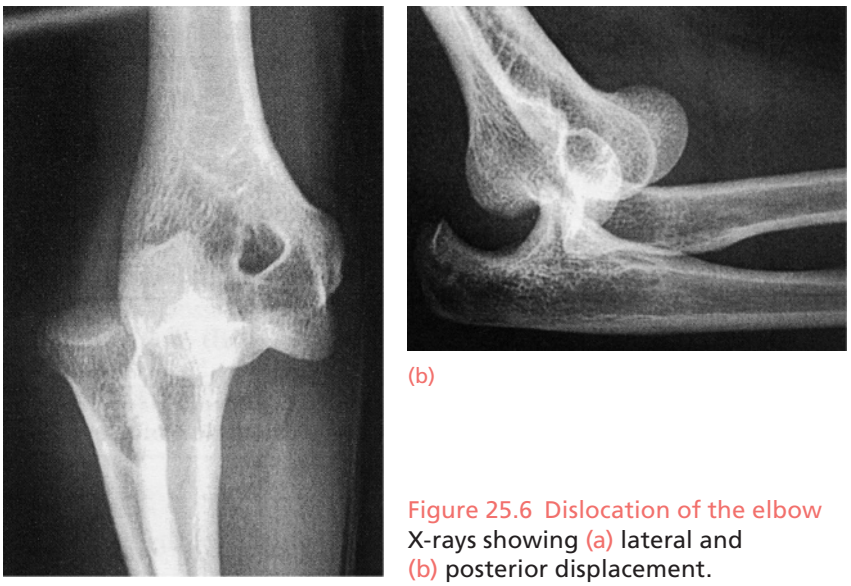
The elbow is a complex hinge, providing sufficient mobility to permit the upper limb to reach through wide ranges of flexion, extension and (in conjunction with the forearm joint) rotation. Yet it must also provide enough stability to support the necessary gripping, pushing, pulling and carrying activities of daily life. Its stability is due largely to the shape and fit of the bones that make up the joint – especially the ulnohumeral component – and this is liable to be compromised by any break in the articulating structures. The surrounding soft-tissue structures also are important, especially the capsular and collateral ligaments and, to a lesser extent, the muscles. Ligament disruption is therefore also a destabilizing factor.

The forearm is normally in slight valgus in relation to the upper arm, the average carrying angle being 10–15 degrees (range 5–25 degrees). When the elbow is flexed, the forearm comes to lie directly upon the upper arm (screw axis of rotation). Doubts about the normality of these features can usually be resolved by comparing the injured with the opposite arm.

With the elbow flexed, the tips of the medial and lateral epicondyles and the olecranon prominence form an isosceles triangle; with the elbow extended, they lie transversely in line with each other.

Mechanism of injury and pathology

The majority of dislocations occur as a result of a fall on an outstretched hand with the elbow in extension often with a valgus force. In sports other mechanisms may occur. The medial ligament will be found to be torn on MRI scan in all cases, but in up to 20% the lateral ligament will be either intact or have only a



(a) (b)

Figure 25.6 Dislocation of the elbow
X-rays showing (a) lateral and (b) posterior displacement.

low-grade partial tear. Approximately 8% of elbows will go on to have problems of recurrent instability. It is not known what the risk factors are but they are likely to be related to associated avulsion of the humeral attachments of the secondary stabilizers of the elbow, i.e. the common flexor and extensor muscles. Simple dislocations may be associated with damage to surrounding nerves and blood vessels, especially if the injury is open – suggesting a more high-energy injury.

Clinical features

The patient supports the elbow in slight flexion. Unless swelling is severe, the deformity is obvious. The bony landmarks (olecranon and epicondyles) may be palpable and abnormally placed. However, in severe injuries the pain and swelling are so marked that examination of the elbow is impossible. Nevertheless, the hand should be examined for signs of vascular or nerve damage.

X-rays

X-ray examination is essential (1) to confirm the presence of a dislocation and (2) to identify any associated fractures (see 'Fracture-dislocations' above).

Treatment

The patient should be fully relaxed with sedation or anaesthesia. Reduction can usually be achieved with gentle traction applied to the supinated forearm as the thumb of the surgeon's other hand is applied to the olecranon process to push it anteriorly and the elbow is taken from an extended to a flexed position.

After the reduction, the elbow should be put through a full range of motion to see whether it is stable. The distal nerves and circulation are checked again and an X-ray is obtained to confirm that the joint is reduced.

The arm is held in a collar and cuff with the elbow flexed above 90 degrees. After 1 week the patient gently moves the elbow while lying supine with the shoulder flexed to 90 degrees and the forearm in neutral rotation. The collar and cuff are discarded when the patient is comfortable. Passive 'stretching' of the elbow is to be avoided. The long-term results are usually good.

Often the clinician deciding on definitive treatment is not the one who performed the reduction and scant information is likely to be available about the elbow stability. MRI will define the extent of the soft-tissue injury. If the soft-tissue injury extends into the lateral structures, there may be a role for examination under anaesthesia and open stabilization of the elbow.

Complications

EARLY

Vascular injury The brachial artery may be damaged. Absence of the radial pulse is a warning. If there are other signs of ischaemia, the injury should be treated as an emergency. Splints must be removed and the elbow should be straightened somewhat. If there is no improvement, an arteriogram is performed; the brachial artery may have to be explored by an appropriately trained surgeon.

Nerve injury The median or ulnar nerve is sometimes injured. Spontaneous recovery usually occurs after 6–8 weeks.

LATE

Stiffness Loss of 20-30 degrees of extension is not uncommon after elbow dislocation; fortunately, this is usually of little functional significance, although the patient may not be happy with the cosmetic impact and should be warned that this is a possibility. The most common cause of stiffness is prolonged immobilization. In the management of all elbow injuries the joint should be moved as soon as possible, with due consideration to stability of the fractures and soft tissues and without undue passive stretching. The use of a hinged brace, designed to control movement, may contribute to stiffness. Hypertonia of biceps and brachioradialis after injury or surgery will limit active extension. Mobilization with the patient lying supine, the shoulder flexed to 90 degrees and the arm supported by the other hand relaxes the muscles and provides gravitational stability of the ulna on the end of the humerus. This simple activity can improve range of movement by 20 degrees immediately.

Persistent stiffness, with more than 30 degrees' loss of extension, or flexion limited to less than 130 degrees, can limit function and can be addressed by capsular release. Surgical intervention should not be rushed as the range of movement may improve up to 6 months from the time of injury.

Heterotopic ossification This may occur in the damaged soft tissues in front of the joint. The precise pathogenesis is unknown but the risk is increased by associated head injury and by delay to surgical intervention. There is a suggestion that passive 'stretch' of the elbow may increase the risk. The clinician should be alert to the possibility in patients who fail to recover movement after injury or surgery.

X-ray examination may be unhelpful initially; soft-tissue ossification is not usually visible until 4-6 weeks after injury. Anti-inflammatory drugs may help to reduce stiffness and can be used as prophylaxis against the formation of heterotopic ossification, but these are contraindicated while fracture healing is ongoing.

Where heterotopic ossification causes significant extrinsic stiffness to the elbow, it can be excised. Historically it was usual practice to wait for the ossification to mature but the evidence base does not support this.

Recurrent dislocation The most common recurrent dislocation is posterolateral rotatory instability due to incompetence of the lateral ligament complex (discussed in Chapter 14). Recurrent ulnohumeral dislocation is rare but may follow simple elbow dislocation and will usually require reconstruction of the lateral ligament complex and possibly the anterior band of the medial ligament too. Instability after neglected fracture-dislocation can occur and may require reconstruction of ligaments and reconstruction or replacement of the radial head. Coronoid reconstruction is required rarely for chronic posteromedial rotatory instability.

Osteoarthritis Secondary osteoarthritis is quite common after severe fracture-dislocations. In older patients, total elbow replacement can be considered.

Unreduced dislocation This is a very challenging problem to address because the joint fills with dense scar tissue, preventing reduction. Open reduction requires extensive soft-tissue release and ligament reconstruction with the use of an external fixator if the elbow remains unstable. Stiffness is likely to follow such an event.

FRACTURES AROUND THE ELBOW IN CHILDREN

The elbow is second only to the distal forearm for frequency of fractures in children. Most of these injuries are supracondylar fractures, the remainder being divided between condylar, epicondylar and proximal radial and ulnar fractures. Boys are injured more often than girls and more than half the patients are under 10 years old.

Mechanism of injury and pathology

The usual accident is a fall directly on the point of the elbow or – more often – on the outstretched hand with the elbow forced into valgus or varus. Pain and swelling are often marked and examination is difficult. X-ray interpretation also has its problems: the bone ends are largely cartilaginous and therefore radiographically incompletely visualized. A good knowledge of the normal anatomy is essential if fracture displacements are to be recognized.

Although all the epiphyses are in some part cartilaginous, the secondary ossific centres can be seen on X-ray; they should not be mistaken for fracture fragments. The average ages at which the ossific centres appear are easily remembered by the mnemonic CRITOL: Capitellum – 2 years; Radial head – 4 years; Internal (medial) epicondyle – 6 years; Trochlea – 8 years; Olecranon – 10 years; Lateral epicondyle – 12 years. Obviously, epiphyseal displacements will not be detectable on X-rays before these ages but soft-tissue swelling may be apparent on X-rays and should raise suspicion of an underlying fracture. Fracture displacement and accuracy of reduction can be inferred from radiographic indices such as Baumann's angle (see Figure 25.8).

SUPRACONDYLAR FRACTURES IN CHILDREN

These are among the most common fractures in children. The distal fragment may be displaced either posteriorly or anteriorly.

Mechanism of injury

Posterior angulation or displacement (95% of cases) suggests a hyperextension injury, usually due to a fall on the outstretched hand. The humerus breaks just above the condyles. The distal fragment is pushed backwards and (because the forearm is usually pronated) twists inwards. The jagged end of the proximal fragment pokes into the soft tissues anteriorly, sometimes injuring the brachial artery or median nerve.

Anterior displacement is rare; it is thought to be due to direct violence (e.g. a fall on the point of the elbow) with the joint in flexion.

Classification

- *Type I* – an undisplaced fracture
- *Type II* – an angulated fracture with the posterior cortex still intact
  - *IIA*: a less severe injury with the distal fragment merely angulated
  - *IIB*: a severe injury; the fragment is both angulated and malrotated
- *Type III* – a completely displaced fracture (although the posterior periosteum is usually preserved, which will assist surgical reduction)
- *Type IV* – an anteriorly displaced fracture

Clinical features

Following a fall, the child is in pain and the elbow is swollen; with a posteriorly displaced fracture the S-deformity if the elbow is usually obvious and the bony landmarks are abnormal. It is essential to feel the pulse distally and check capillary return; passive extension of the flexor muscles should be pain-free otherwise there may be concern regarding ischaemia.

The wrist and the hand should be examined for evidence of nerve injury.

X-rays

The fracture is seen most clearly in the lateral view (Figure 25.7). In an undisplaced fracture the 'fat pad' or 'sail' sign should raise suspicions; this is a triangular lucency in front of and behind the distal humerus like the sails of a yacht, due to the fat pad being pushed up by fluid such as a haematoma. This sign is not specific for distal humerus fractures but should alert the clinician to look for a cause.

In the common posteriorly displaced fracture the fracture line runs obliquely downwards and forwards and the distal fragment is tilted backwards and/or displaced backwards. In the anteriorly displaced fracture the fracture line runs downwards and backwards and the distal fragment is angulated forwards. On a normal lateral X-ray, a line drawn along the anterior cortex of the humerus should cross the middle of the capitellum. If the line is anterior to the capitellum, a type II fracture is suspected.

An anteroposterior view is often difficult to obtain without causing pain and it may need to be postponed until the child has been anaesthetized. It may show that the distal fragment is translated or angulated sideways, and rotated (usually medially). Measurement of Baumann's angle is useful in assessing the degree of medial angulation before and after reduction.

Treatment

If there is even a suspicion of a fracture, the elbow is gently splinted in 30 degrees of flexion to prevent movement and possible neurovascular injury during X-ray examination.

TYPE I: UNDISPLACED FRACTURES

The elbow is immobilized at 90 degrees and neutral rotation in a lightweight splint or cast and the arm is supported by a sling. It is essential to obtain an X-ray 5–7 days later to check that there has been no displacement. The splint is retained for 3 weeks and supervised movement is then allowed.

The capitellum normally angles forwards about 30 degrees; if the capitellum is seen to be in a straight line with the humerus in the lateral X-ray, it will still remodel.

Even with type I fractures, take care to recognize any medial tilt of the fragment on the AP X-ray, otherwise cubitus varus can result. Measure Baumann's angle carefully (Figure 25.8).

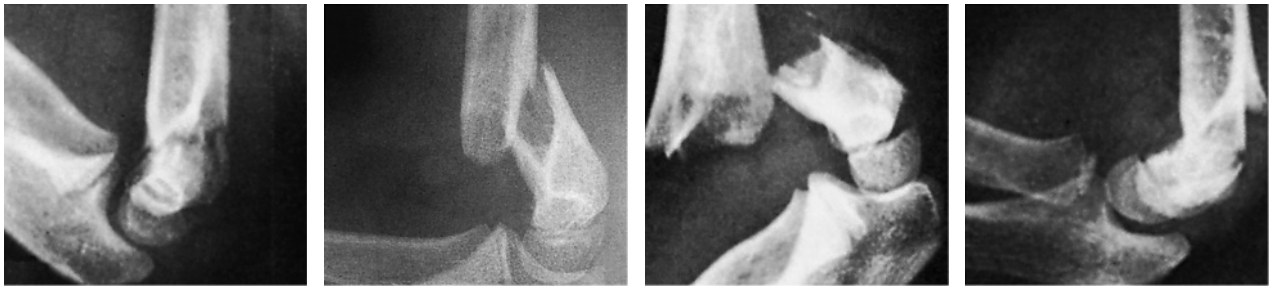


Figure 25.7 Supracondylar fractures X-rays showing supracondylar fractures of increasing severity: (a) undisplaced; (b) distal fragment posteriorly angulated but in contact; (c) distal fragment completely separated and displaced posteriorly; (d) a rarer variety with anterior angulation.

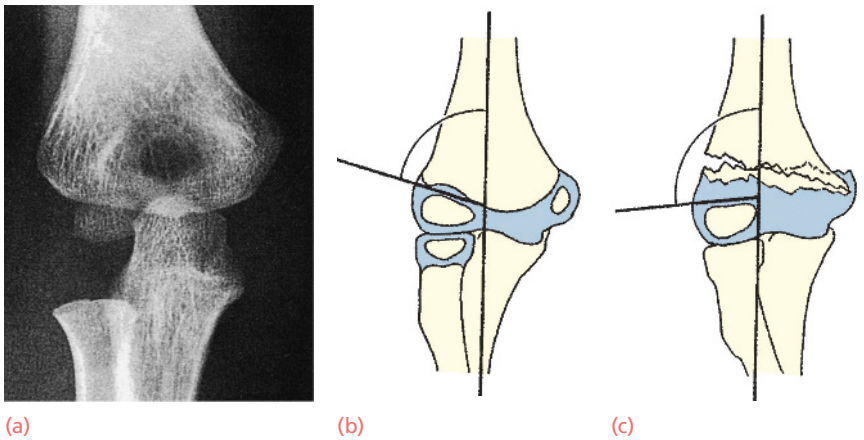


Figure 25.8 Baumann's angle Anteroposterior X-rays are sometimes difficult to make out, especially if the elbow is held flexed after reduction of the supracondylar fracture. Measurement of Baumann's angle is helpful. This is the angle subtended by the longitudinal axis of the humeral shaft and a line through the coronal axis of the capitellar physis, as shown in (a) the X-ray of a normal elbow and the accompanying diagram (b). Normally this angle is less than 80 degrees. If the distal fragment is tilted in varus, the increased angle is readily detected (c).

TYPE IIA: POSTERIORLY ANGULATED FRACTURES – MILD

In these cases swelling is usually not severe and the risk of vascular injury is low. If the posterior cortices are in continuity, the fracture can be reduced under general anaesthesia by the following stepwise manoeuvre (Figure 25.9): (1) traction for 2–3 minutes in the length of the arm with countertraction above the elbow; (2) correction of any sideways tilt or shift and rotation (in comparison with the other arm); (3) gradual flexion of the elbow to 120 degrees, and pronation of the forearm, while maintaining traction and exerting finger pressure on the olecranon to correct the posterior tilt. Then feel the pulse and check the capillary return: if the distal circulation is suspect, immediately relax the amount of elbow flexion until it improves.

X-rays are taken to confirm the reduction, checking carefully to see that there is no varus or valgus angulation and no rotational deformity. The AP view is confusing and unreliable with the elbow flexed. Each column can be assessed by slight internal and external rotation of the humerus to obtain AP oblique views, and Baumann's angle can be assessed on the true AP. *Subtle medial tilt and rotation must be recognized*. If the acutely flexed position cannot be maintained without disturbing the circulation, or if the reduction is unstable (and most of these fractures are unstable), the fracture should be fixed with percutaneous smooth K-wires. The wires should be advanced slowly with low revolutions to avoid generating heat. Configuration of the wires varies but most use either crossed K-wires or two lateral wires. Care must be taken to protect the ulnar nerve and a medial mini-open approach is safest for placement of the medial wire. A backslab should be applied.

Following reduction, the arm is held in a collar and cuff; the circulation should be checked repeatedly during the first 24 hours. An X-ray is obtained after 3–5 days to confirm that the fracture has not slipped. The splint is retained for 3–4 weeks, after which movements are begun. Check X-rays must be obtained on removal of the splint and wires to ensure that adequate position has been maintained.

TYPES IIB AND III: ANGULATED AND MALROTATED OR POSTERIORLY DISPLACED FRACTURES

These are usually associated with severe swelling, are difficult to reduce and are often unstable; moreover, there is a considerable risk of neurovascular injury or circulatory compromise due to swelling. The fracture should be reduced under general anaesthesia as soon as possible, by the method described above, and then held with percutaneous smooth K-wires; this obviates the necessity to hold the elbow acutely flexed. Postoperative management is the same as for type IIA.

OPEN REDUCTION

This is sometimes necessary for (1) a fracture that simply cannot be reduced closed; (2) an open fracture; or (3) a fracture associated with vascular damage. The fracture is exposed from the lateral side, the

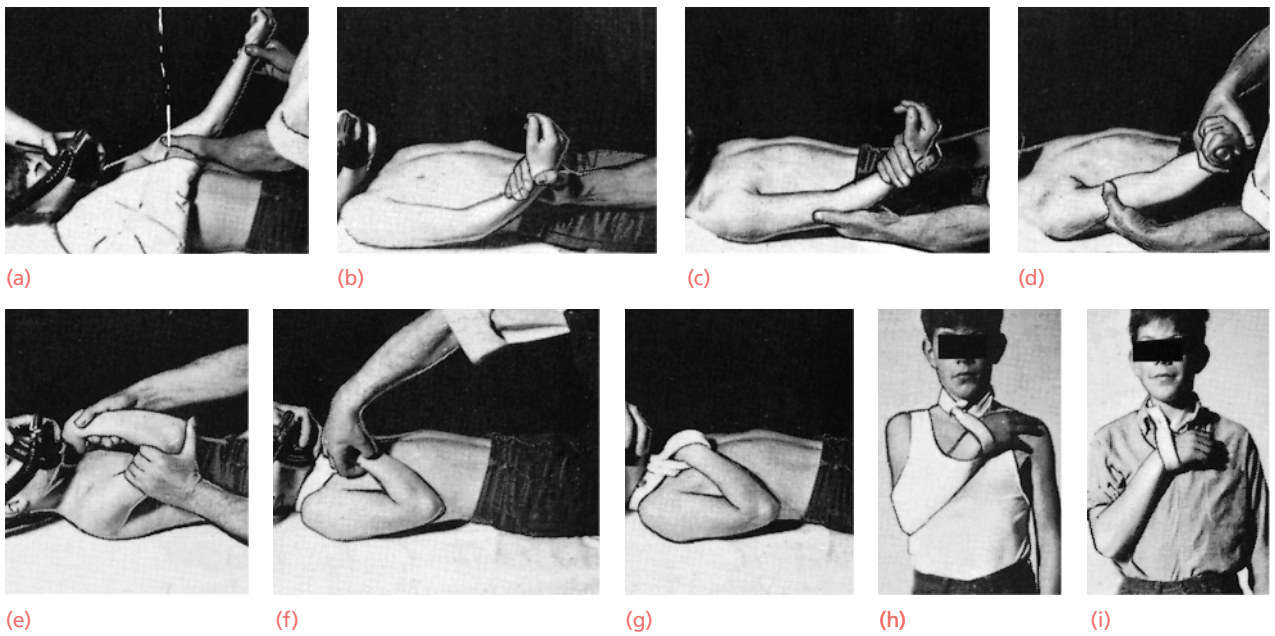


Figure 25.9 Supracondylar fractures – treatment (a) The uninjured arm is examined first; (b) traction of the fractured arm; (c) correcting lateral shift and tilt; (d) correcting rotation; (e) correcting backward shift and tilt; (f) feeling the pulse; the elbow is kept well flexed while X-ray films are taken (g). (h) For the first 3 weeks the arm is kept under the clothes; after this (i) it is outside the clothes.

haematoma is evacuated and the fracture is reduced and held by two K-wires.

CONTINUOUS TRACTION

Traction through a screw in the olecranon, with the arm held overhead, can be used (1) if the fracture is severely displaced and cannot be reduced by manipulation; (2) if, with the elbow flexed 100 degrees, the pulse is obliterated and image intensification is not available to allow pinning and then straightening of the elbow; or (3) for severe open injuries or multiple injuries of the limb. Once the swelling subsides, a further attempt can be made at reduction.

ANTERIORLY DISPLACED FRACTURES

An anteriorly displaced fracture is a rare injury (less than 5% of supracondylar fractures). However, 'posterior' fractures are sometimes inadvertently converted to 'anterior' ones by excessive manipulation.

The fracture is reduced by pulling on the forearm with the elbow semi-flexed, applying thumb pressure over the front of the distal fragment and then extending the elbow fully. Percutaneous smooth pins are used if unstable. A posterior slab is applied and retained for 3–4 weeks. Thereafter the child is allowed to regain flexion gradually.

Complications

EARLY

Vascular injury The great danger of supracondylar fractures is injury to the brachial artery, which, before the introduction of percutaneous pinning, was reported as occurring in more than 5% of cases. Nowadays the incidence is probably less than 1%. Peripheral ischaemia may be immediate and severe, or the pulse may fail to return after reduction. More commonly the injury is complicated by forearm oedema and a mounting compartment syndrome, which leads to necrosis of the muscles and nerves without causing peripheral gangrene. Undue pain plus one positive sign (pain on passive extension of the fingers, a tense and tender forearm, an absent pulse, blunting sensation or reduced capillary return on pressing the finger pulp) demand urgent action.

The flexed elbow must be extended and all dressings removed. If the circulation does not improve, then angiography or Doppler examination is carried out, the vessel is repaired or grafted and a forearm fasciotomy is performed. In extreme cases, operative exploration would be justified on clinical criteria alone. Close early collaboration with vascular or plastic surgical colleagues is required in these cases.

Nerve injury The radial nerve, median nerve (particularly the anterior interosseous branch) or the ulnar nerve may be injured. Tests for nerve function are described in Chapter 11. Fortunately, loss of function is usually temporary and recovery can be expected in 3-4 months. If there is no recovery, the nerve should be explored. However, if a nerve, documented as intact prior to manipulation, is then found to be compromised after manipulation, entrapment in the fracture is suspected and immediate exploration should be arranged.

The ulnar nerve may be damaged by careless pin placement. It is safest to perform a mini-open approach on the medial side of the elbow and identify the nerve before placing the smooth K-wire. If the injury is recognized, and the pin removed, recovery will usually follow.

LATE

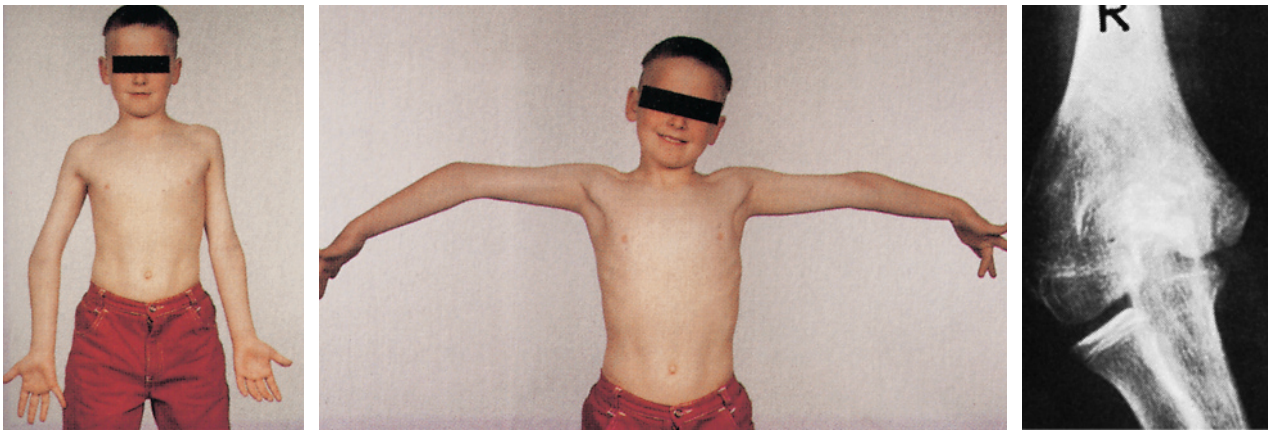
Malunion This is common. However, backward or sideways shifts are gradually smoothed out by modelling during growth and they seldom give rise to visible deformity of the elbow. Forward or backward angulation may limit flexion or extension, but consequent disability is slight.

Uncorrected sideways angulation and rotation are much more important and may lead to varus (or rarely valgus) deformity of the elbow; this is permanent and will not improve with growth (Figure 25.10). The fracture is extraphyseal and so physeal damage should not be blamed for the deformity; usually faulty reduction is responsible. Cubitus varus is disfiguring and cubitus valgus may cause late ulnar nerve palsy. If deformity is marked, it will need correction by supracondylar osteotomy, usually once the child approaches skeletal maturity.

Elbow stiffness and heterotopic ossification Stiffness is an ever-present risk with elbow injuries. Extension in particular may take months to recover, and the patient and parents should be warned that some loss of extension is common but unlikely to affect function. Passive elbow stretch should be avoided as it tends to increase stiffness and may increase the risk of heterotopic ossification, which otherwise is a rare complication.

Fractures of the lateral condyle in children

The lateral condylar (or capitellar) epiphysis begins to ossify during the first year of life and fuses with the shaft at 12–16 years. Between these ages it may be sheared off or avulsed by forceful traction.

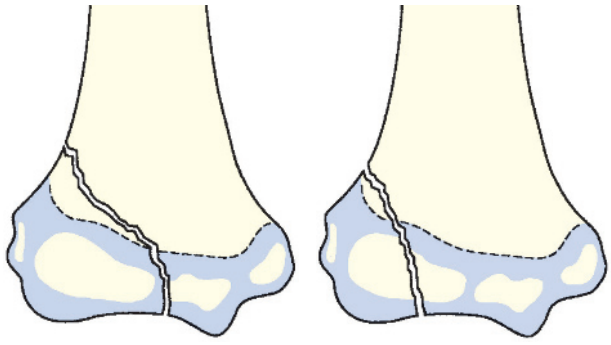


(a) (b) (c)

Figure 25.10 Supracondylar fracture – malunion (a) Varus deformity of the right elbow, due to incomplete correction of the varus and rotational displacements in a supracondylar fracture. (b) It is most obvious when the boy raises his arms, displaying the typical 'gunstock deformity'. (c) X-ray showing the characteristic malunion.

Mechanism of injury and pathology

The child falls on the hand with the elbow extended and forced into varus. A large fragment, which includes the lateral condyle, breaks off and is pulled upon by the attached wrist extensors. Sometimes there is a compression, rather than avulsion, mechanism of injury. The fracture line usually runs along the physis and into the trochlea; less often it continues through the medial epiphysis and exits through the capitellum-trochlea groove (Figure 25.11). It crosses the growth plate and epiphysis so is a Salter–Harris type IV injury. In severe injuries the elbow may dislocate posterolaterally; the condyle is 'capsized' by the pull of the muscle and remains dislocated while the elbow reduces spontaneously.



(a) (b)

Figure 25.11 Physeal fractures of the lateral condoyle (a) The commonest is a fracture starting in the metaphysi and running along the physis of the lateral condyle into the trochlea (Salter–Harris type II injury). (b) Less common is a fracture running right through the lateral condyle to reach the articular surface in the capitulotrochlear groove (Salter–Harris type IV): though uncommon, this latter injury is important because of its potential for causing growth defects.

The extent of this injury is often not appreciated because the condylar epiphysis is largely cartilaginous, and the bone fragment may look deceptively small on X-ray. Displacement can be marked due to muscle pull. The fracture is important for two reasons: (1) it may damage the growth plate; and (2) it always involves the joint. Early recognition and accurate reduction are therefore essential if a poor outcome is to be avoided.

Clinical features

The elbow is swollen and deformed. There is tenderness over the lateral condyle. Passive flexion of the wrist (pulling on the extensors) may be painful.

X-rays

X-ray examination must include oblique views or the full extent of the fracture may be missed. Two types of fracture are recognized and classified by Milch:

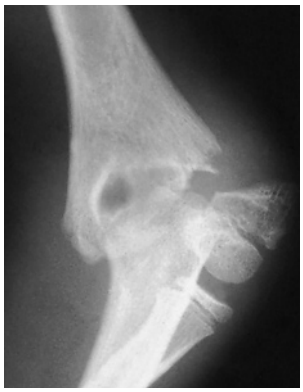
- *Type I* – a fracture lateral to the trochlea: the ulnohumeral joint is not involved and is stable.
- *Type II* – a fracture through the middle of the trochlea. This injury is more common; the elbow is unstable as the lateral restraint of the ulnohumeral joint, the lateral trochlea ridge is carried along with the fragment. The fragment is often grossly displaced and rotated, and it may carry with it a triangular piece of the metaphysis. Remember that the fragment (partly cartilaginous) is much larger than it seems on X-ray.

Treatment

If there is no displacement, the arm can be splinted in a backslab with the elbow flexed 90 degrees, the forearm in neutral rotation and the wrist

extended - this position relaxes the extensor mechanism which attaches to the fragment. However, it is essential to repeat the X-ray after 5 days to make sure that the fracture has not displaced. The splint is removed after 2 weeks and exercises encouraged.

A displaced fracture requires accurate reduction and internal fixation. If the fragment is only moderately displaced (hinged), it may be possible to manipulate it into position by extending the elbow and pressing upon the condyle, and then fixing the fragment with percutaneous pins. If this fails, and for all separated fractures, open reduction and internal fixation with pins or screws is required (Figure 25.12). The arm is immobilized in a cast that is removed along with the pins after 3–4 weeks.



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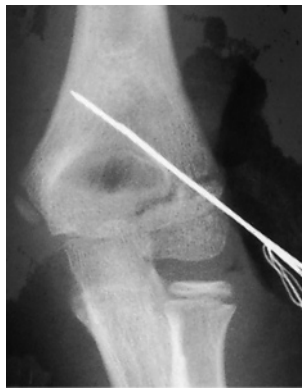


(b)

Figure 25.12 Fractured lateral condyle If displacement is more than 2 mm, open reduction with internal fixation is the treatment of choice.

Complications

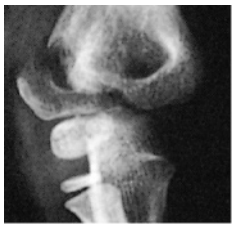
Non-union and malunion If the condyle is left rotated and displaced, non-union is inevitable; with growth the elbow becomes increasingly valgus, and ulnar nerve palsy is then likely to develop. Stiffness and pain can result. Even minor displacements sometimes lead to non-union, and even slight malunion may lead to ulnar palsy in later life; it is for these reasons that open reduction (and internal fixation) is preferred for any displaced fracture. The fracture is a Salter–Harris type IV injury and so imperfect reduction can result in growth arrest. Even if a fracture presents late (e.g. up to 3 months), open reduction and fixation should be attempted, and good results can be achieved (Figure 25.13).

Recurrent dislocation Occasionally, condylar displacement results in posterolateral dislocation of the elbow. The only effective treatment is reconstruction of the bony and soft tissues on the lateral side.

Separation of the medial epicondyle in children

Mechanism of injury and pathology

The medial epicondyle begins to ossify at the age of about 5 years and fuses to the shaft at about 16 years; between these ages it may be avulsed by a severe muscle of ligament strain. The child falls on the outstretched hand with the wrist and elbow extended; the elbow is wrenched into valgus. The unfused epicondylar apophysis is avulsed by tension



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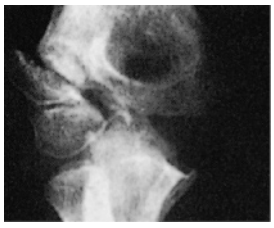

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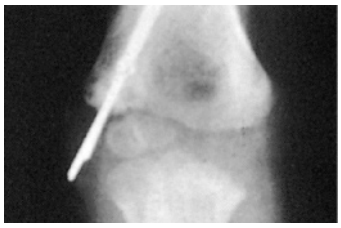

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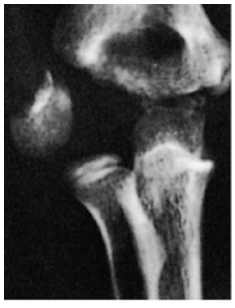

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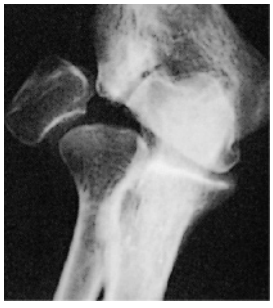

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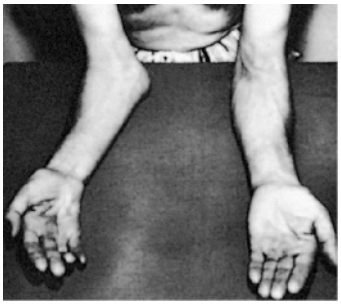


(f)

Figure 25.13 Fractured lateral condyle – complications (a,b) A large fragment of bone and cartilage is avulsed; even with reasonable reduction, union is not inevitable. (c) Open reduction with fixation is often wise. (d) Sometimes the condyle is capsized; if left unreduced, non-union is inevitable (e) and a valgus elbow with delayed ulnar palsy (f) is the likely sequel.

on either the wrist flexor muscles or the medial ligament of the elbow. If the elbow subluxates (even momentarily), the small apophyseal fragment may be dragged into the joint. With more severe injuries, the joint dislocates laterally.

APOPHYSITIS

This is separate pathology. It is a painful traction injury of the medial epicondyle apophysis, and it occurs in young athletes as a result of repeated forced valgus moments of the elbow, such as when bowling a ball or throwing. The condition usually settles with rest.

Clinical features

The diagnosis should be suspected if injury is followed by pain, swelling and bruising to the medial side of the elbow. If the joint is dislocated, deformity is of course obvious. Sensation and power in the fingers should be tested to exclude concomitant ulnar nerve damage.

X-rays

In the AP view the medial epicondylar epiphysis may be displaced distally or rotated; if the joint is dislocated, the fragment lies distal to the lower humerus (Figure 25.14). A lateral view may show the epicondyle looking like a loose body in the joint. If in any doubt, the normal side should be X-rayed for comparison.

Treatment

Minor displacement may be disregarded. This is an extra-articular fracture, so the elbow can be mobilized as soon as the child wishes.

If the epicondyle is trapped in the joint, it must be freed. Manipulation with the elbow in valgus and the wrist hyperextended (to pull on the flexor muscles) may be successful; if this fails, the joint must be opened (the ulnar nerve must be visualized and protected) and the fragments retrieved and fixed back in position.

Displaced fractures that are not trapped in the joint usually do not need to be operated on; however, if there is valgus instability (because the medial collateral ligament complex is attached to the fragment) or marked displacement, then open reduction and pinning, or screw fixation is recommended.

Complications

EARLY

Ulnar nerve damage This is not uncommon. Mild symptoms recover spontaneously; even a complete palsy will usually recover but, if there is the possibility that the nerve is kinked in the joint, exploration should be considered.

LATE

Stiffness Stiffness is common and extension often limited for months; but, provided movement is not forced, it will eventually return.

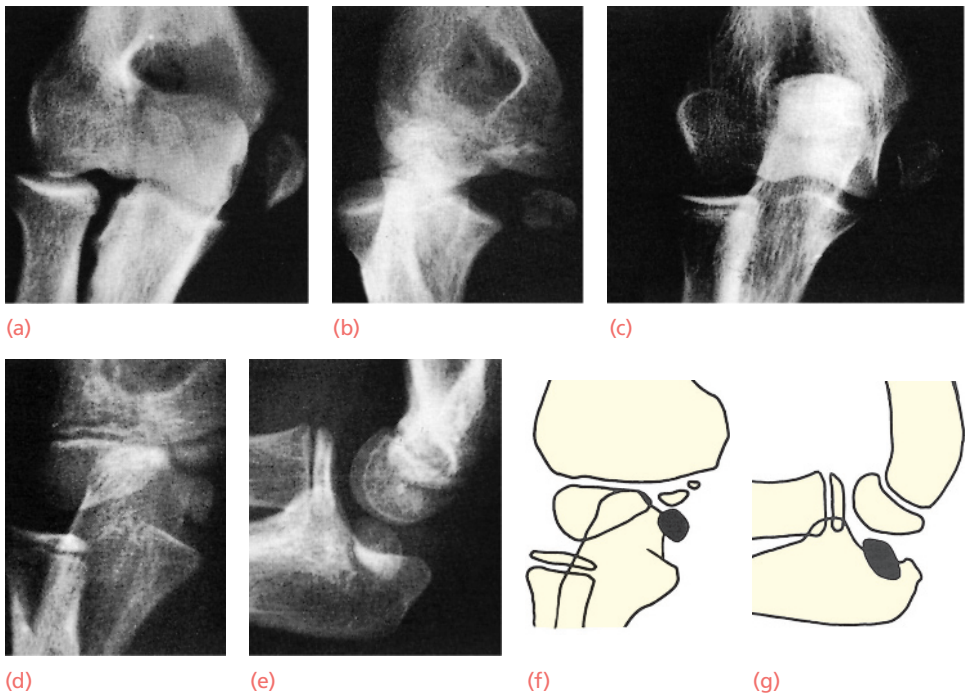


Figure 25.14 Fractured medial epicondyle (a) Avulsion of the medial epicondyle following valgus train. (b) Avulsion associated with dislocation of the elbow; (c) after reduction. Sometimes the epicondylar fragment is trapped in the joint (d,e); the serious nature is then liable to be missed unless the surgeon specifically looks for the trapped fragment, which is emphasized in the tracings (f,g).

Fractures of the medial condyle in children

A fracture of the medial condyle is much less common than either a fracture of the lateral condyle or a separation of the medial epicondylar apophysis.

Mechanism of injury

The injury is usually caused by a fall from height, involving either a direct blow to the point of the elbow or a landing on the outstretched hand with the elbow forced into valgus; in the latter case it would be an avulsion injury. The fracture line runs through the physis, exiting in the trochlea notch or even further laterally, and the medial fragment may be displaced by the pull of the flexor muscle group.

Clinical features and X-rays

This is an intra-articular fracture, resulting in considerable pain and swelling. In older children the metaphyseal component is usually easily visualized on X-ray. However, in younger children much of the medial condylar epiphysis is cartilaginous and therefore not visible on X-ray, so the full extent of the fracture may not be recognized; seeing only the epicondylar ossific centre in a displaced position on the X-ray may mislead the surgeon into thinking that this is only an epicondylar fracture. In doubtful cases an arthrogram or MRI may be helpful.

Treatment

Undisplaced fractures are treated by splintage; X-rays are repeated until the fracture has healed, so as to ensure that it does not become displaced.

Displaced fractures are treated by either closed reduction (sometimes with percutaneous pinning) or open reduction and fixation with smooth pins.

Postoperative management is similar to that of lateral condyle fractures.

Complications

EARLY

Lateral dislocation of the elbow This occasionally occurs with a severe valgus strain and avulsion of the medial condyle. Early reduction of both the dislocation and the fracture, if necessary by open operation and pinning, is important.

Ulnar nerve damage This is not uncommon, but recovery is usual unless the nerve is left kinked in the joint.

LATE

Stiffness of the elbow This is common and extension is often limited for months; but, provided movement is not forced, it will eventually return.

Fracture-separation of the distal humerus physis in children

Up to the age of 7 years the distal humeral epiphysis is a solid cartilaginous segment with maturing centres of ossification. With severe injury it may separate *en bloc*. This is likely to occur with fairly severe violence, such as in birth injuries or child abuse.

Clinical features

The child is in pain and the elbow is markedly swollen. The history may be deceptively uninformative.

X-rays

In a very young child, in whom the bony outlines are still unformed, the X-ray may look normal. All that can be seen of the epiphysis is the pea-like ossification centre of the capitellum; its position should be compared to that of the normal side. Medial displacement of either the capitellar ossification centre or the proximal radius and ulna is very suspicious. In the older child the deformity is usually obvious.

Treatment

If the diagnosis is uncertain, arthrography or ultrasound can help. MRI may be a useful investigation but in very young children this will require a general anaesthetic, a service which may not be available in all hospitals. If the fracture is undisplaced, the elbow is merely splinted for 3 weeks; if it is displaced, the fracture should be accurately reduced and held with smooth percutaneous wires (otherwise there is a high incidence of cubitus varus). The wires are removed at 3 weeks.

Fractured neck of radius in children

Mechanism of injury and pathology

A fall on the outstretched hand forces the elbow into valgus and pushes the radial head against the capitellum. In children the bone fractures through the neck of the radius; in adults the injury is more likely to fracture the radial head.

Clinical features

Following a fall, the child complains of pain in the elbow. There may be localized tenderness over the radial head and pain on rotating the forearm.

X-rays

The fracture line is transverse. It is either situated immediately distal to the physis or there is true

separation of the epiphysis with a triangular fragment of shaft (a Salter–Harris type II injury). The proximal fragment is tilted distally, anteriorly and laterally. Sometimes the upper end of the ulna is also fractured or there may be a posterior dislocation of the elbow. In the younger patient the radial head ossification centre may not be visible and suspicion should be raised in those with marked lateral soft-tissue swelling.

Treatment

In children there is considerable potential for remodelling after these fractures. Up to 30 degrees of radial head tilt and up to 3 mm of transverse displacement are acceptable. The arm is rested in a collar and cuff, and exercises are commenced after a week.

Angulation of more than 30 degrees requires reduction (Figure 25.15). With the patient's elbow extended, traction and varus force are applied; the surgeon then pushes the displaced radial fragment into position with his or her thumb. If this fails, a percutaneous wire can be used to push the fragments back into place. Occasionally, a longitudinal intramedullary flexible nail will need to be passed from the radial styloid to the radial head to stabilize the fracture. Open reduction should be avoided as there is a high risk of avascular necrosis of the radial head. The head of the radius must never be excised in children because this will interfere with the synchronous growth of the radius and ulna.

Fractures that are seen a week or longer after injury should be left untreated (except for light splintage).

Following operation, the elbow is splinted in 90 degrees of flexion for a week or two and then movements are encouraged.

Subluxation of the radial head ('pulled elbow')

In young children the elbow may be injured by pulling on the arm, usually with the forearm pronated.

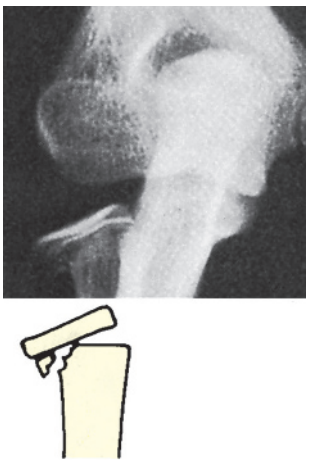


Figure 25.15 Fractured neck of radius in a child Up to 30° of tilt is acceptable. Greater degrees of angulation should be reduced; never excise the radial head in a child.

It is sometimes called subluxation of the radial head; more accurately, it is a subluxation of the annular ligament that slips up over the head of the radius into the radiocapitellar joint.

A child aged 2–3 years is brought with a painful, dangling arm: there is usually a history of the child being jerked by the arm and crying out in pain. The child is reluctant to use the arm, and the forearm is held pronated and extended. There are no X-ray changes.

A dramatic cure is achieved by supinating and then flexing the elbow; the ligament slips back with a snap.

Fractures of the olecranon in children

A fracture of the olecranon in a child is rare. When it does occur, it is usually due to a direct blow onto the tip of the flexed elbow or a fall on the outstretched hand. Most are undisplaced and are treated in a splint for 3–4 weeks. If displaced, they should be reduced and held with K-wires.

FRACTURES OF THE SHAFTS OF THE RADIUS AND ULNA

Mechanism of injury and pathology

Fractures of the shafts of both forearm bones occur quite commonly. A twisting force (usually a fall on the hand) produces a spiral fracture with the bones broken at different levels. An angulating force causes a transverse fracture of both bones at the same level. A direct blow causes a transverse fracture of just one bone, usually the ulna. Additional rotation deformity may be produced by the pull of muscles attached to the radius: they are the biceps and supinator muscles to the upper third, the pronator teres to the middle third, and the pronator quadratus to the lower third. Bleeding and swelling of the muscle compartments of the forearm may cause circulatory impairment and compartment syndrome.

Injuries to the bones of the forearm should be considered intra-articular fractures, because the forearm is a quadrilateral joint – with the proximal distal radioulnar joint at one end and the distal radioulnar joint at the other. Disruption of any one part – the radioulnar joints or the shafts of the long bones – will usually disrupt another part of the quadrilateral ring (Figure 25.16). Malalignment is likely to affect forearm rotation especially in the skeletally mature.

Clinical features

Pain and deformity are the most obvious clinical signs; the pulse must be felt and the hand examined for circulatory or neural deficit. Repeated examination is necessary in order to detect an impending

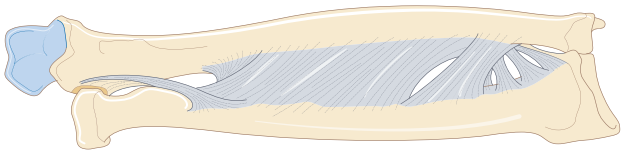


Figure 25.16 The forearm is a quadrilateral joint. A break or angulation in one part will affect another part.

compartment syndrome. Pain out of proportion to the injury is the cardinal sign.

X-rays

Both bones are broken, either transversely and at the same level or obliquely with the radial fracture usually at a higher level. Views of the elbow and wrist must be included to identify any separation of the proximal or distal radioulnar joints. In children, the fracture may be incomplete (greenstick) and only angulated. In adults, displacement may occur in any direction – shift, overlap, tilt or twist. In low-energy injuries, the fracture tends to be transverse or oblique; in high-energy injuries it is comminuted or segmental.

Treatment

CHILDREN

In children, closed treatment is often successful because the tough periosteum tends to guide and then control the reduction. The fragments are held in a well-moulded full-length cast, from just distal to the axilla to the metacarpal shafts (to control rotation). The cast is applied with the elbow at 90 degrees and moulded with three-point fixation with an oval shape over the forearm and moulded in the AP direction over the upper arm. If the fracture is proximal to pronator teres, the forearm is supinated; if it is distal to pronator teres, the forearm is held in neutral. The position is checked by X-ray after a week and, if it is satisfactory, splintage is retained until both fractures are united (usually 6–8 weeks).

Throughout this period hand and shoulder exercises are encouraged. The child should avoid contact sports for a few weeks to prevent refracture.

Occasionally an operation is required, either if the fracture cannot be reduced or if the fragments are unstable. Fractures that are initially displaced 100% or more should be held with percutaneous or intramedullary rods even if they can be reduced closed as there is a high risk of redisplacement (Figure 25.17).

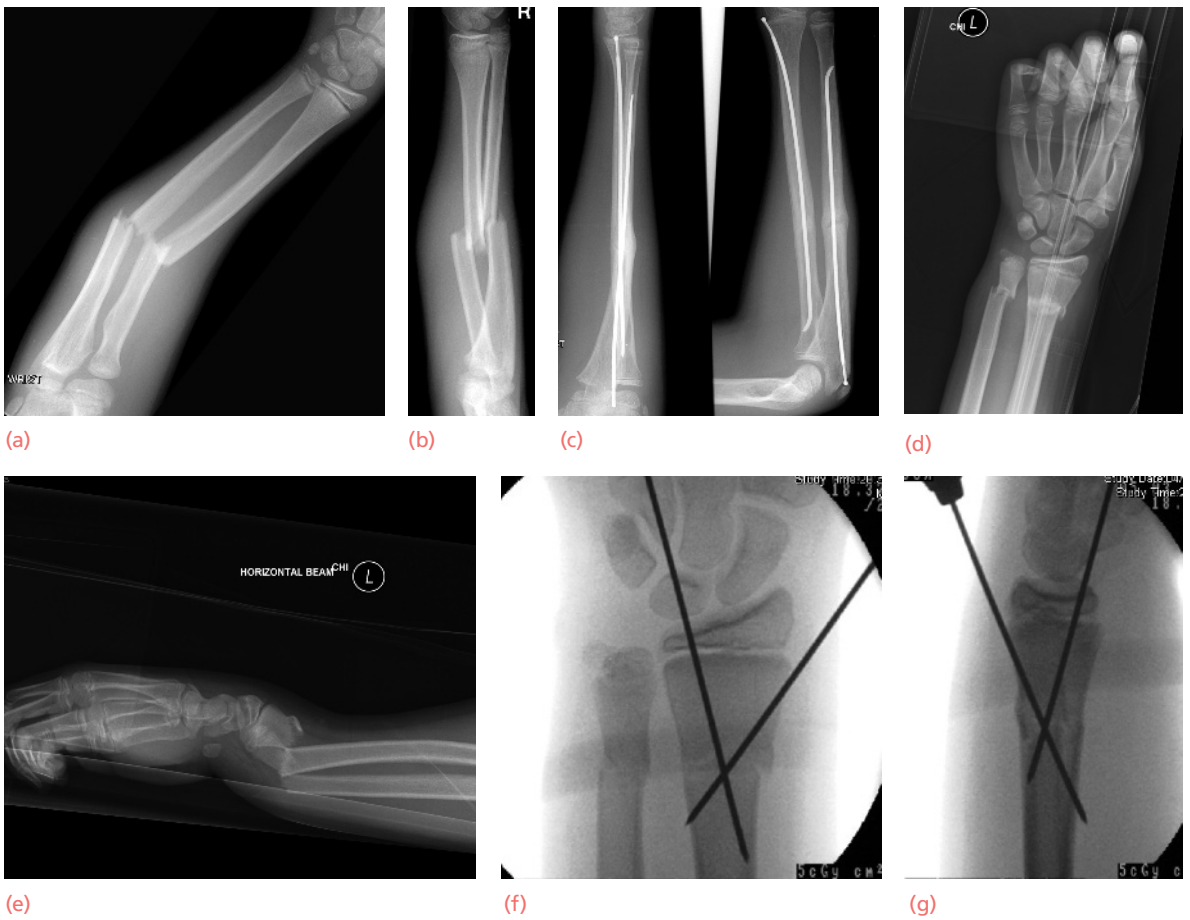


Figure 25.17 Forearm fractures A displaced forearm fracture (a,b) is fixed with flexible intramedullary nails (c). An unstable distal third diaphyseal fracture (d,e) is treated with percutaneous wires (f,g).

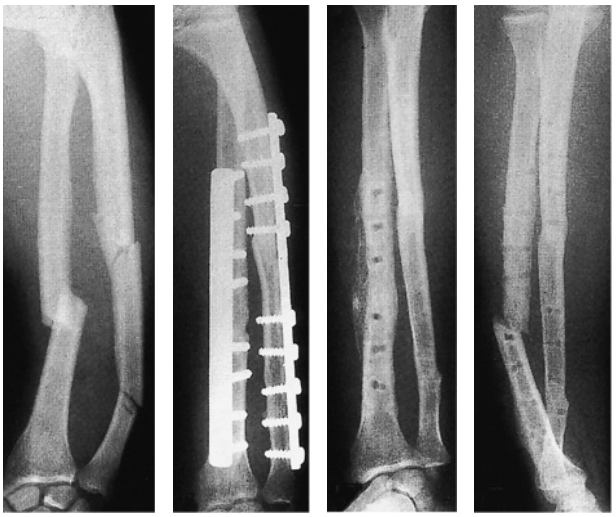
Fixation with flexible intramedullary nails is preferred, but they should be inserted with great care to avoid injury to the growth plates. Alternatively, a plate or K-wire fixation can be used.

Childhood fractures usually remodel well. One can accept 20 degrees of angulation in the distal third of the radius, 15 degrees in the middle third and 10 degrees in the proximal third, as long as there is at least 2 years or more of growth left. One can accept 100% translation as long as there is no more than 1 cm of shortening.

ADULTS

Unless the fragments are in close apposition, reduction is difficult and redisplacement in the cast is almost invariable. So predictable is this outcome that most surgeons opt for open reduction and internal fixation from the outset. The fragments are held by interfragmentary compression with plates and screws (Figure 25.18). Bone grafting is advisable if there is comminution or a loss of continuity in two-thirds of the circumference of the bone. The deep fascia is left open to prevent a build-up of pressure in the muscle compartments, and only the skin is sutured.

After the operation the arm is kept elevated until the swelling subsides, and during this period active exercises of the hand are encouraged. If the fracture is not comminuted and the patient is reliable, early range of movement exercises are commenced but lifting and sports are avoided. It takes 8–12 weeks for the bones to unite. With comminuted fractures or unreliable patients, immobilization in plaster is safer.



(a) (b) (c) (d)

Figure 25.18 Fractured radius and ulna in adults (a,b) These fractures are usually treated by internal fixation with sturdy plates and screws. However, removal of the implants is not without risk. (c,d) In this case, the radius fractured through one of the screw holes.

OPEN FRACTURES

Open fractures of the forearm must be managed meticulously. Antibiotics and tetanus prophylaxis are given as soon as possible; the wounds are copiously washed and nerve function and circulation are checked. At operation the wounds are excised and extended and the bone ends are exposed and thoroughly cleaned. The fractures are primarily fixed with compression screws and plates; if the wounds are absolutely clean, the soft tissues can be closed. If bone grafting is necessary, this is best deferred until the wounds are healed. If there is major soft-tissue loss, the bones are better stabilized by external fixation. The aim is to obtain skin cover as soon as possible; if plastic surgery services are available, these should be enlisted from the outset.

If there is any question of a compartment syndrome, the wounds should be left open and closed 24–48 hours later, with a skin graft if needed.

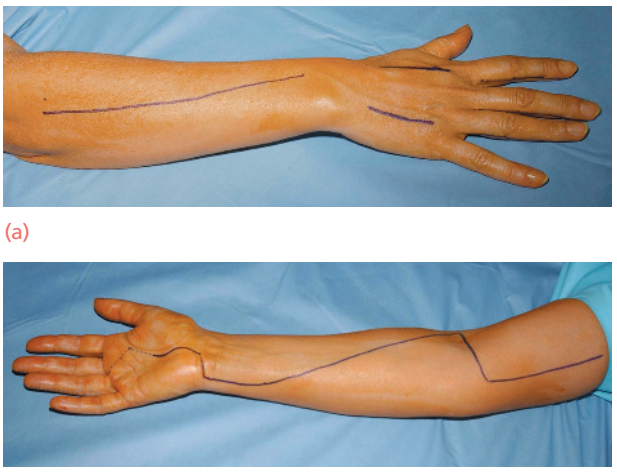
Complications

EARLY

Nerve injury Nerve injuries are rarely caused by the fracture, but they may be caused by the surgeon! Exposure of the radius in its proximal third risks damage to the posterior interosseous nerve where it is covered by the superficial part of the supinator muscle. The proximal fragment of radius may have rotated so the nerve may not be where it is expected. Surgical technique is particularly important here; the anterior Henry approach is safest.

Vascular injury Injury to the radial or ulnar artery seldom presents any problem, as the collateral circulation is excellent.

Compartment syndrome Fractures (and operations) of the forearm bones are always associated with swelling of the soft tissues, with the attendant risk of a compartment syndrome (Figure 25.19). The threat is



(a)

(b)

Figure 25.19 Compartment syndrome Incisions to relieve a compartment syndrome in the forearm.

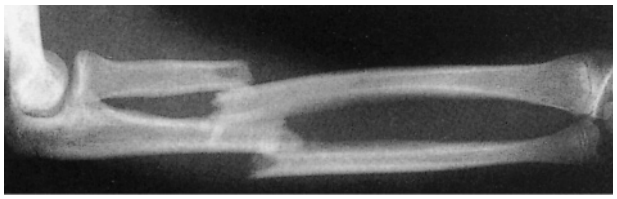
even greater, and the diagnosis more difficult, if the forearm is wrapped up in plaster. A distal pulse does not exclude compartment syndrome! The byword is 'watchfulness'; if there are any signs of circulatory embarrassment, treatment must be prompt and uncompromising.

LATE

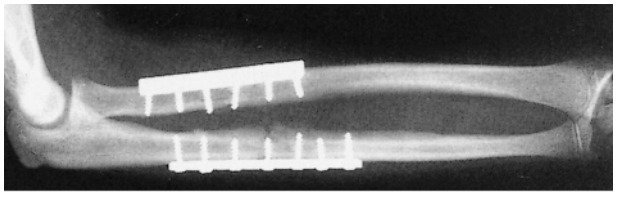
Delayed union and non-union Most fractures of the radius and ulna heal within 8–12 weeks; high-energy fractures and open fractures are less likely to unite. Delayed union of one or other bone (usually the ulna) is not uncommon; immobilization may have to be continued beyond the usual time. Non-union will require bone grafting and internal fixation.

Malunion With closed reduction there is always a risk of malunion, resulting in angulation or rotational deformity of the forearm, cross-union of the fragments (Figure 25.20), or shortening of one of the bones and disruption of the distal radioulnar joint. If pronation or supination is severely restricted, and there is no cross-union, mobility may be improved by corrective osteotomy. However, it can be very difficult to calculate the deformity and subsequent correction.

Complications of plate removal Removal of plates and screws is often regarded as a fairly innocuous procedure. Beware! Complications are common and they include damage to vessels and nerves, infection and fracture through a screw hole.



(a)



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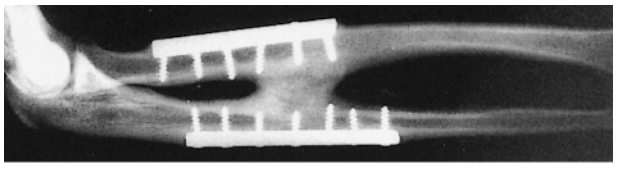


(c)

Figure 25.20 Fractured radius and ulna – cross-union
If the interosseous membrane is severely damaged, even successful plating (a,b) cannot guarantee that cross-union will not occur (c).

Fracture of a single forearm bone

Fracture of the radius alone is very rare and fracture of the ulna alone is uncommon. These injuries are usually caused by a direct blow – the 'nightstick fracture'. They are important for two reasons:

- An associated dislocation may be undiagnosed; if only one forearm bone is broken along its shaft and there is displacement, then either the proximal or the distal radioulnar joint must be dislocated. The entire forearm, elbow and wrist should always be X-rayed.
- Non-union is liable to occur unless it is realized that one bone takes just as long to consolidate as two.

Clinical features and X-rays

Ulnar fractures are easily missed – even on X-ray. If there is local tenderness, a further X-ray a week or two later is wise.

The fracture may be anywhere in the radius or ulna (Figure 25.21). The fracture line is transverse and displacement is slight. In children, the intact bone sometimes bends without actually breaking ('plastic deformation'). To assess an X-ray for this it is important to remember that the ulna is usually straight on a lateral radiograph.

Treatment

ISOLATED FRACTURE OF THE ULNA

The fracture may be undisplaced with good bone apposition, in which case a forearm cast leaving the elbow free can be sufficient. However, it takes about 8 weeks before full activity can be resumed. Angular or rotation alignment will affect forearm rotation; therefore rigid internal fixation will be preferable for many patients to allow earlier activity and reduces the risk of displacement or non-union.

ISOLATED FRACTURE OF THE RADIUS

Radius fractures are prone to rotary and angular displacement; to achieve reduction in children the forearm needs to be supinated for upper third fractures, neutral for middle third fractures and pronated for lower third fractures. The position is sometimes difficult to hold in children and just about impossible in adults; if so, then internal fixation with a compression plate and screws in adults, and preferably intramedullary nails in children, is better.

Middle/distal third fractures of the radius in children These are particularly unstable, being

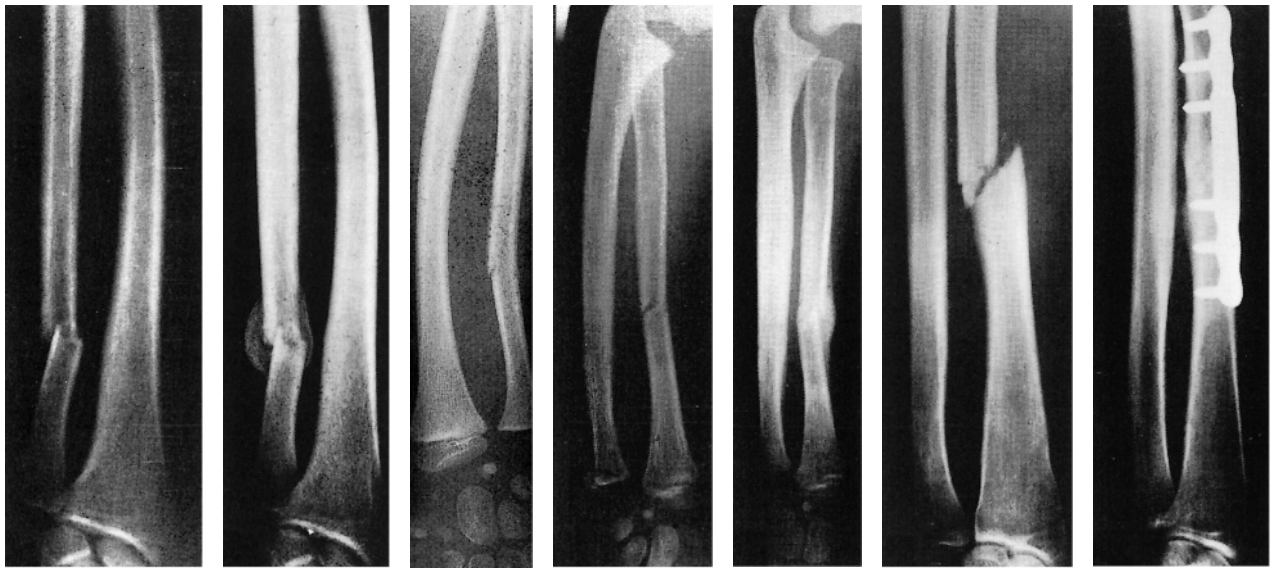


Figure 25.21 Fracture of one forearm bone *Fracture of the ulna:* A fracture of the ulna alone (a) usually joins satisfactorily (b); in children the intact radius may be bowed (c). *Fracture of the radius:* In a child, fracture of the radius alone (d) may join in plaster (e), but in adults a fractured radius (f) is better treated by plating (g).

deformed by the pull of the thumb abductors and pronator quadratus. They can be treated with an above-elbow cast in supination but, failing that, fixation with an intramedullary nail or Kirschner (K-) wires is advisable.

MONTEGGIA FRACTURE-DISLOCATION OF THE ULNA

The injury described by Monteggia in the early 19th century (without benefit of X-rays!) was a fracture of the shaft of the ulna associated with anterior dislocation of the proximal radioulnar joint; the radiocapitellar joint is inevitably dislocated or subluxated as well. More recently the definition has been extended to embrace almost any fracture of the ulna associated with dislocation of the radiocapitellar joint, including transolecranon fractures in which the proximal radioulnar joint remains intact. If the ulnar shaft fracture is angulated with the apex anterior (the commonest type), the radial head is displaced anteriorly and is usually intact; if the fracture apex is posterior, the radial is usually fractured on the capitellum but may be dislocated posteriorly, this carries a worse prognosis. In children, the ulnar injury may be an incomplete fracture (greenstick or plastic deformation of the shaft).

Mechanism of injury

Usually the cause is a fall on the hand; if at the moment of impact the body is twisting, its momentum may forcibly pronate the forearm. The radial head usually dislocates forwards and the upper third of the ulna fractures and bows forwards. Sometimes the causal force is hyperextension.

Clinical features

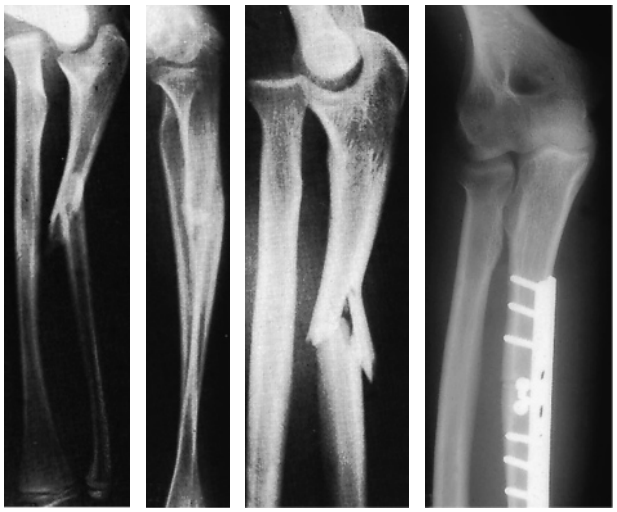
The ulnar deformity is usually obvious but the dislocated head of radius is masked by swelling. A useful clue is pain and tenderness on the lateral side of the elbow. The wrist and hand should be examined for signs of injury to the radial nerve.

X-rays

With isolated fractures of the ulna, it is essential to obtain a true anteroposterior and true lateral view of the elbow. In the usual case, the head of the radius (which normally points directly to the capitellum in every radiographic view) is dislocated forwards, and there is a fracture of the upper third of the ulna with forward bowing. Backward or lateral bowing of the ulna (which is much less common) is likely to be associated with, respectively, posterior or lateral displacement of the radial head. Transolecranon fractures also are often associated with radial head dislocation.

Treatment

The key to successful treatment is to restore the length of the fractured ulna; only then can the dislocated joint be fully reduced and remain stable. In adults, this means an operation through a posterior approach.



(a) (b) (c) (d)

Figure 25.22 Monteggia fracture-dislocation (a) The ulna is fractured and the head of the radius no longer points to the capitellum. In a child, closed reduction and plaster (b) is usually satisfactory; in the adult (c) open reduction and plating (d) is preferred.

The ulnar fracture must be accurately reduced, with the bone restored to full length, and then fixed with a plate and screws. The radial head usually reduces once the ulna has been fixed. Stability must be tested through a full range of flexion and extension. If the radial head does not reduce, or is not stable, open reduction should be performed.

If the elbow is completely stable, then flexion–extension and rotation can be started very soon after surgery. If there is doubt, the arm should be immobilized in plaster with the elbow flexed for 6 weeks.

Complications

Nerve injury Nerve injuries can be caused by over-enthusiastic manipulation of the radial dislocation or during the surgical exposure. Always check for nerve function after treatment. The lesion is usually a neurapraxia, which will recover by itself.

Malunion Unless the ulna has been perfectly reduced, the radial head remains dislocated and limits elbow flexion. In children, the radial head must be reduced and further surgery to correct ulna malalignment to facilitate reduction is performed. With increasing time delay from the index injury, other procedures will be required to stabilize the radial head reduction, unless the delay is substantial, in which case no treatment is advised. In adults, osteotomy of the ulna or perhaps excision of the radial head may be needed.

Non-union Non-union of the ulna should be treated by plating and bone grafting.

Special features in children

The general features of Monteggia fracture-dislocations are similar to those in adults. However, it is important to remember that the ulnar fracture may be incomplete (greenstick or plastic deformation); if this is not detected, and corrected, the child may end up with chronic subluxation of the radial head. Because of incomplete ossification of the radial head and capitellar epiphysis in children, these landmarks may not be easily defined on X-ray and a proximal dislocation could be missed. The X-rays should be studied very carefully and, if there is any doubt, X-rays should be taken of the other side for comparison.

Incomplete ulnar fractures can often be reduced closed, although considerable force is needed to straighten the ulna with plastic deformation. The position of the radial head is then checked; if it is not perfect, closed reduction can be completed by flexing and supinating the elbow and pressing on the radial head. The arm is then immobilized in a cast with the elbow in flexion and supination, for 3 weeks.

Complete fractures are best treated by open reduction and fixation.

GALEAZZI FRACTURE-DISLOCATION OF THE RADIUS

Mechanism of injury

This injury was first described in 1934 by Galeazzi. The usual cause is a fall on the hand, probably with a superimposed rotation force. The radius fractures in its lower third and the inferior radioulnar joint subluxates or dislocates.

Clinical features

The Galeazzi fracture is much more common than the Monteggia. Prominence or tenderness over the lower end of the ulna is the striking feature. It may be possible to demonstrate the instability of the radioulnar joint by 'ballotting' the distal end of the ulna (the 'piano-key sign') or by rotating the wrist. It is important also to test for an ulnar nerve lesion, which may occur.

X-rays

A transverse or short oblique fracture is seen in the lower third of the radius, with angulation or overlap. The distal radioulnar joint is subluxated or dislocated.

Treatment

As with the Monteggia fracture, the important step is to restore the length of the fractured bone. In children,

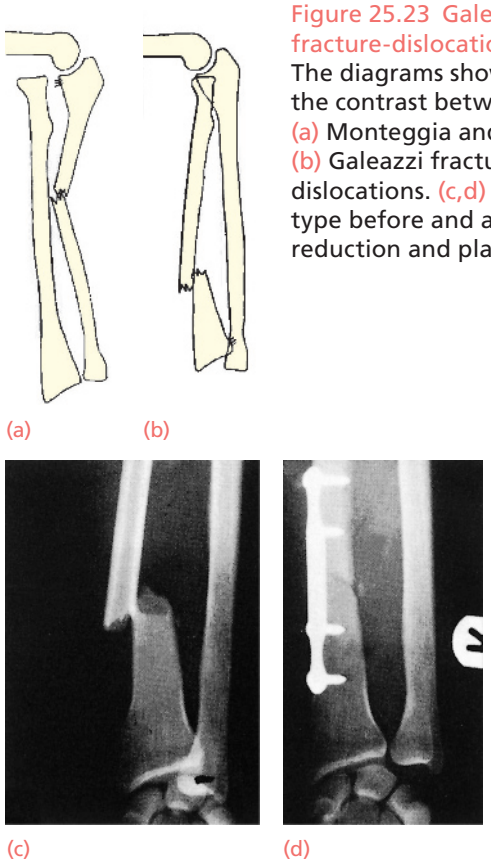


Figure 25.23 Galeazzi fracture-dislocation
The diagrams show the contrast between (a) Monteggia and (b) Galeazzi fracture-dislocations. (c,d) Galeazzi type before and after reduction and plating.

closed reduction is often successful; in adults, reduction is best achieved by open operation and compression plating of the radius. An X-ray is taken to ensure that the distal radioulnar joint is reduced.

There are three possibilities:

- *The distal radioulnar joint is reduced and stable.* No further action is needed. The arm is rested for a few days, then gentle active movements are encouraged. The radioulnar joint should be checked, both clinically and radiologically, during the next 6 weeks.
- *The distal radioulnar joint is reduced but unstable.* The forearm should be immobilized in the position of stability (usually supination), supplemented if required by a transverse K-wire. The forearm is splinted in an above-elbow cast for 6 weeks. If there is a large ulnar styloid fragment, it should be reduced and fixed.
- *The distal radioulnar joint is irreducible.* This is unusual. Open reduction is needed to remove the interposed soft tissues. The triangular fibrocartilage complex (TFCC) and dorsal capsule are then carefully repaired and the forearm immobilized in the position of stability (again, usually supination, supported by a wire if needed) for 6 weeks.

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Injuries of the wrist 26

David Warwick & Adam Watts
Children's sections: Joanna Thomas

FRACTURES OF THE DISTAL RADIUS IN ADULTS

Historically, distal radius fractures have been classified using eponymous terms such as Colles', Smith's or Barton's fracture, but these names can lead to confusion and misunderstanding. There is no classification that completely fulfils the requirement of guiding treatment or informing prognosis.

The distal end of the radius is subject to many different types of fracture, depending on factors such as age, transfer of energy, mechanism of injury and bone quality. Treatment options depend on whether the fracture is intra- or extra-articular and the degree of fragmentation of the joint surface and the metaphysis.

With any of these fractures, the wrist also can suffer substantial ligamentous injury causing instability to the carpus or distal radioulnar joint (DRUJ). These injuries are easily missed because the X-rays may look normal.

Low-energy dorsally displaced fractures ('Colles' fracture')

The injury that Abraham Colles described in 1814 ('*Colles' fracture*') is a transverse fracture of the radius just above the wrist, with dorsal displacement of the distal fragment. It is the most common of all fractures in older people, the high incidence being related to the onset of postmenopausal osteoporosis. Thus the patient is usually an older woman who gives a history of falling on her outstretched hand.

Mechanism of injury and pathological anatomy

Force is applied in the length of the forearm with the wrist in extension. The bone fractures at the corticocancellous junction and the distal fragment collapses into extension, dorsal displacement, radial tilt and shortening. Normal angles are shown in Figure 26.1.

Clinical features

We can recognize the most common fracture pattern (as Colles did long before radiography was invented) by the 'dinner-fork' deformity, with prominence on the back of the wrist and a depression in front. In patients with less deformity there may only be local tenderness and pain on wrist movements.

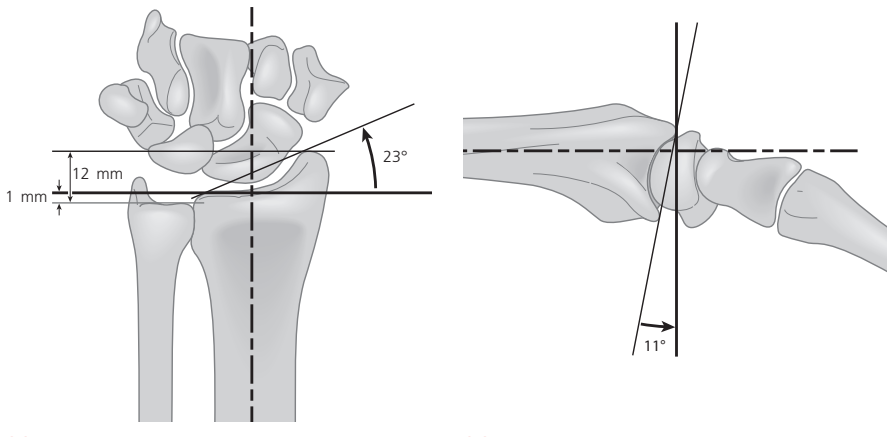


Figure 26.1 Distal radius fracture – normal angles Make sure that the articular congruency is restored.

Imaging

X-rays show there is a transverse fracture of the radius at the corticocancellous junction, and often the ulnar styloid process is broken off. The radial fragment is impacted into radial and backward tilt. Sometimes there is an intra-articular fracture; sometimes it is severely fragmented. If the configuration is not clear, a *CT scan* is very helpful in planning treatment.

Treatment

See Figures 26.2 and 26.3.

UNDISPLACED FRACTURES

If the fracture is undisplaced (or only very slightly displaced), a dorsal splint is applied for a day or two until the swelling has resolved, then the cast is completed. An X-ray is taken at 10–14 days to ensure that the fracture has not slipped; if it has, surgery may be required; if not, the cast can usually be removed after 5 weeks to allow mobilization.

DISPLACED FRACTURES

Displaced fractures must be reduced under anaesthesia (haematoma block, Bier's block, axillary block or general anaesthesia). The hand is grasped and longitudinal traction is applied (sometimes with extension of the wrist to disimpact the fragments); the distal fragment is then pushed into place by pressing on the dorsum while manipulating the wrist into flexion, ulnar deviation and pronation. If it is satisfactory, a dorsal plaster slab

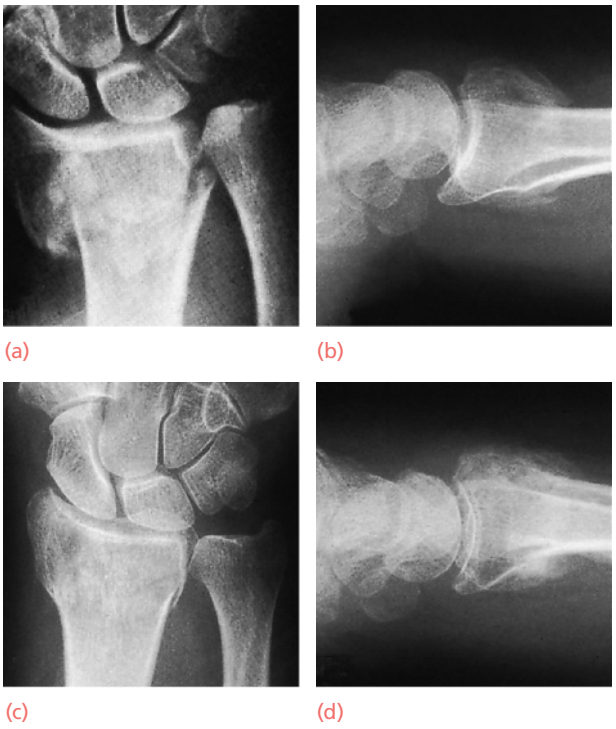


Figure 26.2 Low-energy distal radius fracture (a,b) The low-energy distal radius fracture is both displaced and angulated towards the dorsum and towards the radial side of the wrist. (c,d) Note how, after successful reduction, the radial articular surface faces correctly both distally and slightly volarwards.

Figure 26.3 Distal radius fracture Options include simple plaster (a) or external fixation (b) depending on the amount of comminution, stability of the fracture and patient demands. (c) Typical fixation with K-wires. (d) Dorsally angulated fracture fixed with volar locking plate (e,f).

is applied, extending from just below the elbow to the metacarpal necks and two-thirds of the way round the circumference of the wrist. The position is then checked by X-ray. It is held in position by a crepe bandage. *Extreme positions of flexion and ulnar deviation must be avoided*; 20 degrees in each direction is adequate.

The arm is kept elevated for the next day or two; shoulder, elbow and finger exercises are started as soon as possible. If the fingers become swollen, cyanosed or painful, there should be no hesitation in splitting the bandage.

At approximately 7 days, and if satisfactory then again at about 14 days, fresh X-rays are taken. This is because redisplacement is not uncommon, especially in the elderly after an initial manipulation. If the pattern is particularly unstable, or if there has been substantial slip since the first reduction X-rays but the position is only just acceptable at 14 days, another X-ray at 18–20 days, just prior to the fracture being too 'sticky' to manipulate again, is considered. If the position is probably not compatible with a good outcome and if the risks inherent in surgery are understood, manipulation and fixation with either percutaneous wires or a volar locking plate is undertaken. However, in older patients with low functional demands, modest degrees of displacement should be accepted because (1) outcome in these patients is not so dependent upon anatomical perfection; and (2) fixation of the fragile bone can be very difficult and is not without complications.

The fracture unites in about 6 weeks and, even in the absence of radiological proof of union, wrist exercises can be commenced.

Occasionally the fracture line exits transversely in the central part of the distal radius joint surface; the anterior cortex is intact and the dorsal fragment slips dorsally. These dorsal partial articular shear fractures tend to be more stable in cast but may require surgical stabilization.

IMPACTED OR FRAGMENTED LOW-ENERGY DISTAL RADIUS FRACTURES

With substantial impaction or fragmentation in osteoporotic bone, manipulation and plaster immobilization alone may be insufficient. The fracture can sometimes be reduced and held with percutaneous wires or a volar locking plate but, if impaction is severe, even this may not be enough to hold all the fragments or maintain length; in that case, other techniques, such as dorsal plating, locked intramedullary nails, external fixators, internal plate bridging the radius to the third metacarpal and bone grafts (synthetic or autogenous) are considered.

Volar locking plate or K-wires?

- *Volar locking plates* – There are several volar locking plates available. These are expensive and require surgical skill to apply properly and safely. They are applied to the anterior distal shaft-metaphysis of the radius, approaching the bone beneath pronator quadratus in the safe interval between the flexor carpi radialis and the radial artery, well away from the median nerve and its palmar cutaneous branch. The plate is applied sufficiently distal to support the fracture but proximal enough to avoid abrading the overlying thumb and finger flexors. The screws are fixed to the plate itself and are passed into the relatively stronger subchondral bone distally. These devices, allow stable fixation and thus early mobilization of the forearm.
- *K-wires* – K-wires are cheaper and more readily available. They must be passed with meticulous care to avoid impinging the vulnerable skin nerves and tendons. Two stout wires are the minimum; with softer or more comminuted bone three or four are preferred. They can be passed through the fragments into the radius shaft or through the fracture line as levers 'Kapandji wires'. Wires are not strong enough to allow early mobilization and a supplementary plaster, which will need changing once or twice, is needed.

The final outcomes and complication rates for the two methods appear the same. Early mobilization and thus earlier return to function might tempt certain patients towards a plate if the device is available and affordable.

Outcome

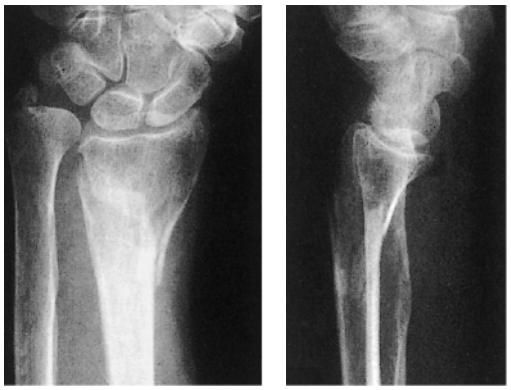
As Colles himself recognized, the outcome of these fractures in an older age group with lower functional demands is usually good, regardless of the cosmetic or the radiographic appearance. Poor outcomes can usually be improved by performing a corrective osteotomy if undertaken before secondary arthritis intervenes. The amount of displacement that can be accepted depends on patient factors such as age, comorbidity, functional demands, handedness and quality of bone, and treatment factors such as surgical skill and implants available. As a rule, poor outcome is associated with:

- loss of radial length by more than 3 mm
- dorsal tilt more than 15 degrees from neutral depending on age and function
- palmar tilt more than 20 degrees from neutral

Early correction should be considered. The tolerances might be less in a younger, higher demand patient.

VOLAR DISPLACED FRACTURE

Smith (a Dubliner, like Colles) described a fracture about 20 years later in which the distal fragment is angulated or displaced volarward ('*Smith's fracture*' – Figure 26.4). It is caused by a fall on the back of the hand and is an unstable injury due to the force generated by the long flexors crossing the wrist. There is a similar fracture with volar displacement which involves just the anterior half of the distal radius with an intact



(a) (b)

Figure 26.4 Smith's fracture (a,b) Here, in contrast to Colles' fracture, the displacement of the lower radial fragment is forwards – not backwards.

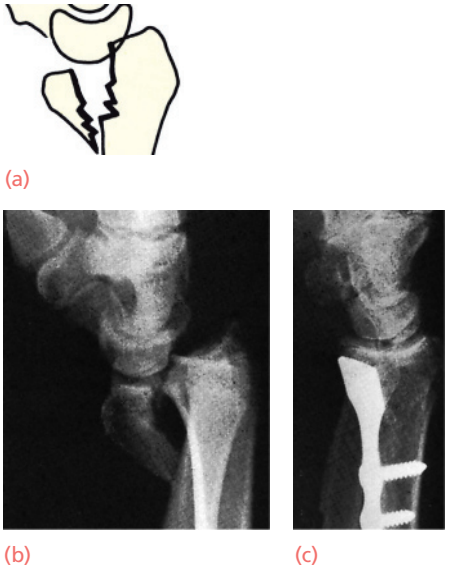
dorsal surface. The carpus slips forward with the anterior block of bone. Alternatively, the volar half of the distal radius surface can shear off; the fragment moves forwards, carrying the carpus with it. This is sometimes known as a '*Barton's fracture*' (Figure 26.5).

Clinical features

The patient presents with a wrist injury, but there is no dinner-fork deformity. Instead, there is a 'garden-spade' deformity.

X-rays

There is a fracture through the distal radial metaphysis; a lateral view shows that the distal fragment is displaced or tilted anteriorly. The entire metaphysis can be fractured, or there can be an oblique fracture exiting at the dorsal or volar rim of the radius.



(a)

(b) (c)

Figure 26.5 Fracture-subluxation (Barton's fracture) (a,b) The true Barton's fracture is a split of the volar edge of the distal radius with anterior (volar) subluxation of the wrist. (c) This has been reduced and held with a small anterior plate.

Treatment

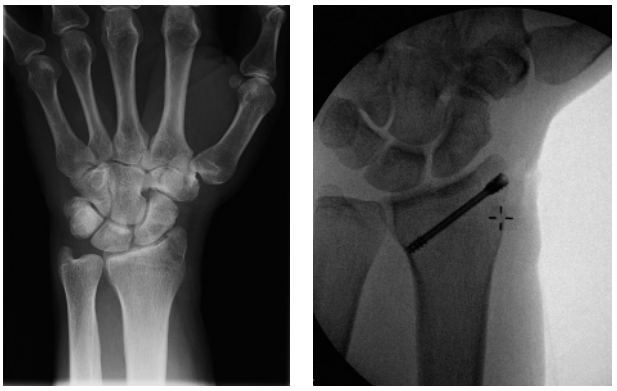
These fractures can be reduced by traction, supination and extension of the wrist, and the forearm immobilized in a cast for 6 weeks, but the risk of redisplacement is high and most advocate early surgical intervention with a volar plate to buttress the distal fragment. X-rays should be taken at 7–10 days to ensure that the fracture has not slipped, even when a plate is used. Particular attention should be paid to fragmentation of the volar-ulnar lip of the distal radius as this can readily displace if not adequately supported.

Fractured radial styloid

This injury is caused by forced radial deviation of the wrist and may occur after a fall, or when a starting handle 'kicks back' – the so-called '*chauffeur's fracture*' (Figure 26.6). The fracture line is transverse or oblique, extending laterally from the articular surface of the radius; the fragment is often undisplaced but the injury is commonly associated with a carpal ligament injury and this should be looked for. The energy transfer which breaks the radial styloid can rupture the scapholunate ligament or fracture the scaphoid. These associated injuries must always be carefully excluded.

Treatment

If there is displacement, it is reduced and the wrist is held in ulnar deviation by a plaster slab round the outer forearm, extending from below the elbow to the metacarpal necks. Imperfect reduction may lead to osteoarthritis; therefore, if closed, the fragment should be perfectly reduced and held with a K-wire or, even better, a buried cannulated screw. Associated ligamentous injuries should be addressed.



(a) (b)

Figure 26.6 Fractured radial styloid (a) X-ray; (b) fixation with cannulated percutaneous screw.

Fragmented intra-articular fractures in young adults

In the young adult, a fragmented intra-articular fracture is a high-energy injury (Figure 26.7). A poor outcome will probably result unless intra-articular congruity, fracture alignment and length are restored, and movements are started as soon as possible. For these patients, anatomical perfection is probably not essential but it is a proper ambition, and certainly a higher standard must be set than would be accepted for the typical osteoporotic fracture in an elderly, low-demand individual (Figure 26.8). In addition to the usual posteroanterior and lateral X-rays, oblique views and often CT scans are useful to show the fragment alignment.

Simple manipulation under anaesthetic and cast may be successful. X-rays are needed at about 7 days and, if the fracture is still undisplaced, at about 14 days. If the anatomy is not restored, then either closed reduction with percutaneous wires or an open reduction may well be necessary. All the fragments must be reduced into a good position and held in a stable manner. Volar locking plates are particularly useful. Note that there is often a separate ulnar palmar fragment which must be anatomically reduced (Figure 26.9); this can be a technical challenge,

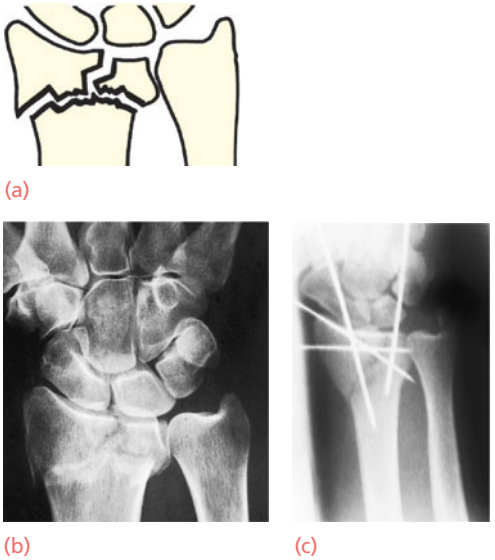


Figure 26.7 Comminuted fracture of the distal radius The 'die-punch fragment' of the lunate fossa of the distal radius (a,b) must be perfectly reduced and fixed; here this has been achieved by closed reduction and percutaneous K-wire fixation (c). The wires can be used as 'joy sticks' to manipulate the fragment back before fixation.

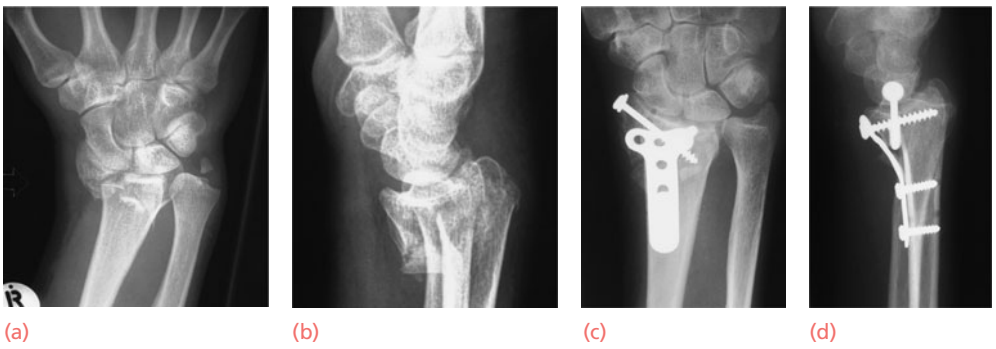


Figure 26.8 High-energy injuries in younger patients Perfect reduction is required.

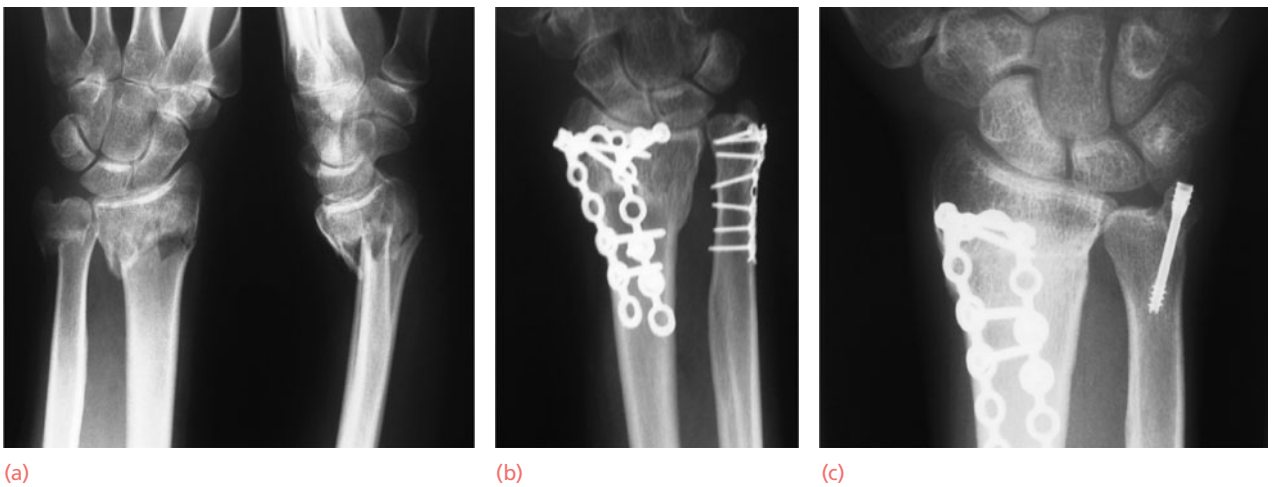


Figure 26.9 Don't forget the ulna (a) Fracture of radius and ulna, both unstable. (b) Both bones fixed. (c) Ulnar styloid fracture fixed to prevent instability of distal radioulnar joint.

needing quite a high degree of surgical skill; special implants are available to address this fragment.

Complications of distal radius fractures

See Figure 26.10.

EARLY

Circulatory problems The circulation in the fingers must be checked regularly; Elevation, early finger movement and avoiding undue flexion of the elbow are important to encourage venous drainage. The bandage holding the splint may need to be split or loosened.

Nerve injury Compression of the median nerve in the carpal tunnel is fairly common. If it occurs soon after injury and if the symptoms are mild, this may resolve with release of the dressings and elevation. If symptoms are severe or persistent, the transverse ligament should be divided. The cutaneous nerves (especially the superficial radial nerve) are at risk with clumsy K-wire insertion; the median nerve and the palmar cutaneous branch can be damaged during volar plate fixation with an inept exposure or clumsy retraction.

Complex regional pain syndrome (CRPS) This condition (previously known as reflex sympathetic dystrophy or Sudeck's atrophy) is probably quite common, but fortunately it seldom progresses to the full-blown picture. There may be swelling and tenderness of the finger joints, and vasomotor changes such as marbling of the skin, altered sweating and temperature sensitivity. The risk of this condition may be reduced by conscientious finger exercises, avoiding tight dressings and taking regular high-dose vitamin C.

Ulnar corner pain and instability Ulnar corner pain is very common after a distal radius fracture. The twisting and bending forces that displace the radius will be transferred to the attachment of the radioulnar ligaments (triangular fibrocartilage) into the base of the ulnar styloid. This is displayed as the obvious ulnar styloid fracture on about 50% of X-rays or, if the insertion is torn rather than the styloid avulsed, with persisting ulnar corner pain. Occasionally the avulsion is so significant that the DRUJ is unstable. If so, reattachment of the ulnar styloid with a screw or a wire loop is needed.

In the absence of instability, ulnar corner pain usually settles after several months even if there is a fractured styloid. If pain fails to settle, a steroid injection or arthroscopic surgery may help.

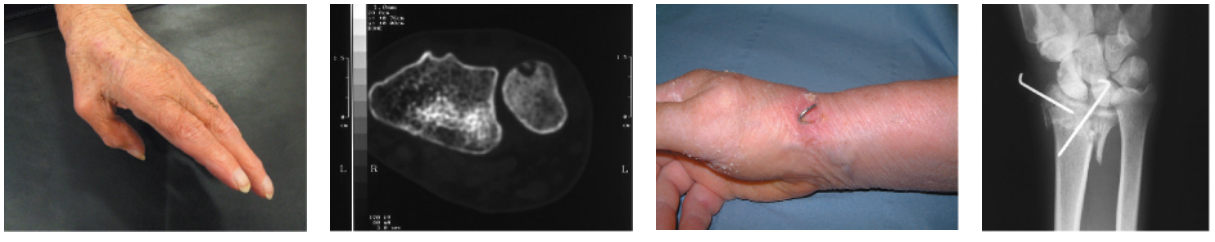
Associated injuries of the carpus Injuries of the carpus are easily overlooked while attention is focused on the radius. Carpal injuries, especially if there is a radial styloid fracture, must be excluded by careful clinical and X-ray examination, occasionally supplemented by MRI or arthroscopy.

Redisplacement There is a tendency for radius fractures to redisplace, especially those shearing fractures which run transversely into the joint line, fractures with volar tilt or fractures which have been manipulated or are particularly comminuted. Careful follow-up with X-rays is needed for about 14 days, and occasionally longer if the fracture is particularly unstable. Some of the most unstable fractures are best stabilized surgically from the outset.

LATE

Malunion Malunion is common, either because reduction was not complete or because displacement within the plaster, and sometimes even despite surgical fixation, was overlooked. The appearance is unsightly. Problems include loss of rotation (due to DRUJ incongruity), weakness, stiffness and pain (from intra-articular incongruity or malalignment of the midcarpal joint). In most cases treatment is not necessary but, if symptoms are intrusive, correction of the radius anatomy by osteotomy can be very effective. The distal end of the ulna should not be excised to restore rotation, except in the most frail. Otherwise intractable weakness and pain can result.

Delayed union and non-union Non-union of the radius is rare unless there is infection or severe bone loss with instability. Surgical treatment is complex



(a) (b) (c) (d)

Figure 26.10 Colles' fracture complications (a) Rupture of extensor pollicis longus; (b) malunion – CT scan showing incongruity of the DRUJ; (c) infected K-wire; (d) failed fixation as the wires have cut through the osteoporotic bone.

and often a fusion is needed. The ulnar styloid process often joins by fibrous tissue only and remains painful and tender for several months. Treatment is rarely required.

Tendon rupture Rupture of extensor pollicis longus (EPL) occasionally occurs a few weeks after an apparently trivial undisplaced fracture of the lower radius. Tendon transfer (using the spare index finger extensor) works well. The flexor pollicis longus (FPL) tendon is vulnerable with a misplaced volar plate; a dorsal plate or a protruding screw in a volar plate can abrade EPL and the other finger extensors.

Carpal instability The patient may present years later with chronic carpal instability if a substantial ligament injury was overlooked at the time of the original fracture.

Secondary osteoarthritis Fractures that disrupt the joint congruency may eventually lead to secondary osteoarthritis. While *radiological* changes are quite common, *symptomatic* arthritis is remarkably rare after distal radius fractures. Symptoms of pain and stiffness develop slowly but disability is often not severe. Splints, painkillers and steroid injections may help. Occasionally a partial wrist fusion (e.g. radius-scaphoid-lunate) or total wrist replacement might be offered; in someone with low functional demands who understands the risk of loosening, a wrist replacement is an option.

DISTAL FOREARM FRACTURES IN CHILDREN

The distal radius and ulna are among the most common sites of childhood fractures. The break may occur through the distal radial physis or in the metaphysis of one or both bones. Metaphyseal fractures are often incomplete or greenstick (Figure 26.11).

Mechanism of injury

The usual injury is a fall on the outstretched hand with the wrist in extension; the distal fragment is forced posteriorly. However, sometimes the wrist is in flexion and the fracture is angulated anteriorly. Lesser force may do no more than buckle the metaphyseal cortex (a type of compression fracture, or 'torus' fracture).

Clinical features

There is usually a history of a fall, though this may be passed off as one of many childhood spills. The wrist is painful, and often quite swollen; sometimes there is an obvious 'dinner-fork' deformity.

X-rays

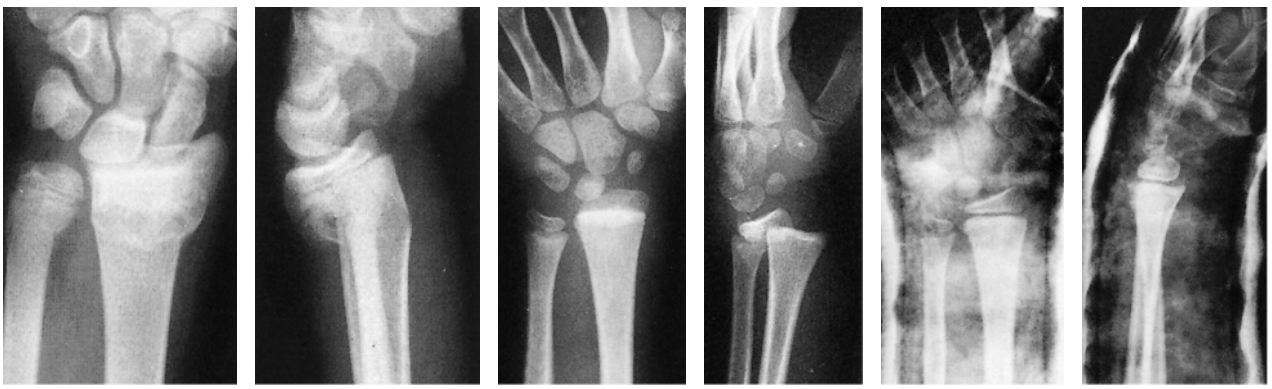
The precise diagnosis is made on the X-ray appearances. There are four types:

- *Physeal fractures* are usually Salter–Harris type II, with the epiphysis shifted and tilted backwards and radially. Type V injuries are unusual and are more likely diagnosed in retrospect when premature epiphyseal fusion occurs.
- *Incomplete fractures:*
  - *torus (buckle) fractures:* appear as a mere 'buckle' on a cortex
  - *greenstick fractures:* where one cortex is broken and the other is just bent.
- *Undisplaced fractures.*
- *Adult-pattern displaced fractures.*

Treatment

PHYSEAL FRACTURES

Physeal fractures are reduced, under anaesthesia, by pressure on the distal fragment. The arm is



(a) (b) (c) (d) (e) (f)

Figure 26.11 Distal forearm fractures in children (a,b) In older children the fracture is usually slightly more proximal than a true Colles', and often merely a greenstick or buckling injury. (c,d) In young children physeal fractures are usually Salter–Harris type I or II. In this case, accurate reduction has been achieved (e,f).

immobilized in a full-length cast with the wrist slightly flexed, the ulnar deviated and the elbow at 90 degrees. The cast is retained for 4 weeks. These fractures very rarely interfere with growth. Even if reduction is not absolutely perfect, further growth and modelling will obliterate any deformity. Patients seen more than 2 weeks after injury are best left untreated because manipulation at this stage may damage the growth plate.

UNDISPLACED OR BUCKLE/TORUS FRACTURES

These are stable injuries, so named because the force applied to the bone causes one side to compress, leading to the other side of the bone bending or buckling. This type of injury requires immobilization in a splint (not necessarily a formal plaster cast). The splint or plaster can be removed 3–4 weeks post injury and the child can be discharged with advice for finger and wrist movement.

GREENSTICK FRACTURES

Greenstick fractures are incomplete fractures where one side of the bone is bent (usually the side of the bone experiencing a compressive force), but the other side of the bone (usually the side experiencing tension from the force applied) is fractured. They are usually easy to reduce but may redisplace in the cast in 10% of cases. Some degree of residual angulation can be accepted: in children under 10, up to 30 degrees; in children over 10, up to 15 degrees. If the deformity is greater, the fracture is reduced by thumb pressure and the arm is immobilized with three-point fixation in a full-length cast with the wrist and forearm in neutral and the elbow flexed 90 degrees. The cast is changed and the fracture re-X-rayed after 1 week; if it has redisplaced, a further manipulation can be carried out and held in place with a K-wire. The cast is finally discarded after 6 weeks.

COMPLETE FRACTURES

These can be embarrassingly difficult to reduce, especially if the ulna is intact. The fracture is manipulated in much the same way as an adult dorsally displaced distal radius fracture; however, it is often necessary to exaggerate the deformity to ensure that the periosteum is disimpacted before attempting the reduction. The reduction is checked by X-ray and a full-length cast is applied with the wrist in neutral and the forearm supinated. After 2 weeks, a check X-ray is obtained; the cast is kept on for 6 weeks. If the fracture slips, especially if the ulna is intact, it should be stabilized with a percutaneous K-wire. Wiring should be considered in all cases where the fracture fragment is displaced 100% or more as the risk of redisplacement is very high.

Complications

EARLY

Forearm swelling and threatened compartment syndrome This dire combination can be prevented by avoiding over-forceful or repeated manipulations, splitting the plaster, elevating the arm for the first 24–48 hours and encouraging finger exercises.

LATE

Malunion This late sequel is uncommon in children under 10 years of age. Deformity of as much as 30 degrees will straighten out with further growth and remodelling over the next 1–2 years. This should be carefully explained to the worried parents.

Radioulnar discrepancy Premature fusion of the radial epiphysis may result in bone length disparity and subluxation of the radioulnar joint. If this is troublesome, the radius can be lengthened and, if the child is near to skeletal maturity, the ulnar physis can be fused surgically.

CARPAL INJURIES

Fractures and dislocations of the carpal bones are common. They vary greatly in type and severity. *These should never be regarded as isolated injuries; the entire carpus suffers*, and sometimes, long after the fracture has healed, the patient still complains of pain and weakness in the wrist.

The commonest carpal injuries, after the distal radius fracture of course, are:

- simple self-limiting sprains of the joint capsule and interosseous ligaments
- more serious unstable tears of the joint capsule and interosseous ligaments
- fracture of a carpal bone (usually the scaphoid)
- injury of the triangular fibrocartilage complex (TFCC) and distal radioulnar joint
- dislocations of the lunate or the bones around it.

Clinical assessment

Following a fall, the patient complains of pain in the wrist. There may be swelling or deformity of the joint. Tenderness should be carefully localized; undirected prodding will confuse both the patient and the examiner. The blunt end of a pencil is helpful in testing for point tenderness. For scaphoid fractures, all of the following should be performed but only one might be positive:

- anatomical snuffbox tenderness, although this is non-specific
- pain on pushing the scaphoid tubercle backwards, which is more specific
- pain on axial loading of the thumb
- pain on passive ulnar deviation.

When the scapholunate ligament is injured, tenderness is present just beyond Lister's tubercle; for lunate dislocation, in the middle of the wrist; for triquetral injuries, beyond the head of the ulna; for hamate fractures, at the base of the hypothenar eminence; for TFCC injuries, over the dorsum of the ulnocarpal joint. Movements are often limited (more by pain than by stiffness) and they may be accompanied by a palpable catch or an audible clunk.

Imaging

X-rays are the key to diagnosis. There are three golden rules:

- Accept only high-quality films.
- If the initial X-rays are 'normal', treat the clinical diagnosis and immobilize the wrist. 10–15% of scaphoid fractures are not visible on initial X-rays.
- Early MRI reduces uncertainty and streamlines care.

If MRI is not available, repeated X-rays are needed 2 weeks later (as shifting of the bones and resorption at the fracture line can make the fracture more apparent). If there is still doubt after a further 2 weeks, X-ray again.

Initially, four standard views are obtained:

- PA with wrist in ulnar deviation
- lateral
- semi-pronated oblique
- semi-supinated oblique.

An AP view with the fist clenched can be added if there is a suspicion of a scapholunate injury.

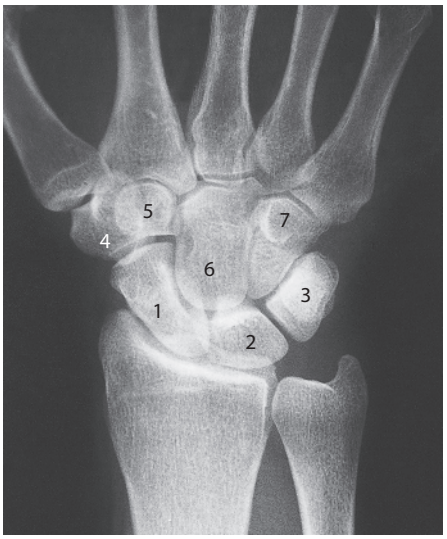


Figure 26.12 X-ray appearance of the normal carpus
X-ray of a normal wrist showing the shape and disposition of the eight carpal bones: 1 scaphoid; 2 lunate; 3 triquetrum overlain by pisiform; 4 trapezium; 5 trapezoid; 6 capitate; and 7 hamate.

The examiner should be familiar with the normal X-ray anatomy of the carpus (Figures 26.12 and 26.13) in all the standard views, so that he or she can visualize a three-dimensional picture from the two-dimensional, overlapping images of the carpal bones in carpal injuries (Figure 26.14).

In the anteroposterior X-ray note the shape of the carpus, whether the individual bones are clearly outlined and whether there are any abnormally large gaps suggesting disruption of the ligaments. The scaphoid may be fractured; or it may have lost its normal bean shape and look squat and foreshortened, sometimes with an inner circular density (the

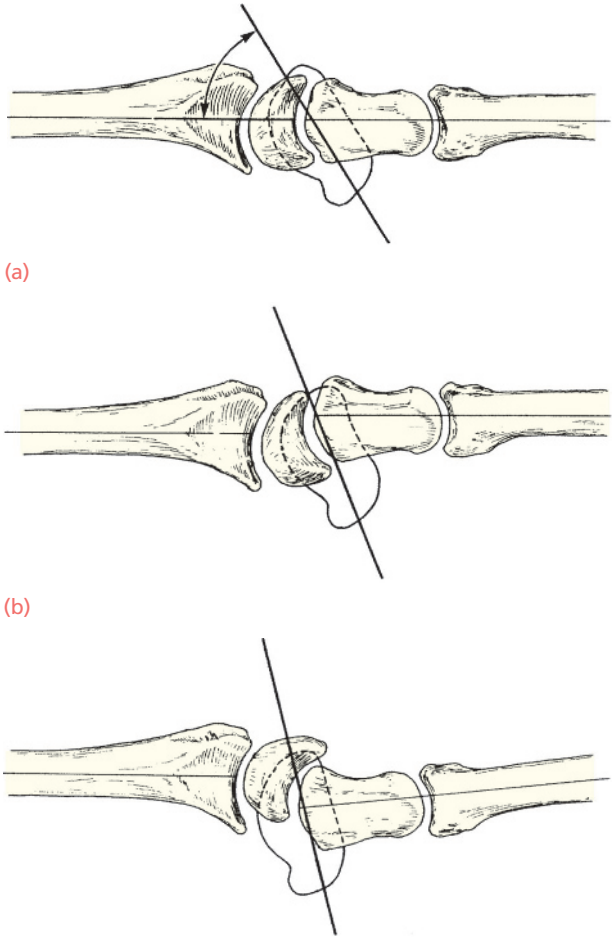


Figure 26.13 Carpal instability – X-ray patterns
(a) Normal lateral view. The radius, capitate and middle metacarpal lie in a straight line and the scaphoid axis is angled at 45° to the line of the radius. (b) Dorsal intercalated segmental instability (DISI). The lunate is tilted dorsally and the scaphoid is tilted somewhat volarwards; the axes of the capitate and metacarpals now lie behind (dorsal to) that of the radius. (c) Volar intercalated segmental instability (VISI). The lunate and scaphoid are tilted somewhat volarwards and the capitate and metacarpals lie anterior (volar) to the radius.

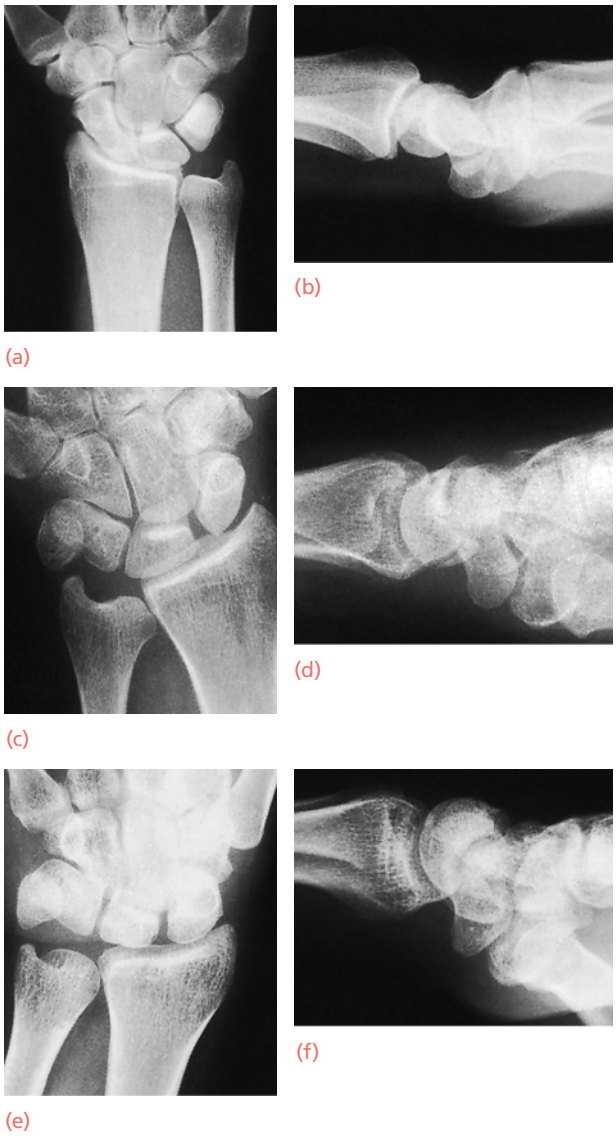


Figure 26.14 Carpal injuries (a,b) Normal appearances in AP and lateral X-rays. (c,d) Following a 'sprained wrist' this patient developed persistent pain and weakness. X-rays showed (c) scapholunate dissociation and (d) dorsal rotation of the lunate (the typical DISI pattern). (e,f) This patient, too, had a sprained wrist. The AP and lateral X-rays show foreshortening of the scaphoid and volar rotation of the lunate (VISI).

cortical ring sign) – features of an end-on view when the bone is hyperflexed because of damage to the restraining scapholunate ligament. The lunate is normally quadrilateral in shape but, if it is dislocated, it looks triangular.

In the lateral X-ray the axes of the radius, lunate, capitate and third metacarpal are co-linear, and the scaphoid projects at an angle of about 45 degrees to this line. With traumatic instability the linked carpal segments collapse (like the buckled carriages of a derailed train). Two patterns are recognized: dorsal intercalated segment instability (DISI), in which the lunate is torn from the scaphoid and tilted backwards; and volar intercalated segment instability (VISI), in which the lunate is torn from the triquetrum and turns towards the palm; the capitate shows a complementary dorsal tilt. There may be a flake fracture off the back of a carpal bone (usually the triquetrum) that indicates avulsion of the dorsal radiocarpal ligament.

Special X-ray studies are sometimes helpful: a *carpal tunnel view* may show a fractured hook of hamate, and *motion studies* in different positions may reveal a subluxation. A *radioisotope scan* will confirm a wrist injury although it may not precisely localize it.

MRI is sensitive and specific (especially for detecting undisclosed fractures or Kienböck's disease), but unless the most modern machines are used and very fine cuts are taken, it may miss TFCC and interosseous ligament tears where the addition of arthrography may be helpful.

Arthroscopy

Wrist arthroscopy is probably the best way of demonstrating TFCC or interosseous ligament tears if there is any doubt from imaging.

Principles of management

'Wrist sprain' should not be diagnosed unless a more serious injury has been excluded with certainty. Even with apparently trivial injuries, ligaments are sometimes torn and the patient may later develop carpal instability.

If the X-rays are normal but the clinical signs strongly suggest a carpal injury, the wrist should be immobilized while the underlying diagnosis is found. An MRI scan is ideal (preferably with arthrography) to show or exclude a major fracture or ligament injury. If MRI is not available, a bone scan will exclude a substantial bone injury but is not particularly specific if it is positive. Arthroscopy is considered if there is some uncertainty about a scapholunate or lunotriquetral ligament injury. The aim is to treat as soon as possible if there is an injury, or to avoid an unnecessary period of immobilization and time from work. If these tests are not readily available, the patient should be re-examined repeatedly until the symptoms settle or a firm diagnosis is made.

The more common lesions are dealt with below.

Fractured scaphoid

Scaphoid fractures account for almost 75% of all carpal fractures although they are rare in the elderly and in children.

Mechanism of injury and pathological anatomy

The scaphoid lies obliquely across the two rows of carpal bones and is also in the line of loading between the thumb and forearm. The combination of forced carpal movement and compression, as in a fall on the dorsiflexed hand, exerts severe stress on the bone and it is liable to fracture. Fractures occur in three anatomical locations: distal tubercle, waist and proximal pole. Some fractures, especially distal oblique and waist fractures, are unstable, which predisposes to non-union or malunion.

The blood supply of the scaphoid arises from the dorsal distal pole. This means that the proximal pole has a poor blood supply and is less likely to heal than the distal pole.

Clinical features

The appearance may be deceptively normal, but the astute observer can usually detect fullness in the anatomical snuffbox; the scaphoid must be very carefully examined as described above with particular attention to pain on axial compression and ulnar distraction, as well as tenderness over the tubercle and snuffbox.

X-rays

ACUTE FRACTURES

The four standard scaphoid views (described above) must be examined very carefully – often a recent fracture shows only in the oblique view. Usually the fracture line is transverse, and through the narrowest part of the bone (waist), but it may be more proximally situated (proximal pole fracture). Sometimes only the tubercle of the scaphoid is fractured (Figure 26.15).

It is very important to look for subtle signs of displacement or instability, including obliquity of the fracture line, opening of the fracture line, angulation of the distal fragment and foreshortening of the scaphoid image. Repeat X-rays may be needed after 2 weeks (and sometimes even later) before the fracture line shows.

DELAYED UNION AND NON-UNION

If union is delayed, the fracture line becomes more apparent and a 'hard' border can develop, sometimes even making it seem as if there is an extra carpal bone. As the non-union establishes itself, cavitation appears on either side of the break. In the distal pole and waist, the distal fragment tends to flex forwards and the proximal pole tends to tilt backwards. This gives the appearance of a round scaphoid tubercle on

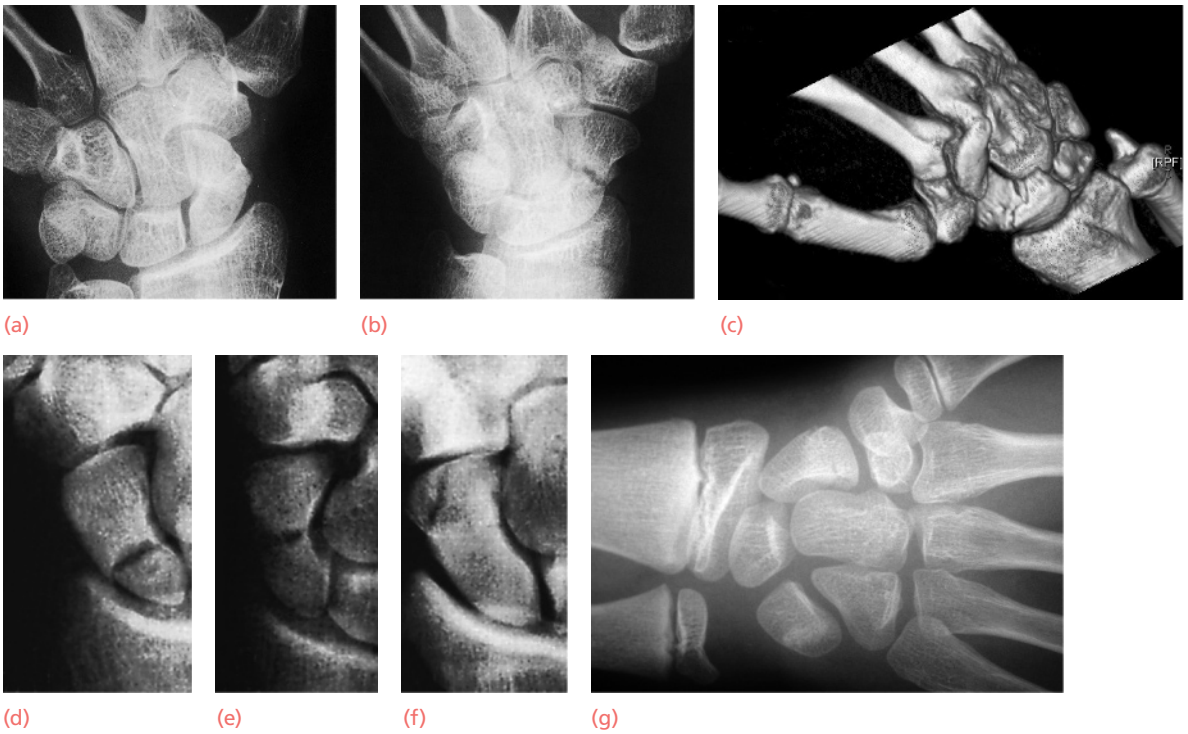


Figure 26.15 Fractures of the scaphoid – diagnosis (a) The initial AP view often fails to show the fracture; (b) always ask for a 'scaphoid series', including two oblique views. If the clinical features are suggestive of a fracture, immobilize the wrist and repeat the X-ray 2 weeks later when the fracture is more likely to be apparent. (c) A CT scan is useful for showing the fracture configuration. The fracture may be through (d) the proximal pole, (e) the waist, or (f) the scaphoid tubercle. Occasionally these fractures are seen in children (g).

the PA view ('ring sign') and a dorsally tilted proximal pole and lunate on the lateral view (the so-called DISI). Non-union of the proximal pole does not lead to malalignment but does lead to crumbling and relative sclerosis of the proximal pole – pathognomonic or *avascular necrosis*.

Treatment

See Figure 26.16.

FRACTURE OF THE SCAPHOID TUBERCLE

These are treated in a cast for 4–6 weeks. Usually there are no complications but occasionally there is a non-union needing excision of a small fragment or grafting of a larger fragment.

UNDISPLACED WAIST FRACTURES

These can be treated in two ways: *plaster* or *percutaneous fixation*.

Plaster Around 90% of waist fractures should heal in plaster (a neutral forearm cast from the upper forearm to just short of the metacarpophalangeal joints of the fingers; the thumb is not incorporated). It is retained (and if necessary repaired or renewed) for 6–8 weeks.

The plaster is removed and the wrist examined clinically and radiologically. If there is no tenderness and the X-ray shows full healing, the wrist is left free; a CT scan is the most reliable means of confirming union if in doubt.

If the scaphoid is tender, or the fracture is still visible on X-ray, the cast is reapplied for a further 4 weeks. At that stage, one of two pictures may emerge: (1) the wrist is painless and the fracture has healed – the cast can be discarded; or (2) the X-ray shows signs of delayed healing (bone resorption and cavitation around the fracture) – union can be hastened by bone grafting and internal fixation.

Percutaneous fixation This should be considered for those patients who do not want to endure

Figure 26.16 Fractures of the scaphoid – treatment (a) Scaphoid plaster – the thumb is optional. (b,c) CT scan showing collapse, which needs a graft from the front to wedge it open. (d) Delayed union, treated successfully by (e) bone grafting and screw fixation. (f) Waist fracture treated with distal radius graft. (g) Non-union with avascular necrosis and secondary osteoarthritis can be treated by (h) scaphoid excision and four-corner fusion or (i) proximal row carpectomy.

prolonged plaster immobilization and who want to get back to work or sport earlier (but they must still avoid impact or heavy load on the wrist until the fracture is healed). A screw is passed through a small incision in the front of the scaphoid tubercle. Special cannulated screws and technical perfection are essential.

DISPLACED FRACTURES

These can also be treated in plaster, but the outcome is less predictable: it may not heal or may heal in a poor position. It is better to reduce the fracture (closed if possible, otherwise open) and to fix it with a compression screw. This should increase the likelihood of union and reduce the time of immobilization.

PROXIMAL POLE FRACTURES

These may heal in plaster, and probably will if left long enough. However, the risk of non-union and the disadvantages of prolonged immobilization are such that early surgical fixation (through a small dorsal incision) should be considered.

Complications

Avascular necrosis The proximal fragment may die, especially with small proximal pole fractures which are further from their blood supply, and then at 2–3 months it appears dense on X-ray. Although revascularization and union are theoretically possible in some cases, this is unpredictable and may take many years. For small proximal pole fragments, the bone can be stabilized by a very small screw; if there is space, a bone graft from the distal radius is inserted. There may be a greater chance of healing if the graft is attached to an arterial pedicle. Alternatively, the fragment can be grafted through the arthroscope and stabilized with fine K-wires. If this fails, the dead fragment can be excised but the wrist tends to collapse after this procedure and arthritis develops. A better option would be a proximal row carpectomy (to remove the entire proximal row of carpal bones) or a scaphoid excision and four-corner fusion (i.e. remove the scaphoid and fuse the capitate–hamate–triquetrum–lunate).

Non-union Sometimes a patient is seen for the first time with a 'sprain' but X-rays show an old, un-united fracture with sclerosed edges that may be several years old. 3–4 weeks in plaster may suffice to make him or her comfortable once again, and no further treatment is required.

In older patients with low demands, and those who are completely asymptomatic, the non-union may be left untreated if the patient agrees after full discussion.

In the more acute stage, by 3 months (and sometimes earlier) it may be obvious that the fracture will not unite. Surgery should be advised, especially in the younger, more vigorous patient, because this probably reduces the chance of later, symptomatic osteoarthritis. There are various options, depending on the fracture configuration and the surgeon's skill:

- If the scaphoid has folded into a flexed 'humpback' shape, then it is approached from the front and a wedge of corticocancellous iliac crest graft is inserted to restore the shape of the bone. The graft is fixed with a buried screw and/or K-wires. If the scaphoid has not collapsed, the graft is inserted into a trough carved into the front or back of the scaphoid and again stabilized with a screw or wires.
- A vascularized graft may increase the union rate, especially if there is avascular necrosis. Usually these use a small block of distal radius bone based on a tiny artery close to the scaphoid. Free microvascular grafts have been suggested.
- Arthroscopic grafting and fixation is also an option for those with the equipment and skill.

Osteoarthritis Non-union or avascular necrosis may lead to secondary osteoarthritis of the wrist. If the arthritis is localized to the distal pole, excising the radial styloid may help. As the arthritis progresses, changes appear in the scaphocapitate joint then the capitate-lunate joint. The lunate-radius joint is never affected, thus allowing salvage procedures – either proximal row carpectomy or four-corner fusion.

Fractures of other carpal bones

Fractures of other carpal bones are all rare; they can be easily overlooked. Careful examination, and meticulous inspection of X-rays, especially after a significant fall or blow, is mandatory. If in any doubt, CT or MRI is invaluable. Examples are shown in Figure 26.17.

Triquetrum

Avulsion of the dorsal ligaments is not uncommon; analgesics and splintage for a few weeks are all that is required. Occasionally the body is fractured; it usually heals after 4–6 weeks in plaster although displaced fractures (seen on CT) need fixation.

Hamate

A fracture of the *hook of hamate* follows a direct blow to the palm of the hand (typically a poor golf shot). These fractures cannot be seen on routine X-rays; a carpal tunnel view, CT or MRI is needed. The fracture does not heal readily; if symptoms are prolonged, then the fragment is excised, taking care not to damage the ulnar nerve. Fractures of the *body* are rare. They are also difficult to define on plain X-rays. If the CT scan shows a displaced fracture, fixation may be needed. Fractures of the *distal dorsal lip* are associated with CMC dislocations and are discussed in Chapter 27.

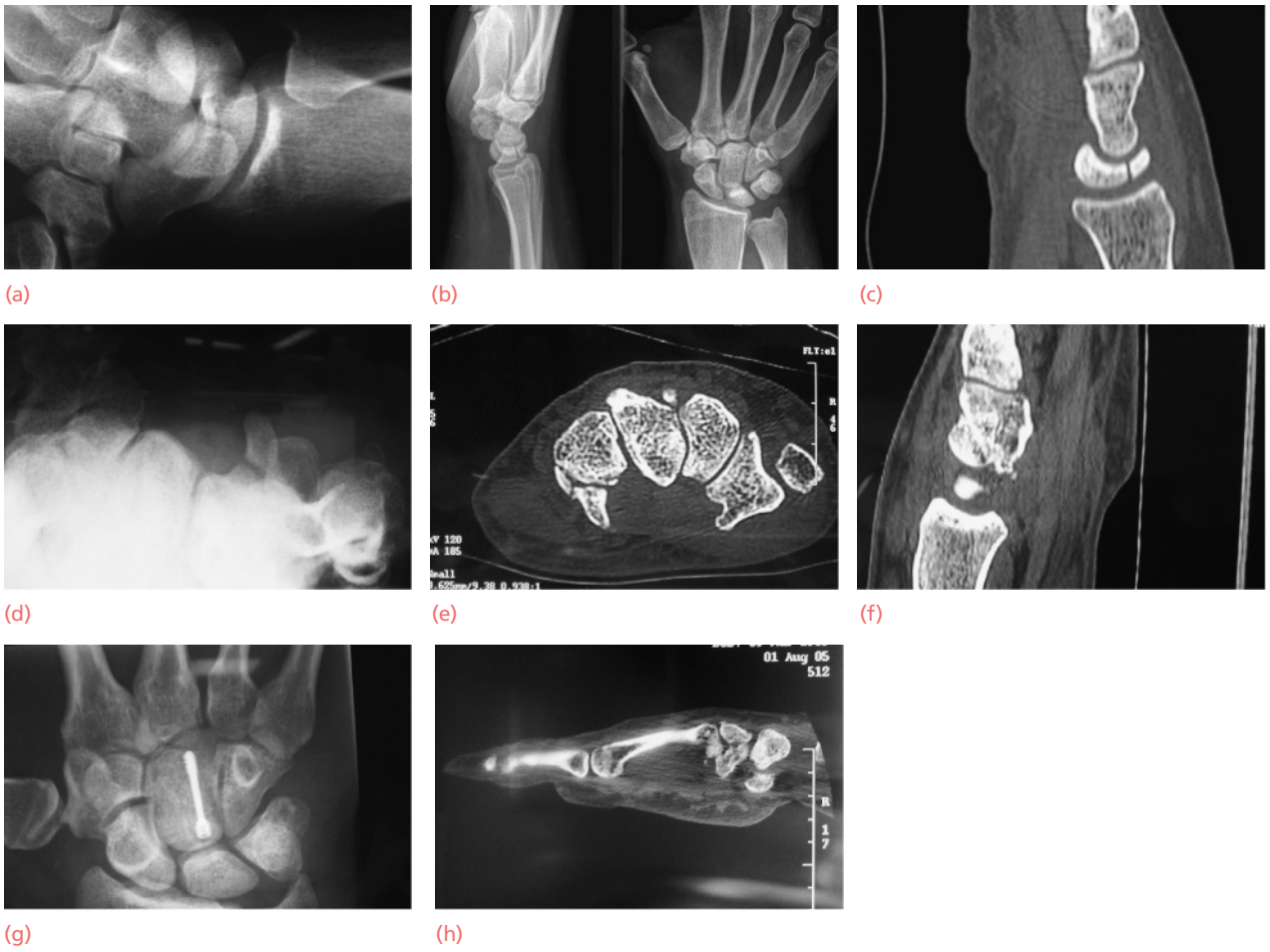


Figure 26.17 Fractures of other carpal bones (a) Fracture of the body of trapezium; (b,c) lunate fractures; (d) hook of hamate fracture; (e) hook of hamate CT; (f) capitate fracture fixed with a screw (g); (h) fracture of body of hamate.

Trapezium

The *body* of the trapezium can be fractured if the shaft of the first metacarpal impacts onto it; the *ridge* (to which the transverse carpal ligament attaches) can be fractured by a direct blow. The latter fracture can usually be seen on a CT scan or carpal tunnel view rather than standard X-rays. The body fracture may need open reduction and internal fixation if displaced; the ridge fracture usually settles with a cast for 4 weeks.

Capitate

The capitate is relatively protected within the carpus. However, in severe trauma the wrist can be fractured; the distal fragment can rotate, in which case open reduction and internal fixation are required.

Lunate

Fractures of the lunate are rare and follow a hyperextension injury to the wrist or compression. Sometimes the lunate itself may be abnormal prior to the injury (Kienböck's disease). There is a real risk of non-union; undisplaced fractures should be immobilized in a cast for 6 weeks; displaced fractures should be reduced and fixed with a screw.

ULNAR-SIDE WRIST INJURIES
(see also Chapter 15)

After a fall, the *distal radioulnar joint* is often injured with a radial fracture; it can also be damaged in isolation, particularly after hyperpronation. *The triangular fibrocartilage complex* can be torn, the *ulnar styloid* avulsed or the articular surfaces of the DRUJ damaged. With sudden supination (e.g. playing tennis or golf) the *extensor carpi ulnaris* (ECU) can be pulled out of its sheath.

Clinical features

The patient complains of pain and often clunking. The ulnar corner must be examined very thoroughly.

There is tenderness over the injured site (e.g. the DRUJ, the ulna fovea – the dent at the front of the base of the ulna styloid – or the ECU sheath) and pain on rotation of the forearm. The distal ulna may be unstable; the *piano-key sign* is elicited by holding the patient's forearm pronated and pushing sharply forwards on the head of the ulna. Remember, though, that the ulna is the fixed point and it is the radius that is unstable. The ECU may be seen and felt to clunk out of its groove on rotation.

Imaging and arthroscopy

A lateral *X-ray* in pronation and supination shows incongruity of the DRUJ. The AP view may show an avulsed ulnar styloid. *Arthrography*, *CT*, *MRI* and *arthroscopy* may be needed to confirm the diagnosis.

Treatment

An acute dislocation of the DRUJ usually resolves if the arm is held in supination for 6 weeks; occasionally a K-wire is needed to maintain the reduction. If the dislocation is irreducible, this may be due to trapped soft tissue, which will have to be removed. Chronic instability may require reconstructive surgery.

An unstable TFCC tear should be reattached. A displaced fracture at the base of the ulnar styloid, if painful and associated with instability of the radioulnar joint, should be fixed with a small screw. An acute ECU sheath tear can be treated with either repair or an above-elbow cast in pronation for 6 weeks.

CARPAL DISLOCATIONS, SUBLUXATIONS AND INSTABILITY

The wrist functions as a system of intercalated segments or links, stabilized by the scaphoid (which acts as a bridge between the proximal and distal rows of the carpus) and the ligaments either side of the lunate (the scapholunate ligament and the lunotriquetral ligament). Fractures and dislocations of the carpal bones, or even simple ligament tears and sprains, may seriously disturb this system so that the links collapse into one of several well-recognized patterns (see Chapter 16).

Lunate and perilunate dislocations

A fall with the hand forced into dorsiflexion may tear the tough ligaments that normally bind the carpal bones. The lunate usually remains attached to the radius and the rest of the carpus is displaced backwards (*perilunate dislocation*). Usually the hand immediately snaps forwards again but, as it does so, the lunate may be levered out of position to be displaced anteriorly (*lunate dislocation*). Sometimes the scaphoid remains attached to the radius and the force of the perilunar dislocation causes it to fracture through the waist (*trans-scaphoid perilunate dislocation*).

Clinical features

The wrist is painful and swollen and is held immobile. If the carpal tunnel is compressed, there may be paraesthesia or blunting of sensation in the territory of the median nerve and weakness of palmar abduction of the thumb.

X-rays

Most dislocations are perilunate (Figure 26.18). In the AP view the carpus is diminished in height and the bone shadows overlap abnormally. One or more of the carpal bones may be fractured (usually the scaphoid and radial styloid). If the lunate is dislocated, it has a characteristic triangular shape instead of the normal quadrilateral appearance.

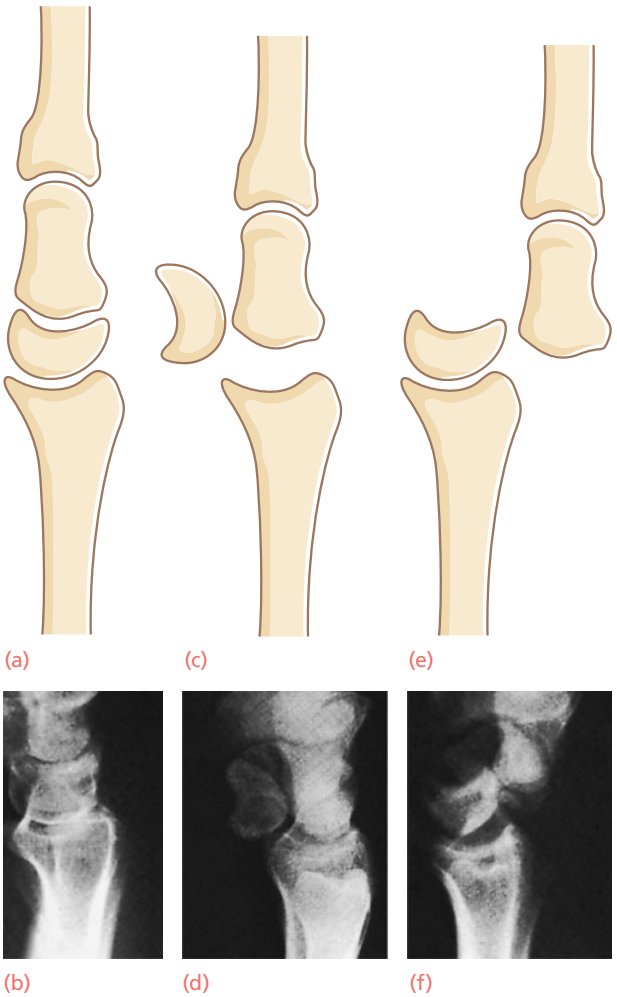


Figure 26.18 Lunate and perilunate dislocations (a,b) Lateral X-ray of normal wrist; (c,d) lunate dislocation; (e,f) perilunate dislocation.

In the lateral view it is easy to distinguish a lunate from a perilunate dislocation. The *dislocated lunate* is tilted forwards and is displaced in front of the radius, while the capitate and metacarpal bones are in line with the radius. With a *perilunate dislocation* the lunate is tilted only slightly and is not displaced forwards, and the capitate and metacarpals lie behind the line of the radius (DISI pattern); if there is an associated *scaphoid fracture*, the distal fragment may be flexed.

Treatment

CLOSED REDUCTION

The surgeon pulls strongly on the dorsiflexed hand; then, while maintaining traction, then slowly flexes the wrist forwards, at the same time squeezing the lunate backwards with their other thumb. These manoeuvres usually effect reduction; they also prevent conversion of a perilunate to a lunate dislocation. A plaster splint is applied holding the wrist neutral. Percutaneous K-wires may be needed to hold the reduction.

OPEN REDUCTION

Reduction is imperative and, if closed reduction fails, or if a later X-ray shows that the wrist has collapsed into the familiar DISI pattern, open reduction is needed. The carpus is exposed by an anterior approach which has the advantage of decompressing the carpal tunnel. A dorsal approach may also be used, which has the advantage of allowing repair of the torn scapholunate ligament. While an assistant pulls on the hand, the lunate is levered into place and kept there by a K-wire, which is inserted through the lunate into the capitate (Figure 26.19). If the scaphoid and/or radial styloid are fractured, this too can be reduced and fixed with a headless compression screw or K-wires. Where possible, the torn ligaments should be repaired. At the end of the procedure, the wrist is splinted in a plaster cast, which is retained for 6 weeks. Finger, elbow and shoulder exercises are practised throughout this period. The K-wires are removed at 6 weeks.

This injury is frequently accompanied by severe compression of the median nerve, which should be released.

Scapholunate dissociation

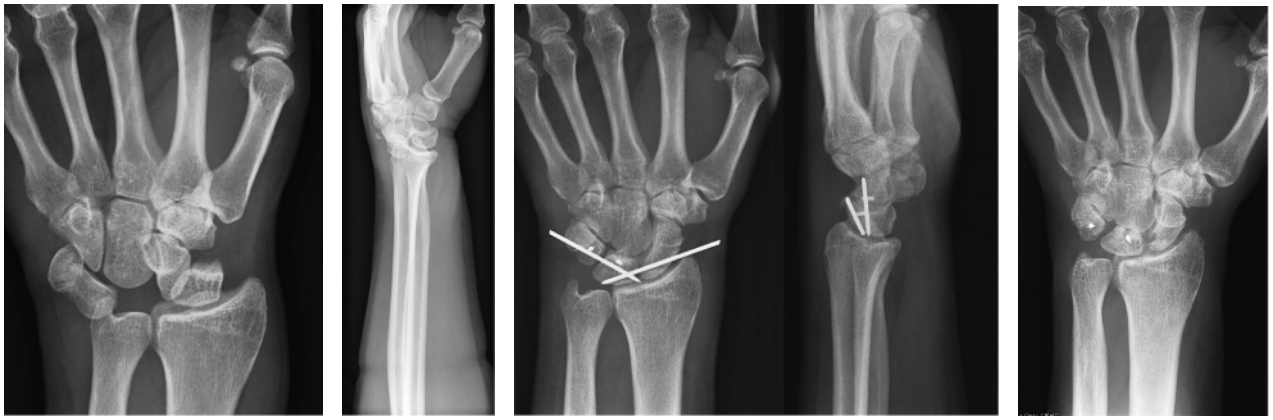
Clinical features

A fall on the outstretched hand, rather than breaking a bone, may tear the all-important scapholunate ligament. There is pain and swelling with tenderness over the dorsum just distal to Lister's tubercle. Pushing backwards on the scaphoid tubercle is very painful and, if performed while moving the wrist radialwards and ulnarwards, it can elicit a clunk ('Watson's test').

X-rays

These may, in a significant unstable tear, show an excessively large gap between the scaphoid and the lunate. The scaphoid may appear foreshortened, with a typical cortical ring sign. In the lateral view, the lunate is tilted dorsally and the scaphoid anteriorly (DISI pattern). In lesser injuries, a clenched-fist view forces the two bones apart and may reveal the injury.

In children the injury is extremely rare but may be misdiagnosed due to the asymmetric ossification seen in the scaphoid particularly. If suspected radiographically, a competent clinical examination will usually reassure the clinician.



(a) (b) (c) (d)

Figure 26.19 Perilunate dislocation (a,b) Lunate still in its original position while the rest of the carpus is dislocated around it. (c) The dislocation has been reduced and held with K-wires. (d) The lunotriquetral ligament is reattached with ligament anchors.

Treatment of acute injuries

If the patient is seen early (i.e. less than 4 weeks after injury), the scapholunate ligament should be repaired directly with interosseous sutures, and protected by K-wires for 6 weeks and a cast for 8–12 weeks. If seen 4–24 weeks after the injury, the ligament is unlikely to heal. In chronic lesions without secondary osteoarthritis, a ligament reconstruction may be attempted but the results are unpredictable. If there are significant symptoms (especially if arthritis develops) a proximal row carpectomy, four-corner fusion or radiosapholunate arthrodesis should be considered. More information is given in Chapter 15.

Lunotriquetral dissociation

This is a relatively rare injury, much less common than the scapholunate tear. A fall on the ulnar side of the hand followed by ulnar carpal pain, weakness of grip and tenderness distal to the head of the ulna should suggest disruption of the lunotriquetral ligaments. The physical and X-ray signs may be subtle so the injury is often not diagnosed in the early stage.

X-rays sometimes show a gap or step between the triquetrum and the lunate, with a VISI carpal collapse pattern in the lateral view (i.e. the lunate and scaphoid tilted forwards).

Treatment of acute tears

Acute tears should be repaired with interosseous sutures (Figure 26.19d), supported by temporary K-wires for 6 weeks and a cast for 8–12 weeks. In chronic injuries, a ligament substitution (e.g. a slip of extensor carpi ulnaris) or a limited intercarpal fusion may be considered. For more information, see Chapter 15.

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Injuries of the hand 27

David Warwick

Hand injuries – the commonest of all injuries – are important out of all proportion to their apparent severity, because of the need for perfect function. Nowhere else do painstaking evaluation, meticulous care and dedicated rehabilitation yield greater rewards. The outcome is often dependent upon the judgement of the doctor who first sees the patient.

If there is skin damage, the patient should be examined in a clean environment with the hand displayed on sterile drapes.

A brief but searching history is obtained; often the mechanism of injury will suggest the type and severity of the trauma. The patient's age, occupation and 'handedness' should be recorded.

Superficial injuries and severe fractures are obvious, but deeper injuries are often poorly disclosed. It is important in the initial examination to assess the circulation, soft-tissue cover, bones, joints, nerves and tendons.

X-rays should include at least three views (postero-anterior, lateral and oblique), and with finger injuries the individual digit must be X-rayed.

GENERAL PRINCIPLES OF TREATMENT

Most hand injuries can be dealt with under local or regional anaesthesia; a general anaesthetic is only rarely required.

Circulation If the circulation is threatened, it must be promptly restored, if necessary by direct repair or vein grafting.

Swelling Swelling must be controlled by elevating the hand and by early and repeated active exercises.

Splintage Incorrect splintage is a potent cause of stiffness; it must be appropriate and it must be kept to a minimum length of time. If a finger has to be splinted, it may be possible simply to tape it to its neighbour so that both move as one; if greater security is needed, only the injured finger should be splinted. If the entire hand needs splinting, this must always be in the 'position of safety/safe immobilization' – with the metacarpophalangeal joints flexed at least 70 degrees and the interphalangeal joints almost straight (Figure 27.1c). Sometimes an external splint, to be effective, would need to immobilize undamaged fingers or would need to hold the joints of the injured finger in an unfavourable position (e.g. flexion of the interphalangeal joints). If so, internal fixation may be required (K-wires, screws or plates).

Skin cover Skin damage needs thorough wound washout followed by suture, skin grafting, local flaps, pedicled flaps or (occasionally) free flaps. Treatment of the skin takes precedence over treatment of the fracture.

Nerve and tendon injury Generally, the best results will follow primary repair of tendons and nerves. Occasionally grafts are required.

FINGER METACARPAL FRACTURES

The metacarpal bones are vulnerable to blows and falls upon the hand, or the longitudinal force of the boxer's punch. Injuries are common and the bones may fracture at their *base*, in the *shaft*, through the *neck* or at their *head* (Figure 27.2).

Figure 27.1 Splintage of the hand Three positions of the hand: (a) the position of relaxation, (b) the position of function (ready for action) and (c) the position of safe immobilization, with the ligaments taut.

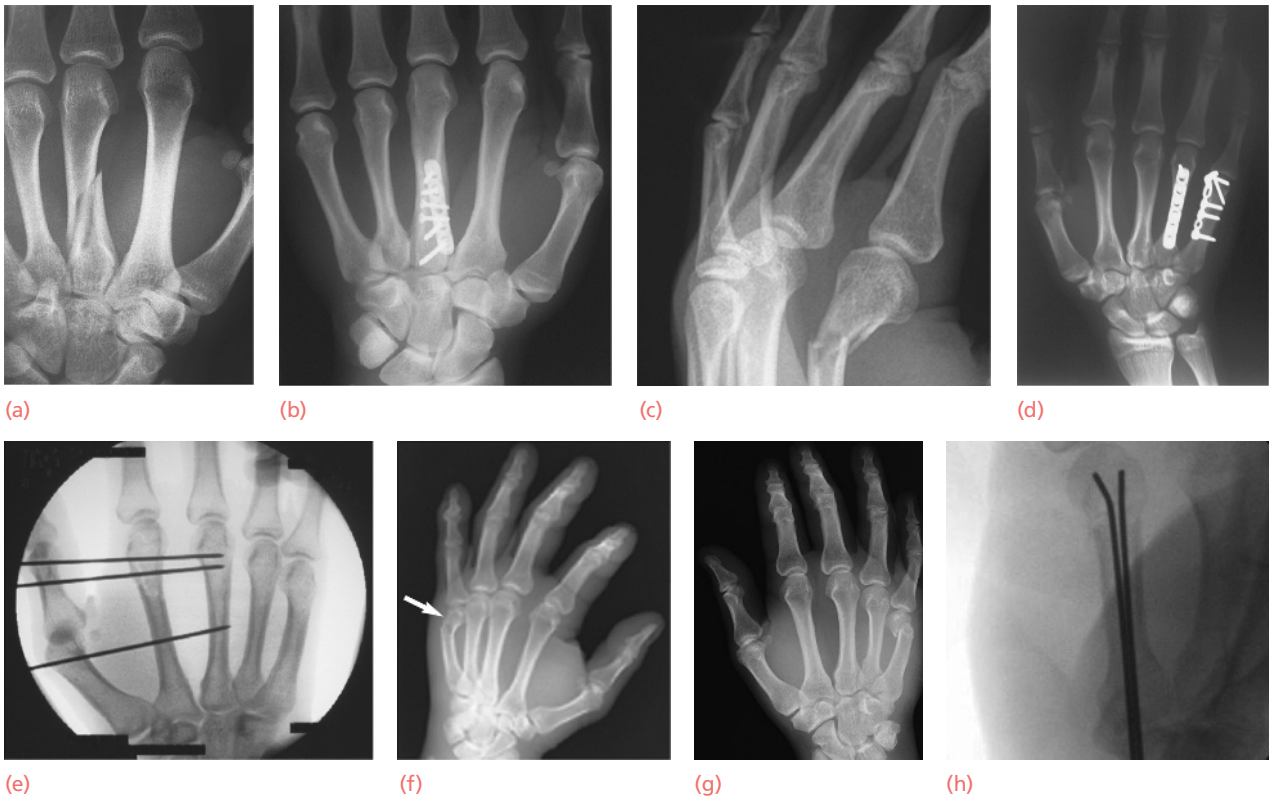


Figure 27.2 Metacarpal fractures (a) A spiral fracture (especially an 'inboard' one) can be adequately held by the surrounding muscles and ligaments but internal fixation (b) allows early mobilization. A displaced shaft fracture (c), especially an 'outboard' one, can be held by a plate or transverse wires to allow early mobilization (d); multiple metacarpal fractures should be fixed with rigid plates for wires (e). A boxer's fracture (f) should usually not be treated. A more displaced fracture (g) can be treated with intramedullary wires (h).

Angular deformity is usually not very marked and, even if it persists, it does not interfere much with function. Rotational deformity, however, is serious. Close your hand with the distal phalanges extended, and look: the fingers converge across the palm to a point above the thenar eminence; malrotation of the metacarpal (or proximal phalanx) will cause that finger to diverge and overlap one of its neighbours. Thus, with a fractured metacarpal it is important to regain normal rotational alignment.

The fourth and fifth metacarpals are more mobile at their base than the second and third, and therefore they are better able to compensate for residual angular deformity.

Fractures of the thumb metacarpal usually occur near the base and pose special problems. They are dealt with separately below.

Fractures of the metacarpal shaft

A direct blow may fracture one or several metacarpal shafts transversely, often with associated skin damage. A twisting or punching force may cause a spiral fracture of one or more shafts. There is local pain and swelling, and sometimes a dorsal 'hump'.

Treatment

OBLIQUE OR TRANSVERSE FRACTURES WITH SLIGHT DISPLACEMENT

These fractures require no reduction. Splintage also is unnecessary, but a firm crepe bandage may be comforting; this should not be allowed to discourage the patient from active movements of the fingers, which should be practised assiduously. As the patient moves the fingers, the fracture may shorten until the intercarpal ligaments between the metacarpal necks tighten, thus limiting further shortening and rotational deformity. After 4–5 weeks callus forms and the fracture becomes stable.

TRANSVERSE FRACTURES WITH CONSIDERABLE DISPLACEMENT

Fractures of this type can be reduced by traction and pressure then held with a plaster cast, but they are usually unstable and should be fixed surgically with compression plates to allow immediate mobilization. If plates are not available, percutaneous K-wires are placed transversely through the neighbouring undamaged metacarpals or up through the medulla, supported by a cast for 4–5 weeks.

SPIRAL FRACTURES

Spiral fractures are liable to rotate. They can be managed non-operatively if there is no rotational displacement and the patient accepts a shorter knuckle profile but, otherwise, they should be perfectly reduced and fixed with lag screws and a plate to allow immediate mobilization. If rigid fixation is not available, percutaneous wires protected by a plaster for 4–5 weeks is an alternative.

Fractures of the metacarpal neck

A blow may fracture the metacarpal neck, usually of the fifth finger (the 'boxer's fracture') and occasionally one of the others. There may be local swelling, with flattening of the knuckle. X-rays show an impacted transverse fracture with volar angulation of the distal fragment.

Treatment

The main function of the *fifth and fourth fingers* is firm flexion ('power grip') and, as can be readily demonstrated on a normal hand, there is 'spare' extension available at the metacarpophalangeal (MCP) joint and CMC joint. Therefore in these digits, a flexion deformity of up to 70 degrees can be accepted; as long as there is no rotational deformity and as long as the patient accepts a dropped knuckle profile, a good outcome can be expected. The hand is immobilized in a gutter splint with the MCP joint flexed and the interphalangeal (IP) joints straight until discomfort settles – a week or two – and then the hand is mobilized. In the *index and middle fingers*, which function mainly in extension, no more than 15 degrees of flexion at the fracture is acceptable.

If the fracture needs reduction, this can be done under a local block. The reduced finger is held with a gutter splint moulded at three points to support the fracture; the MCP joints are flexed and the IP joints are straight. Unfortunately, these fractures are usually fairly unstable because of the tone of the flexor and intrinsic tendons together with the palmar comminution of the fracture and so they usually redisplace; fixation should be considered. Plates are not really suitable because the fracture is so distal that they impinge on the collateral ligaments and the metacarpal heed. A 'bouquet' of two or three bent wires passed distally through a hole in the metacarpal base is particularly effective.

Complications

Malunion Malunion with volar angulation of the distal fragment is poorly tolerated if this occurs in the second or third rays. The patient may be aware of a bump in the palm from the prominent metacarpal head and the digit may take on a 'Z' appearance as the knuckle joint hyperextends to compensate for the deformity.

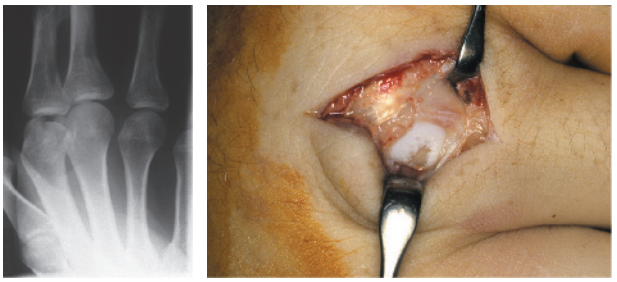
Fractures of the metacarpal head

These fractures occur after a direct blow. They are often quite comminuted and sometimes 'open'. They are associated with punching and every care must be taken to ensure there is no communication between the skin and the fracture. Even a small apparently superficial wound will usually connect with the joint because the tightly compressed layers of skin, fat, tendon, capsule and synovium are penetrated with the knuckle fully flexed, yet the wound is usually examined with the knuckle straight – the layers overlap and the communication is not appreciated. Thorough urgent washout of the highly aggressive mouth flora with copious volumes of saline is needed. Displaced head fragments can be difficult to visualize – a CT scan is very helpful. Operative reduction and fixation with small headless buried screws is ideal (Figure 27.3). Occasionally the joint is so badly damaged that primary replacement is considered (Silastic, pyrocarbon or polythene–metal).

Fractures of the metacarpal base

Excepting fractures of the thumb metacarpal, these are usually stable injuries which can be treated by ensuring that rotation is correct and then splinting the digit in a volar slab extending from the forearm to the proximal finger joint. The splint is retained for 3 weeks and exercises are then encouraged.

Displaced intra-articular fractures of the base of the fourth or fifth metacarpal may cause marked incongruity of the joint. This is a mobile joint and it may therefore be painful. The fracture should be reduced by traction on the little finger and then held with a percutaneous K-wire or compression screw. In the long term, if painful arthritis supervenes, treatment would be with either arthrodesis or joint excision.



(a) (b)

Figure 27.3 Fracture of the metacarpal head
(a) Depressed head fracture which was reduced and held with buried mini-screws. (b) A 'fight-bite', with metacarpal head damage from an opponent's tooth. Urgent thorough washout is needed.

FRACTURES OF THE THUMB METACARPAL

Three types of fracture are encountered: impacted fracture of the metacarpal base; Bennett's fracture-dislocation of the carpometacarpal (CMC) joint; and Rolando's comminuted fracture of the base (Figure 27.4).

Fractures of the base

A fall on the back of the thumb, or a misplaced punch, can break the base of the first metacarpal. Localized swelling and tenderness are found, and X-rays show a transverse fracture just beyond the CMC joint, with outward bowing and impaction.

Treatment

If the angulation is *less than about 30 degrees* and the fragments are impacted, the thumb is rested in a plaster of Paris cast extending from the forearm to just short of the interphalangeal thumb joint with the thumb fully abducted and extended. The cast is removed after 3–4 weeks and the thumb is mobilized.

If the angulation is *greater than about 30 degrees*, then the reduced thumb web span will be noticeable and so the fracture should be reduced. This can be done by pulling on the abducted thumb and correction of the bowing by levering the metacarpal outwards. While a plaster might hold the fracture, it usually collapses forward again and so a percutaneous K-wire is usually inserted and supported by a cast for 5 weeks; even better, a low-profile plate can be used to allow early movement.

Bennett's fracture-dislocation

This fracture, too, occurs at the base of the first metacarpal bone and is commonly due to falling or punching; however, the fracture is oblique, extends into the CMC joint and is very unstable because of the strong pull of the abductor pollicis longus tendon that remains attached to the shaft of the metacarpal.

On examination, the thumb looks short and the thumb base is swollen and very tender. X-rays show that a small triangular fragment has remained in contact with the medial edge of the trapezium, while the remainder of the thumb has subluxated proximally, pulled upon by the abductor pollicis longus tendon.

Treatment

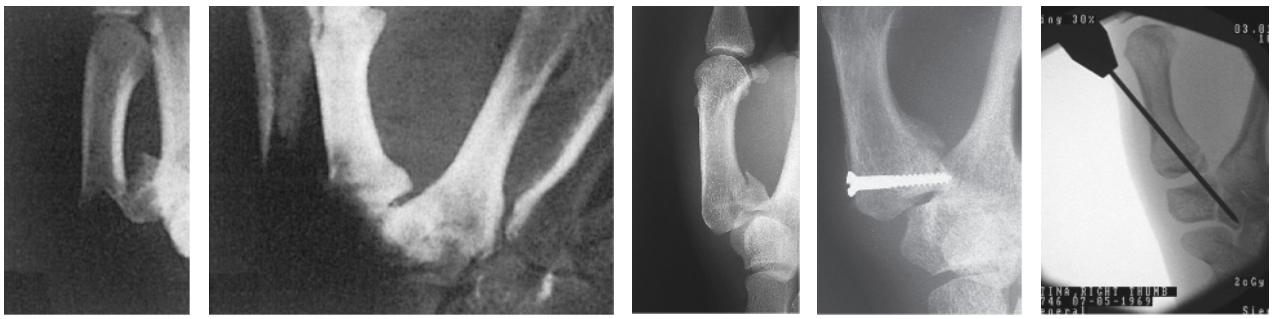
It is widely supposed (with little evidence) that perfect reduction is essential. It should, however, be attempted and can usually be achieved by pulling on the thumb, abducting it and extending it. Reduction can then be held in one of two ways: *plaster* or *internal fixation*.

Plaster Plaster may be applied with a felt pad over the fracture, and the first metacarpal held abducted and extended (usually best achieved by *flexing* the MCP joint). However, plaster only works if it is applied with great skill, and the pressure required to maintain a reduction can cause skin damage; it has, therefore, generally been abandoned in favour of surgery.

Surgical fixation This is achieved by passing a K-wire across the metacarpal base into the carpus. If the fragment is large and cannot be reduced and held with a wire, then open reduction and fixation with a lag screw is effective.

Rolando's fracture

This is an intra-articular comminuted fracture of the base of the first metacarpal with a T or Y configuration. Closed reduction and K-wiring or open reduction and plate fixation can be used. With more severe comminution, external fixation is needed.



(a) (b) (c) (d) (e)

Figure 27.4 Fractures of the first metacarpal base A transverse fracture (a) can be reduced and held in plaster (b). Bennett's fracture-dislocation (c) is best held with a small screw (d) or a percutaneous K-wire (e).

METACARPAL FRACTURES IN CHILDREN

Metacarpal fractures are less common in children than in adults. In general they also present fewer problems: the vast majority can be treated by manipulation and plaster splintage; angular deformities will almost always be remodelled with further growth. However, rotational alignment is as important as it is in adults.

Bennett's fracture is rare but, when it does occur, it usually requires open reduction. This is, by definition, a Salter–Harris type III fracture-separation of the physis; it must be accurately reduced and fixed with a K-wire.

FRACTURES OF THE PHALANGES

The fingers are usually injured by direct violence, and there may be considerable swelling or open wounds. Injudicious treatment may result in a stiff finger which, in some cases, can be worse than no finger.

Fractures of the proximal and middle phalanges

The phalanx may fracture in various ways:

- *transverse fractures of the shaft* – often with forward angulation
- *spiral fracture of the shaft* – from a twisting injury
- *comminuted fracture* – usually due to a crush injury and often associated with significant tendon damage and skin loss
- *avulsion* of a small fragment of bone (representing avulsion of a collateral ligament, central slip or volar plate).
- *metaphyseal fracture at the base of the proximal phalanx* – commonly seen in osteopaenic bone; the shaft is pulled into extension
- *distal condyle fractures* – most commonly seen in children; one or both condyles can break off and rotate backwards.

Treatment

UNDISPLACED FRACTURES

These can be treated by 'functional splintage'. The finger is strapped to its neighbour ('buddy/neighbour strapping') and movements are encouraged from the outset. Splintage is retained for 3–4 weeks, but during this time it is wise to check the position by X-ray in case displacement has occurred; if so surgery may be needed.

DISPLACED FRACTURES

Displaced fractures must be reduced and immobilized. *It is essential to check for rotational* correction by:

- noting the convergent position of the finger when the MCP joint is flexed
- seeing that the fingernails are all in the same plane.

The treatment depends on the fracture pattern (Figure 27.5). Some just need simple manipulation and can then be held in a splint. *Basal fractures* with extension are manipulated and held with a dorsal blocking splint with the MCP joint at 90 degrees. *Angulated basal fractures* are manipulated with a pencil between the digits as a lever and then held with neighbour strapping which pulls the injured finger to the next one. *Spiral fractures* are held with 'derotation taping' to the next digit, using tension in the tape to unwind the fracture. *Transverse fractures* may be held in a gutter splint or neighbour splint.

If a reduction cannot be achieved, or if it is unstable and the position slips, surgery is needed. The technique depends upon the configuration of the fracture. *A plate and screws* provide stable fixation and thus early movement, but stiffness due to tendon adhesion is common. *K-wires* are less invasive and are perfect for some fractures but they can cause stiffness by stapling the extensor tendon to the bone. For some fractures of the proximal phalanx, a wire passed across the metacarpal neck into the proximal phalanx, with the MCP flexed to 90 degrees, avoids the stiffness that can be associated with crossed wires or a plate. Other techniques include *percutaneous lag screw fixation* (for spiral fractures and distal condyle fractures). *External fixation* may be needed for comminuted fractures.

Childhood phalangeal fractures

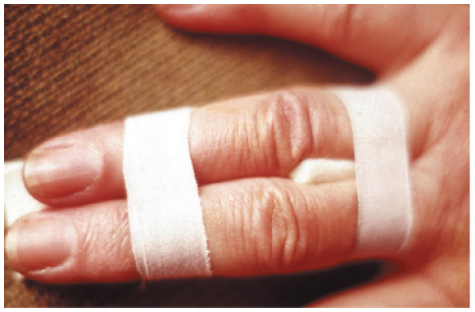
In older children the shaft may break; closed reduction and early mobilization will usually suffice. If the growth plate is damaged, perfect reduction and temporary K-wire fixation is needed. Distal condylar fractures are more common in children; these are discussed below in the section on 'Joint injuries'.

Fractures of the terminal phalanx

The terminal phalanx, small though it is, is subject to five different types of fracture (Figure 27.6).

Fracture of the tuft

The tip of the finger may be struck by a hammer or caught in a door, and the bone shattered. The fracture is disregarded and treatment is focused on controlling



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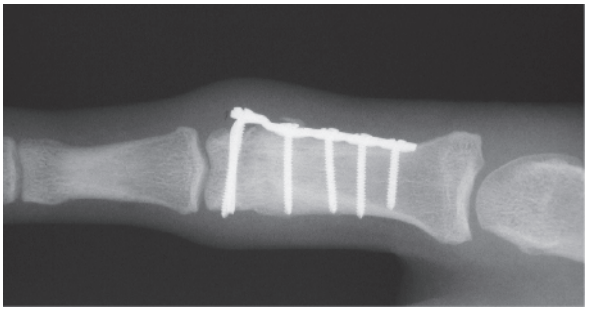

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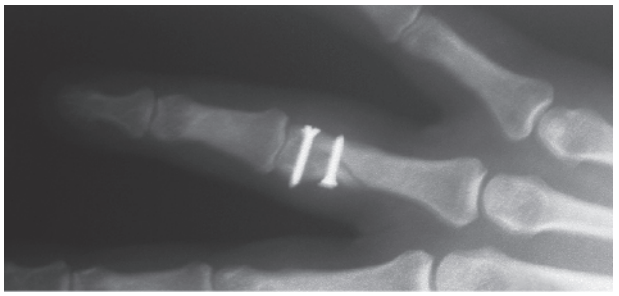

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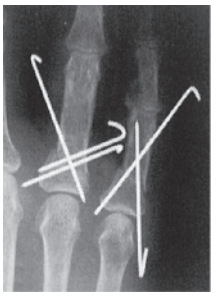

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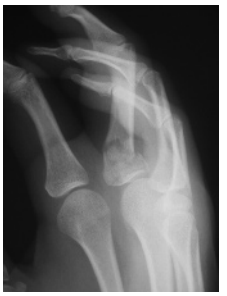

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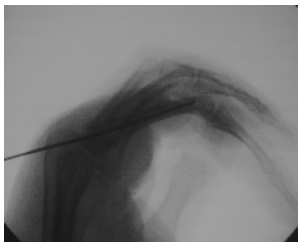
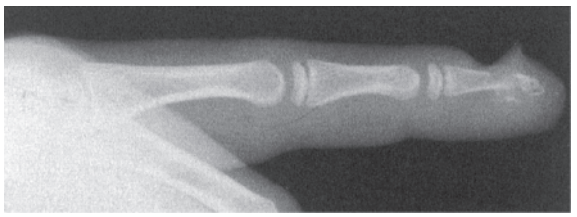


(f)

Figure 27.5 Phalangeal fractures These can be treated, depending on the 'personality' of the fracture, experience of the surgeon and equipment available, with neighbour strapping (a), plate fixation (b), percutaneous screw fixation (c) or percutaneous wires (d). Unstable fracture at the base of the proximal phalanx (e) fixed with wire across the flexed MCP joint (f) (picture courtesy of Professor G. Giddins).



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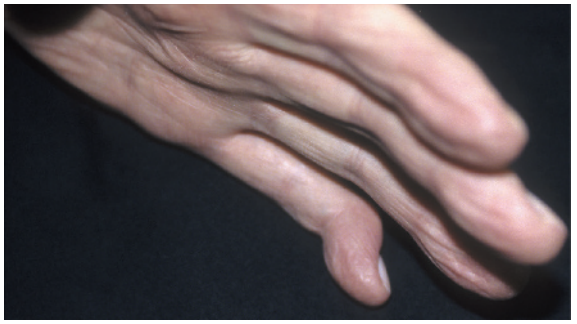

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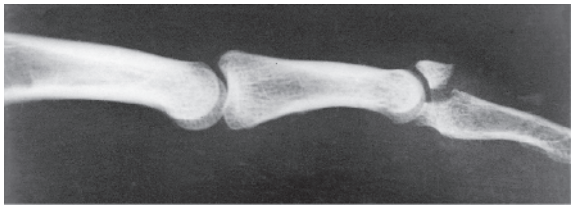

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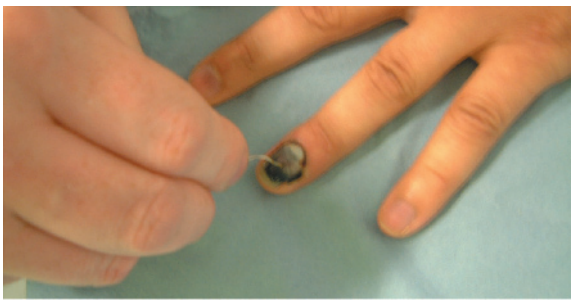

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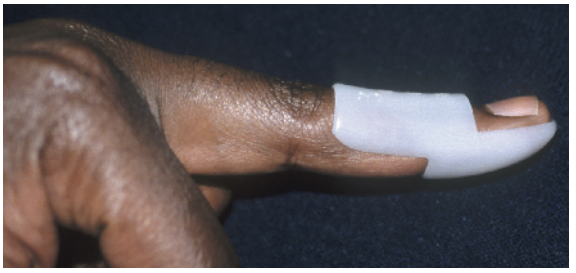


(d)

Figure 27.6 Distal phalangeal injury A fracture of the tuft (a), caused by a hammer blow, is treated by a protective dressing. The subungual haematoma should be evacuated using a red-hot paper clip tip (b) or a small drill. A mallet finger (c) is best treated with a splint for 6 weeks (d). Mallet fractures (e) are also better splinted – surgery can make the outcome worse.

swelling and regaining movement. The painful haematoma beneath the finger nail should be drained by piercing the nail with a hot paper clip. If the nail bed (matrix) is torn and cosmesis is important, it should be meticulously repaired under magnification.

Mallet finger injury

After a sudden flexion injury (e.g. stubbing the tip of the finger) the distal phalanx droops and cannot be straightened actively. Three types of injury are recognized:

- rupture of the most distal part of the extensor tendon
- avulsion of a small flake of bone from the base of the terminal phalanx with the joint line unaffected
- avulsion of a large dorsal bone fragment, sometimes with subluxation of the distal interphalangeal joint (DIPJ).

TREATMENT

The DIPJ should be immobilized in slight hyperextension, using a special mallet-finger splint which fixes the distal joint but leaves the proximal joints free.

Tendinous avulsions These usually occur painlessly. The splint should be kept in place constantly for 8 weeks and then only at night for another 4 weeks. Even if there has been a delay of 3–4 weeks after injury, this prolonged splintage is usually successful.

Bone avulsions These are also treated in a splint, but 6 weeks should suffice as bone heals quicker than tendon. Operative treatment is generally avoided, even for large bone fragments, unless there is subluxation. Surgery carries a high complication rate (wound failure, metalwork problems) without evidence that the outcome is improved. However, if there is subluxation, K-wires or small screws are used to fix the fragment in place.

COMPLICATIONS OF MALLET FINGER

Non-union This is usually painless and treatment is not needed.

Persistent droop About 85% of mallet fingers recover full extension. If there is a persistent droop, this can be treated by tendon repair supported by K-wire fixation of the joint, but the results are often disappointing. The alternatives would be either a central slip tenotomy which can rebalance the finger extensor mechanism or, if there is persisting painful incongruity, a joint arthrodesis, best achieved with a buried intramedullary double-pitch screw.

Swan neck deformity Imbalance of the extensor mechanism can cause this in lax-jointed individuals. Again, a central slip tenotomy is straightforward and can give a very good result.

Fracture of the terminal shaft

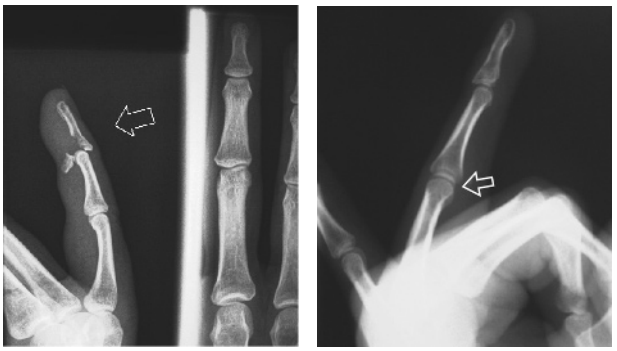
Undisplaced fractures of the shaft need no treatment apart from analgesia. If angulated, they should be reduced and held with a longitudinal K-wire through the pulp for 4 weeks. The nail is often dislocated from its fold; if so, it must be thoroughly cleaned (there are lots of bacteria in the nail fold), carefully tucked back in and held with a suture in each corner.

Avulsion of the flexor tendon

This injury is caused by sudden hyperextension of the distal joint, typically when a game player catches his finger on an opponent's shirt (hence the nickname 'rugger jersey finger'. The ring finger is most commonly affected. The flexor digitorum profundus tendon is avulsed, either rupturing the tendon itself or taking a fragment of bone with it (Figure 27.7). If the bone fragment is small, or if only the tendon is ruptured, it can recoil into the palm. If the lesion is detected within a few days (and the diagnosis is easily missed if not thought about), then the tendon can be reattached. If the diagnosis is much delayed, repair is likely to be unsuccessful. Two-stage tendon reconstruction is possible but difficult, and the finger may end up stiff. For late cases, tenodesis or fusion of the distal joint is often preferable.

Physeal fracture

The basal physis can break, usually producing a Salter–Harris type I fracture (Seymour fracture). The nail may be dislocated from its fold and the germinal matrix can be trapped in the fracture. The injury is easily overlooked if the finger is very swollen. The nail must be cleaned and carefully replaced into its bed.



(a) (b)

Figure 27.7 Flexor tendon avulsion (a) Large fragment and (b) smaller fragment lodged in front of the PIP joint.

JOINT INJURIES

Any finger joint may be injured by a direct blow (often the overlying skin is damaged), or by an angulation force, or by the straight finger being forcibly stubbed. The affected joint is swollen, tender and too painful to move. X-rays may show that a fragment of bone has been sheared off or avulsed.

CARPOMETACARPAL DISLOCATION

See Figure 27.8.

The thumb

The thumb is most frequently affected; clinically the injury then resembles a Bennett's fracture-dislocation; however, X-rays reveal proximal subluxation or dislocation of the first metacarpal bone without a fracture. The displacement is easily reduced by traction and hyperpronation, but reduction is unstable and can be held only by a K-wire driven through the metacarpal into the carpus. The wire is removed after 5 weeks but a protective splint should be worn for 8 weeks because of the risk of instability.

If chronic instability does occur, this is treated prior to arthritis developing, by using part of the flexor carpi radialis tendon to reconstruct the ruptured and incompetent palmar ligament of the CMC joint.

Finger carpometacarpal joints

These can occur after punching or falling on the fist. Usually the fifth and fourth CMCJs are involved; rarely all the CMCJs are damaged in a high-energy motorcycle collision. Often there is an associated fragment of bone from the hamate. The hand swells up rapidly and the diagnosis is easily missed unless a true lateral X-ray is carefully examined. Closed manipulation is usually successful, although a K-wire is recommended to prevent the joint from dislocating again.

Late presentation Late presentation or secondary arthritis is treated by joint fusion. However, if just the fifth CMC joint is involved, a neat operation is to fuse the base of the fourth to the fifth metacarpal and then excise the articular surface of the fifth. This will maintain movement at the fourth CMC, so allowing the ulnar side of the hand to 'cup' around during grip.

METACARPOPHALANGEAL DISLOCATION

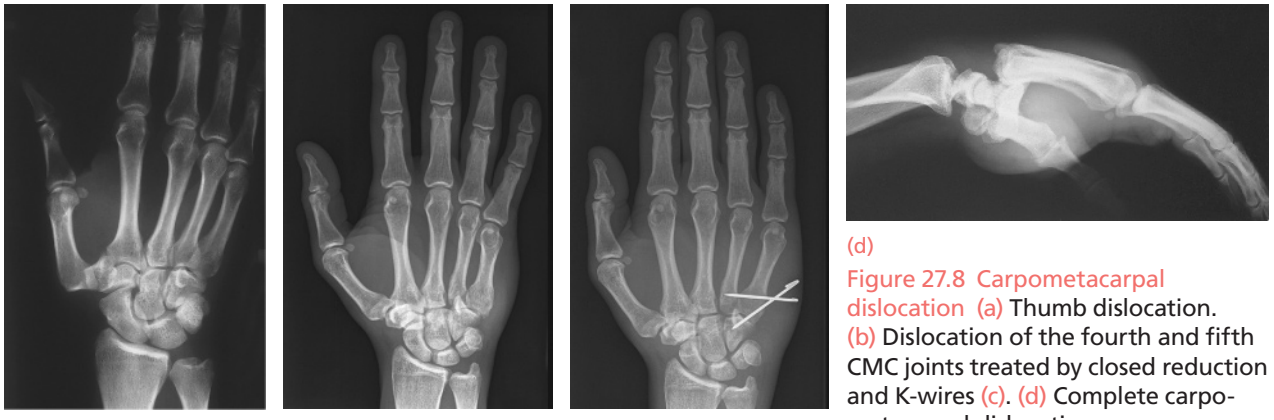
Usually the thumb is affected (Figure 27.9a), sometimes the fifth finger, and rarely the other fingers. The entire digit is suddenly forced into hyperextension and the capsule and muscle insertions in front of the joint may be torn. There are two types of dislocation: *simple* and *complex*.

Simple dislocation

The digit is extended about 75 degrees. It is easily reduced by traction, firstly in hyperextension then pulling the finger around. The finger is strapped to its neighbour and early mobilization is encouraged.

Complex dislocation

The avulsed palmar plate sits in the joint, blocking reduction. Furthermore, the metacarpal head can be clasped between the flexor tendon and lumbrical tendon. The digit is extended only about 30 degrees and there is usually a telltale dimple in the palm. Very occasionally the fracture can be reduced closed by hyperextending the MCP joint and flexing the IP joints to release the clasp. If this fails, open reduction is required. A dorsal approach is safest. After reduction the joint is stable and should be mobilized in a neighbour splint.



(a) (b) (c) (d)

Figure 27.8 Carpometacarpal dislocation (a) Thumb dislocation. (b) Dislocation of the fourth and fifth CMC joints treated by closed reduction and K-wires (c). (d) Complete carpometacarpal dislocation.

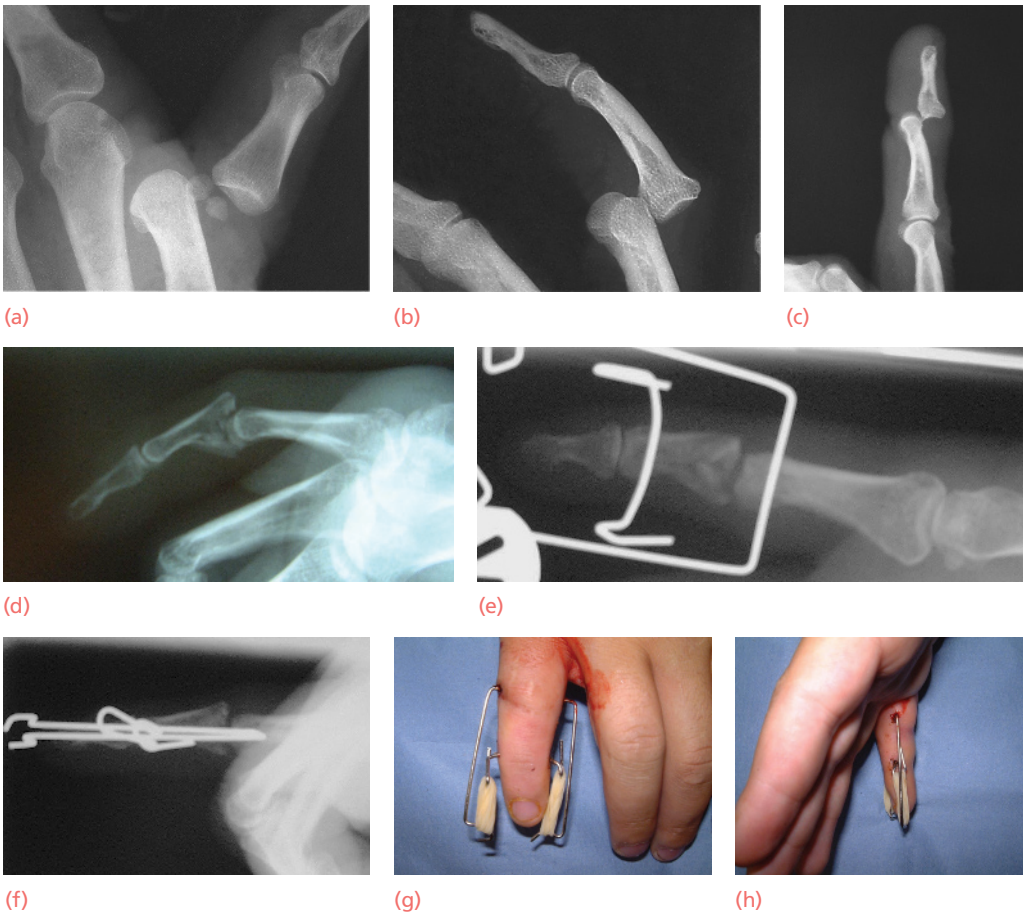


Figure 27.9 Finger dislocation (a) Metacarpophalangeal dislocation in the thumb occasionally buttonholes and needs open reduction; (b,c) interphalangeal dislocations are easily reduced (and easily missed if not X-rayed!). A comminuted pilon fracture (d) is best treated by dynamic external fixation (e,f,g,h).

Chronic instability in the thumb MCP joint

This can be treated by a sesamoid arthrodesis so long as arthritis has not yet intervened. The abductor sesamoid is fused to the underside of the metacarpal neck. This preserves some flexion yet prevents hyperextension. An alternative is formal arthrodesis. The use of a low-profile compression plate allows early mobilization. The functional result is usually very good.

Interphalangeal joint dislocation

DIP joint dislocation is rare; PIP joint dislocation is more common. The dislocation is easily reduced by pulling (Figure 27.9b). The joint is strapped to its neighbour for a few days and movements are begun immediately. The lateral X-ray may show a small flake of bone at the front of the base of the middle phalanx, representing a palmar plate avulsion; this should be ignored. The patient must be warned that it can take many months (and sometimes forever) for the spindle-like swelling of the joint to settle and for full extension to recover.

If closed reduction is successful and the joint is stable, an extension blocking splint or temporary transarticular wire is used. If it cannot be reduced or remains unstable, screw fixation or a transarticular wire is used. If there is very marked comminution and instability, a spring-loaded external fixator to distract the joint yet allow it to move congruously can be remarkably effective. An alternative, especially when the anterior fragments are too badly damaged, is to expose the joint from the palmar surface, excise the damaged fragments and then reattach the palmar plate into the base of the proximal phalanx ('palmar-plate arthroplasty').

'Pilon' fractures of the middle phalanx

These are quite common injuries and can be very troublesome. The head of the proximal phalanx impacts into the base of the middle phalanx, causing the latter to splay open in several pieces. These injuries are best treated with dynamic distraction using a spring-loaded external fixator which rotates around the head of the proximal phalanx and disimpacts the distal fragment. The results can be surprisingly good.

Condylar fracture

The convex distal joint surface of the phalanges can be fractured, usually by an angulation force. One or

both condyles may be fractured. If the fragment is not displaced, it is best to disregard the fracture, strap the finger to its neighbour and concentrate on regaining movement. An X-ray should be taken after a week to ensure there is no displacement.

If the fracture is displaced, there is a risk of permanent angular deformity and loss of movement at the joint. The fracture should be anatomically reduced, either closed or by open operation and fixed with small K-wires or mini-screws. The finger is splinted for a few days and then supervised movements are commenced.

Volar fracture-dislocation

These are relatively unusual. The middle phalanx dislocates forwards. The central slip insertion into the back of the base of the middle phalanx is torn, often with a fragment of bone. If the fragment is large enough, fixation with a small screw is undertaken; if there is a tiny but displaced fragment, reattachment with a suture loop or bone anchor is performed. If there is no fracture or an undisplaced fracture, splinting alone will suffice but X-rays must be taken at 1 and 2 weeks to ensure that the joint has not slipped. The rehabilitation must be meticulous to allow early movement of the tendon mechanism. The PIP is splinted straight for 4 weeks but the DIP joint is moved actively and passively immediately to keep the lateral bands moving separately from the damaged central slip.

LIGAMENT INJURIES

Proximal interphalangeal ligaments

Partial or complete tears of the proximal interphalangeal ligaments are quite common, due to forced angulation of the joint. *Mild sprains* require no treatment but with more severe injuries the finger should be splinted in extension for 2–3 weeks then mobilized with neighbour splinting to the finger adjacent to the injured ligament. If the joint is frankly unstable, especially the index and middle radial ligaments, which oppose load from the thumb, repair is considered.

Occasionally, the bone to which the ligament is attached is avulsed; if the fragment is markedly displaced (and large enough), it should be reattached. The patient must be warned that the joint is likely to remain swollen and slightly painful for at least 6–12 months. If the instability persists – which is rare – it can be treated by a thin strip of tendon graft or, in the index or middle finger PIP, fusion is considered to allow stable reliable pain-free pinch against the thumb.

Metacarpophalangeal joints

The radial collateral ligament of the index finger is most vulnerable, although with a suitable force any ligament can be injured. The tension of the ligament is tested with the MP joint flexed – if extended, even a normal ligament is very lax!

In children, the injury may be accompanied by a Salter–Harris type III fracture at the base of the proximal phalanx.

A large bone fragment, if displaced, can be reattached from a palmar approach, using a tension-band suture or small screw. Smaller fragments are treated by splintage with the MP joints flexed.

Ulnar collateral ligament of the thumb metacarpophalangeal joint ('gamekeeper's thumb'; 'skier's thumb')

In former years, gamekeepers who twisted the necks of little animals ran the risk of tearing the ulnar collateral ligament of the thumb metacarpophalangeal joint, either acutely or as a chronic injury. Nowadays this injury is seen in skiers who fall onto the extended thumb, forcing it into hyperabduction (Figure 27.10). A small flake of bone may be pulled off at the same time. The resulting loss of stability may interfere markedly with prehensile (pinching) activities.

The ulnar collateral ligament inserts partly into the palmar plate. In a *partial rupture*, only the ligament proper is torn and the thumb is unstable in flexion but still more or less stable in full extension because the palmar plate is intact. In a *complete rupture*, both the ligament proper and the palmar plate are torn and the thumb is unstable in all positions. If the ligament pulls away from its distal attachment to the base of the proximal phalanx, it will not heal unless it is repaired; this is because the proximal end gets trapped in front of the adductor pollicis aponeurosis (the Stener lesion – Figure 27.10e).

Clinical assessment

On examination there is tenderness and swelling precisely over the ulnar side of the thumb metacarpophalangeal joint. *An X-ray is essential, to exclude a fracture before carrying out any stress tests.* Laxity is often obvious but, if in doubt, the joint can be examined under local anaesthetic. If there is no undue laxity (compare with the normal side) in both extension and 30 degree flexion, a serious injury can be excluded. If there is more obvious laxity, there is probably a complete rupture, which will require operative repair.

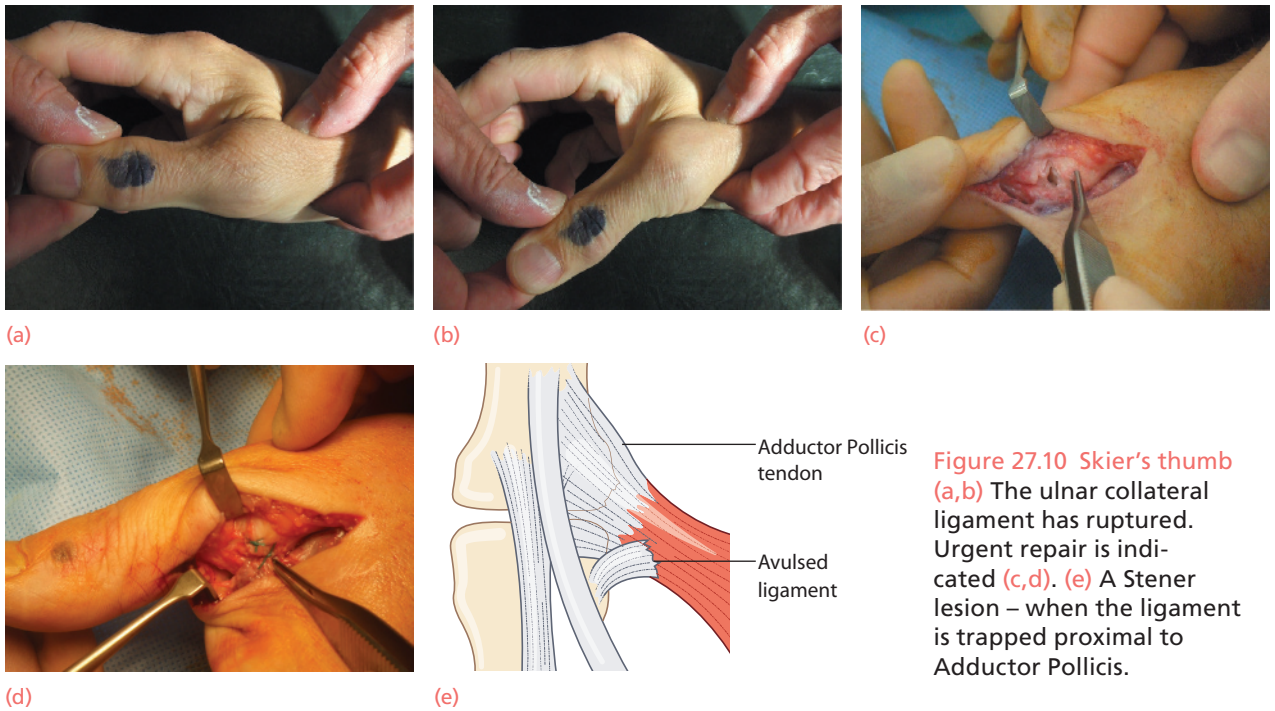


Figure 27.10 Skier's thumb (a,b) The ulnar collateral ligament has ruptured. Urgent repair is indicated (c,d). (e) A Stener lesion – when the ligament is trapped proximal to Adductor Pollicis.

Treatment

PARTIAL TEARS

These can be treated by a short period (2–4 weeks) of immobilization in a splint followed by increasing movement. Pinch should be avoided for 6–8 weeks.

COMPLETE TEARS

Complete tears need operative repair. Care should be taken during the exposure not to injure the superficial radial nerve branches. The Stener lesion is found at the proximal edge of the adductor aponeurosis. The aponeurosis is incised and retracted to expose the ligament and capsule; the torn structures are then carefully repaired.

Postoperatively, the joint is immobilized in a thumb splint for 6 weeks but it can be moved early in the flexion–extension plane as the ligament is isometric (i.e. the same length in flexion and extension). The thumb interphalangeal joint should be left free from the outset to avoid the adductor aponeurosis becoming adherent (which would limit flexion).

A *neglected tear* leads to weakness of pinch. In early cases without articular damage, stability may be restored by using a free tendon graft. If this fails, or if the joint is painful, MP joint arthrodesis is reliable and leaves minimal functional deficit.

In children, the injury may be accompanied by a Salter–Harris type III fracture through the physis. This should be reduced and fixed with smooth K-wires, which should not cross the growth plate.

CLOSED TENDON INJURIES

Flexor tendons

The flexor tendon may avulse, with or without a bone fragment, from the distal phalanx of the flexor tendon (*rugger jersey finger*). This is described above. *Rheumatoid arthritis* is a potent cause of tendon rupture with synovial thickening and invasion, especially within flexor sheaths.

A *volar plate* placed too distally can abrade the FPL tendon, causing loss of thumb-tip flexion. Sharp bone from *osteoarthritis* can lacerate the adjacent flexor tendons. Pisotriquetral arthritis (lacerating the little finger flexors) and STT arthritis (lacerating the FPL tendon and known as Mannerfelt syndrome)) are the usual culprits.

Management requires removal of the cause (usually metalwork or arthritis) then reconstruction with either a tendon transfer or a two-stage tendon graft (discussed below). Primary repair is usually not suitable as the tendon will have been abraded over the length of its excursion against the offending metal or bone.

Extensor tendons

Mallet finger (avulsion from the distal phalanx) is described above. *Rheumatoid arthritis* can rupture the extensors over the back of the unstable DRUJ or within the extensor retinaculum. Osteoarthritis of the DRUJ can abrade the finger extensors (Vaughan Jackson syndrome). *Metalwork* (either the edge of a

dorsal plate or the tip of a protruding screw inserted with a volar plate) can abrade the thumb or finger extensors. EPL can rupture within its tight compartment after an undisplaced distal radius fracture.

Management of extensor tendon rupture is usually the same as that described for flexors – removal of the cause then transfer or graft.

The sagittal bands are transverse structures which maintain the extensor tendon in a central position over the back of the MCP joint. A sagittal band can suddenly rupture, either spontaneously in elderly people especially with rheumatoid arthritis, or after a punch. The extensor tendon is caught in flexion to the ulnar side of the MCP joint. The finger cannot be actively elevated but can be passively centralized. Treatment is either a splint for 6 weeks in extension or early repair.

OPEN INJURIES OF THE HAND

Over 75% of work injuries affect the hands; inadequate treatment costs the patient (and society) dear in terms of functional disability.

Clinical assessment

Open injuries comprise tidy or 'clean' cuts, lacerations, crushing and injection injuries, burns and pulp defects (Figure 27.11).

The precise *mechanism of injury* must be understood. Was the instrument sharp or blunt? Clean or dirty? The position of the fingers (flexed or extended) at the time of injury will influence the relative damage to the deep and superficial flexor tendons. A history of high-pressure injection predicts major soft-tissue damage, however innocuous the wound may seem. What are the patient's occupation, hobbies and aspirations? Is he or she right-handed or left-handed?

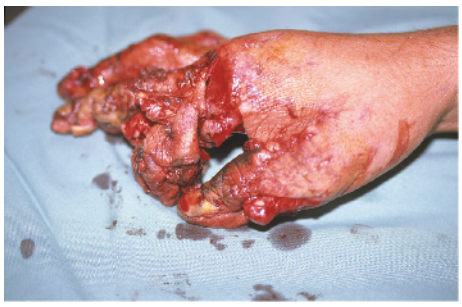
Examination should be gentle and painstaking. *Skin damage* is important, but it should be remembered that even a tiny, clean cut may conceal nerve or tendon damage.

The circulation to the hand and each digit must be assessed. The Allen test can be applied to the hand as a whole or to an individual finger. The radial and ulnar arteries at the wrist are simultaneously compressed by the examiner while the patient clenches his or her fist for several seconds before relaxing; the hand should now be pale. The radial artery is then released; if the hand flushes, it means that the radial blood supply is intact. The test is repeated for the ulnar artery. An injured finger can be assessed in the same way. The digital arteries are occluded by pinching the base of the finger. When blood is squeezed out of the finger, the pulp will become noticeably pale; one digital artery is then released and the pulp should pink up; the test is repeated for the other digital artery.

Sensation is tested in the territory of each nerve. *Two-point discrimination* may be reduced in partial injuries. In children, who are more difficult to examine, the *plastic pen* test is helpful: if a plastic pen is brushed along the skin, it will tend to 'stick' due to the normal thin layer of sweat on the surface; absence of sweating (due to a nerve injury) is revealed by noting that the pen does not adhere as it should (compared to the normal side). Another observation is that the skin in the territory of a divided nerve will not *wrinkle* if immersed in water.

Tendons must be examined with similar care. Start by testing for 'passive tenodesis'. When the wrist is extended passively, the fingers automatically flex in a gentle and regular cascade; when the wrist is flexed, the fingers fall into extension. These actions rely upon the balanced tension of the opposing flexor and extensor tendons to the fingers; if a tendon is cut, the cascade will be disturbed.

Active movements are then tested for each individual tendon (Figure 27.12). Flexor digitorum profundus is tested by holding the proximal finger joint straight and instructing the patient to bend the distal joint. Flexor digitorum superficialis is tested by the examiner holding all the fingers together out straight, then releasing one and asking the patient to bend the proximal joint. Holding the fingers out straight 'immobilizes' all the deep flexors (including that of the finger being tested), which have a common muscle belly. However, in the index finger this test is not 100% reliable because the deep flexor is sometimes



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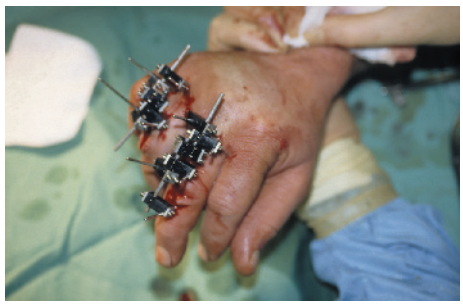


(b)

Figure 27.11 Open injuries (a) A mangled hand; (b) open finger fracture treated with external fixation.

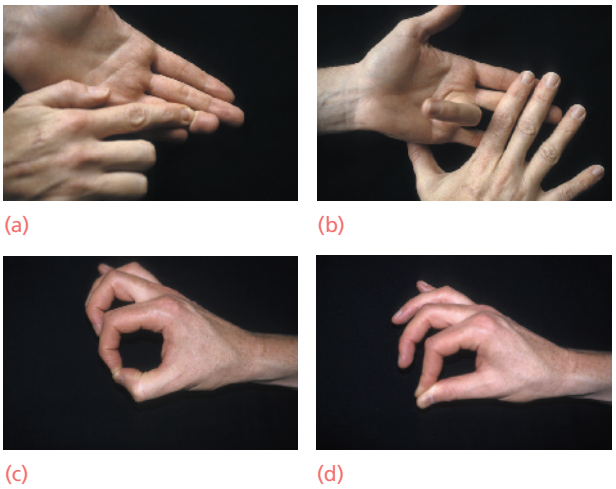


Figure 27.12 Testing the flexor tendons Testing for (a) flexor digitorum profundus (FDP) lesser fingers, (b) flexor digitorum superficialis (FDS) lesser fingers, (c) FDP index, (d) FDS index.

separate. It is better to ask the patient to make a 'circle' between thumb and index (FDP intact) and a 'buttonhole' (FDS intact).

If a tendon is only partly divided, it will still work although it may be painful. In full-thickness skin lacerations, if there is any doubt about the integrity of the tendons, the wound should be explored.

X-rays may show fractures, foreign bodies, air or paint.

Primary treatment

PREOPERATIVE CARE

If the wound is contaminated, it should be rinsed thoroughly with sterile saline; antibiotics should be given as soon as possible. Prophylaxis against tetanus may also be needed. The hand is lightly splinted and the wound is covered with an iodine-soaked dressing. For clean wounds, antibiotics should be omitted to avoid the plague of antibiotic resistance that the unfettered use of antibiotics will bring.

WOUND EXPLORATION

Under general or regional anaesthesia, the wound is cleaned and explored. A pneumatic tourniquet is essential unless there is a crush injury where muscle viability is in doubt. Skin is too precious to waste and only obviously dead skin should be excised. For adequate exposure the wound may need enlarging, but incisions must not directly cross a skin crease or an interdigital web – they must be oblique to avoid skin contracture (Figure 27.13). Through the enlarged wound, loose debris is picked out, dead muscle is excised and the tissues are thoroughly irrigated with saline. A further assessment of the extent of the injury is then undertaken.

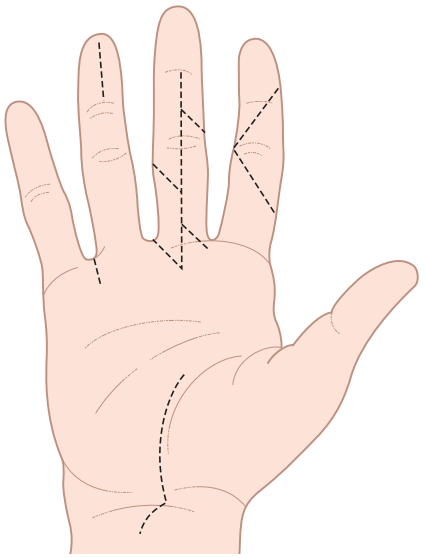


Figure 27.13 Hand incisions 'Permissable' incisions in hand surgery. Incisions must not cross a skin crease or an interdigital web or else scarring may cause contracture and deformity.

Fractures are reduced and held appropriately (splintage, K-wires, external fixator or plate and screws) unless there is some specific contraindication.

NERVE AND VESSEL REPAIR

Joint capsule and ligaments are repaired.

Artery and vein repair may be needed if the hand or finger is ischaemic. This is done with the aid of an operating microscope. Any gap should be bridged with a vein graft.

Severed nerves are sutured under an operating microscope (or at least loupe magnification) with thin inert sutures. If the repair cannot be achieved without tension, a nerve graft (e.g. from the posterior interosseous nerve at the wrist or sural nerve in the leg) should be performed. More recently, dissolvable nerve guides have been used to bridge the gap in digital nerves, allowing biological regeneration across the gap.

EXTENSOR TENDON REPAIR

Extensor tendon repair is not as easy and the results not as reliable as some have suggested. Repair and postoperative management should be meticulous.

FLEXOR TENDON REPAIR

Flexor tendon repair is even more challenging, particularly in the region between the distal palmar crease and the flexor crease of the proximal interphalangeal joint where both the superficial and deep tendons run together in a tight sheath (Zone II – an area that before the advent of reliable suture techniques and postoperative rehabilitation, used to be called more

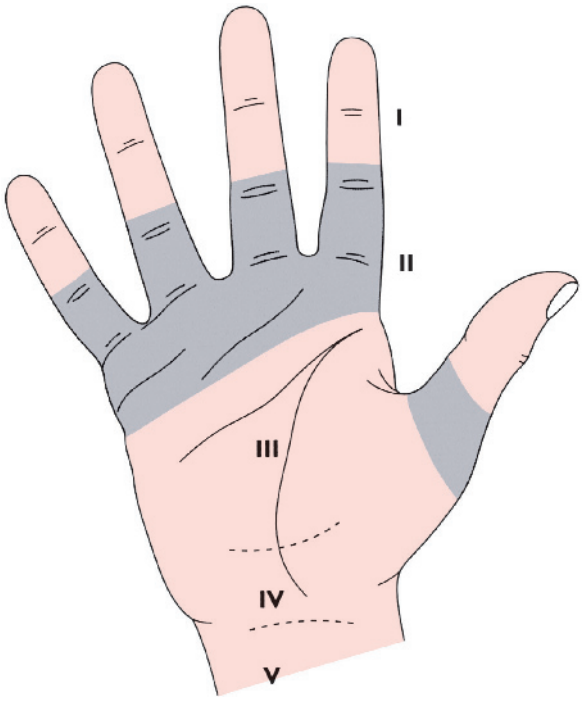


Figure 27.14 The zones of injury I – Distal to the insertion of flexor digitorum superficialis. II – Between the opening of the flexor sheath (the distal palmar crease) and the insertion of flexor superficialis. III – Between the end of the carpal tunnel and the beginning of the flexor sheath. IV – Within the carpal tunnel. V – Proximal to the carpal tunnel.

dramatically 'no man's land') (Figure 27.14). Primary repair by a trained surgeon with fastidious postoperative supervision by a trained and experienced therapist gives the best outcome. If the necessary facilities are not available, the wound should be washed out and loosely closed, and the patient transferred to a special centre. A delay of several days, with a clean wound, is unlikely to affect the outcome when balanced by a more skilled repair.

The tendon repair must be strong and accurate enough to allow early mobilization so that the tendons can glide freely and independently from each other and the sheath. Four strands of locked core suture are placed without handling the tendon any more than is absolutely necessary; this is supplemented by a continuous circumferential suture which strengthens the repair and smoothes it, thus making the gliding action through the sheath easier. The A2 and A4 pulleys must be repaired or reconstructed, otherwise the tendons will bowstring (Figure 27.15). Cuts above the wrist (Zone V), in the palm (Zone III) or distal to the superficialis insertion (Zone I) generally have a better outcome than injuries in the carpal tunnel (Zone IV) or flexor sheath (Zone II). Division of the superficialis tendon noticeably weakens the hand and a

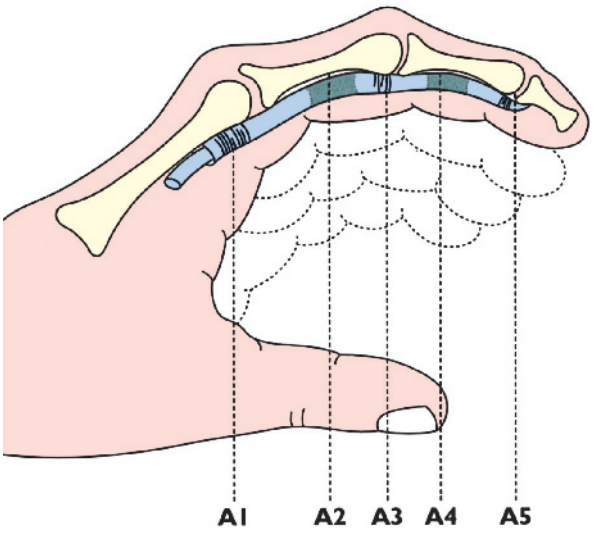


Figure 27.15 The flexor tendon sheath and pulleys Fibrous pulleys – designated A1 to A5 – hold the flexor tendons to the phalanges and prevent bowstringing during movement. A1, A3 and A5 are attached to the palmar plate near each joint; A2 and A4 have a crucial tethering effect and must always be preserved or reconstructed.

swan neck deformity can develop in those with lax ligaments. At least one slip should therefore always be repaired.

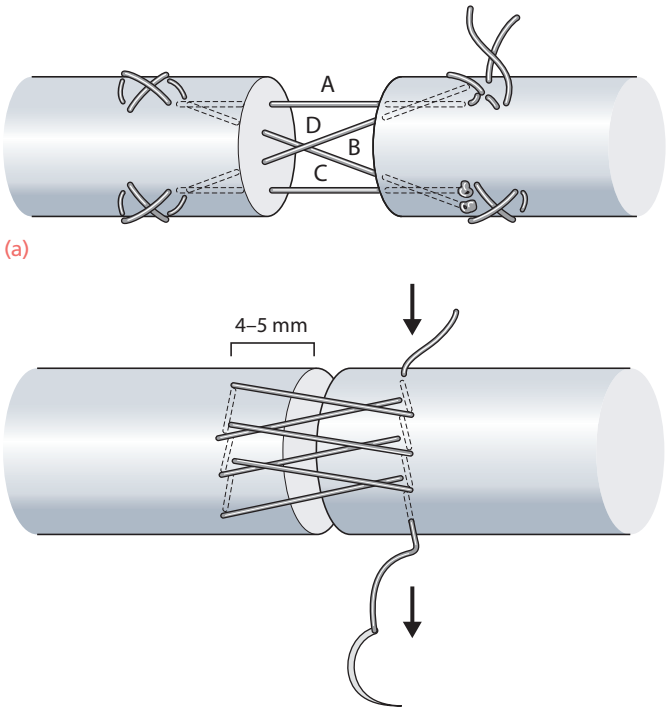
Amputation of a finger as a primary procedure should be avoided unless the damage involves many tissues and is clearly irreparable. Even when a finger has been amputated by the injury, the possibility of reattachment should be considered (see below).

Ring avulsion is a special case. When a finger is caught by a ring, the soft tissues are sheared away from the underlying skeleton. Depending on the amount of damage, skin reattachment, microvascular reconstruction or even amputation may be required.

CLOSURE

The tourniquet is deflated and bipolar diathermy is used to stop bleeding. Haematoma formation risks wound failure, infection and stiffness. Unless the wound is contaminated, the skin is closed – either by direct suture without tension (Figure 27.16) or, if there is skin loss, by skin grafting or skin flaps. Skin grafts are conveniently taken from the inner aspect of the upper arm. If tendon or bare bone is exposed, this must be covered by a rotation or pedicled flap. Sometimes a severely mutilated finger is sacrificed and its skin used as a rotation flap to cover an adjacent area of loss.

Pulp and finger-tip injuries In full-thickness wounds without bone exposure, the wound should be thoroughly cleaned and then covered with a



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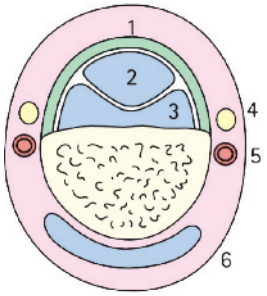


(c)

Figure 27.16 Flexor tendon repair A core 4-strand suture (a) is supplemented by circumferential sutures (b). (c) The relationship of the important structures in 'no man's land': 1 the tendon sheath; 2 flexor digitorum profundus; 3 flexor digitorum superficialis; 4 digital nerve; 5 artery; 6 extensor tendon.

non-adherent dressing. This is left well alone for 7 days; the accumulation of fluid beneath the dressing is not usually a sign of infection and antibiotics should be avoided. The wound is inspected only infrequently, then recovered with the non-adherent dressing, until it heals.

If the open area is greater than 1 cm in diameter, healing will be quicker with a split-skin or full-thickness graft, but the residual pulp cover may not be as satisfactory as a wound that has been left to heal naturally by granulation and re-epithelialization.

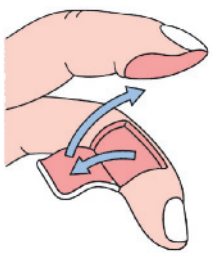
If bone is exposed and length of the digit is important for the individual patient, an advancement flap or neurovascular island flap should be considered. The precise type of flap depends on the orientation of the cut (Figure 27.17). Otherwise, primary cover can be achieved by shortening the bone and tailoring the skin flaps ('terminalization').

In young children, the finger-tips recover extraordinarily well from injury and they should be treated with dressings rather than grafts or terminalization. *Thumb length should never be sacrificed lightly* and every effort should be made to provide a long, sensate digit.

Nail-bed injuries Nail-bed injuries are often seen in association with fractures of the terminal phalanx. If appearance is important, meticulous repair of the nail bed under magnification, replacing any loss with a split thickness nail bed graft from one of the toes, will give the best cosmetic result. In children, these injuries are associated with a physeal fracture.

DRESSING AND SPLINTAGE

The wound is covered with a single layer of paraffin gauze and ample wool roll. A light plaster slab



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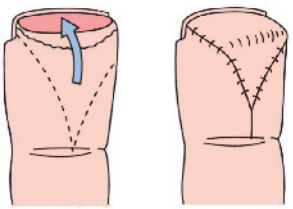

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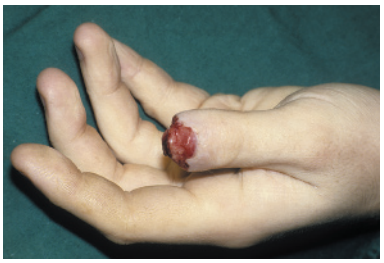
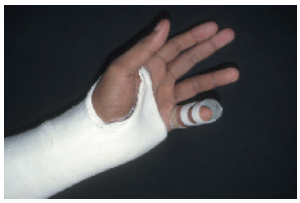


(c)

Figure 27.17 Pulp and finger-tip injuries (a) Cross-finger flap for a palmar oblique finger-tip injury with exposed bone. (b) V-to-Y advancement for a transverse finger-tip injury with exposed bone. Thumb tip loss (c) must always be reconstructed – never amputate.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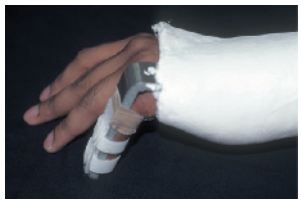
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Figure 27.18 Splintage Always splint in the safe position (wrist slightly extended, MCP joint lint flexed, PIP extended. Immobilize only the affected ray if there is a metacarpal or phalangeal injury.

holds the wrist and hand in the *position of safety* (wrist extended, metacarpophalangeal joints flexed to 90 degrees, interphalangeal joints straight, thumb abducted) (Figure 27.18). This is the position in which the metacarpophalangeal and interphalangeal ligaments are fully stretched and fibrosis therefore least likely to cause contractures. *Failure to appreciate this point is the commonest cause of irrecoverable stiffness after injury.*

- *After primary flexor tendon suture*, the wrist is held with a dorsal splint in about 20 degrees of flexion and the MCP joints in full flexion to take tension off the repair (too much wrist flexion invites wrist stiffness and carpal tunnel symptoms) but the interphalangeal joints must remain straight. There should be minimal restriction by dressings at the front of the fingers, otherwise the resistance can precipitate rupture of the tendon.
- *After extensor tendon repair*, the metacarpophalangeal joints are flexed to only about 30 degrees so that there is less tension on the repair; the wrist is extended to 30 degrees and the interphalangeal joints remain straight.

Postoperative management

IMMEDIATE AFTERCARE

Following an operation, the hand is kept elevated in a roller towel or high sling. If the latter is used, the sling must be removed several times a day to exercise the elbow and shoulder. Too much elbow flexion can stop venous return and make swelling worse. Antibiotics are continued as necessary.

REHABILITATION

Movements of the hand must be commenced within a few days at most. Splintage should allow as many joints as possible to be exercised, consistent with protecting the repair. Most extensor tendon injuries are splinted for about 4 weeks. Dynamic splintage can be used, particularly for injuries at the level of the extensor retinaculum and the metacarpophalangeal joint. Various protocols are followed for flexor tendon injuries, including passive, active or elastic-band assisted flexion. Early movement promotes tendon healing and excursion. In all cases the risk of rupture is balanced against the need for early mobilization. Close supervision and attention to detail are essential.

Once the tissues have healed, the hand is increasingly used for more and more arduous and complex tasks, especially those that resemble the patient's normal job, until he or she is fit to start work; if necessary, his or her work is modified temporarily. If secondary surgery is required, tendon or nerve repair is postponed until the skin is healthy, there is no oedema and the joints have regained a normal range of passive movement.

Replantation

With modern microsurgical techniques and appropriate skill, amputated digits or hands can be replanted. An amputated part should be wrapped in sterile saline gauze and placed in a plastic bag, which is itself placed in watery ice. The 'cold ischaemic time' for a *finger*, which contains so little muscle, is about 30 hours, but the 'warm time' less than 6 hours. For a *hand or forearm*, the cold ischaemic time is only about 12 hours and the warm time much less.

After resuscitation and attention to other potentially life-threatening injuries, the patient and the amputated part should be transferred to a centre where the appropriate surgical skills and facilities are available.

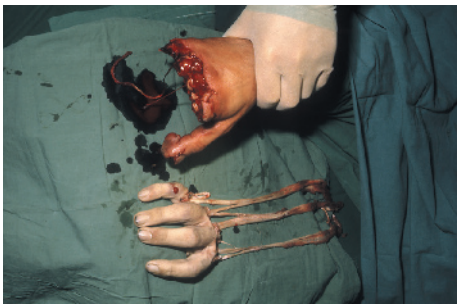
INDICATIONS

The decision to replant depends on the patient's age, his or her social and professional requirements, the condition of the part (whether clean-cut, mangled, crushed or avulsed), and the warm and cold ischaemic time. Furthermore, and perhaps most importantly, it depends on whether the replanted part is likely to give better function than an amputation.

The *thumb* should be replanted whenever possible. Even if it functions only as a perfused 'post' with protective sensation, it will give useful service. *Multiple digits* also should be replanted, and in a child even a single digit. *Proximal amputations* (through the palm, wrist or forearm) likewise merit an attempt at replantation.

RELATIVE CONTRAINDICATIONS

Single digits do badly if replanted. There is a high complication rate, including stiffness, non-union,



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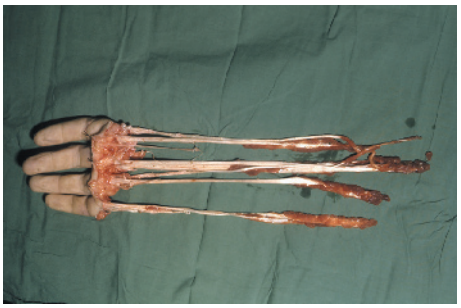

(b)

Figure 27.19 Avulsion This is not replantable.

poor sensation, and cold intolerance; a replanted single finger is likely to be excluded from use. The exception is an amputation beyond the insertion of flexor digitorum superficialis, when a cosmetic, functioning finger-tip can be retrieved. Severely *crushed, mangled or avulsed* parts may not be replantable (Figure 27.19); and parts with a *long ischaemic time* may not survive. *General medical disorders* or *other injuries* may engender unacceptable risks from the prolonged anaesthesia needed for replantation.

BURNS

Generally, hand burns should be dealt with in a specialized unit.

Flame and fluid burns

Superficial burns are covered with moist non-adherent dressings; the hand is elevated and finger movements are encouraged. *Partial-thickness burns* can usually be allowed to heal spontaneously; the hand is dressed with an antimicrobial cream and splinted in the position of safety.

Full-thickness burns will not heal. Devitalized tissue should be excised; the wound is cleaned and dressed and 2–5 days later skin-grafted. Full-thickness circumferential burns may need early escharotomy to preserve the distal circulation. Skin flaps are sometimes needed in sites such as the thumb web which are prone to contracture. The hand should be splinted in the position of safety; K-wires may be needed to maintain this position.

Electric burns

These may cause extensive damage and thrombosis which become apparent only after several days. The patient may, of course, need resuscitation (treating cardiac anomalies and myoglobinuria). The arm needs to be monitored and fasciotomy with debridement of dead tissue is often needed.

Chemical burns

These should be irrigated copiously for 20–30 minutes, usually with water or saline but sometimes with a specific reagent (calcium gluconate for hydrogen fluoride burns, soda lime or magnesium solution for hydrochloric acid, mineral oil for sodium).

INJECTION INJURIES

Oil, grease, solvents, hydraulic fluid or paint injected under pressure are damaging because of tension, toxicity or both. The thumb or index finger is usually involved. Substances can gain entry even through intact skin. Air or lead paint may show on X-ray.

Immediate decompression and removal of the foreign substance offers the best hope. This means an extensive dissection. The outcome is often poor, with amputation sometimes being necessary.

FROSTBITE

Frostbite requires special treatment (Figure 27.20). The limb is rewarmed in a water bath at 40–42 °C for 30 minutes. Oedema is minimized by elevation, and blisters are drained. Digits sometimes need amputation.

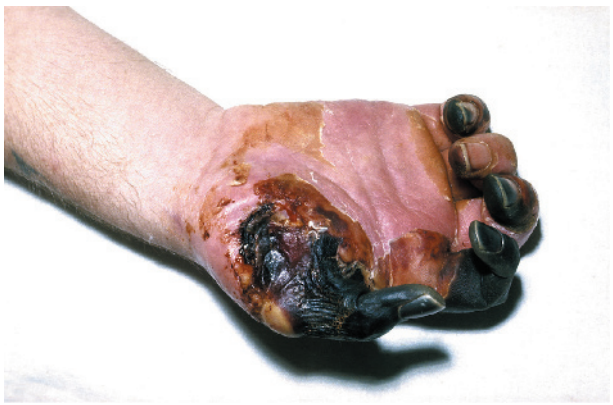

Figure 27.20 Frostbite

DELAYED REPAIR

The primary treatment of hand injuries should always be carried out with an eye to any future reconstructive procedures that might be necessary. These are of three kinds:

- secondary repair or replacement of damaged structures
- amputation of fingers
- reconstruction of a mutilated hand.

Skin

If the skin cover is unsuitable for primary closure or has broken down, it is replaced by a graft or flap. As always, the skin creases must be respected. Contractures are dealt with by Z-plasty, skin grafting, local flaps, regional flaps or free flaps. When important volar surfaces such as the thumb or index tip are insensate, a flap of skin complete with its neurovascular supply may be transposed.

Split-thickness skin contracts and so full-thickness grafts are preferred. The upper inner arm can provide a fair amount of skin leaving a reasonable cosmetic defect. Larger amounts of skin can be harvested from the groin or abdomen. Bear in mind that grafts will not adhere to raw tendon or bone.

Tendons

Primary suture may have been contraindicated by wound contamination, undue delay between injury and repair, massive skin loss or inadequate operating facilities. In these circumstances secondary repair or tendon grafting may be necessary.

Flexor tendons

In a late-presenting *injury of the profundus tendon with an intact superficialis*, advancement of a retracted tendon can cause a flexion deformity of the entire finger. Tendon grafting also is risky: the finger could end up even stiffer. Unless the patient's work or hobby demands flexion of the distal joint and maximum power in the finger, fusion or tenodesis of the distal interphalangeal joint is a more reliable option.

If *both the superficialis and profundus tendons* have been divided and have retracted, a tendon graft is needed. Full passive joint movement is a prerequisite.

In the unlikely event that the pulley system is in good condition and there are no adhesions, the tendons are excised from the flexor sheath and replaced with a tendon graft (palmaris longus, plantaris or a toe extensor). Rehabilitation is the same as for a primary repair.

If the pulleys are damaged, the skin cover poor, the passive range of movement limited or the sheath scarred, a two-stage procedure is preferred. The tendons are excised and the pulleys reconstructed with extensor retinaculum or excised tendon. A Silastic rod is sutured to the distal stump of the profundus tendon and left free proximally in either the palm or distal forearm. Rehabilitation is planned to maintain a good passive range of movement. A smooth gliding surface forms around the rod. At least 3 months later, the rod is removed through two smaller incisions and a tendon graft (palmaris longus, plantaris or a lesser toe extensor) is sutured to the proximal and distal stumps of flexor digitorum profundus. Rehabilitation is the same as that for a primary repair.

Tenolysis

This is sometimes needed after flexor tendon repair in Zone II. A poor excursion is not infrequent because of adhesions between the tendons and the sheath. There is some active movement – indicating that the tendon is intact – but not enough for good function. The passive range of movement should be good if the tenolysis is to succeed. The tendons are painstakingly freed through small windows in the flexor sheath. Postoperatively an intensive programme of movement is essential, otherwise there will be even more scar tissue than before and the tenolysis will have made matters worse.

Tenolysis of the extensor tendons is commonly needed after a fracture of the proximal phalanx when the tendons have a propensity to get stuck to the metalwork and bone.

Nerves

Late-presenting nerve injuries must be carefully assessed. The results of repair deteriorate with time, particularly for motor nerves where the end plate begins to fail and the muscle begins to fibrose. If several months have passed, tendon transfer may be a more reliable alternative. If nerve repair is attempted, the scar is excised and the stumps pared back until healthy nerve is found proximally and distally; a nerve graft or tubular nerve guide is usually needed to avoid tension at the suture line.

Joints

The proximal interphalangeal joint is most prone to a flexion contracture. Active and passive exercises can be supplemented by serial static splints or dynamic splints. If stiffness persists, once the pain has subsided (and that can take several months) surgery is considered. Under a local anaesthetic, the contracted

proximal edge of the palmar plate – the so-called 'check rein ligament' – is excised. Early active and passive mobilization to maintain the correction is essential. Unstable joints are best fused; stable but painful joints can be fused (especially the DIP joints and the index or middle PIP joints) or replaced (especially the ring and little PIP joints).

Bones

Malunion, especially if rotational, may require treatment. Non-union is very uncommon but, if present, grafting may be required. Extensor tendons may stick to bone, most commonly after plate fixation of the proximal phalanx. Plate removal and tenolysis is followed by diligent active and passive movements: a fair result is usually achieved.

AMPUTATION

Indications

A finger is amputated only if it remains painful or unhealed, or if it is a nuisance (i.e. the patient cannot bend it, straighten it or feel with it), and then only if repair is impossible or uneconomic. There can be significant issues with amputation, not least of which are cold intolerance, painful neuroma and psychological distress.

Technique

THUMB

The length of the amputation stump must be maximized in the thumb where every millimetre is worth preserving to bring the tip competently against the index finger. Even a stiff or deformed thumb is worth keeping. This can be achieved by dressings, local advancement flaps or neurovascular island flaps, or by a cross-finger flap from the back of the index finger.

FINGER-TIP AMPUTATION ('TERMINALIZATION')

In the other digits, the loss of length can be minimized with similar techniques. Although a cross-finger flap is fairly straightforward and provides good skin cover, sensation is limited and a flexion contracture can develop in the donor finger. The final choice depends on the patient's requirements and the surgeon's skill.

DIGITS

The index finger should usually not be amputated at the PIP level as the stump tends to interfere with thumb span; amputation at the base is usually functionally more acceptable.

The middle and ring fingers should not be amputated through the knuckle joint because cosmetically this is unsatisfactory and small objects will fall through the gap ('incontinence of grip'). If the proximal phalanx can be left, the appearance is still abnormal but function is better. The extensor tendon must never be sutured to the flexor tendon; this will act as a tether on the common belly of flexor digitorum profundus and prevent the other digits from flexing fully (the 'quadriga effect', named after the four reins in a chariot). If the middle phalanx is amputated distal to the flexor digitorum superficialis insertion, the profundus tendon continues to pull, but now through the lumbrical, making the proximal interphalangeal joint paradoxically extend rather than flex. This irritating anomaly is avoided by suturing the superficialis stump to the flexor sheath or by dividing the lumbrical.

For more proximal injuries, the entire finger with most of its metacarpal may be amputated; the hand is weakened but the appearance is usually satisfactory. If the middle ray is amputated through the metacarpal, the index finger may 'scissor' across it in flexion; this can be overcome by dividing the adjacent index metacarpal and transposing it to the stump of the middle metacarpal.

LATE RECONSTRUCTION

A severely mutilated hand should be dealt with by a hand expert. Certain options may be considered in exceptional cases. If the thumb has been lost, the options include pollicization (shortening and rotating the index finger to oppose the other fingers), microvascular free transfer of the second toe (Figure 27.21) and osteoplastic reconstruction (a cortical bone graft surrounded by a skin flap). If all the fingers have been lost but the thumb is present, a new finger can similarly be constructed with either a free toe transfer or an osteoplastic digit.

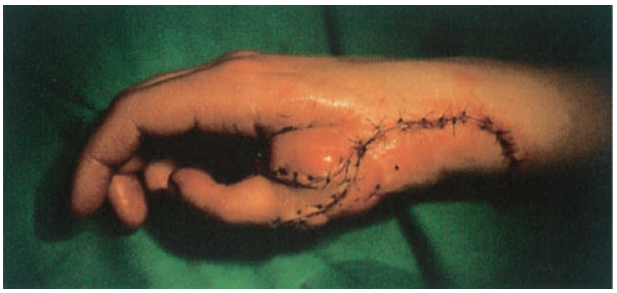

Figure 27.21 Late reconstruction The second toe has been transferred to replace the thumb, which was severed in an accident.

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Injuries of the spine 28

Robert Dunn & Nicholas Kruger

PATHOPHYSIOLOGY OF SPINE INJURIES

Stable and unstable injuries

A stable injury is one in which the vertebral components will not be displaced by normal physiological loads, whereas an unstable injury is one in which there is a significant risk of displacement and damage to the neural tissues. Historically, many classification systems were developed to describe thoracolumbar spinal injuries. In the 1960s Holdsworth and later Kelly and Whitesides described a two-column concept of spinal stability. This was superseded in 1983 by the three-column theory of Denis which became the most widely used system of classification of spinal injuries (Figure 28.1).

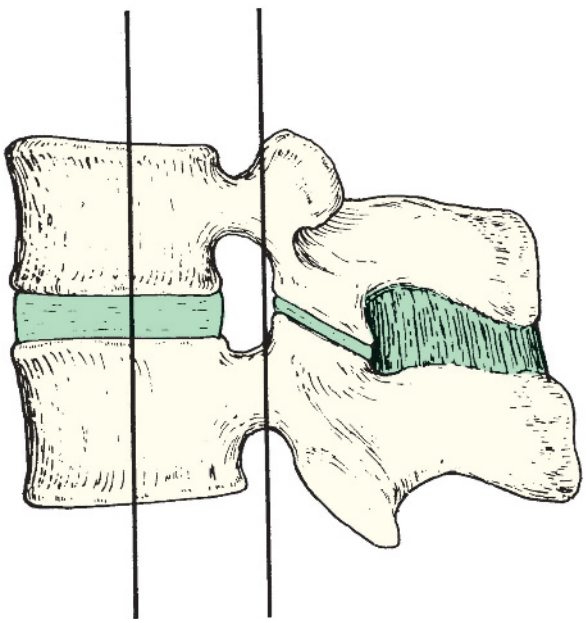


Figure 28.1 Structural elements of the spine The vertical lines show the Denis classification of the structural elements of the spine. The three elements are: the posterior complex, the middle component and the anterior column. This concept is particularly useful in assessing the stability of lumbar injuries.

This classification has now been replaced by the AO/ASIF (Arbeitsgemeinschaft für Osteosynthesefragen/Association for the Study of Internal Fixation) system which reverts back to the original two-column theory, the *anterior column* consisting of vertebral body and disc and the *posterior column* consisting of pedicles, laminae, facets and posterior ligamentous complex.

- *Type A injuries* – anterior column compression fractures which tend to be stable
- *Type B injuries* – involve anterior and posterior columns with distraction; these are unstable
- *Type C injuries* – double-column injuries with rotation or sheer; these are unstable.

Fortunately, only 10% of spinal fractures are unstable and less than 5% are associated with cord damage.

Pathophysiology

PRIMARY CHANGES

Physical injury may be limited to the vertebral column, including its soft-tissue components, and varies from ligamentous strains to vertebral fractures and fracture-dislocations. The spinal cord and/or nerve roots may be injured, either by the initial trauma or by ongoing structural instability of a vertebral segment, causing direct compression, severe energy transfer, physical disruption or damage to its blood supply.

SECONDARY CHANGES

During the hours and days following a spinal injury biochemical changes may lead to more gradual cellular disruption and extension of the initial neurological damage.

Mechanism of injury

There are three basic mechanisms of injury: *traction (avulsion)*, *direct injury* and *indirect injury*.

TRACTION (AVULSION) INJURY

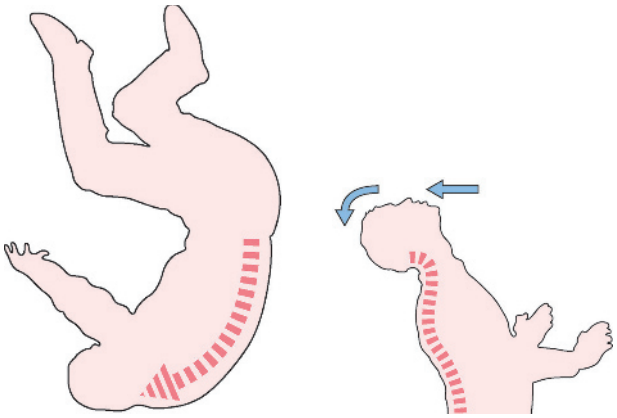
In the lumbar spine resisted muscle effort may avulse transverse processes; in the cervical spine the seventh spinous process can be avulsed ('clay-shoveller's fracture').

DIRECT INJURY

Penetrating injuries to the spine, particularly from firearms and knives, are becoming increasingly common. There rarely cause vertebral column instability but do commonly result in direct neurological injury.

INDIRECT INJURY

This is the most common cause of significant spinal damage; it occurs most typically in a fall from a height when the spinal column collapses in its vertical axis, or else during violent free movements of the neck or trunk (Figure 28.2). A variety of forces may be applied to the spine (often simultaneously): axial compression, flexion, lateral compression, flexion-rotation, shear, flexion-distraction and extension.



(a) (b)

Figure 28.2 Mechanism of injury The spine is usually injured in one of two ways: (a) a fall onto the head or the back of the neck; or (b) a blow on the forehead, which forces the neck into hyperextension.

NOTE: *Insufficiency fractures* may occur with minimal force in bone which is weakened by osteoporosis or a pathological lesion.

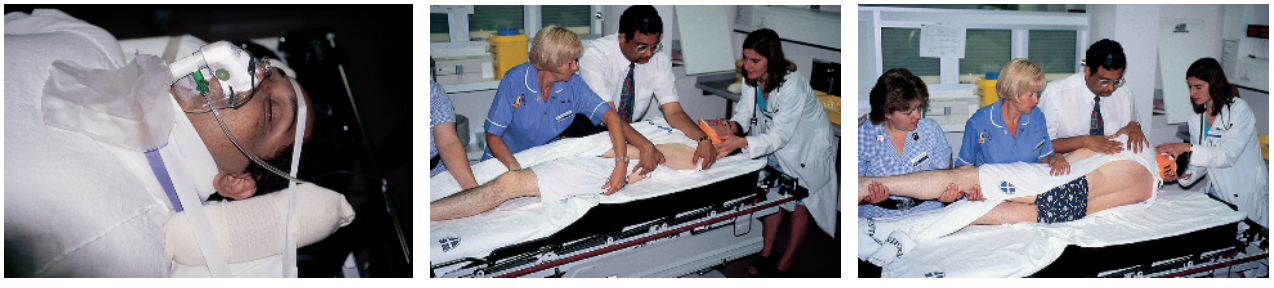
Healing

Spinal injuries include both bony and soft-tissue (ligaments, facet joint capsule and intervertebral disc) elements. Bony injuries tend to heal but the patient may be left with kyphosis or loss of height. Ligamentous injuries seldom heal to a stable state and will potentially lead to progressive kyphosis, chronic pain and further neurological sequelae.

EARLY MANAGEMENT

Assessment and resuscitation according to a recognized protocol, such as the ATLS® protocol, precedes the assessment of the spinal injury. Adequate oxygenation and perfusion helps minimize secondary spinal cord injury. Spinal precautions need to be followed until the patient has been resuscitated and other life-threatening injuries have been managed. Immobilization continues until spinal injury has been excluded by both clinical and radiological assessment.

Temporary immobilization with a semi-rigid collar and sandbags may be used when transferring a spinal injury patient into a CT or MRI scanner. *Log-rolling* with spinal precautions is mandatory to avoid further injury, and the patient should be transferred to a pressure care mattress as soon as possible to avoid bedsores (Figure 28.3).



(a) (b) (c)

Figure 28.3 Spinal injuries – early management (a) Quadruple immobilization: the patient is on a backboard, the head is supported by sandbags and held with tape across the forehead, and a semi-rigid collar has been applied. (b,c) The log-rolling technique for exposure and examination of the back.

DIAGNOSIS

History

A high index of suspicion is essential since symptoms and signs may be minimal. The history is important and, with any high-energy injury such as high-speed traffic accidents or falls from heights, the emphasis should be on excluding a vertebral column injury. Unconscious and polytraumatized patients need to be considered as having an unstable spinal injury until proven otherwise. Any history of trauma with neck/back pain or neurological symptoms needs careful examination and investigation to exclude spinal injury.

Examination

The head and face are thoroughly inspected for bruises or grazes which could indicate indirect trauma to the cervical spine (Figure 28.4). Note the attitude of the head; torticollis may allude to an underlying axial cervical spine injury. The neck is inspected for deformity, bruising or penetrating injury. The patient is 'log-rolled' (i.e. turned over 'in one piece') to avoid movement of the vertebral column. The spine is inspected from occiput to coccyx for deformity, penetrating injury, haematoma or bruising. The spinous processes are palpated for tenderness, haematoma, gap or a step which may indicate instability. A digital rectal examination completes the log-roll, and anal tone, sensation and voluntary 'pinch' are documented.

GENERAL EXAMINATION – 'SHOCK'

Early examination of the severely injured patent is considered in Chapter 22. The ABC sequence of advanced trauma life support (ATLS®) always takes precedence. Three types of shock may be encountered in patients with spinal injury.

Hypovolaemic shock This is suggested by tachycardia, peripheral shutdown and, in later stages, hypotension.

Neurogenic shock This reflects loss of the sympathetic pathways in the spinal cord; the peripheral vessels dilate, causing hypotension, but the heart, with no sympathetic innervation, remains bradycardic. The combination of paralysis, warm and well-perfused peripheral areas, bradycardia and hypotension with a low diastolic blood pressure suggests neurogenic shock. Overenthusiastic use of fluids can cause pulmonary oedema; atropine and vasopressors may be required.

'Spinal shock' The physiological dysfunction following structural injury rarely lasts for more than 48 hours. Below the level of the injury, the muscles are flaccid, the reflexes are absent and sensation is lost. While the primitive reflexes (anal 'wink' and the bulbocavernosus reflex) are absent, there is spinal shock and the neurological level cannot be accurately determined. The bulbocavernosus reflex is commonly elicited by tugging on the Foley catheter during a digital rectal exam. If an anal contraction occurs, the bulbocavernosus reflex is present and the patient is out of spinal shock. A complete spinal cord lesion cannot be diagnosed in the presence of spinal shock, as the neurological level may still improve.

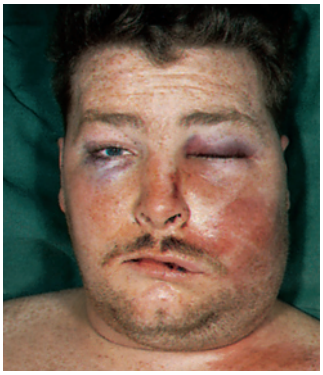
NEUROLOGICAL EXAMINATION

A full neurological examination is carried out in every case; this may have to be repeated several times during the first few days. Each dermatome, myotome and reflex is tested (Figure 28.5; Tables 28.1 and 28.2).

Cord longitudinal column functions are assessed: corticospinal tract (posterolateral cord, ipsilateral motor power), spinothalamic tract (anterolateral cord, contralateral pain and temperature) and posterior columns (ipsilateral proprioception).

Sacral sparing should be tested for. Preservation of active great toe flexion, active anal squeeze (on digital examination) and intact perianal sensation suggest a partial rather than complete lesion. Further recovery may occur.

The unconscious patient is difficult to examine; a spinal injury must be assumed until proven



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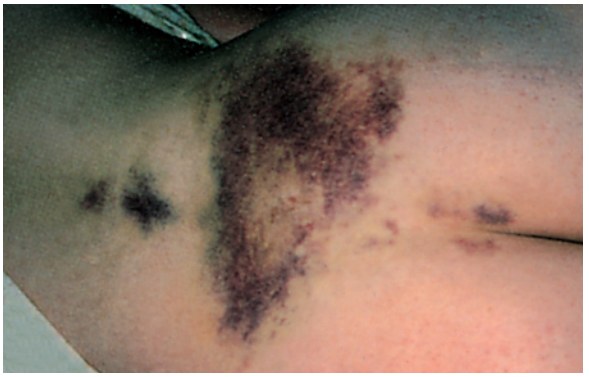


(b)

Figure 28.4 Spinal injuries – suspicious signs First appearances do matter. (a) With severe facial bruising, always suspect a hyperextension injury of the neck. (b) Bruising over the lower back should raise the suspicion of a lumbar vertebral fracture.

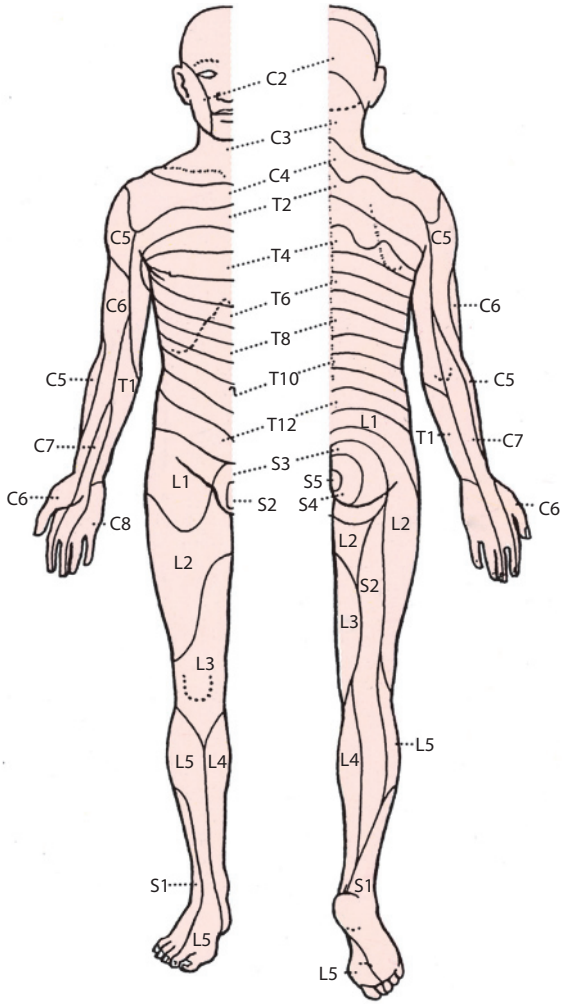


Figure 28.5 Spine injuries – neurological examination
Dermatomes supplied by the spinal nerve roots.

otherwise. Clues to the existence of a spinal cord lesion are a history of a fall or rapid deceleration, a head injury, diaphragmatic breathing, a flaccid anal sphincter, hypotension with bradycardia and a pain response above, but not below, the clavicle. Priapism is an important finding since it occurs almost immediately with 'complete' cord injuries and settles in a few hours. Loss of sympathetic control causes uncontrolled arterial inflow directly into the penile sinusoidal spaces with a subsequent high-flow priapism.

The neurological examination should be recorded on the ASIA scoring system, which is the standard neurological classification of spinal cord injury (Figure 28.6).

IMAGING

- *X-ray* examination of the spine is mandatory for all accident victims complaining of pain or stiffness in the neck or back or peripheral paraesthesiae, all patients with head injuries or severe facial injuries (cervical spine), patients with rib fractures or severe seat-belt bruising (thoracic spine), and those with severe pelvic or abdominal injuries (thoracolumbar spine). This is performed during the secondary survey.
- Accident victims who are unconscious should have spine imaging as part of the routine workup.
- Elderly people and patients with known vertebral pathology (e.g. ankylosing spondylitis) may suffer fractures after comparatively minor back injury. The spine should be X-rayed even if pain is not marked.
- In addition to AP and lateral X-ray views, open-mouth views are needed for the upper two cervical vertebrae. The cervicothoracic junction, if poorly visualized, will need a 'swimmer's view' with the arm abducted. 'Flexion-and-extension' X-rays are not requested initially.
- 'Difficult' areas – such as the upper cervical spine, the cervicothoracic junction and the upper thoracic segments, which are often obscured by shoulder and rib images – are imaged with *CT* or *MRI*. Odontoid fractures are best appreciated on CT sagittal and coronal views. CT is ideal for showing structural damage to individual vertebrae and displacement of bone fragments into the canal (Figure 28.7), and it helps define complex fracture patterns. Helical CT scanning allows rapid acquisition times, high-definition images and multiplanar reconstruction views including 3D reconstructions. It has a higher radiation exposure than plain X-rays but its advantages make it the imaging modality of choice in modern trauma centres.

Table 28.1 Tests for nerve root motor function

Nerve root	Test
C5	Elbow flexion
C6	Wrist extension
C7	Wrist flexion, finger extension
C8	Finger flexion
T1	Finger abduction
L1,2	Hip abduction
L3,4	Knee extension
L5, S1	Knee flexion
L5	Great toe extension
S1	Great toe flexion

Table 28.2 Root values for tendon reflexes

Root value	Tendon reflex
C5	Biceps
C6	Brachioradialis
C7	Triceps
L3,4	Quadriceps
L5,S1	Achilles tendon

Patient Name _____

Examiner Name _____ Date/Time of Exam_____

ASIA American Spinal Injury Association

ISCOS

STANDARD NEUROLOGICAL CLASSIFICATION OF SPINAL CORD INJURY

MOTOR

KEY MUSCLES (scoring on reverse side)

	R	L	
C5	☐	☐	Elbow flexors
C6	☐	☐	Wrist extensors
C7	☐	☐	Elbow extensors
C8	☐	☐	Finger flexors (distal phalanx of middle finger)
T1	☐	☐	Finger abductors (little finger)

UPPER LIMB TOTAL (MAXIMUM) ☐ + ☐ = ☐ (25) (25) (50)

Comments:

	R	L	
L2	☐	☐	Hip flexors
L3	☐	☐	Knee extensors
L4	☐	☐	Ankle dorsiflexors
L5	☐	☐	Long toe extensors
S1	☐	☐	Ankle plantar flexors

Voluntary anal contraction (Yes/No) ☐

LOWER LIMB TOTAL (MAXIMUM) ☐ + ☐ = ☐ (25) (25) (50)

SENSORY

KEY SENSORY POINTS

	LIGHT TOUCH R	LIGHT TOUCH L	PIN PRICK R	PIN PRICK L
C2				
C3				
C4				
C5				
C6				
C7				
C8				
T1				
T2				
T3				
T4				
T5				
T6				
T7				
T8				
T9				
T10				
T11				
T12				
L1				
L2				
L3				
L4				
L5				
S1				
S2				
S3				
S4-5				

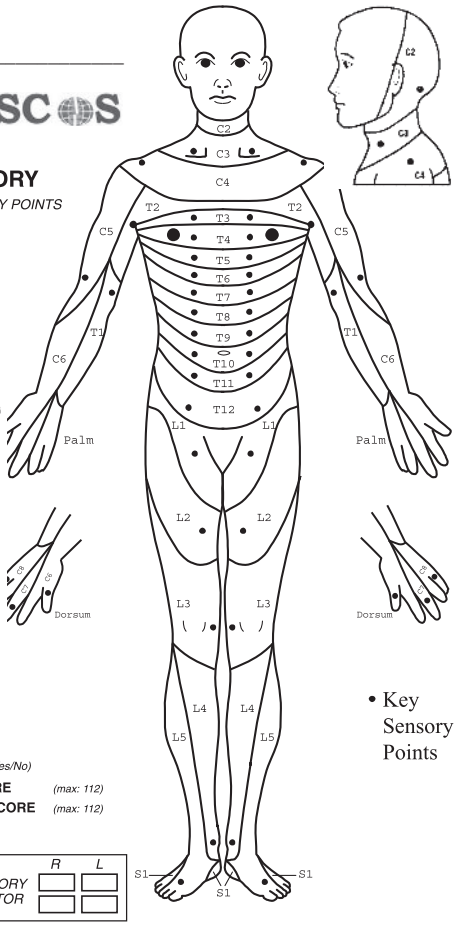
0 = absent
1 = impaired
2 = normal
NT = not testable

Any anal sensation (Yes/No) ☐

TOTALS (MAXIMUM) (56) (56) (56) (56) ☐ + ☐ = ☐

PIN PRICK SCORE ☐ (max: 112)

LIGHT TOUCH SCORE ☐ (max: 112)



NEUROLOGICAL LEVEL (The most caudal segment with normal function)	SENSORY R ☐ L ☐ / MOTOR R ☐ L ☐	COMPLETE OR INCOMPLETE? (Incomplete = Any sensory or motor function in S4-S5) ☐	ZONE OF PARTIAL PRESERVATION (Caudal extent of partially innervated segments)	SENSORY R ☐ L ☐ / MOTOR R ☐ L ☐
		ASIA IMPAIRMENT SCALE ☐		

REV 03/06

MUSCLE GRADING

0 total paralysis

1 palpable or visible contraction

2 active movement, full range of motion, gravity eliminated

3 active movement, full range of motion, against gravity

4 active movement, full range of motion, against gravity and provides some resistance

5 active movement, full range of motion, against gravity and provides normal resistance

5* muscle able to exert, in examiner's judgement, sufficient resistance to be considered normal if identifiable inhibiting factors were not present

NT not testable. Patient unable to reliably exert effort or muscle unavailable for testing due to factors such as immobilization, pain on effort or contracture.

ASIA IMPAIRMENT SCALE

☐ **A = Complete**: No motor or sensory function is preserved in the sacral segments S4-S5.

☐ **B = Incomplete:** Sensory but not motor function is preserved below the neurological level and includes the sacral segments S4-S5.

☐ **C = Incomplete:** Motor function is preserved below the neurological level, and more than half of key muscles below the neurological level have a muscle grade less than 3.

☐ **D = Incomplete:** Motor function is preserved below the neurological level, and at least half of key muscles below the neurological level have a muscle grade of 3 or more.

☐ **E = Normal:** Motor and sensory function are normal.

CLINICAL SYNDROMES (OPTIONAL)

☐ Central Cord
☐ Brown-Sequard
☐ Anterior Cord
☐ Conus Medullaris
☐ Cauda Equina

STEPS IN CLASSIFICATION

The following order is recommended in determining the classification of individuals with SCI.

1. Determine sensory levels for right and left sides.
2. Determine motor levels for right and left sides.
   Note: in regions where there is no myotome to test, the motor level is presumed to be the same as the sensory level.
3. Determine the single neurological level.
   This is the lowest segment where motor and sensory function is normal on both sides, and is the most cephalad of the sensory and motor levels determined in steps 1 and 2.
4. Determine whether the injury is Complete or Incomplete (sacral sparing).
   If voluntary anal contraction = **No** *AND all S4-5 sensory scores* = **0** *AND any anal sensation* = **No**, *then injury is COMPLETE. Otherwise injury is incomplete.*
5. Determine ASIA Impairment Scale (AIS) Grade:

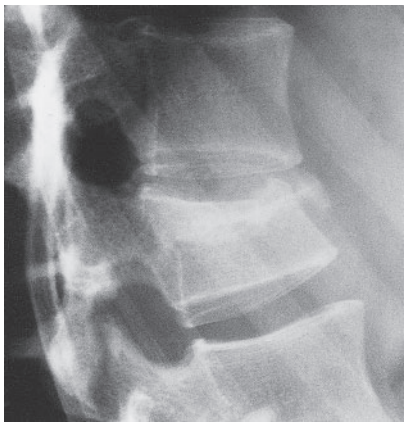
Is injury Complete?
NO ↓ — If **YES**, AIS=A Record ZPP (For ZPP record lowest dermatome or myotome on each side with some (non-zero score) preservation)

Is injury motor incomplete?
YES ↓ — If **NO**, AIS=B (Yes=voluntary anal contraction OR motor function more than three levels below the motor level on a given side.)

Are at least half of the key muscles below the (single) neurological level graded 3 or better?
NO ↓ AIS=C
YES ↓ AIS=D

If sensation and motor function is normal in all segments, AIS=E
Note: AIS E is used in follow up testing when an individual with a documented SCI has recovered normal function. If at initial testing no deficits are found, the individual is neurologically intact; the ASIA Impairment Scale does not apply.

Figure 28.6 The ASIA score sheet



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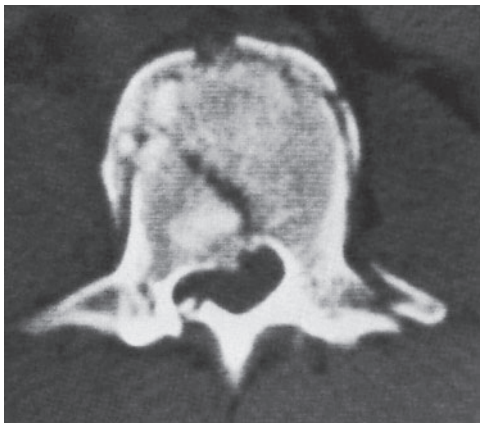


(b)

Figure 28.7 X-ray diagnosis (a) This X-ray showed the fracture, but it needed a CT scan (b) to reveal the large fragment encroaching on the spinal canal.

- MRI is the investigation of choice for displaying intervertebral discs, ligamentum flavum and neural structures, and it is indicated for all patients with neurological signs and those who are considered for surgery. MRI may help with decisions about spinal stability by demonstrating disruption to the posterior ligamentous complex.

PRINCIPLES OF DEFINITIVE TREATMENT

The *objectives of treatment* are:

- to preserve neurological function
- to minimize a perceived threat of neurological compression
- to stabilize the spine
- to rehabilitate the patient.

The *indications for urgent surgical stabilization* are:

- an unstable fracture with incomplete neurological deficit or progressive neurological deficit with imaging confirming neural compression
- an unstable fracture in a polytraumatized patient (relative indication).

Stability

'Clinical instability' is defined as a loss in the ability of the spine, under physiologic loads, to maintain relationships between vertebrae in such a way that there is neither damage nor subsequent irritation to the spinal cord or nerve roots. In addition, there is no development of incapacitating deformity or pain due to structural changes.

Three factors need to be assessed in deciding on clinical stability:

- *Morphology of the injury* – From imaging, the anterior column is assessed for kyphosis, collapse and coronal translation.
- *Integrity of the posterior ligamentous complexes from clinical examination and/or imaging* – Clinical assessment will reveal bony tenderness or a gap in the interspinous processes which suggests loss of the posterior tension band. This may be supported by MRI findings. An intact posterior tension band is a major determinant of stability of the vertebral column.
- *Presence of neurological deficit from clinical examination* – Neurological deficit is more common in unstable injuries and often lowers the threshold for surgical stabilization.

Bone injuries can be initially unstable but will stabilize with time due to bony healing over several weeks and may be managed non-operatively, depending on socioeconomic factors and surgical expertise. Spinal injuries with predominantly discoligamentous injuries are unlikely to stabilize and will require surgical stabilization.

Patients with no neurological injury

STABLE INJURIES

These may be managed with bed rest until pain and muscle spasm subside. For pain control, bracing may be used.

UNSTABLE INJURIES

Unstable fractures will unite over 3 months and may involve part bed rest/traction and part bracing, depending on the injury. In the cervical spine this should be done as soon as possible by traction, using tongs or a halo device attached to the skull. If the halo is attached to a body cast, the combination can be used as an external fixator for prolonged immobilization out of bed (see below). Prolonged bed rest, however, is not always socioeconomically viable and surgical stabilization may be offered.

Surgical stabilization is standard for unstable spinal column injuries. Patients can be mobilized earlier with reduction in morbidity from prolonged bed rest.

Deformity correction and maintenance is improved and shorter hospitalization times decrease the overall socioeconomic cost.

Patients with a neurological injury

Once out of spinal shock, the full extent of the neurological injury is assessed. Spinal injury patients need a multidisciplinary team to manage their multisystem physiological impairment and malfunction, including the spinal injury. Intensive care units (ICUs) or high care units are required during the acute injury phase. Whenever feasible, the patient should be transferred to a spinal injury centre as soon as possible after injury in order to receive early appropriate surgical care, multidisciplinary involvement and rehabilitation.

STABLE SPINAL INJURIES

These patients can be treated conservatively and rehabilitated as soon as possible. The most common causes are penetrating injuries from stabs, bullets and projectiles. These cause direct neurological injury and almost never vertebral column instability.

UNSTABLE SPINAL INJURIES

Conservative treatment can be used where surgical skills and resources are not available. It is an inferior option due to problems with prolonged bed rest, hospitalization costs, delayed rehabilitation and risks of progressive spinal deformity and pain. Discoligamentous injuries will not stabilize and require surgery to avoid progressive deformity.

Surgical stabilization has been shown to be safe, and it results in shorter hospitalization periods, reduced healthcare costs, earlier rehabilitation and improved neurological outcomes.

Complete spinal cord injuries do not benefit from decompression, and surgical goals are to establish spinal column alignment and stability.

In '*incomplete*' cord injuries with spinal cord compression, decompression is required. There is a growing trend for early surgery supported by an increasing volume of animal and human studies reporting improved neurological outcomes. More recently, large multicentre trials have shown improved neurological outcomes with surgery within 24 hours.

The one clear indication for urgent surgery is progressive neurological deterioration, with imaging (MRI or CT scan) confirming cord compression.

'MEDICAL TREATMENT' OF CORD INJURIES

From the mid 1980s to 1990s several large trials evaluated the use of corticosteroids following acute spinal cord injuries. Enthusiasm for steroids was tempered by study design faults, lack of demonstrated functional improvements in outcomes and increasing evidence for the complications associated with large doses of corticosteroids. For these reasons corticosteroids have fallen out of favour and are not routinely recommended for spinal cord injury. Research is continuing into tissue oxygenation, perfusion and various neuroprotective and neuroregenerative agents.

TREATMENT METHODS

Cervical spine

Collars *Soft collars* offer little support and their use is restricted to minor sprains for the first few days after injury. *Semi-rigid collars* are widely used in the acute setting but are not adequate for unstable injury patterns. *Four-poster braces* are more stable, applying pressure to the mandible, occiput, sternum and upper thoracic spine. They can be uncomfortable and, with bedbound patients, may cause suboccipital pressure sores.

Tongs A pin is inserted into the outer table on each side of the skull; these are mounted on a pair of tongs and traction is applied to reduce the fracture or dislocation and to maintain the reduced position (Figure 28.8).

Halo ring At least four pins are inserted into the outer table of the skull and a ring is applied. The use of titanium pins and a graphite ring allows an MRI scan to be performed. The halo ring can be used for initial traction and reduction of the fracture or dislocation, and it can then be attached to a plaster vest or a fitted body orthosis (Figure 28.9). Proper positioning and torque-pressure of the pins is essential. Bear in mind that the use of a halo vest carries a significant risk of complications such as pin loosening, pin-site infection and (in elderly patients) respiratory distress.

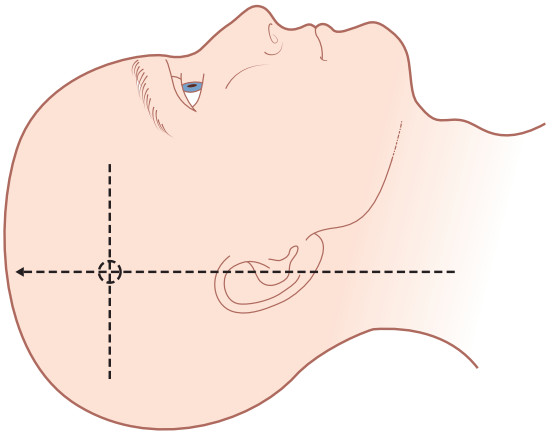
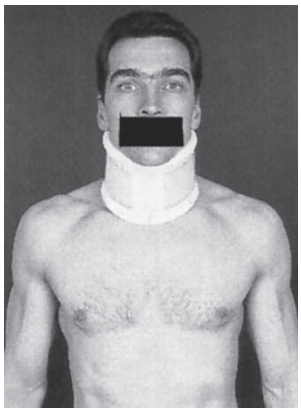


Figure 28.8 Cone's calliper pin location The intersection of a vertical line through the external auditory meatus and a horizontal line 1 cm above the ear pinna marks the location for the pin insertion.



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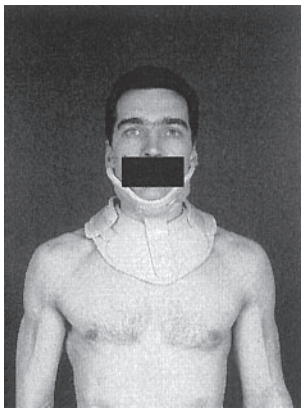

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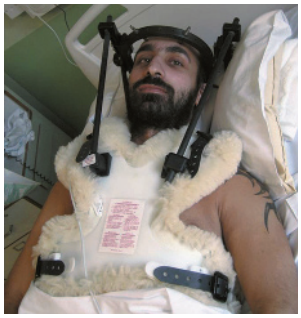


(c)

Figure 28.9 Spine injuries treatment (a) Standard cervical collar; (b) more rigid variety; (c) halo body cast.

Fixation Various operative procedures are available, depending on the level and pattern of injury. *Odontoid fractures* can be fixed with lag screws, *burst fractures* can be decompressed through an anterior approach, and *facet dislocations* can be reduced through a posterior approach. The spine can be stabilized anteriorly with plate and/or cage constructs or posteriorly with interspinous wiring, lateral mass screws or pedicle screws.

Thoracolumbar spine

Beds Special beds are used in the management of spinal injuries. They are designed to avoid pressure sores (with special mattresses or the facility to turn the patient frequently).

Brace A thoracolumbar brace avoids flexion by three-point fixation. It is suitable for some burst fractures, seat-belt injuries and compression fractures. Recent studies have found bracing unnecessary for stable compression fractures.

Decompression and stabilization The aim of surgery is to reduce the fracture, hold the reduction and decompress the neural elements. The surgical approach can be either anterior or posterior.

The anterior approach is suitable for fractures with persistent anterior neural compression or to augment posterior fixation. The spine is exposed through a transthoracic, transdiaphragmatic or transperitoneal approach depending on the level of the fracture. The vertebral body is removed for canal decompression and reconstructed with bone graft (rib, fibula or iliac crest) or a cage.

The posterior approach is most commonly used for most fracture patterns. Some fractures can be reduced indirectly from posterior by distraction with instrumentation and pedicle screw systems are ideal for deformity correction. Bone graft is required so that a biological fusion can supplement the implants.

CERVICAL SPINE INJURIES

The patient will usually give a history of a fall from a height, a diving accident or a vehicle accident in which the neck is forcibly moved. In an unconscious patient, an unstable cervical spine injury needs to be excluded. An abnormal position of the neck is suggestive (Figure 28.10), and careful palpation may elicit tenderness. Movement is best postponed until the neck has been X-rayed. Pain or paraesthesia in the limbs is significant, and the patient should be examined for evidence of spinal cord or nerve root damage.

IMAGING

Plain X-rays must be of high quality and should be inspected methodically (Figures 28.11 and 28.12).

- In the anteroposterior view the lateral outlines should be intact, and the spinous processes and tracheal shadow should be in the midline. An open-mouth view is necessary to show C1 and C2 (for odontoid and lateral mass fractures).
- In the lateral view the smooth lordotic curve should be followed, tracing four parallel lines formed by the front of the vertebral bodies, the back of the bodies, the posterior borders of the lateral masses and the bases of the spinous processes; any irregularity suggests a fracture or displacement. Forward

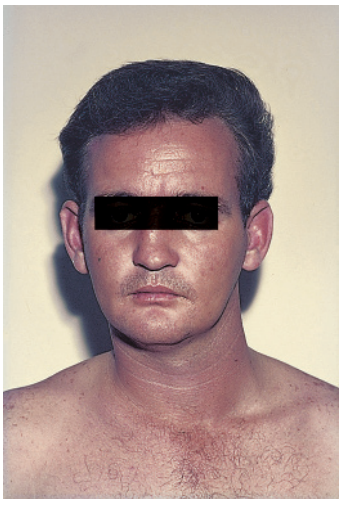
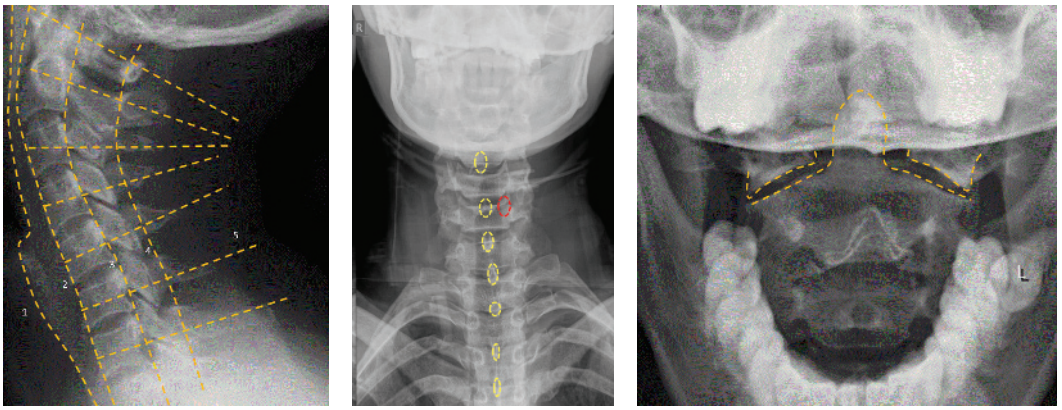
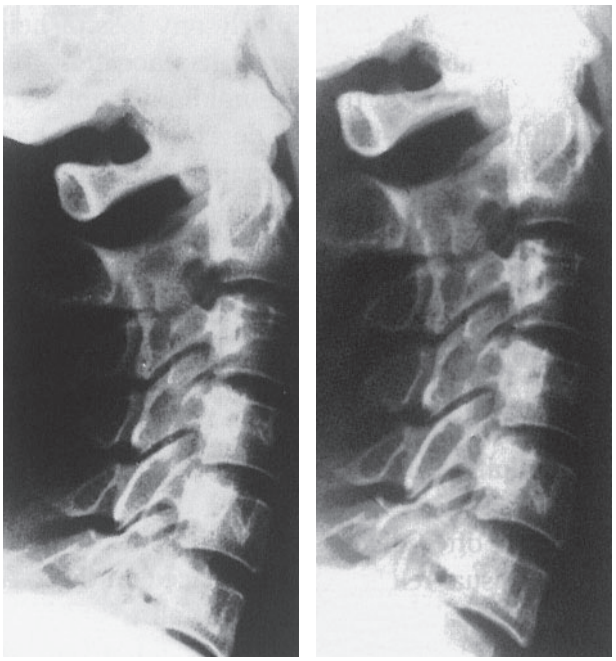


Figure 28.10 Cervical spine injury Look at the position of this patient's neck. He complained of pain and stiffness after a fall. X-ray examination revealed an odontoid fracture.



(a) (b) (c)

Figure 28.11 Cervical spine – normal X-ray findings (a) In the lateral projection, the radiological lines are: 1 soft-tissue line; 2 anterior vertebral body line; 3 posterior vertebral body line; 4 spinolaminar line. 5 The lines of convergence are drawn down the spinous processes and should converge posteriorly; these help identify posterior element disruption. (b) AP view confirms alignment of the spinous processes. (c) Open-mouth view should show a centrally placed dens with congruous and symmetrical C1/C2 lateral mass joints.



(a) (b)

Figure 28.12 Cervical spine injuries – X-ray diagnosis (a) Following an RTC this patient had a painful neck and consulted her doctor three times; on each occasion she was told 'The X-rays are normal.' But count the vertebrae! There are only six in this film. (b) When a shoulder 'pull-down view' was obtained to show the entire cervical spine, a dislocation of C6 on C7 could be seen at the very bottom of the film.

shift of the vertebral body by 25% suggests a unilateral facet dislocation and by 50% a bilateral facet dislocation.

- The lateral view must include all seven cervical vertebrae and the upper half of T1, otherwise an injury at the cervicothoracic junction could be missed. If the cervicothoracic junction cannot be seen, the lateral view should be repeated while the patient's shoulders are pulled down. If this fails, then a 'swimmer's view' is obtained. If this, too, fails, a CT scan is required.
- The distance between the odontoid peg and the posterior aspect of the anterior arch of the atlas should be no more than 3 mm in adults and 5 mm in children.
- Compare the shape of each vertebral body with that of the others; note particularly any loss of height, fragmentation or backward displacement of the posterior border of the vertebral body.
- Examine the soft-tissue shadows. The retropharyngeal space may contain a haematoma; the prevertebral soft-tissue shadow should be less than 5 mm in thickness above the level of the trachea and less than one vertebral body's width in thickness below. The interspinous space may be widened after ligament rupture.

Diagnostic pitfalls in children

Children are often distressed and difficult to examine; more than usual reliance may be placed on the X-rays. It is well to recall some common pitfalls.

- *An increased atlantodental interval* (up to 5 mm) may be quite normal; this is because the skeleton is incompletely ossified and the ligaments relatively lax during childhood. There may also be apparent subluxation of C2 on C3 (*pseudosubluxation*).
- *An increased retropharyngeal space* can be caused by crying. Growth plates and synchondroses can be mistaken for *fractures*. The normal synchondrosis at the base of the dens has usually fused by the age of 6 years, but it can be mistaken for an undisplaced fracture; the spinous process growth plates

also resemble fractures; and the growth plate at the tip of the odontoid can be taken for a fracture in older children.

- *SCIWORA* is an acronym for spinal cord injury without obvious radiographic abnormality. Normal radiographs in children do not exclude the possibility of spinal cord injury.

Upper cervical spine

Occipital condyle fracture

This is usually a high-energy fracture and associated skull or cervical spine injuries must be sought. The diagnosis is likely to be missed on plain X-ray examination and CT is essential. Impacted and undisplaced fractures can be treated by brace immobilization for 8–12 weeks. Displaced fractures are best managed by using a halo vest or by operative fixation.

Occipitocervical dislocation

This high-energy injury is almost always associated with other serious bone and/or soft-tissue injuries, including arterial and pharyngeal disruption, and the outcome is often fatal. The diagnosis can sometimes be made on the lateral cervical radiograph: the tip of the odontoid should be no more than 5 mm in vertical alignment and 1 mm in horizontal alignment from the basion (anterior rim of the foramen magnum). Greater distances are allowable in children. CT scans are more reliable where the distance between the dens and the clivus should be less than 12 mm. The injury is likely to be unstable and requires immediate reduction (without traction!) and stabilization with a halo vest, pending surgical treatment. After appropriate attention to the more serious soft-tissue injuries and general resuscitation, the dislocation should be surgically stabilized by means of an occipitocervical instrumented fusion posteriorly if severe (Figure 28.13), or a 6–8-week period in the halo vest.

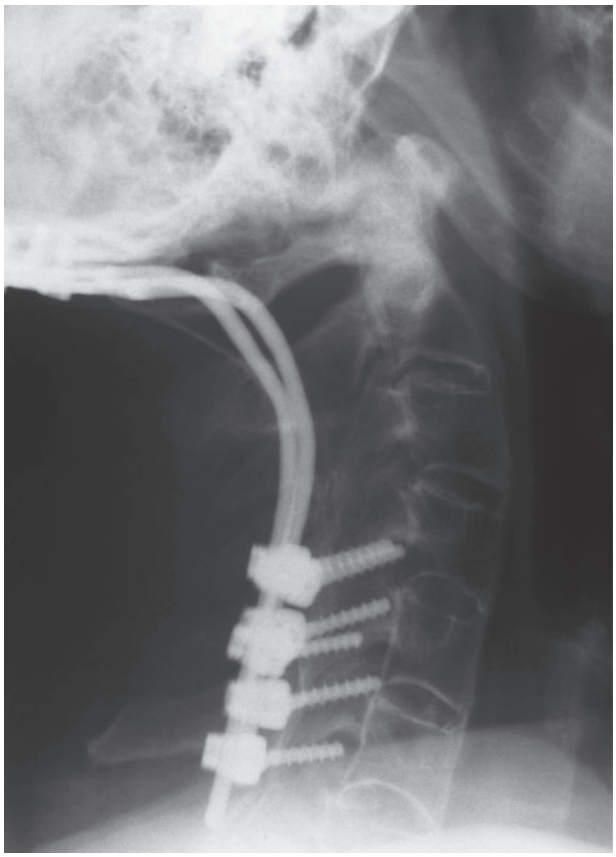


Figure 28.13 Occipitocervical fusion X-ray showing one of the devices used for internal fixation in occipitocervical fusion operations.

C1 ring fracture

An axial load may result in a burst fracture of the ring of the atlas (Jefferson's fracture – Figure 28.14). There is seldom a neurological injury. The fracture is identified on the open-mouth X-ray view with displacement of C1 relative to C2, i.e. overhand of the lateral processes. A fracture may be visible on the lateral view. A CT scan will confirm and define the fracture. If undisplaced, the injury is stable and the patient is managed with a semi-rigid collar.

If the C1 lateral masses have a combined overhang on C2 of more than 7 mm on the open-mouth view, the transverse ligament is likely to be ruptured (rule of Spence), making the injury unstable. It will require surgical intervention. A hyperextension injury can fracture either the anterior or posterior arch of the atlas. These injuries are usually stable and are managed with a semi-rigid collar until union occurs. Fractures of the atlas are associated with injury elsewhere in the cervical spine in up to 50% of cases.

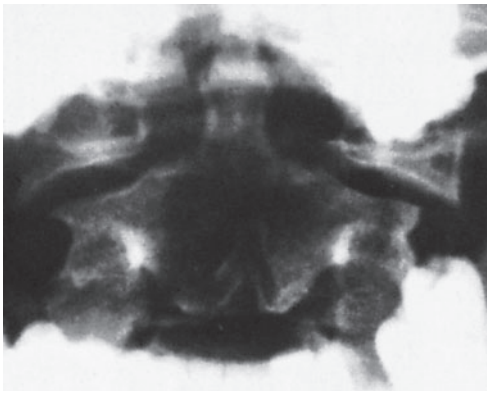


Figure 28.14 Fracture of C1 ring Jefferson's fracture – bursting apart of the lateral masses of C1.

C2 traumatic spondylolisthesis

This is frequently incorrectly referred to as a 'hangman's fracture' (Figure 28.15). Judicial hanging caused the fracture and death by spinal cord injury due to a distractive force. This is not frequently seen today but rather a hyperextension mechanism from a force to the forehead and the 'Forsyth mechanism' causing a resultant anterior translation at C2. This results in a pars fracture, sometimes extending into the posterior body wall and a C2/3 disc disruption. Neurological injury is unusual as the pars fractures expand the canal. Stability is determined by the C2/3 disc status and facet integrity.

Undisplaced fractures which are shown to be stable on supervised flexion–extension views (less than 3.5 mm of C2/3 subluxation) can be treated in a semi-rigid orthosis until united (usually 6–12 weeks). Fractures with more than 3.5 mm displacement but no kyphotic angulation may need reduction with inline traction followed by a semi-rigid collar and infrequently surgery dependent on patient compliance. Rarely there is associated C2/3 facet dislocation which will require open reduction and stabilization.

C2 odontoid process fracture

Odontoid fractures are uncommon. They usually occur as flexion injuries in young adults after high-velocity accidents. They also occur in elderly, osteoporotic people as a result of low-energy trauma in which the neck is forced into hyperextension, such as a fall onto the face or forehead. There is seldom neurological injury due to the large canal size at this level.

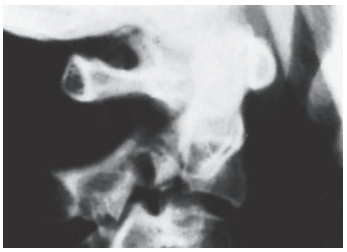


Figure 28.15 Fracture of C2 – traumatic spondylolisthesis
Fracture of the pars interarticularis of C2.

CLASSIFICATION

Odontoid fractures have been classified by Anderson and D'Alonzo (Figure 28.16):

- *Type I* – an avulsion fracture of the tip of the odontoid process due to traction by the alar ligaments. The fracture is stable (above the transverse ligament) and unites without difficulty.
- *Type II* – a fracture at the junction of the odontoid process and the body of the axis. This is the most common (and potentially the most dangerous) type. The fracture is unstable and prone to non-union.
- *Type III* – a fracture through the body of the axis. The fracture is stable and almost always unites with immobilization.

CLINICAL FEATURES

The complaints may be non-specific and easily missed. The diagnosis is confirmed by high-quality X-ray examination or CT.

IMAGING

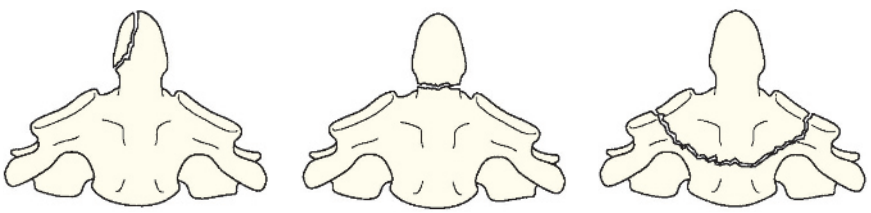
The lateral *X-ray* will identify the obviously displaced fracture (Figure 28.17). A normal dens tilts posteriorly and, if it appears straight or anteriorly tilted, fracture should be considered. The open-mouth view will also demonstrate the fracture. *CT* is the mainstay of diagnosis and classification.

TREATMENT

Type I fractures Isolated fractures of the odontoid tip are stable. They need no more than immobilization in a rigid collar until discomfort subsides.

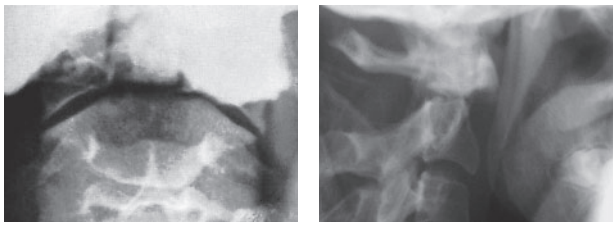
Type II fractures These are often unstable and prone to non-union, especially if displaced more than 4 mm, tilted more than 11 degrees, in older patients and in smokers.

In the young patient with an undisplaced fracture, union is anticipated. Management can be non-operative with a period of traction followed by collar or halo vest. In the very elderly, a collar is appropriate due to the poor risk-to-benefit ratio of surgery. In the patient where non-union is anticipated, surgery can be carried out to insert an anterior screw from the C2 body into the dens using a lag technique to compress the fracture site.



(a) (b) (c)

Figure 28.16 Odontoid fractures – classification (a) *Type I* – fracture through the tip of the odontoid process. (b) *Type II* – fracture at the junction of the odontoid process and the body. (c) *Type III* – fracture through the body of the axis. Neurological symptoms occur in a significant number of cases.



(a) (b)

Figure 28.17 Fractured odontoid process
(a) Anteroposterior 'open-mouth' X-ray showing a type II odontoid fracture. (b) Lateral X-ray of the same patient.

If the fracture pattern is not conducive (e.g. reverse oblique, comminuted) or the fracture is irreducible, a posterior C1–2 instrumented fusion is indicated (Figure 28.18).

Type III fractures Undisplaced fractures are treated in a halo vest for 8–12 weeks. If displaced, attempts should be made at reducing the fracture by halo traction, which will allow positioning of the neck in either flexion or extension, depending on whether the displacement is forward or backward; the neck is then immobilized in a halo vest for 8–12 weeks. For elderly patients with poor bone, a collar may suffice, though this carries a higher risk of non-union.

Lower cervical spine

Fractures of the cervical spine from C3 to C7 (subaxial cervical spine) tend to produce characteristic fracture patterns, depending on the mechanism of injury. Many classification systems were developed including the Ferguson and Allen classification in the 1980s. This gained popularity, and subaxial cervical spine injuries were grouped into compressive flexion, vertical compression, distractive flexion, compressive extension, distractive extension and lateral flexion injuries. Each of these groups was allocated a numerical value based in increasing severity. The patterns range from stable injuries to unstable, depending on the degree of displacement and the extent of the posterior ligamentous injury.

More recently the Subaxial Cervical Spine Injury Classification (SLIC) scoring system (Vaccaro) was developed to guide management of cervical injuries.

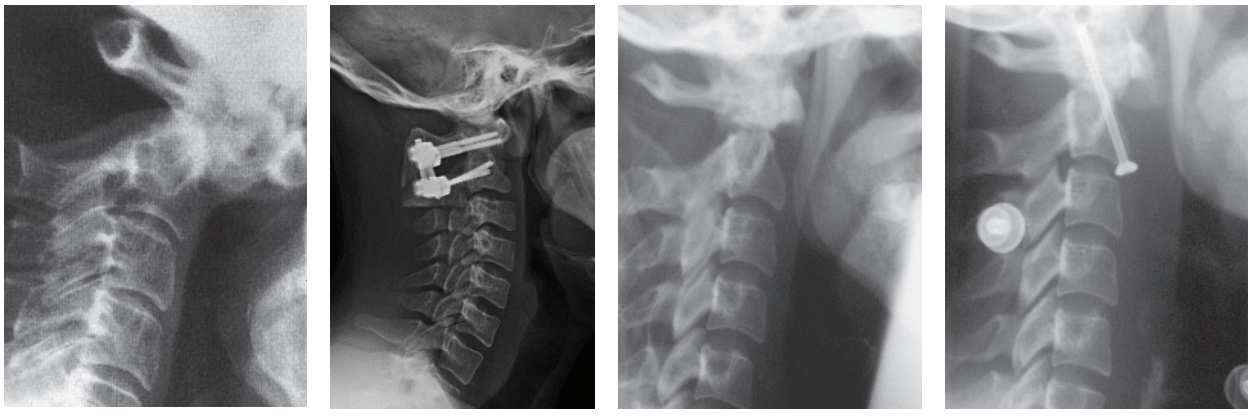
The SLIC scoring system (Table 28.3) incorporates three main characteristics:

- *Morphology of the injury* – This is based on the available imaging, and consists of compression, distraction and translation/rotation of the vertebral bodies.
- *Discoligamentous complex (DLC)* – This considers the integrity of the intervertebral disc, anterior and posterior ligamentous structures. They are defined as disrupted, intact and indeterminate. The DLC is considered directly proportional to stability.
- *Neurological status of the patient* – This is linked with the severity of the injury.

The SLIC classification is gaining popularity, but the mechanistic descriptive classifications of subaxial cervical spine injuries are still prevalent and are discussed here.

Flexion injuries

These include distractive flexion and compressive flexion injuries. Depending on the instantaneous axis of rotation during the injury, there is a spectrum from posterior ligament disruption ranging to anterior column compression.



(a) (b) (c) (d)

Figure 28.18 Fractured odontoid – treatment (a) A severely displaced type II odontoid fracture. (b) The Harms technique of C1/C2 fixation with C1 lateral mass screws and C2 pedicle screws. (c) An undisplaced type II fracture, which was suitable for (d) anterior screw fixation.

Table 28.3 The SLIC scale

Characteristic	Points
Morphology	
No abnormality	0
Compression	1
Burst	+1 = 2
Distraction	3
Disrupted	4
Discoligamentous complex (DLC)	
Intact	0
Indeterminate	1
Disrupted	2
Neurological status	
Intact	0
Root injury	1
Complete cord injury	2
Incomplete cord injury	3
Continuous cord compression in setting of neurological deficit	+1

Wedge compression fracture (compressive flexion mechanism)

This results in a wedge compression fracture of the vertebral body (Figure 28.19). With intact posterior elements the injury is stable. Clinical examination of the posterior elements and flexion X-rays will confirm stability.

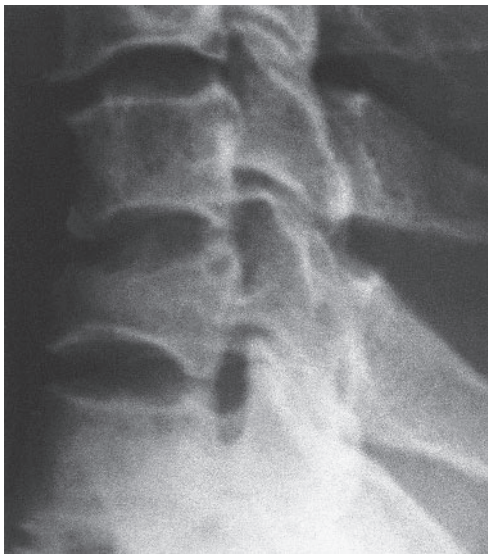


Figure 28.19 Cervical compression fracture
A wedge compression fracture of a single cervical vertebral body. This is a stable injury because the posterior elements are intact. Compare and contrast with Figure 28.21.

A note of warning: The X-ray should be carefully examined to exclude posterior displacement of the vertebral body fragment, i.e. features of a burst fracture (see below) which is potentially unstable. If there is the least doubt, an axial CT or MRI should be obtained. If there is clinical and radiological confirmation of stability, a semi-rigid collar is worn for 6–12 weeks.

Burst and 'teardrop' fractures (compressive flexion mechanism)

These severe injuries are due to axial compression of the cervical spine with flexion, usually in diving or athletic accidents (Figure 28.20). If the vertebral body is crushed in neutral position of the neck, the result is a '*burst fracture*'. With an increasing flexion moment, the anteroinferior fragment of the vertebral body is sheared off, producing the eponymous '*teardrop*' on the lateral X-ray.

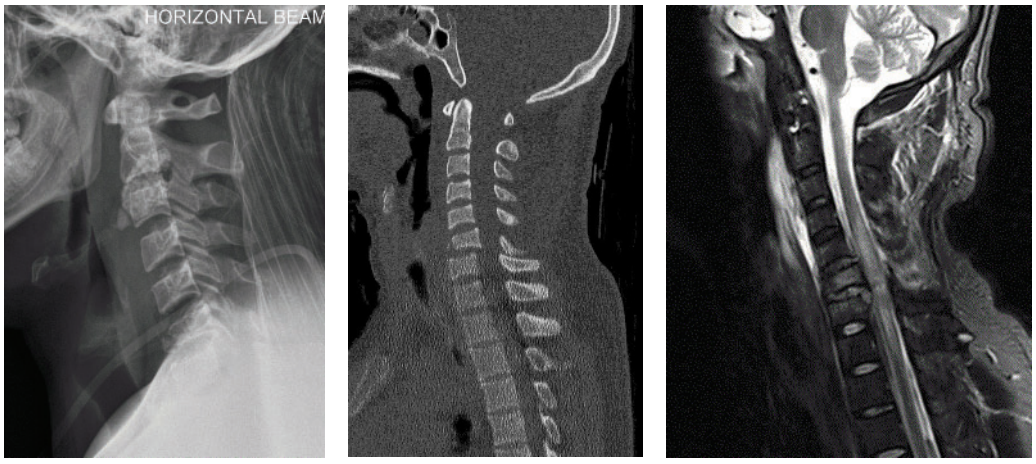
In both types of fracture there is a risk of posterior displacement of the vertebral body fragment and spinal cord injury.

Plain *X-rays* show either a crushed vertebral body (burst fracture) or a flexion deformity with a triangular fragment separated from the anteroinferior edge of the fractured vertebra (the innocent-looking 'teardrop'). These are highly unstable injuries due to the antero column disruption and need immobilization. *CT scan* will demonstrate the bony injuries and canal compression while MRI will confirm the disc injury with the teardrop fracture and posterior ligamentous complex disruption.

TREATMENT

Conservative treatment of these injuries is a poor option due to the high degree of instability and likelihood of subsequent kyphosis and spinal cord compression. These patients are often quadriplegic and prolonged recumbence in traction is not cost-effective and carries significant morbidity.

Surgical management is the treatment of choice. The primary goal is stabilization and neurological decompression as a secondary goal since the cord injury is often already established. The patient is initially stabilized in traction, optimized medically with respect to spinal cord and concomitant injuries and appropriately imaged (CT and MRI). Surgery should be on the next available theatre list where anterior corpectomy, strut bone grafting (iliac crest) and plate fixation is usually adequate. If there is severe posterior disruption and the anterior reconstruction is tenuous, posterior stabilization may be added.



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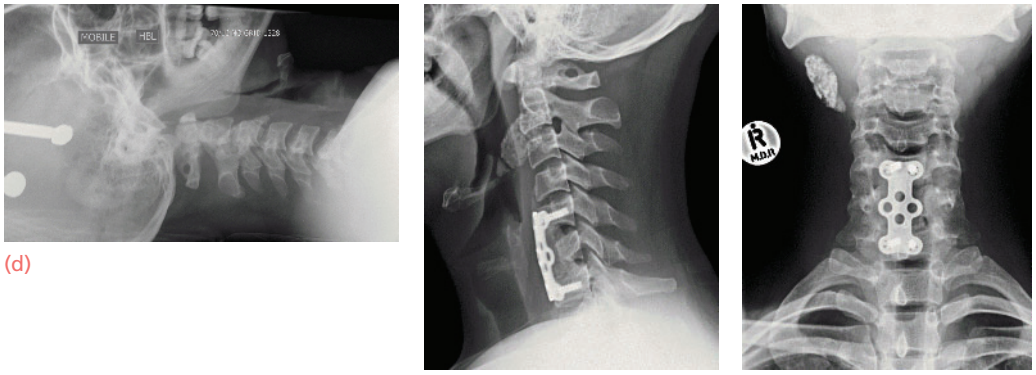


(d) (e) (f)

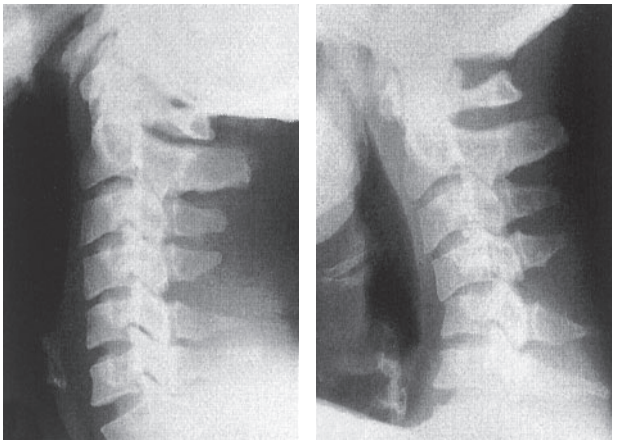
Figure 28.20 Teardrop fracture (a) An innocuous-looking fracture at C6 with a large pre-vertebral swelling heralds the 'teardrop' fracture; note the mild retrolisthesis; (b) CT details the interspinous widening and retrolisthesis of C6; (c) MRI reveals the posterior interspinous ligament disruption and spinal cord injury. (d) Cervical traction helps maintain alignment until surgery. (e,f) Surgical stabilization with corpectomy and anterior cervical plating.

Posterior ligament injury, dislocations and fracture-dislocations (distractive flexion mechanism)

These range from simple posterior ligament injury to subsequent facet subluxation, dislocation and vertebral body dislocation. Structures fail sequentially from posterior to anterior with progressive instability and increasing risk of neurological injury.

Posterior ligament injury Posterior spinous tenderness or gap should alert the clinician to this injury. *X-ray* may reveal only an increased interspinous gap (Figure 28.21). Flexion views should confirm stability unless prevented by pain, in which case assume there is a potential injury and manage in a semi-rigid cervical collar with repeat flexion views after 10 days. More than 11 degrees of angulation or more than 3.5 mm anterior translation on the flexion view is considered unstable and may need surgical stabilization if conservative management fails.

Bilateral facet joint dislocations (jumped facets) These are severe flexion injuries with complete disruption of the posterior ligamentous complex and spinal instability, often with cord damage. Occasionally facet fractures occur. Lateral *X-rays* show disruption of the radiological lines, anterior vertebral translation more than half a body width and spinous processes aligned on AP views (no rotation).



(a) (b)

Figure 28.21 Cervical spine – posterior ligament injury (a) The film taken in extension shows no displacement of the vertebral bodies, but there is an unduly large gap between the spinous processes of C4 and 5. (b) With the neck slightly flexed, the subluxation is obvious. NB: Flexion–extension views are potentially dangerous and should be used only in specific situations under direct supervision of an experienced surgeon.

Unilateral facet dislocations (UFDs) These are distractive flexion–rotation injuries with a single facet joint dislocation and cord injury is less common. On lateral *X-ray* the vertebra displaces less than one-half a

body width and rotation of the facets is seen cephalad to the injury level resulting in the typical 'bow-tie' sign. On an AP X-ray the alignment of the spinous processes is distorted. Left untreated, these patients may develop neck pain and nerve root symptoms and they remain potentially unstable.

Management of unifacet injuries and bilateral dislocations is the same. The dislocation must be reduced as a matter of urgency due to ongoing cord compression. Neurological outcomes are improved with earlier reduction, especially in low-energy injuries such as sporting accidents. Depending on the clinical skills and resources, this can be done with closed cervical reduction or open reduction in theatre. Closed cervical reduction is the most rapid method and requires only X-rays, an awake and cooperative patient and traction equipment. The risk of permanent neurological worsening with closed reduction is less than 1% and the risk of a transient worsening of the neurological status is 2–4%.

Once reduced, the injured level can then be surgically stabilized on the next available theatre list. There is no role for conservative management. These are discoligamentous injuries; occasionally the disc may ankylose, but this is the exception and there remains the risk of chronic neck pain and redislocation.

TREATMENT

See Figure 28.22.

Pre-reduction imaging *X-rays* will diagnose the injury and are mandatory. *CT* is useful to exclude a facet fracture not appreciable on plain X-rays which may prevent closed reduction.

The *MRI scan* will demonstrate disc, cord and ligament injuries as well as the facet dislocation. There is no correlation between disc prolapse on pre- and post-reduction MRI scans and clinical status. In the setting of a low-energy dislocation and severe or deteriorating neurological level, the emphasis should be on urgent closed reduction and MRI is not a prerequisite.

Closed cervical reduction Skull traction is used, starting with 5 kg and increased step-wise by similar amounts up to about 30 kg; intravenous muscle relaxants and a bolster beneath the shoulders may help. The patient needs to be awake and cooperative throughout, and neurological examination should be repeated after each incremental step along with lateral X-rays. If neurological symptoms or signs worsen, the reduction is abandoned. The reduction process is helped by 20-degree flexion of the neck (raising the swan-neck pulley). Once the facets are perched or reduced, the neck is extended and the weights reduced to 5 kg

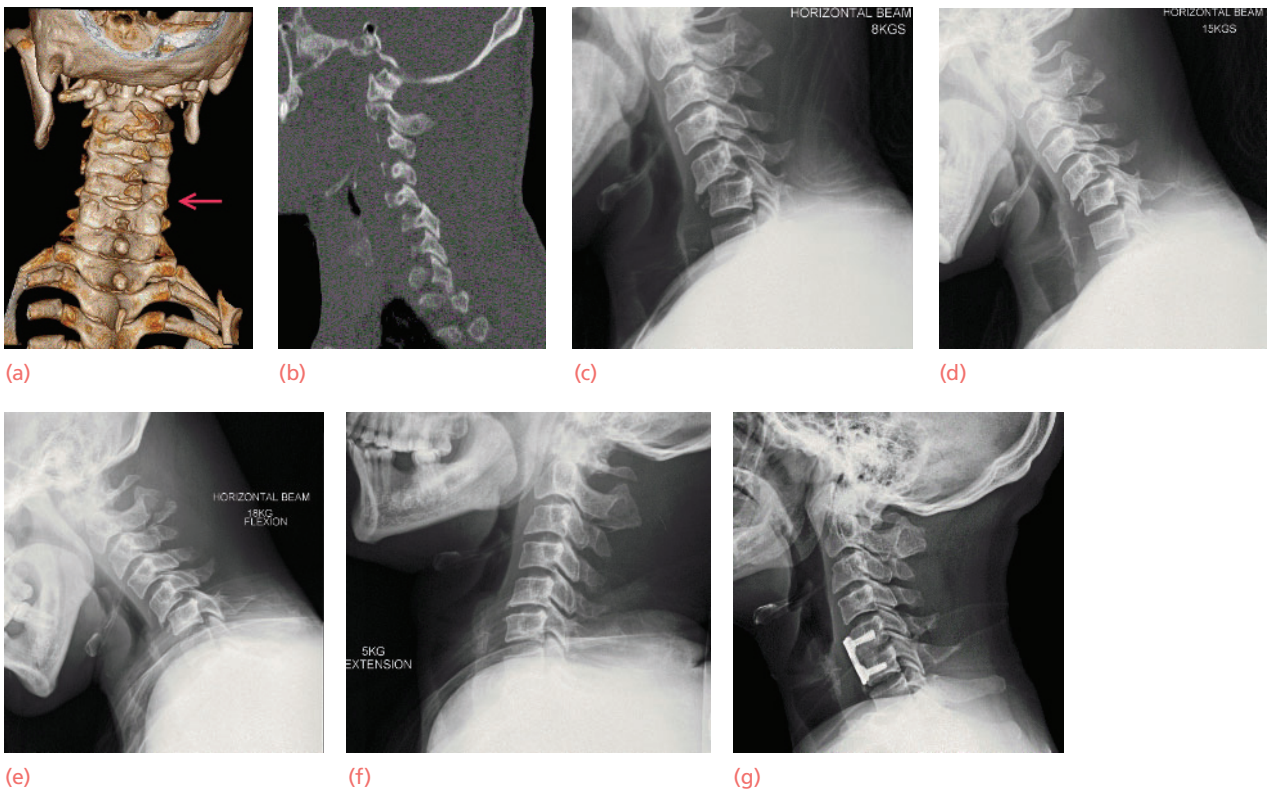


Figure 28.22 Cervical unilateral facet dislocation (UFD) stages in reduction (a) 3D CT scan showing right C5/C6 UFD; (b) CT sagittal view confirms single facet dislocation without fracture; (c) initial traction at 8 kg; (d) increasing weight under X-ray control; (e) flexion of 20–30 degrees aids reduction when the facet is perched; (f) after reduction the neck is extended and weight reduced to 5 kg for maintenance; (g) definitive surgical stabilization with an anterior plate.

(a double mattress allows neck extension). After reduction, an MRI can be requested to plan for surgery.

Surgical open reduction Practically this takes longer to accomplish since a CT or MRI is normally required, theatre takes time to set up and preparation takes longer than closed reduction. Surgical open reduction, however, is required in cases of failure of closed reduction (20% of cases) or where there are contraindications to closed reduction.

Surgical stabilization The options are anterior, posterior or combined approaches to reduce and stabilize the dislocation. With anterior approaches, the disc is removed from the canal and the facets indirectly reduced by manipulation of the vertebrae. An interbody fusion is done with bone graft and a plate. With posterior approaches, the facets are directly reduced and fixed with lateral mass screws, which have largely replaced the less robust interspinous wiring techniques (Figure 28.23). Potential anterior disc compression is not addressed and this approach is indicated mainly for patients with high-energy complete spinal cord injuries in whom the primary goal is vertebral column stability.

Combined anterior-posterior approaches are occasionally employed in patients with neurologically incomplete lesions or those with normal neurology when there is anterior cord compression but anterior open reduction fails. In this case, anterior decompression takes priority, then the patient is repositioned and posterior direct reduction and fixation is performed.

Hyperextension injury (distraction or compression combined with extension)

Hyperextension strains of soft-tissue structures are common and may be caused by comparatively mild acceleration forces. Bone and joint disruptions, however, are rare. The more severe injuries are suggested by the history and the presence of facial bruising or lacerations. With compressive extension, the posterior bone elements are compressed and may fracture. When the disc and vertebral body remain intact, these injuries tend to be stable.

With distractive extension, the anterior structures fail in tension, with tearing of the anterior longitudinal ligament (Figure 28.24) or an avulsion fracture of the anterosuperior or anteroinferior edge of the

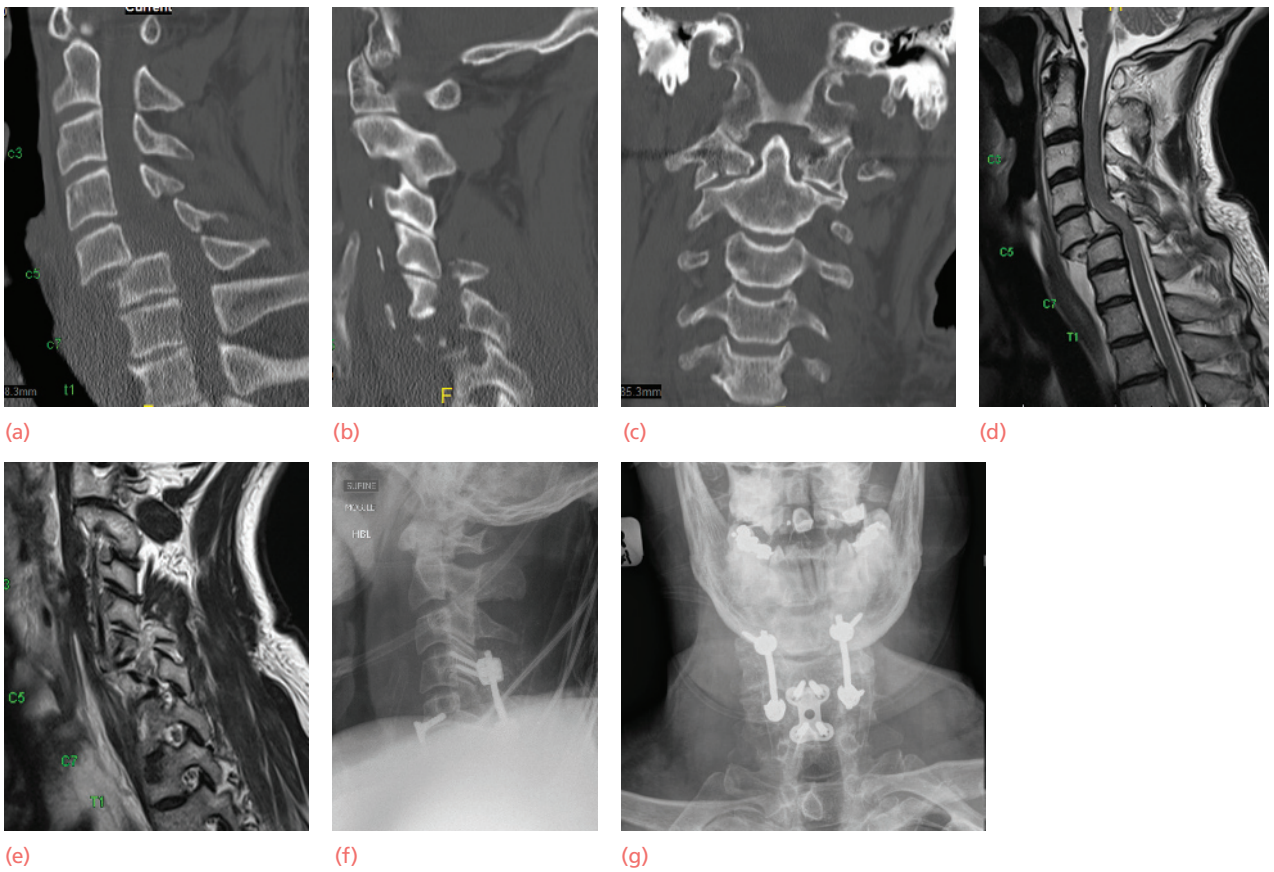
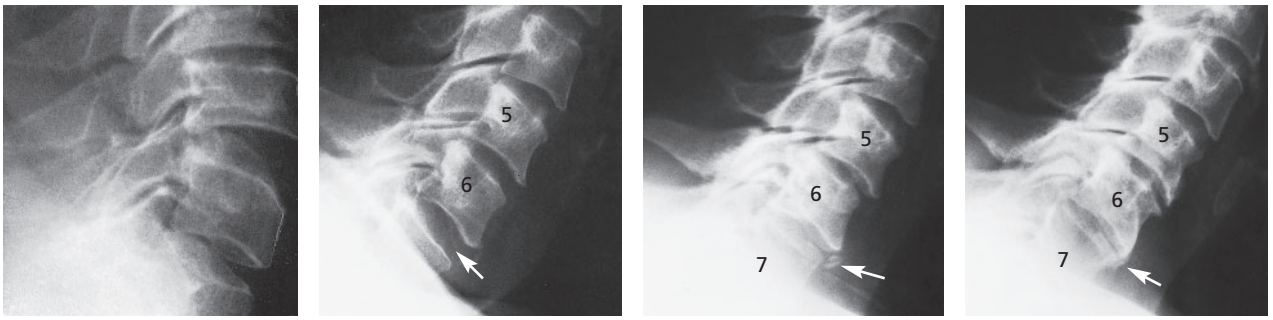


Figure 28.23 Cervical fracture-dislocation (a,b) CT scan in a cyclist with C5/C6 vertebral body listhesis and facet fracture-dislocation. (c) C1 ring fracture in the same patient shows the value of CT in detecting non-contiguous injuries which may be missed on X-rays. (d,e) MRI shows compression and cord injury and is useful to assess the facets on the parasagittal views. (f,g) Due to the facet fractures, anterior-only stabilization was not adequate and posterior fixation was added.



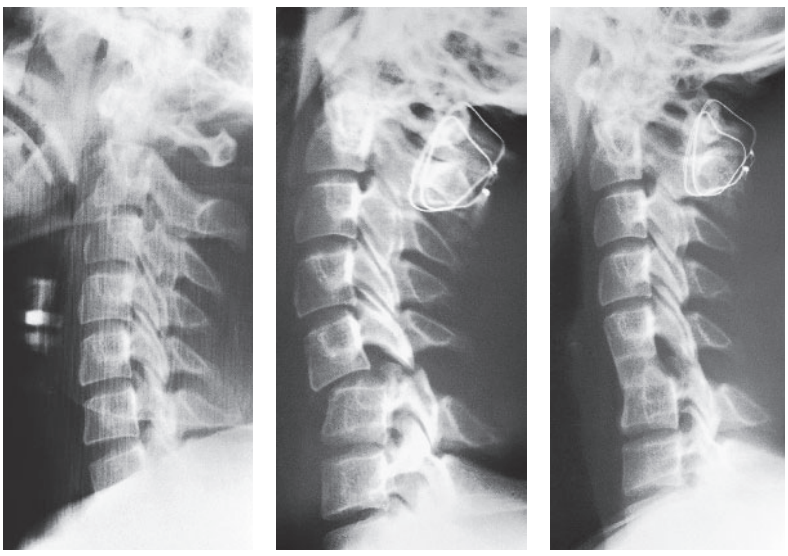
(a) (b) (c) (d)

Figure 28.24 Hyperextension injuries (a) The anterior longitudinal ligament has been torn; in the neutral position the gap will close and reduction will be stable, but a collar or brace will be needed until the soft tissues are healed. (b) X-ray in this case showed a barely visible flake of bone anteriorly at the C6/7 disc space. (c) A month later the traction fracture at C6/7 was more obvious, as was the disc lesion at C5/6. (d) A year later C6/7 has fused anteriorly; the patient still has neck pain due to the C5/6 disc degeneration.

vertebral body and opening up of the anterior part of the disc space. These are more common in cervical spondylosis where there is a relatively rigid cervical column. In these patients, the cord can be pinched between the bony spurs or disc and the posterior ligamentum flavum, causing an acute central cord syndrome (quadriplegia, sacral sparing and more upper-limb than lower-limb deficit, a flaccid upper-limb paralysis and spastic lower-limb paralysis). In stable injuries these may be treated conservatively in a semi-rigid collar for 6–8 weeks. Unstable injuries or those with cord compression are treated by surgical stabilization.

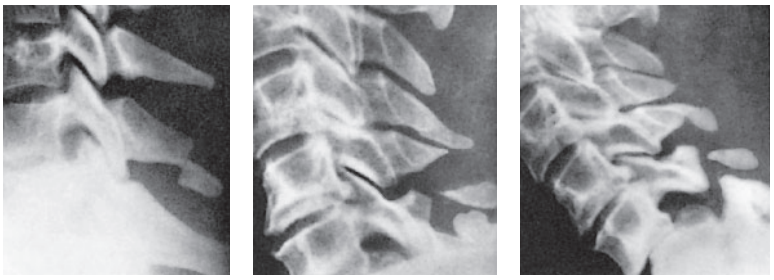
Double injuries

With high-energy trauma the cervical spine may be injured at more than one level. Discovery of the most obvious lesion is no reason to drop one's guard. Two salutary examples are shown in Figures 28.25 and 28.26.



(a) (b) (c)

Figure 28.25 Double cervical injuries (a) This patient with a neck injury was suspected of having an odontoid fracture. This was confirmed and a posterior stabilization was performed. Only when the brace was removed and he started flexing his neck did the X-ray show an obvious subluxation lower down (b). This was treated by anterior fusion (c).



(a) (b) (c)

Figure 28.26 Avulsions (a) The clay-shoveller's fracture. Jerking the neck backwards has resulted in avulsion of one of the spinous processes – a benign injury. (b) This patient might be thought to have a similar fracture, but a subsequent flexion film (c) shows the serious nature of the injury – a severe fracture-dislocation.

Avulsion injury of the spinous process

Fracture of the C7 spinous process may occur with severe voluntary contraction of the muscles at the back of the neck; it is known as the *clay-shoveller's fracture*. The injury is painful but harmless. No treatment is required; as soon as symptoms permit, neck exercises are encouraged.

Cervical disc herniation

Acute post-traumatic disc herniation may cause severe pain radiating to one or both upper limbs, and neurological symptoms and signs ranging from mild paraesthesia to weakness, loss of a reflex and blunted sensation. Rarely, a patient presents with full-blown paresis. The diagnosis is confirmed by MRI. Sudden paresis will need immediate surgical decompression. With lesser symptoms and signs, conservative treatment is recommended. Failure to improve will require anterior discectomy and interbody fusion.

Neurapraxia of the cervical cord

Accidents causing sudden, severe axial loading with the neck in hyperflexion or hyperextension are occasionally followed by transient pain, paraesthesia and weakness in the arms or legs, all in the absence of any X-ray or MRI abnormality. Symptoms may last for as little as a few minutes or as long as 2–3 days. The condition has been called neurapraxia of the cervical cord and is ascribed to pinching of the cord by the bony edges of the mobile spinal canal and/or local compression by infolding of the posterior longitudinal ligament or the ligamentum flavum. Congenital narrowing of the spinal canal may be a predisposing factor.

Treatment consists of reassurance (after full neurological investigation) and graded exercises to improve strength in the neck muscles.

Whiplash injury (sprained neck/cervical acceleration–deceleration injury)

Soft-tissue sprains of the neck are so common after motor vehicle accidents that they now constitute a veritable epidemic. There is usually a history of a low-velocity rear-end collision in which the occupant's body is forced against the car seat while his or her head flips backwards and then recoils in flexion. This mechanism has generated the imaginative term '*whiplash injury*', which has served effectively to enhance public apprehension at its occurrence. However, similar symptoms are often reported with flexion and rotation injuries. Women are affected more often than men, perhaps because their neck muscles are more gracile. There is disagreement about the exact pathology but it has been suggested that the anterior longitudinal ligament of the spine and the capsular fibres of the facet joints are strained and in some cases the intervertebral discs may be damaged in some unspecified manner. There is no correlation between the amount of damage to the vehicle and the severity of complaints from occupants.

Clinical features

Often the victim is unaware of any abnormality immediately after the collision. Pain and stiffness of the neck usually appear within the next 12–48 hours, or occasionally only several days later. Pain sometimes radiates to the shoulders or interscapular area and may be accompanied by other, more ill-defined, symptoms such as headache, dizziness, blurring of vision, paraesthesia in the arms, temporomandibular discomfort and tinnitus. Neck muscles are tender and movements often restricted; the occasional patient may present with a 'skew neck'. Other physical signs – including neurological defects – are uncommon.

X-ray examination may show loss of cervical lordosis, a sign of muscle spasm; or this finding may be a normal variant for the age group. *MRI* is not indicated except when there are neurological signs. For purposes of comparison, the severity grading system proposed by the Quebec Task Force on whiplash-associated disorders is useful (Table 28.4).

Differential diagnosis

The diagnosis of sprained neck is reached largely by a process of exclusion, i.e. the inability to demonstrate any other credible explanation for the patient's symptoms. X-rays should be carefully scrutinized to avoid missing a vertebral fracture or a midcervical subluxation. The presence of neurological signs such as muscle weakness and wasting, a depressed reflex or definite loss of sensibility should suggest an acute disc lesion and is an indication for MRI. Seat-belt injuries often accompany neck sprains. They do not always cause

Table 28.4 Proposed grading of whiplash-associated injuries

Grade	Clinical pattern
0	No neck symptoms or signs
1	Neck pain, stiffness and tenderness No physical signs
2	Neck symptoms and musculoskeletal signs
3	Neck symptoms and neurological signs
4	Neck symptoms and fracture or dislocation

bruising of the chest, but they can produce pressure or traction injuries of the suprascapular nerve or the brachial plexus, either of which may cause symptoms resembling those of a whiplash injury. The examining doctor should be familiar with the clinical features of these conditions.

Treatment

Collars are more likely to hinder than help recovery. Simple pain-relieving measures, including analgesic medication, may be needed during the first few weeks. However, the emphasis should be on graded exercises, beginning with isometric muscle contractions and postural adjustments, and then going on gradually to active movements and lastly movements against resistance. The range of movement in each direction is slowly increased without subjecting the patient to unnecessary pain. Many patients find osteopathy and chiropractic treatment to be helpful.

Progress and outcome

The natural history of whiplash injury is reflected in the statistics appearing in the medical literature on this subject. Many people who are involved in road collisions do not seek medical attention at all; this is particularly the case in countries where medical and legal costs are not compensated. Some patients start improving within a few weeks and reports in the medical literature suggest that 50–60% eventually make a full recovery; in most cases symptoms diminish after about 3 months and go on improving over the next year or two; however, 2–5% continue to complain of symptoms and loss of functional capacity more or less indefinitely.

Negative prognostic indicators have been suggested to be increasing age, severity of symptoms at the outset, prolonged duration of symptoms and the presence of pre-existing intervertebral disc degeneration. Other factors that presage a poor outcome are a history of pre-accident psychological dysfunction, unduly frequent attendance with unrelated physical complaints, a record of unemployment and a general tendency to underachievement. It should be borne in mind that outcome studies are almost invariably based on a selected group of patients, namely those who attend for medical treatment after the accident, and little is known of the natural progress in the thousands of people who experience similar injuries and either do not develop symptoms or do not report them. A recent review of the literature suggests that initial pain and anxiety about the outcome of the injury are predicitive of outcome, but the evidence of any other associations is very weak.

Whiplash-associated disorder (chronic whiplash syndrome)

Those patients who, in the absence of any objective clinical or imaging signs, continue almost indefinitely to complain of pain, restriction of movement, loss of function, depression and inability to work constitute a sizeable problem in terms of medical resources, compensation claims, legal costs and – not least – personal suffering. As yet, no convincing evidence of a new pathological lesion has been adduced to account for this long-lasting disorder and it cannot be said with certainty how much of it is due to a physical abnormality and how much is an expression of a behavioural disorder.

THORACOLUMBAR INJURIES

Most injuries of the thoracolumbar spine occur in the transitional area – T11 to L2 – between the somewhat rigid upper and middle thoracic column and the flexible lumbar spine. The upper three-quarters of the thoracic segments are also protected to some extent by the rib cage, and fractures in this region tend to be mechanically stable. However, the spinal canal in that area is relatively narrow so cord damage is not uncommon and, when it does occur, it is usually complete. The spinal cord actually ends at L1 and below that level it is the lower nerve roots that are at risk.

Pathogenesis

Pathogenic mechanisms fall into three main groups:

- *low-energy insufficiency fractures* – arising from comparatively mild compressive stress in osteoporotic bone
- *minor fractures of the vertebral processes* – due to compressive, tensile or torsional strains
- *high-energy fractures or fracture-dislocations* – due to major injuries sustained in motor vehicle collisions, falls or diving from heights, sporting events, horse-riding and collapsed buildings.

It is mainly in the third group that one encounters neurological complications, but lesser fractures also sometimes cause nerve damage.

The common mechanisms of injury are *compression*, *rotation/translation* and *distraction*.

COMPRESSION

Compression injuries occur where there is failure of the vertebral body under axial loading. In its simplest form there is 'wedging' of the anterior body with localized kyphosis. With increasing forces posterior body wall collapse occurs with retropulsion of fragments into the canal and interpedicular widening.

Occasionally lateral compression occurs with a local scoliotic deformity. These fractures are usually stable unless there is posterior ligamentous disruption. With more than 50% loss of height there is usually disruption of the posterior tension band with instability. Under the AO classification, these fractures are described as type A injuries, when there is posterior ligament disruption they fall into the type B category.

The *posterior ligamentous complex (PLC)* comprises the supraspinous and interspinous ligaments, ligamentum flavum and facet joint capsules. This group of structures creates a posterior tension band which plays a role in limiting spinal flexion, rotation and translation. Their integrity is determined by clinical examination of the injured spine and confirmed on radiological imaging. A gap or tenderness on palpation is indicative of PLC disruption which, when combined with anterior column disruption, implies spinal instability.

ROTATION/TRANSLATION

This occurs with shear or torsional forces and injuries are mostly unstable. Commonly associated with neurological injury, they are identified by pedicle or spinous process malalignment on AP films. The facet joints, if not fractured, are dislocated. These are type C injuries under the AO system.

DISTRACTION

This is a '*Chance fracture*', as described by Chance in 1948, or 'seat-belt' injury, caused by spinal flexion with a distraction moment (as with a lap seat belt). Disruption of the posterior column structures may be bony, ligamentous or a combination of both. Up to 50% of Chance fractures have associated intra-abdominal injuries. These are unstable injuries due to the loss of the posterior tension band and usually require surgical fixation.

Examination

Patients complaining of back pain following an injury or showing signs of bruising and tenderness over the spine, as well as those suffering head or neck injuries, chest injuries, pelvic fractures or multiple injuries elsewhere, should undergo a careful examination of the spine for posterior tenderness or interspinous gap and a full neurological examination, including rectal examination to assess sphincter tone, voluntary contraction and perianal sensation.

Imaging

X-RAYS

The *anteroposterior X-ray* may show loss of height or splaying of the vertebral body with a crush fracture. Widening of the distance between the pedicles at one level, or an increased distance between two adjacent spinous processes, is associated with posterior column damage. The *lateral view* is examined for alignment, bone outline, structural integrity, disc space defects and soft-tissue shadow abnormalities. Always look carefully for evidence of fragment retropulsion towards the spinal canal identified by convexity of the normal concave posterior body wall. Plain X-rays, while showing the lower thoracic and lumbar spine quite clearly, are less revealing of the upper thoracic vertebrae because the scapulae and shoulders get in the way.

HELICAL CT SCANNING

This is now routine in most major trauma units. Not only are they more reliable than X-rays in showing bone injuries throughout the spine, and indispensable if axial views are necessary, but they also eliminate the delay, discomfort and neurological risk associated with multiple attempts at 'getting the right views' with plain X-rays.

MRI

MRI is indicated with neurological deficit and will illustrate injury to the disc, longitudinal ligaments and posterior ligament complex.

Treatment

Treatment depends on: (1) the type of anatomical disruption; (2) whether the injury is stable or unstable; (3) whether there is neurological involvement; and (4) the presence or absence of concomitant injuries. Details are discussed under each fracture type.

Minor injuries

Fractures of the transverse processes

The transverse processes can be avulsed with sudden muscular activity. Isolated injuries need no more than symptomatic treatment (Figure 28.27). More ominous than usual is a fracture of the transverse process of L5; this should alert one to the possibility of a vertical shear injury of the pelvis.

Fractures of the pars interarticularis

A stress fracture of the pars interarticularis should be suspected if a gymnast, athlete or weightlifter complains of the sudden onset of back pain during the course of strenuous activity. The injury is often ascribed to a disc prolapse, whereas in fact it may be a stress fracture of the pars interarticularis (*traumatic spondylolysis*). This is best seen in oblique *X-rays*, but a thin fracture line is easily missed; a week or two later, an *isotope bone scan* may show a 'hot' spot.

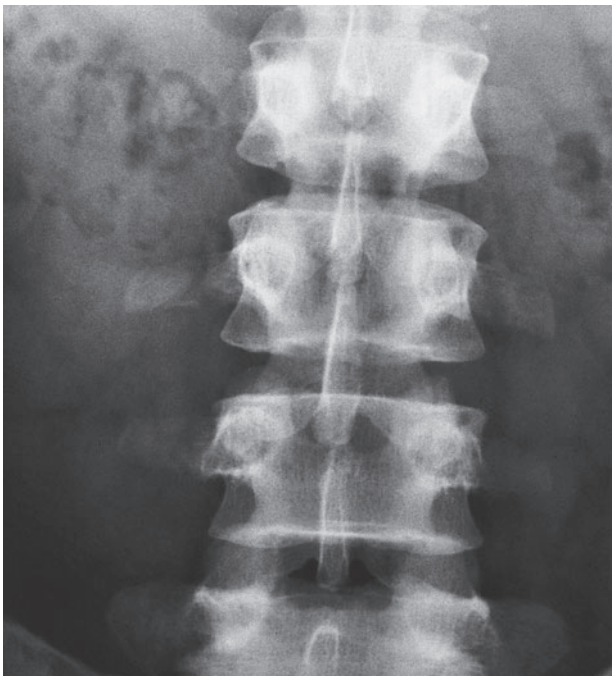


Figure 28.27 Thoracolumbar injuries – minor fractures
Fracture of the transverse processes on the right at L3 and L4.

CT scan will demonstrate the fracture and also differentiate between an acute and a chronic fracture. Often a chronic pars non-union is brought to light after investigation for acute back pain. Bilateral fractures occasionally lead to spondylolisthesis.

TREATMENT

An acute fracture usually heals spontaneously, provided the patient is prepared to forego sporting activities for several months. Unilateral pars fractures usually go on to union with conservative treatment. Pars fractures, if confirmed on CT to be chronic with sclerosis and 'cold' on isotope scan, have little chance of healing with rest alone (Figure 28.28).

MAJOR INJURIES

Flexion–compression injury

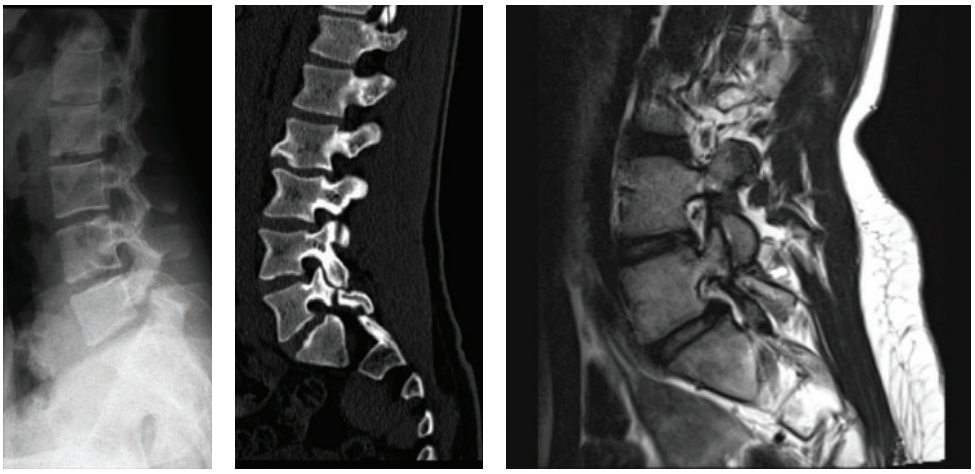
This is by far the most common vertebral fracture and is due to severe spinal flexion, although in osteoporotic individuals fracture may occur with minimal trauma. The posterior ligaments usually remain intact but, if anterior collapse is marked, they may be damaged by distraction. Examination includes careful palpation of the spinous processes for tenderness or a gap. CT shows that the posterior part of the vertebral body, pedicles, laminae and spinous processes are intact. Pain may be quite severe but the fracture is usually stable. Neurological injury is extremely rare.

TREATMENT

Patients with minimal wedging and a stable fracture pattern are kept in bed for a week or two until pain subsides and they are then mobilized; no support is needed. *Those with moderate wedging (loss of 20–40% of anterior vertebral height) with no instability* (disruption of the PLC) can be allowed up after a week, wearing a thoracolumbar brace or a body cast applied with the back in extension (Figure 28.29). At 3 months, flexion–extension X-rays are obtained with the patient out of the orthosis; if there is no instability, the brace is discarded. If the deformity increases and neurological signs appear, or if the patient cannot tolerate the orthosis, surgical stabilization is indicated.

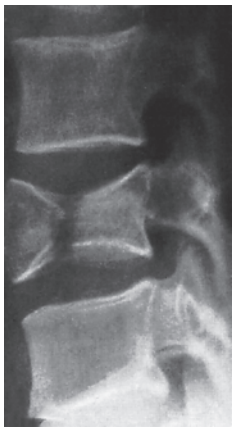
With loss of anterior vertebral height greater than 40%, it is likely that the posterior ligaments are damaged by distraction and the injury is unstable. This can be confirmed clinically and on MRI scan (PLC disruption). Posterior fixation is the preferred treatment.

In the rare cases of patients with a wedge compression fracture and neurological impairment, treatment will depend on the stability of the injury and the risk of kyphotic deformity progression. Neurological deficit



(a) (b) (c)

Figure 28.28 Pars defect (a) X-ray shows L5 pars defect; (b) corticalization and 'rounding off' of the fracture on CT indicates chronicity and non-union. (c) Occasionally the pars defect is visible on MRI scan, but MRI is better served to evaluate disc degeneration and other causes of pain such as nerve root compression.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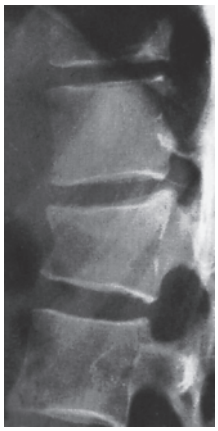
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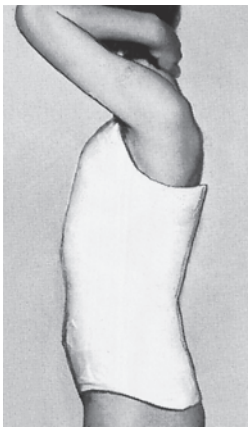
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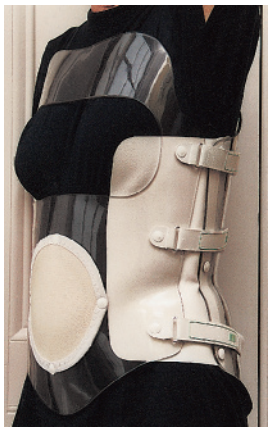
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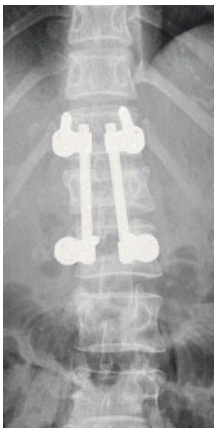
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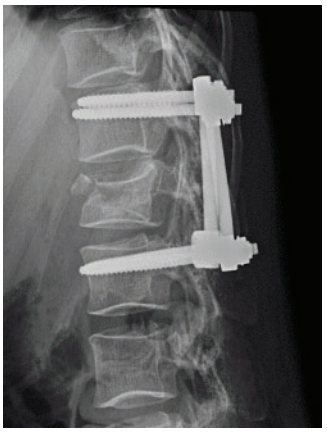

(f)

Figure 28.29 Wedge-compression fractures (a) Central compression fracture of the vertebral body and (b) anterior wedge-compression fracture with less than 20% loss of vertebral body height. If the fracture is stable without injury to the posterior ligamentous complex, it can be managed conservatively with 8–12 weeks in (c) a plaster 'jacket' or (d) a lightweight removable orthosis. (e,f) Unstable compression fractures may need posterior internal fixation if there is disruption of the PLC.

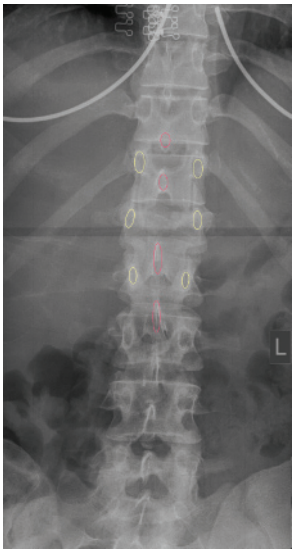
implies likelihood of spinal instability and MRI scan is indicated. If there is PLC disruption, these fractures are fixed with posterior segmental instrumentation.

Axial compression or burst injury

Severe axial compression may 'explode' the vertebral body, causing failure of the anterior vertebral column. The posterior part of the vertebral body is shattered and fragments of bone and disc may be displaced into the spinal canal. The injury is often stable, depending on the posterior ligamentous complex integrity. Anteroposterior *X-rays* may show spreading of the vertebral body with an increase of the interpedicular distance (Figure 28.30). Posterior displacement of bone into the spinal canal (retropulsion) can be appreciated at the posterosuperior border of the vertebral body where the normal concavity of the posterior body wall becomes convex. A *CT scan* will demonstrate the vertebral body comminution and retropulsion of fragments into the canal.

TREATMENT

If there is minimal anterior wedging and the fracture is stable (PLC intact) with no neurological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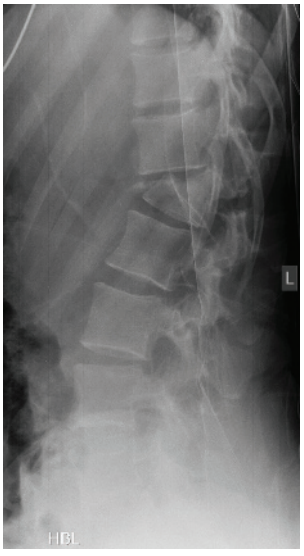
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Figure 28.30 Lumbar burst fracture An 18-year-old female fell from a trampoline sustaining L1 burst fracture, ASIA D. (a) AP X-ray showing pedicle widening and interspinous gap; (b) lateral X-ray showing kyphosis at L1 with mild retrolisthesis.

damage, the patient is kept in bed until the acute symptoms settle (usually under a week) and is then mobilized in a thoracolumbar brace or body cast which is worn for about 12 weeks. Even if CT shows that there is considerable compromise of the spinal canal, provided there are no neurological symptoms or signs, non-operative treatment is still appropriate in stable injuries. Fragments in the canal usually remodel with time.

A prospective randomized trial comparing operative and non-operative treatment of stable thoracolumbar burst fractures with no neurological impairment found no difference in the long-term results in the two groups, but complications were more frequent in the surgical group.

Stability of these fractures is determined by the fracture pattern, presence of neurological injury and disruption of the PLC. Essentially, with PLC disruption, most fractures require posterior pedicle screw fixation, which is the standard of care. Fractures with more than 50% canal compromise on CT and/or neurological deficits also tend to be unstable, requiring fixation. With increasing vertebral body comminution, the risk of posterior-alone fixation failure increases and anterior column reconstruction becomes required. Modern pedicle screw systems, however, are less likely to fail and simple posterior fixation is normally adequate.

Canal decompression can be indirect or direct. *Indirect decompression* is achieved with pedicle screw fixation and posterior distraction, utilizing the intact posterior longitudinal ligament (PLL) to reduce retropulsed fragments from the canal (Figure 28.31). When the posterior wall fragment flips 180 degrees with the cancellous aspect facing the canal ('reversed cortical sign'), this indicates disruption of the PLL and indirect

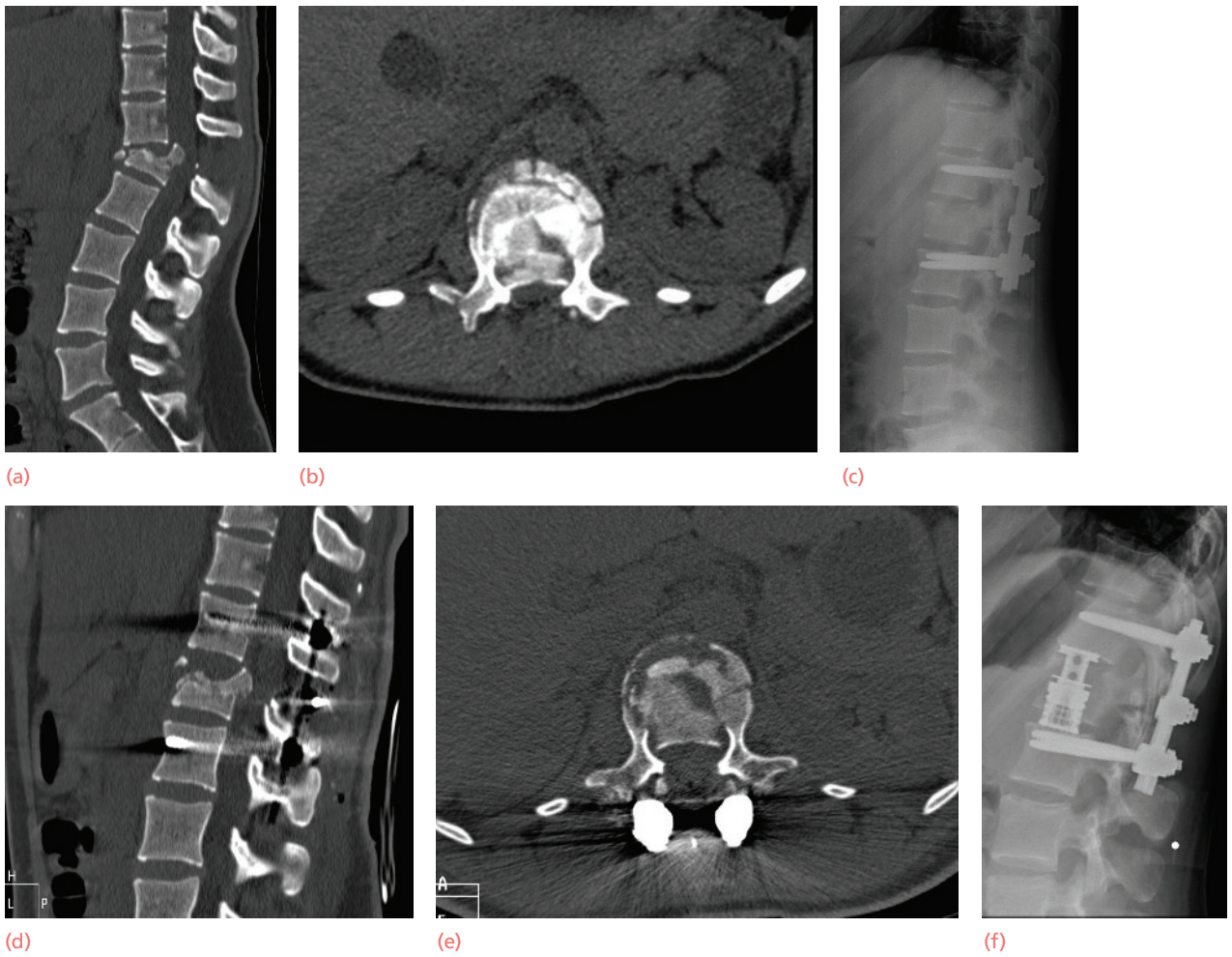


Figure 28.31 Burst fracture – treatment (a,b) L1 burst fracture in an 18-year-old female; CT demonstrates a large interspinous gap (PLC disruption) and fragments retropulsed into the canal. (c) Simple posterior fixation with Shantz screws allows kyphosis correction and distraction with indirect canal decompression and anterior column restoration. (d,e) Persistent neurological symptoms prompted repeat CT showing remaining significant fragment in the canal. (f) Anterior decompression and reconstruction with an expandable cage resolved the neurological deficit.

reduction is not possible. In this case *direct decompression* would be required if indicated via the anterior approach.

When required, a retroperitoneal or transthoracic approach is used for anterior decompression in neurologically incomplete injuries. With complete spinal cord injuries, the emphasis is on vertebral column stabilization rather than decompression, and posterior fixation would usually achieve this. Occasionally there is severe vertebral body fragmentation and the anterior column can be reconstructed anteriorly with bone strut grafts or cages.

Chance injury (jack-knife injury or seat-belt fracture)

Combined flexion and posterior distraction may cause the mid-lumbar spine to jack-knife around an axis that is placed anterior to the vertebral column. This is seen most typically in *lap seat-belt injuries*, where the body is thrown forward against the restraining strap. There is little or no crushing of the vertebral body, but the posterior and middle columns fail in distraction; thus these fractures are unstable in flexion. The tear passes transversely through the bones or the ligament structures, or both. The most perfect example of tensile failure is the injury described by Chance in 1948, in which the split runs through the spinous process, the transverse processes, pedicles and the vertebral body. The split may run through bone or discoligamentous structures only or a combination of both. Neurological damage is uncommon, although the injury is (by definition) unstable. In 50% of these injuries, there are associated intra-abdominal injuries.

X-rays may show horizontal fractures in the pedicles or transverse processes, and in the AP view the apparent height of the vertebral body may be increased. In the lateral view there may be opening up of the disc space posteriorly. In pure ligamentous injuries *MRI* will demonstrate the line of disruption not appreciated on X-ray or CT scan.

TREATMENT

The Chance fracture (being an 'all-bone' injury) heals rapidly and can be managed conservatively with bed rest and bracing. Posterior fixation is cost-effective and allows early mobilization. Ligamentous Chance injuries will not stabilize and need posterior instrumented fusion (Figure 28.32).

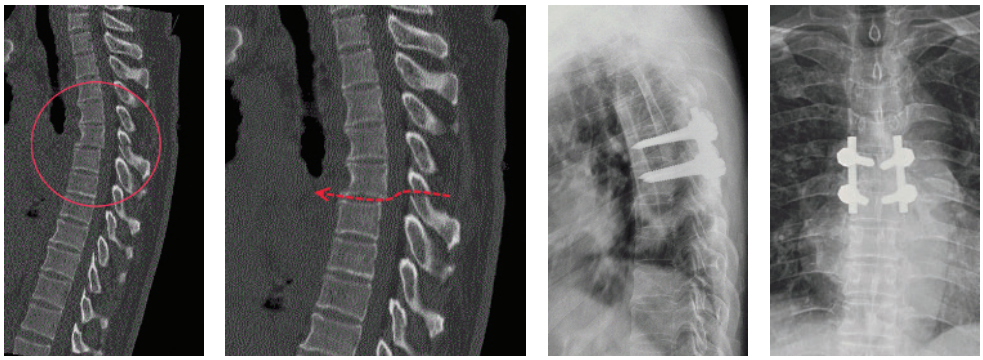
Fracture-dislocation

Segmental displacement may occur with various combinations of flexion, compression, rotation and shear. All three columns are disrupted and the spine is grossly unstable. These are the most dangerous injuries and are often associated with neurological damage to the lowermost part of the cord or the cauda equina.

The injury most commonly occurs at the thoracolumbar junction. There may be coronal and sagittal loss of alignment on *X-rays*. Fractures can involve the vertebral body, pedicles, articular processes and laminae; there may be varying degrees of subluxation or even bilateral facet dislocation. Often there are associated fractures of transverse processes or ribs. *CT* is helpful in demonstrating the degree of spinal canal occlusion.

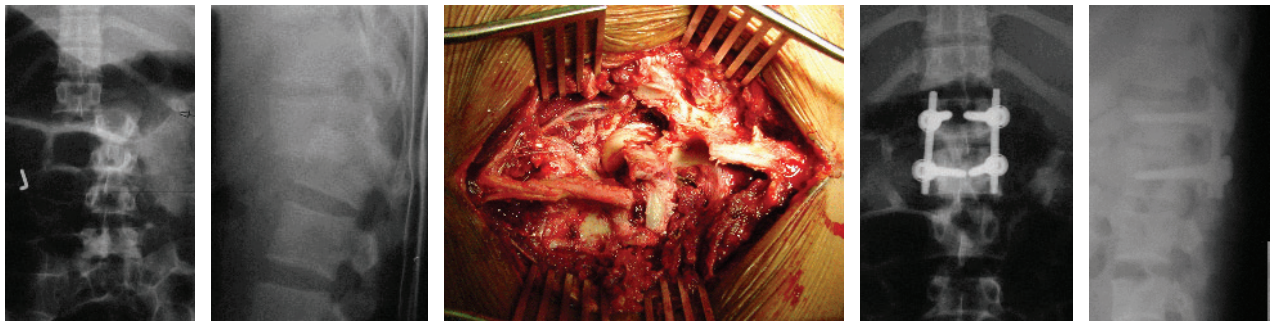
TREATMENT

There is little place for conservative management of these highly unstable fractures. Most patients have neurological deficits, and surgical stabilization allows easier nursing care and results in lower patient morbidity, decreased hospitalization costs and earlier rehabilitation (Figure 28.33). Fixation of these fractures often requires two vertebral levels instrumented above and below the injury due to the degree of instability related to the shear component and facet disruption. Neurological outcome is mostly dictated by the initial neural injury rather than subsequent efforts at decompression.



(a) (b) (c) (d)

Figure 28.32 Chance injuries (a) This innocuous-looking lamina fracture is a Chance fracture. (b) The injury forces run posteriorly from the lamina through the disc anteriorly, making it a mixed bony-ligamentous Chance injury. (c,d) Surgical stabilization with posterior instrumentation.



(a) (b) (c) (d) (e)

Figure 28.33 Thoracolumbar fracture-dislocation (a,b) High-energy fracture-dislocation at L1/L2, ASIA A. (c) Intra-operative findings reveal the extensive soft-tissue disruption associated with these high-energy injuries, which require surgical stabilization. (d,e) Short segment posterior fixation was adequate since the facets were intact. There is no requirement for decompression with complete cord injury.

NEURAL INJURIES

In spinal injuries the displaced structures may damage the cord or the nerve roots, or both; cervical lesions may cause quadriplegia, thoracolumbar lesions paraplegia. The damage may be partial or complete. Three varieties of lesion occur: neurapraxia, cord transection and root transection.

Neurapraxia

Motor paralysis (flaccid), burning paraesthesia, sensory loss and visceral paralysis below the level of the cord lesion may be complete, but within minutes or a few hours recovery begins and soon becomes full. The condition is most likely to occur in patients who, for some reason other than injury, have a small-diameter anteroposterior canal; there is, however, no radiological evidence of recent bony damage.

Cord transection

Motor paralysis, sensory loss and visceral paralysis occur below the level of the cord lesion; as with cord concussion, the motor paralysis is at first flaccid. This is a temporary condition known as cord shock, but the injury is anatomical and irreparable. After a time the cord below the level of transection recovers from the shock and acts as an independent structure; i.e. it manifests reflex activity. Within 48 hours the primitive anal wink and bulbocavernosus reflexes return. Within 4 weeks of injury tendon reflexes return and the flaccid paralysis becomes spastic, with increased tone, increased tendon reflexes and clonus; flexor spasms and contractures may develop with inadequate management.

Root transection

Motor paralysis, sensory loss and visceral paralysis occur in the distribution of the damaged roots. Root transection, however, differs from cord transection in two ways: recovery may occur and residual motor paralysis remains permanently flaccid.

ANATOMICAL LEVELS

Cervical spine Cord transection cephalad to C3 is fatal because all the respiratory muscles are paralysed (phrenic nerve C3–C5). With injury below the C5 vertebra, the upper limbs are partially spared and characteristic deformities result.

Between T1 and T10 vertebrae The first lumbar cord segment in the adult is at the level of the T10 vertebra. Consequently, cord transection at that level spares the thoracic cord but isolates the entire lumbar and sacral cord, with paralysis of the lower limbs and viscera. The lower thoracic roots may also be transected but they are of relatively little importance.

Below T10 vertebra The cord forms a slight bulge (the conus medullaris) between the T10 and L1 vertebrae and tapers to an end at the interspace between the L1 and L2 vertebrae. The L2–S4 nerve roots arise from the conus medullaris and stream downwards in a bunch (the cauda equina) to emerge at successive levels of the lumbosacral spine. Spinal injuries above the T10 vertebra therefore cause cord transection, those between the T10 and L1 vertebrae cause cord and nerve root lesions, and those below the L1 vertebra cause only root lesions.

The *sacral roots* innervate:

- sensation in the 'saddle' area (S3, S4), a strip down the back of the thigh and leg (S2) and the outer two-thirds of the sole (S1)
- motor power to the muscles controlling the ankle and foot
- the anal and penile reflexes, plantar responses and ankle jerks
- bladder and bowel continence.

The *lumbar roots* innervate:

- sensation to the groin and entire lower limb other than that portion supplied by the sacral segment
- motor power to the muscles controlling the hip and knee
- the cremasteric reflexes and knee jerks. It is essential, when the bony injury is at the thoracolumbar junction, to distinguish between cord transection with root escape and cord transection with root transection. A patient with root escape is much better off than one with cord and root transection.

INCOMPLETE CORD INJURY SYNDROMES

Persistence of any sensation distal to the injury (perianal pinprick is most important) suggests an incomplete lesion.

The commonest is the *central cord syndrome* where the initial flaccid weakness is followed by lower motor neuron paralysis of the upper limbs with upper motor neuron (spastic) paralysis of the lower limbs, and intact perianal sensation (sacral sparing). Bladder control may or may not be preserved from an early stage.

With the less common *anterior cord syndrome* there is complete paralysis and anaesthesia but deep pressure and position sense are retained in the lower limbs (dorsal column sparing). The *posterior cord syndrome* is rare; only deep pressure and proprioception are lost.

The *Brown-Séquard syndrome* (due to cord hemisection) is usually associated with penetrating thoracic injuries. There is loss of motor power on the side of the injury and loss of pain and temperature sensation on the opposite side. Most of these patients improve and regain bowel and bladder function and some walking ability.

Management of traumatic paraplegia and quadriplegia

Early surgical stabilization of these patients facilitates nursing care, mobilization and earlier transfer to specialized rehabilitation units where a multidisciplinary team approach is required.

The strategy is outlined below.

Skin

Pressure sores can develop in a few hours; prevention is with regular 2-hourly log-rolling, pressure care and pressure-reducing mattresses. Patient and carer education is required for lifelong pressure care.

Bladder and bowel

Bladder training is begun as early as possible. Although retention is complete to begin with, partial recovery may lead to either an autonomic bladder which works reflexively or an expressible bladder which is emptied by manual suprapubic pressure. A few patients are left with a high volume of residual urine after emptying the bladder. They need special investigations, including cystography and cystometry; transurethral resection of the bladder neck or sphincterotomy may be indicated but should not be performed until at least 3 months of bladder training have been completed. The bowel is more easily trained, with the help of enemas, aperients and abdominal exercises.

Muscles and joints

The paralysed muscles, if not treated, may develop severe flexion contractures. These are usually preventable by moving the joints passively through their full range twice daily. Later, splints may be necessary. Callipers are usually necessary to keep the knees straight and the feet plantigrade. The callipers are removed at intervals during the day while the patient lies prone, and while he or she is having physiotherapy. The upper limbs must be trained until they develop sufficient power to enable the patient to use crutches and a wheelchair.

If flexion contractures have been allowed to develop, tenotomies may be necessary. Painful flexor spasms are rare unless skin or bladder infection occurs. They can sometimes be relieved by tenotomies, neurectomies, rhizotomies or the intrathecal injection of alcohol. Heterotopic ossification is a common and disturbing complication. It is more likely to occur with high lesions and complete lesions. It may restrict or abolish movement, especially at the hip. Once the new bone is mature, it should be considered for excision if it interferes with function.

Tendon transfers

Some function can be regained in the upper limb by the use of tendon transfers. The aim with patients who have a low cervical cord injury is to use the limited number of functioning muscles in the arm to provide a primitive pinch mechanism (normally powered by C8 or T1 which, being below the level of injury, are lost). One must establish which muscles are working, which are not and which are available for transfer.

- *If only deltoid and biceps are working (C5, C6)* – a posterior-deltoid to triceps transfer using interposition tendon grafts will replace the lost C7 function of elbow extension; this will enable the patient to orient his or her hand in space.
- *If brachioradialis (C6) is working* – this can be transferred to become a wrist extensor (since its prime function as an elbow flexor is duplicated by biceps). A primitive thumb pinch can be achieved by the Moberg procedure in which the thumb interphalangeal joint is fused and the basal joint of

the thumb is tenodesed with a loop of the redundant flexor pollicis longus. On active extension of the wrist, the basal joint of the thumb is passively flexed.

- *If extensor carpi radialis longus and brevis (C7) are both available* – one of them can be transferred into the flexor pollicis longus to provide active thumb flexion (normally supplied by C8).

FURTHER READING

American Spinal Injury Association. *International Standards for Neurological Classification of Spinal Cord Injury*, 2002 revision. Chicago, IL: American Spinal Injury Association; 2002.

Denis F. The three-column spine and its significance in the classification of acute thoracolumbar spinal injuries. *Spine* 1983; **8**: 817–31.

Fehlings MG, Vaccaro A, Wilson JR, *et al.* Early versus delayed decompression for traumatic cervical spinal cord injury: results of the Surgical Timing in Acute Spinal Cord Injury Study (STASCIS). *PLoS ONE* 2012; **7**(2): e32037.

Hugenholtz H, Cass DE, Dvorak MF. High-dose methylprednisolone for acute closed spinal cord injury: Only a treatment option. *Can J Neurol Sci* 2002; **29**: 227–35.

Sarrami P, Armstrong E, Naylor JM, Harris IA. Factors predicting outcome in whiplash injury: a systematic meta-review of prognostic factors. *J Orthop Traumatol* 2016 Oct 13. [Epub ahead of print.]

Spitzer WO, Skovron ML, Salmi LR, *et al.* Scientific monograph of the Quebec Task Force on whiplash-associated disorders: redefining whiplash and its management. *Spine* 1995; **20**(8): 1S–73S.

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Injuries of the pelvis 29

Gorav Datta

Injuries to the pelvis may result in pelvic ring or acetabular fractures, or both. Fractures of the pelvis account for less than 5% of all skeletal injuries, but they are particularly important due to the potential risk of severe blood loss. Like other serious injuries, they demand a combined approach by experts in various fields. The majority of severe pelvic injuries occur in road traffic accidents where there is a high velocity of injury; over 10% of these patients will have associated visceral injuries, and in this group the mortality rate is in excess of 10%.

Surgical anatomy

The pelvic ring is made up of the two innominate bones and the sacrum, articulating in front at the symphysis pubis and posteriorly at the sacroiliac joints. This structure transmits weight from the trunk to the lower limbs and provides protection for the pelvic viscera, vessels and nerves. Think of the pelvis as a junction box containing the lumbosacral plexus of nerves, a rich supply of blood vessels, together with vital organs. There is no inherent bony stability of the pelvis. Unlike a hinge joint or a ball-and-socket joint, the pelvic bones do not articulate as part of a stable construct dictated by their shape. The stability of the pelvic ring depends upon the integrity of the strong ligaments that bind the three segments together across the symphysis pubis anteriorly and the sacroiliac joints posteriorly. At the sacroiliac joints, anterior stability is provided by the anterior sacroiliac ligaments and the iliolumbar ligaments (Figure 29.1a), posterior stability by the posterior sacroiliac ligaments (Figure 29.1b), the sacrococcygeal ligaments, and the sacrotuberous and sacrospinous ligaments (Figure 29.1c). The superior pubic ligament and the arcuate pubic ligament give rise to stability at the symphysis pubis. These form a construct resembling a suspension bridge. As long as the bony ring and the ligaments are intact, load-bearing is unimpaired.

The major branches of the common iliac arteries and veins arise within the pelvis between the level of the sacroiliac joint and the greater sciatic notch.

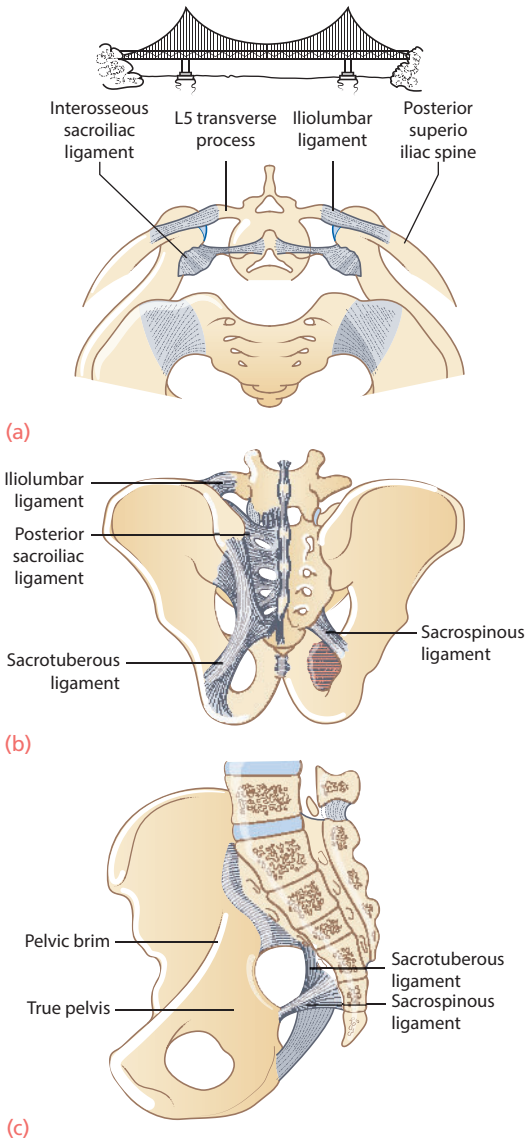


Figure 29.1 Posterior ligamentous stabilizers of the pelvis (a) Superior view; the anterior sacroiliac ligament and the iliolumbar ligament. (b) Posterior view; the posterior sacroiliac ligament, sacrotuberous ligament and sacrospinous ligament. (c) Lateral view; the sacrotuberous ligament and sacrospinous ligament.

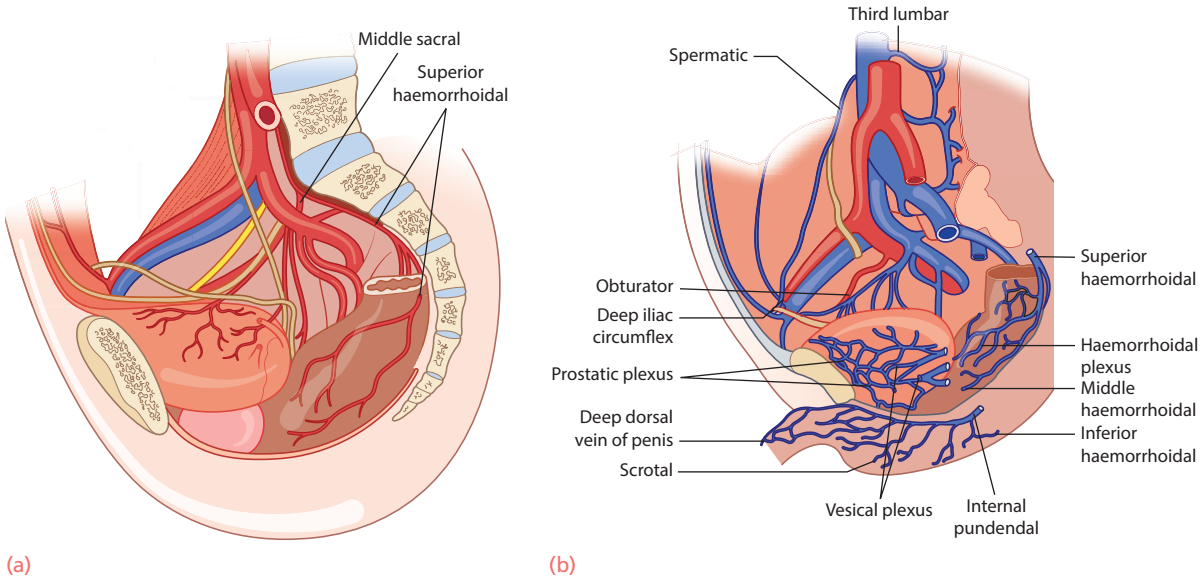


Figure 29.2 Vascular anatomy of the pelvis (a) Major arteries of the pelvis; (b) major veins of the male pelvis.

The internal iliac vessels supply the pelvic viscera, the external iliac vessels continue in their journey to supply the lower limbs (Figure 29.2). These vessels are vulnerable in fractures through the posterior part of the pelvic ring. There is a rich low-pressure venous plexus posteriorly, which is especially prone to injury if there is bony disruption around the sacroiliac joints. It is bleeding from this plexus that comprises the major blood loss in pelvic haemorrhage.

The lumbosacral plexus (Figure 29.3) is at risk with posterior pelvic injuries. The L5 and S1 nerve roots are the most commonly damaged nerves in pelvic ring injuries, and the sciatic nerve is most commonly damaged in acetabular fractures.

The bladder lies behind the symphysis pubis. The trigone is held in position by the lateral ligaments of the bladder and, in the male, by the prostate. The prostate lies between the bladder and the pelvic floor. The urethra is much more mobile and shorter in females, and it is less prone to injury.

In severe pelvic injuries the membranous urethra is damaged when the prostate is forced backwards while the urethra remains static. The male urethra is the commonest urogenital organ to be damaged.

The pelvic colon, with its mesentery, is a mobile structure and therefore not readily injured. However, the rectum and anal canal are more firmly tethered to the urogenital structures and the muscular floor of the pelvis and are therefore vulnerable in pelvic fractures. Fortunately, this is rare.

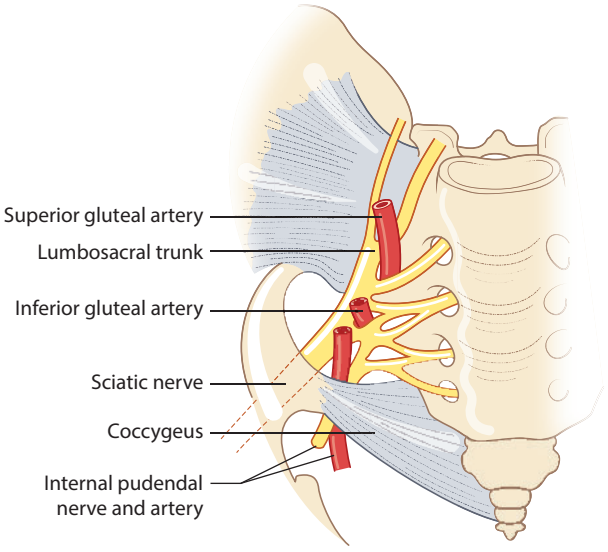
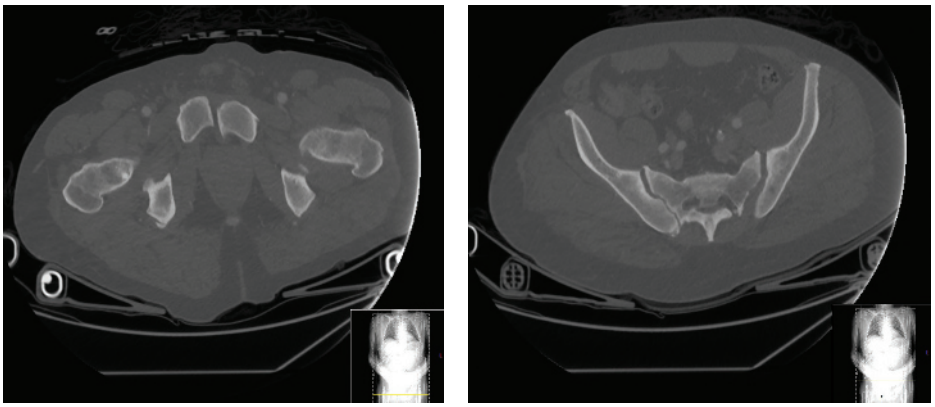


Figure 29.3 The lumbosacral plexus

Clinical assessment

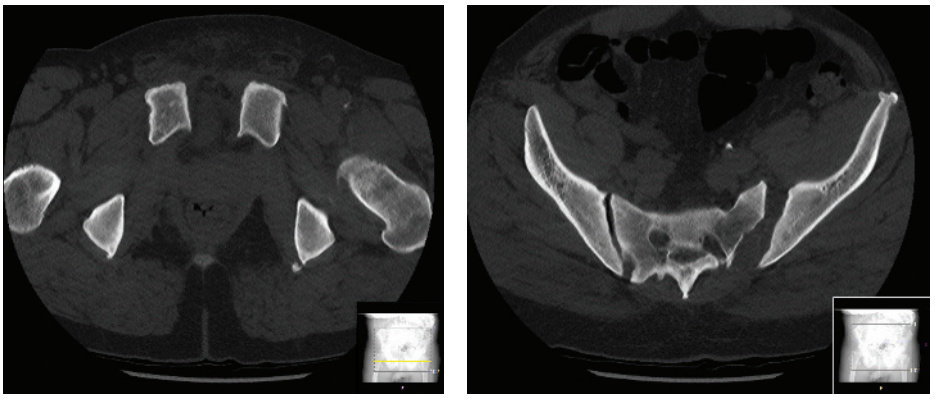
A fracture of the pelvis should be suspected in any multiply injured patient. Nowadays, the application of a pelvic binder at the scene of the accident is mandatory in any suspected pelvic injury. Patients should be managed according to the ATLS protocol (Chapter 22). The delivery of care to a multiply injured patient is best provided in a Major Trauma Centre. Patients with a suspected pelvic fracture may be transferred to a Level 1/Major Trauma Centre directly from the accident site. The history, taken from the patient, a witness or the ambulance crew, is essential as the mechanism of the injury can be predictive of the type of pelvic fracture sustained.

There may be swelling and bruising of the lower abdomen, the thighs, the perineum, the scrotum or the vulva. The abdomen should be carefully palpated. Guarding or tenderness suggests the possibility of intraperitoneal bleeding. Repeated assessment of an unstable pelvis will lead to disruption of any clot formed; this may worsen haemorrhage. The pelvis is palpated and gently compressed to determine if



(a) (b)

Figure 29.4 Pelvic ring fracture (a) The pubic symphysis and (b) the sacroiliac joints with the binder applied. There is a pubic symphysis injury which can be seen with the step across the symphysis, with a left sacroiliac joint injury which can be appreciated with the subtle widening of the left side compared to the right.



(a) (b)

Figure 29.5 Pelvic ring fracture The same patient as in Figure 29.4 with the binder removed. Note the now much more significant widening of (a) the pubic symphysis and (b) the left sacroiliac joint.

there is tenderness consistent with a fracture, but the pelvis is not 'sprung' or vigorously tested for stability to avoid provoking further bleeding. Generally, pelvic binders are kept on until an experienced surgeon can make an assessment of pelvic stability using imaging. Pelvic binders can provide false reassurance by completely reducing a pelvic fracture, therefore the clinical assessment remains a critical part of the assessment of the patient and X-rays should be taken following the removal of the binder to ensure no displacement of the pelvis has occurred (Figures 29.4 and 29.5).

A ruptured bladder should be suspected in patients who do not void or in whom a bladder is not palpable after adequate fluid replacement (Figure 29.6). Palpation is often difficult because of abdominal wall haematoma. Bladder rupture may be intraperitoneal or extraperitoneal. Intraperitoneal rupture may be associated with massive haemorrhage.

Neurological examination is very important; there may be damage to the lumbosacral plexus. If the patient is unconscious, an assessment must be made at a later date when the patient has awoken.

Imaging of the bony pelvis

During the initial survey of severely injured patients, a plain anteroposterior (AP) X-ray of the pelvis is obtained at the same time as the chest X-ray.

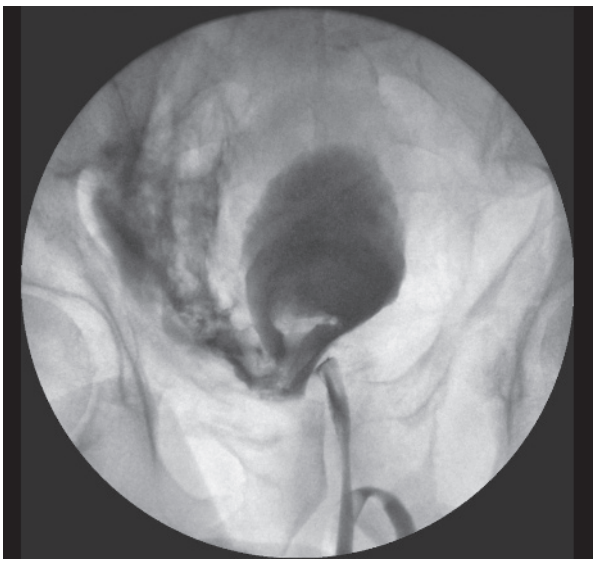


Figure 29.6 Bladder rupture Retrograde cystourethrogram showing a bladder rupture – contrast is leaking out on the right side of the pelvis.

An AP radiograph of the pelvis (Figure 29.7) should be carefully inspected, systematically looking in each of the five zones of injury:

1 *The sacroiliac joint* area is inspected for any diastasis or sacral fracture.
2 *The ilium* is inspected for any fracture.

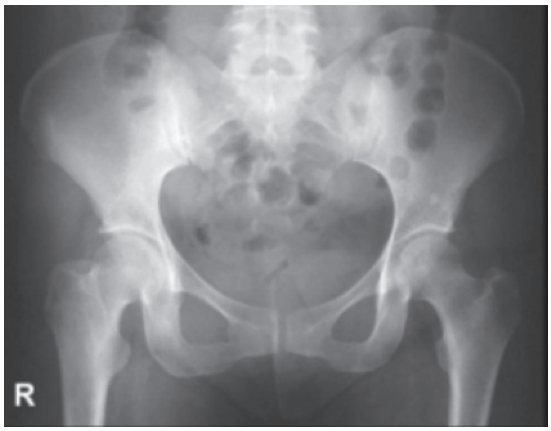


Figure 29.7 Anteroposterior radiograph of the pelvis
Examine each of the five areas carefully.

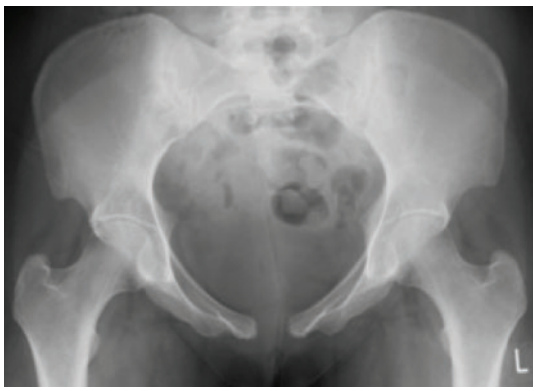
3 *The teardrop* is inspected. This is a radiological feature which correlates to the non-articular floor of the acetabulum, and close inspection in this area will reveal any acetabular fracture.
4 *The obturator foramen* is inspected for any fracture of the superior or inferior pubic ramus.
5 *The symphysis pubis* is examined for any fracture or diastasis.

Specialized radiographs include the use of pelvic inlet and outlet views (Figure 29.8). These are taken at approximately 30–40 degrees in a caudal and cephalic angle and are used to assess the pelvic ring. The inlet view provides an axial view of the sacrum and sacroiliac joints, and the outlet view provides a true anteroposterior view of the sacrum and pubic symphysis areas. These are very useful if CT scanning is not available. These images are also routinely used for serial follow-up imaging of patients with a pelvic ring injury.

Specialized radiographs are also taken to assess acetabular fractures in more detail. Judet views (taken at 30 degrees obliquely) comprise an obturator and iliac oblique view (Figure 29.9). The obturator oblique view shows the anterior column of the acetabulum, and the iliac oblique view shows the posterior column and anterior wall of the acetabulum.

There are six lines that aid the diagnosis and classification of acetabular fractures (Figure 29.10):

1 anterior wall of the acetabulum
2 posterior wall of the acetabulum
3 roof or dome of the acetabulum
4 iliopectineal line – this corresponds to the anterior column of the acetabulum



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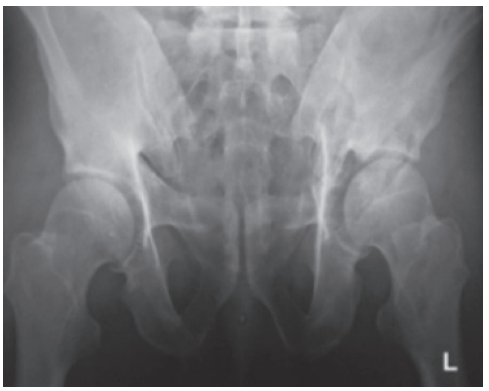
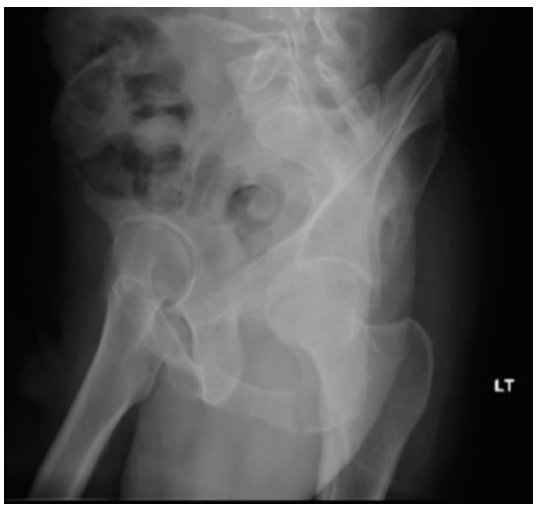


(b)

Figure 29.8 Radiographic views of the pelvis
(a) Inlet view;
(b) outlet view.



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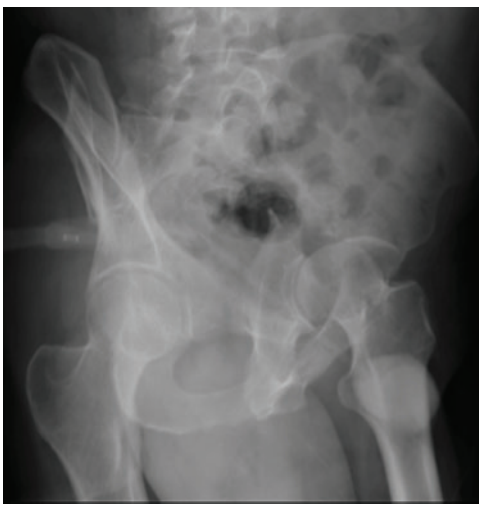

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Figure 29.9 Radiographic views of the acetabulum
(a) Obturator oblique view of the left acetabulum;
(b) iliac oblique view of the left acetabulum.

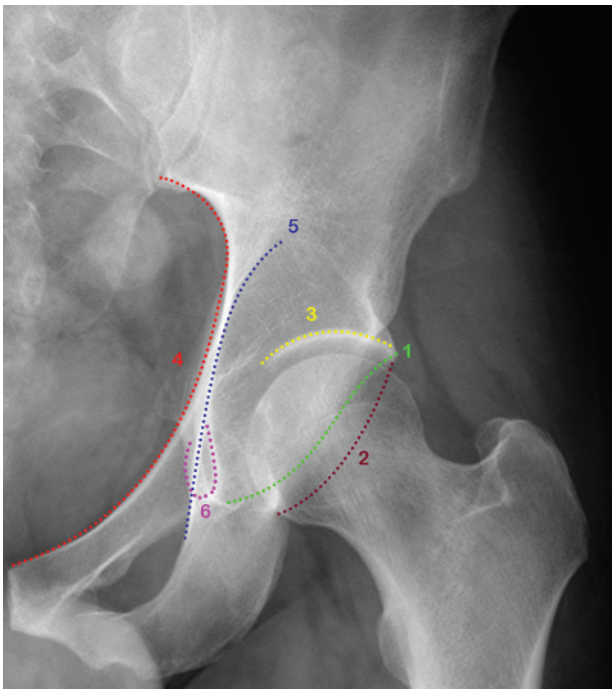


Figure 29.10 Pelvic X-ray assessment The six lines of the acetabulum: 1 anterior wall of the acetabulum; 2 posterior wall of the acetabulum; 3 roof or dome of the acetabulum; 4 iliopectineal line – this corresponds to the anterior column of the acetabulum; 5 ilioischial line – this corresponds to the posterior column of the acetabulum; and 6 teardrop.

5 ilioischial line – this corresponds to the posterior column of the acetabulum
6 teardrop.

In every acetabular fracture, systematically look at each line to see which is broken. Deciding which line is intact and which is broken helps to interpret the Letournel classification.

In Level 1/Major Trauma Centres, after initial resuscitation, patients are now taken early to the CT scanning unit for a full 'trauma CT scan'. This comprises a CT scan of the head, neck, chest, abdomen and pelvis. Contrast is often also given. This is very helpful in excluding a bladder rupture or urethral injury. CT scanning provides a detailed anatomical view of the posterior structures, which are not seen well on conventional radiographs. CT scans can be reformatted to provide Judet views, multiplanar reconstructions (axial, coronal and sagittal images), and 3D reconstructions (Figure 29.11). Using CT scanning, 3D printing techniques can be used to print plastic moulds of pelvic fractures. This provides assistance in preoperative planning and plate contouring prior to fixation.

MRI is rarely used to assess the bony pelvis. Its use is mainly confined to imaging tumours of the bony pelvis or soft tissues, and it can be very helpful in visualizing fragility fractures of the pelvis in elderly patients.

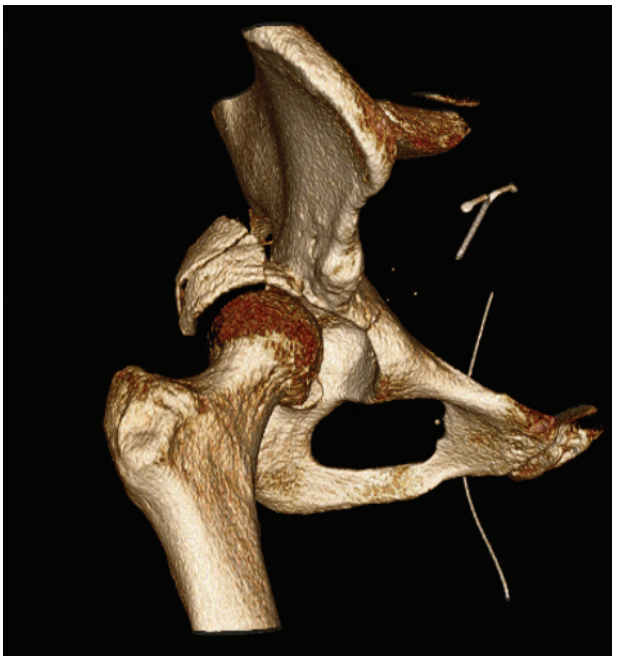


Figure 29.11 3D CT reconstruction of the pelvis
A dislocated hip with a posterior wall fracture.

Imaging of the urinary tract

Contrast is usually administered at the time of the 'trauma CT scan'. This will demonstrate any bladder rupture or urethral injury (providing no catheter is *in situ*). It provides a complete picture of the upper urinary tract too. A retrograde cystourethrogram is sometimes performed if there is suspicion of a urethral or bladder injury (see Figure 29.6).

PELVIC FRACTURES

ISOLATED PELVIC FRACTURES

Pelvic ring fractures and acetabular fractures are usually the result of a high-energy injury. However, isolated pelvic fractures can and do occur.

Avulsion fractures

The most common avulsion injuries seen around the pelvis are of the anterior inferior iliac spine (rectus femoris origin) and the ischial tuberosity (hamstring origin). The bone is pulled off by violent muscle contraction; this is usually seen in sportsmen and women and athletes. Other patterns include the sartorius pulling off the anterior superior iliac spine and the adductor longus a piece of the pubis. All are essentially muscle injuries, needing only rest for a few days and muscle rehabilitation if the bony fragment is small. If there is a large bony fragment with displacement, however, operative fixation may be necessary for an optimal functional result.

Stress fractures

Fractures of the pubic rami are fairly common in osteoporotic bone. More difficult to diagnose are insufficiency fractures around the sacroiliac joints (SIJs); this is an uncommon cause of 'sacroiliac' pain in elderly osteoporotic patients. MRI is very helpful for the diagnosis of posterior insufficiency fractures. Stress fractures are also seen in the superior and inferior pubic rami in slim individuals and long-distance runners. Patients may present with groin pain lasting a few weeks or months and radiographs will reveal the fracture, which becomes more apparent when callus formation occurs during healing. Consideration should be given to checking vitamin D levels in these patients to exclude deficiency. Patients usually heal with rest; rarely a painful non-union may persist which requires surgery.

Pelvic ring fractures

Pelvic ring fractures are usually due to a high-energy injury. Think of the pelvis as a 'polo mint'. It is impossible to break a polo mint in one place. The same principle applies to the normal bony pelvic ring. If there is an anterior ring injury, always look for the associated posterior fracture or joint disruption. Anteriorly the symphysis pubis or pubic rami will be disrupted, and posteriorly there will either be a sacroiliac joint displacement or sacral fracture.

To fully appreciate the causation and management of pelvic fractures, it is important to have an understanding of the classification of pelvic fractures. Two classifications have stood the test of time and are used together as they each provide valuable information. The Young and Burgess is a classification of pelvic ring injuries based on the mechanism of injury, and the Tile classification provides an assessment of stability of the pelvis. The mechanism of injury (Young and Burgess) is predictive of the severity of the injury (blood loss) and also guides the surgeon on how to correct any deformity or displacement of the fracture, and stability (Tile) guides the surgeon as to whether an injury needs operative fixation.

Young and Burgess classification

ANTEROPOSTERIOR COMPRESSION (APC)

This type of injury results from a front-on force transmitted through the pelvis. Initially the anterior structures open up (symphysis pubis) and, as the energy increases, the posterior structures (sacroiliac joint) are injured. The APC injury (or open-book injury) is commonly seen in motorcyclists who straddle the bike and are subject to a sudden deceleration at the point of impact. The injury is also seen in horse riders when the horse rears up, the rider falls off, and the horse falls backwards between the rider's legs. Note that the degree of displacement of the pubic symphysis is that present when there is no pelvic binder on.

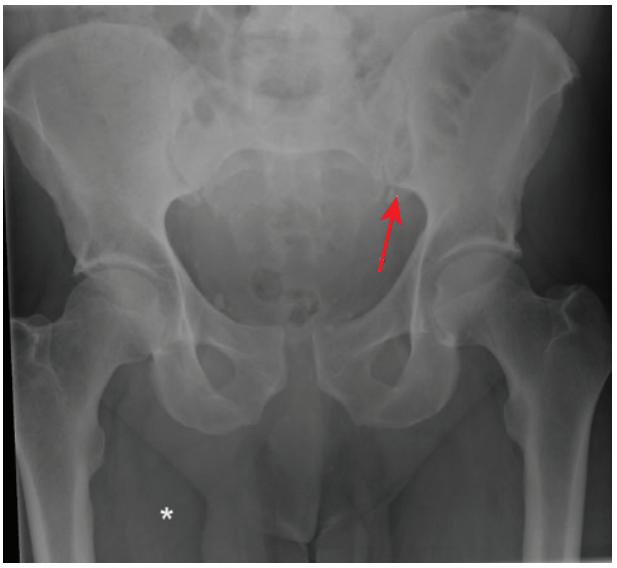


Figure 29.12 APC II pelvic fracture A diastasis (widening of the pubic symphysis) anteriorly and widening of the left SIJ (arrow) can be seen.

- *APC I* – less than 2.5 cm of widening at the pubic symphysis
- *APC II* – symphysis widening of more than 2.5 cm with an anterior widening of a sacroiliac joint; the posterior ligaments are intact (Figure 29.12)
- *APC III* – widening of the symphysis of more than 2.5 cm with a dislocation of a sacroiliac joint (see Figure 29.5).

LATERAL COMPRESSION (LC)

Lateral compression injuries result from the force applied to and transmitted from the side of the pelvis. This is commonly seen in pedestrians hit by an automobile, or from a side-on impact where an automobile is hit (T-boned). In the milder form one side of the hemipelvis is affected and, as the energy increases, the injury is transmitted to and affects the opposite side.

- *LC I* – rami fracture and ipsilateral anterior sacral alar fracture (Figures 29.13 and 29.14)
- *LC II* – rami fracture and ipsilateral posterior ilium fracture dislocation
- *LC III* – ipsilateral lateral compression and contralateral APC pattern injury (windswept pelvis).

VERTICAL SHEAR (VS)

Vertical shear injuries are usually seen after a fall from height, landing on one leg leading to one hemipelvis being driven up (Figure 29.15a).

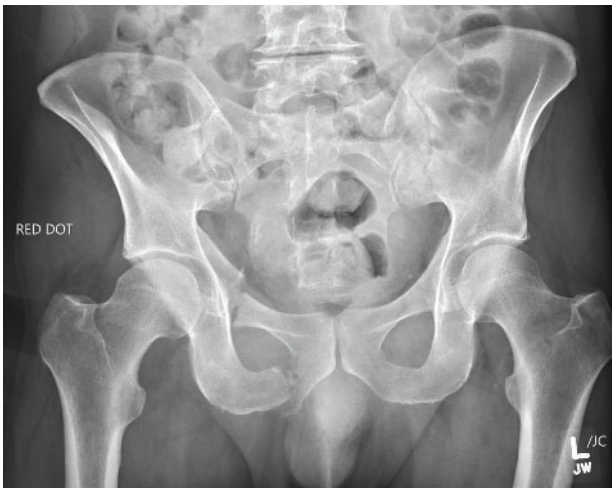


Figure 29.13 Right-sided superior and inferior pubic ramus fracture

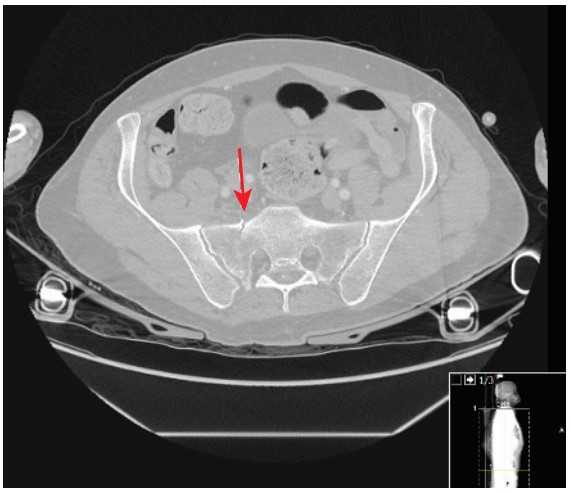
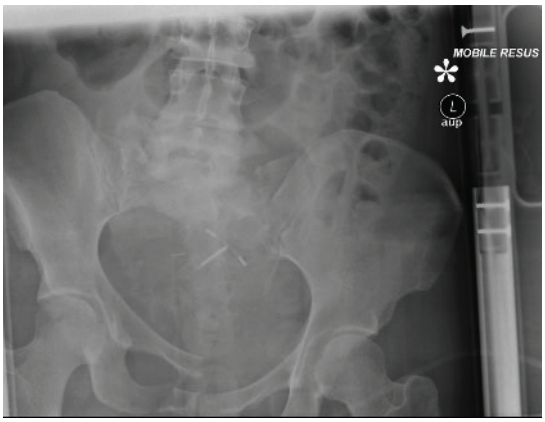


Figure 29.14 Right-sided sacral alar fracture



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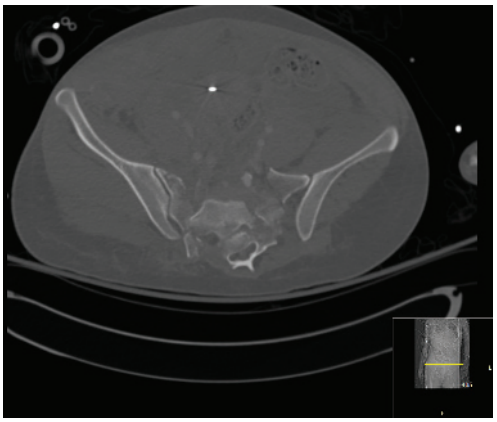


(b)

Figure 29.15 Vertical shear fracture (a) Note how the right hemipelvis is higher than the left. (b) CT scan of the same patient showing complete displacement of the posterior structures.

This leads to complete disruption of all posterior structures (Figure 29.15b).

COMBINED

This injury results from a combination of APC, LC and VS. It may occur when an individual is ejected from an automobile or motorcycle.

Tile classification

The Tile classification gives an accurate assessment of pelvic stability (Figure 29.16). This is vital for the clinician as it guides whether the patient may require surgery and whether the patient can safely mobilize with their injury. The classification is relatively simple: fractures are stable (type A), partially stable (type B) or unstable (type C). There are many subgroups, but a simplified version is supplied below:

- A:
  - A1 – fracture not involving pelvic ring (e.g. avulsion or iliac wing fracture)
  - A2 – iliac wing fracture or anterior rami fractures
  - A3 – transverse sacral fracture
- B:
  - B1 – unilateral anterior disruption of posterior structures (SIJ widening or sacral fracture)
  - B2 – unilateral SIJ joint fracture / subluxation (anterior ring rotation)
  - B3 – bilateral SIJ / sacral fracture / subluxation
- C:
  - C1 – complete unilateral posterior disruption
  - C2 – complete unilateral posterior disruption with contralateral partial disruption
  - C3 – complete bilateral posterior disruption.

At first glance both classifications appear daunting. Use the Young and Burgess first. In many cases the mechanism of action is given in the history and one can predict the type of fracture pattern seen. Then use the Tile classification to decide whether the injury is stable or not.

It is the APC III, LC III, VS and Combined mechanism injuries that are associated with massive blood loss. In all of these cases, the pelvic ring is completely unstable (type C). Open fractures of the pelvis are also prone to major bleeding, particularly if they remain unreduced, hence the need for early application of a pelvic binder.

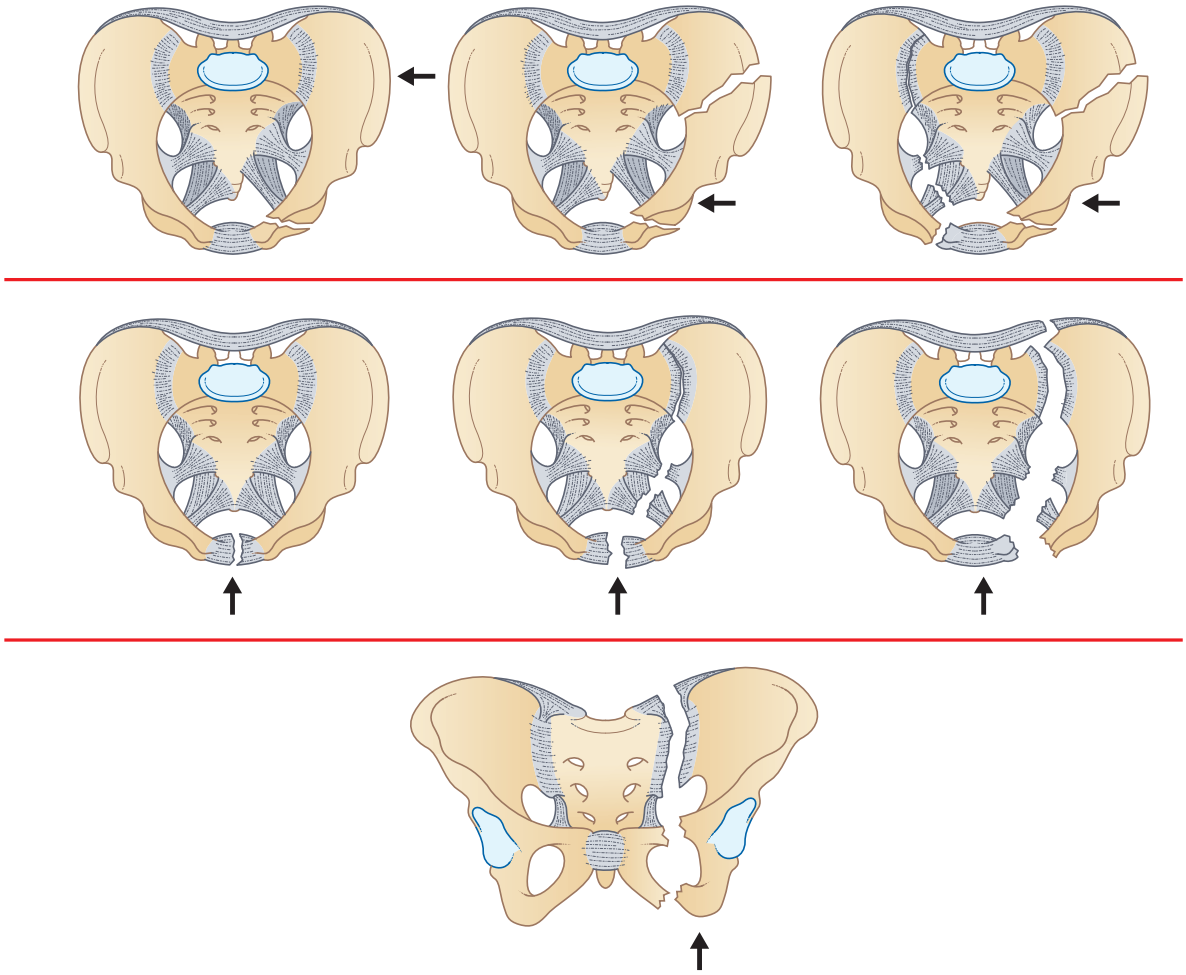


Figure 29.16 Tile classification of pelvic fractures

Treatment of pelvic ring fractures

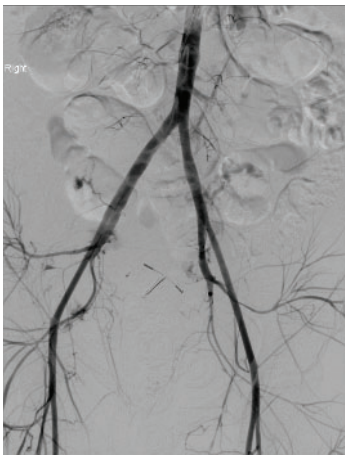
Many pelvic ring injuries are sustained in incidents involving high energy. Patients may have sustained multiple injuries. The initial management must follow the ATLS protocol to the injured patient. Part of this protocol involves immediate resuscitation of the patient, management of haemorrhagic shock, and early institution of blood products.

PELVIC BINDERS

The pelvic binder is now used routinely in many countries and applied at the accident scene by paramedics in any suspected case of a pelvic injury. Appropriate application of the binder is key. The binder should be applied at the level of greater trochanters of the hips (not the iliac crest). It is effective in closing the pelvic volume, and providing temporary stability (see Figures 29.4 and 29.5). Ideally, it should not be left on for more than 24 hours as pressure sores can develop. If the facility or expertise to stabilize an unstable pelvis that is the cause of haemodynamic instability is not present within 24 hours, the binder should be released and the pressure areas checked, it can then be reapplied if pressure areas are intact and not threatened. Most pelvic binders have a ratchet locking system so the person applying them knows when they are tight enough. Sometimes, a pelvis can be 'over-reduced', causing too much internal rotation of the hemipelvis if there is complete posterior disruption. If this is seen on X-ray or CT scan, the binder can be loosened until an acceptable position is achieved. Even if the pelvis is over-reduced, the pelvic volume has been reduced, hence limiting blood loss and therefore this is preferable to a binder not being applied or one being applied too loosely. The use of a pelvic binder has virtually eliminated the use of an external fixator to provide immediate pelvic stability.

MANAGEMENT OF THE PATIENT IN EXTREMIS

Massive blood loss caused by a pelvic fracture is, fortunately, rare. Knowing how to manage the patient appropriately will save a life. If a binder is *in situ*, and there is persistent haemodynamic instability despite resuscitation with blood products, immediate haemorrhage control is required. Other causes and sites of haemorrhage must first be excluded (chest, abdomen,



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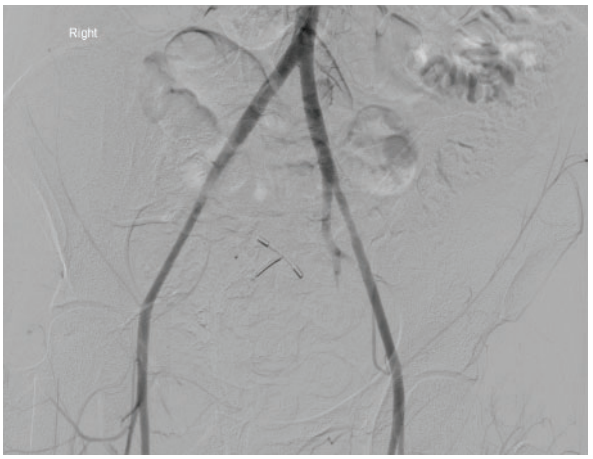


(b)

Figure 29.17 Pelvic angiography (a) The vascular tree in the pelvis can be seen. (b) Post-embolization, note how the internal iliac vessels are now occluded and the only patent arteries are the external iliac vessels.

external bleeding). If there is no time for a trauma CT, an AP X-ray of the pelvis should be taken; a useful rule of thumb is that there cannot be haemodynamic instability without mechanical instability; the radiograph is likely to demonstrate a Tile B or C fracture.

Two options exist; these depend on the experience of those available and the protocol that your unit follows in this situation:

1 *Angiography and embolization* – Immediate transfer to the angiography suite for targeted embolization is undertaken (Figure 29.17). This method controls arterial bleeding only – the major vessels are the internal iliac artery and superior gluteal artery. Haemorrhage in the unstable pelvic fracture is predominantly bleeding from low-pressure veins, with a smaller contribution from arterial bleeding and bleeding bone. The venous bleeding will not be controlled by embolization. One of the potential consequences of embolization is muscle necrosis (gluteals) as a result of superior gluteal vessel occlusion. Unselected embolization is associated with a significant morbidity secondary to ischaemia distally and should only be used as a last resort.
2 *Immediate transfer to the operating theatre for pre-peritoneal packing* – An external fixator is swiftly applied, and the pelvis is opened via the Stoppa approach; the rectus abdominis muscle is divided in the midline. At least six large abdominal packs are inserted, three either side of the midline: one posteriorly, one in the mid-pelvis, and another anteriorly. The external fixator should be sited and constructed to allow access not only for this approach but also for a trauma laparotomy to be performed.

The author's preference is for pre-peritoneal packing to control haemorrhage. The patient can be transferred for angiography following the operative procedure if they remain haemodynamically unstable.

CONSERVATIVE MANAGEMENT

If the pelvic injury is of a stable configuration (Tile A), conservative management is generally indicated unless the injury is open. Bed rest or traction is not favoured for the treatment of pelvic injuries due the complications of being immobile in hospital for such prolonged periods (venous thromboembolism, chest sepsis, pressure sores). Early mobilization with the use of walking aids is favoured; patients may partially weight-bear as tolerated on the affected side. Common fractures treated conservatively include avulsion fractures and the LC I type fracture.

OPERATIVE FIXATION

The principle of operative fixation is to convert an unstable pelvic ring to a stable one. Specialized tools are required for pelvic surgery. These include the use of a radiolucent operating table, as well as traction devices to facilitate the reduction. Pubic symphysis diastasis is treated with open reduction and internal fixation with plates and screws

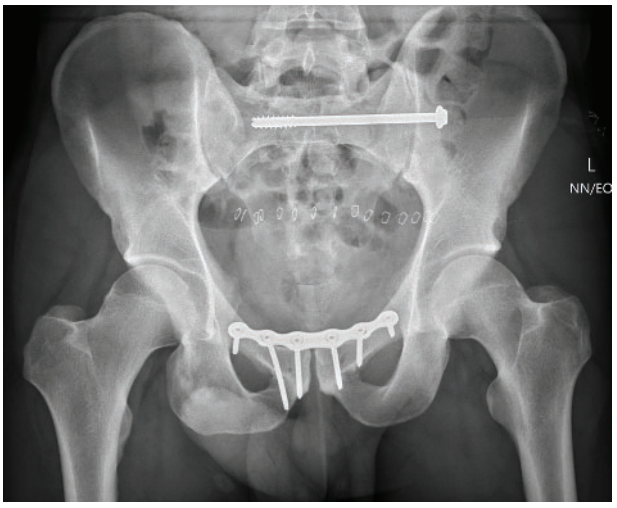


Figure 29.18 Post-operative pelvic fracture fixation
Symphysis pubis plating with a left iliosacral screw.

(Figure 29.18). Sacroiliac dislocations, subluxations and sacral fractures are reduced and stabilized. Reduction of a hemipelvis involves a combination of traction and rotation to correct the deformity. This can be performed in a closed manner or via an open reduction. Percutaneous iliosacral screws are passed to stabilize sacral fractures and sacroiliac joint injuries following reduction. Sometimes, in the case of a displaced sacral fracture, open reduction and internal fixation is performed via a posterior approach (Figure 29.19).

UROGENITAL INJURIES

Bladder and urethral injury is the commonest associated injury in pelvic fractures. A posterior urethral tear is diagnosed by retrograde urethrography. Involvement of a urologist is important to manage these injuries correctly. Urethral tears are usually treated conservatively with catheterization for a few weeks. If a soft, silicone 16F catheter (in adults) cannot be passed by a single, gentle attempt, a suprapubic catheter is required. If the catheter is passed and drains blood-stained urine, a retrograde cystogram via the catheter is mandated. If only blood is drained or the catheter will not pass, a retrograde urethrogram should be performed. If there is a urine leak from the bladder or urethra, the pelvic fracture should be treated as open with antibiotics for 72 hours and debridement and fixation as soon as the patient is stable. Intraperitoneal rupture of the bladder requires emergency laparotomy and direct repair; extraperitoneal bladder rupture may be treated conservatively unless the pelvic fracture is unstable, in which case it should be repaired at the time of fixation.

Complications

Urethral stricture Urethral stricture may arise as a result of a scarring from a tear that has healed. On rare occasions extrinsic pressure from fracture healing has been seen to cause urethral narrowing.

Impotence Damage to vascular and nervous structures supplying the genitalia is as high as 30% in pelvic fractures. It is especially common following symphyseal separation. This can be debilitating, particularly for younger patients. Early referral to a sexual dysfunction/andrological clinic should be made.

Venous thromboembolism (VTE) The risk of VTE is especially high following pelvic surgery. Trauma increases the risk; in a multiply injured patient undergoing repeated visits to the operating theatre, it may be wise to insert a vena cava filter. VTE prophylaxis is essential in all patients: they should receive TED stockings, mechanical compression and pharmacological prophylaxis prior to surgery (unless there is major bleeding). Those patients undergoing surgery should receive prolonged pharmacological treatment postoperatively. Options include low-molecular-weight heparin for 6 weeks postoperatively; some units administer warfarin for 3 months following surgery.

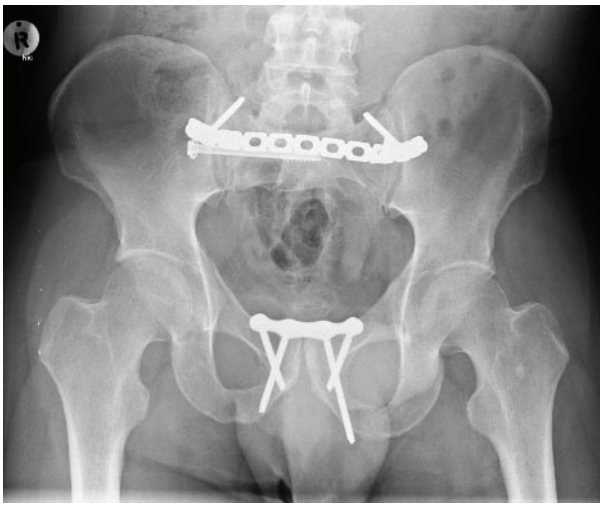


Figure 29.19 Post-operative pelvic fracture fixation
Symphysis pubis plating and posterior plating of the SIJ.

Nerve injury Nerve injury can arise as a result of the initial injury, or as a result of surgery. During placement of the S1 iliosacral screw, the L5 nerve root is at risk of being injured (as it runs anteriorly to the sacral ala). It is essential to use the correct views during screw placement (inlet, outlet and lateral views) to ensure the screw is not too anterior (L5 nerve root at risk) and not too posterior (sacral canal at risk). If S1 screw placement is too inferior, it will traverse the S1 foramen and injure the S1 nerve root. Always check and record nerve function preoperatively and postoperatively.

Infection Infection is a rare but serious complication following surgery. The risk is increased in open fractures, and in cases of bowel and bladder injury. In these cases, definitive fixation using external fixation is sometimes the best way to manage the patient to minimize this risk.

Non-union This is sometimes seen following conservative management. It usually results in a pubic rami non-union and may not always be symptomatic. Sacral non-union does occur following operative fixation, but this risk is minimized by an accurate reduction.

ACETABULAR FRACTURES

Acetabular fractures occur when the femoral head is driven into the acetabulum. It is the direction

of the force (i.e. the position of the leg at impact) that determines the fracture pattern. Displaced fractures result in hip joint incongruency; this will lead to osteoarthritis. Acetabular fractures may occur in conjunction with pelvic ring fractures, or in isolation. These fractures were studied by Emile Letournel in the 1950s. He developed the specialized imaging (Judet views), the classification, and the ilioinguinal approach for surgical fixation. The acetabulum is thought of an inverted Y shape (Figure 29.20), linked to the sacroiliac joint by the sciatic buttress. The longer part of the 'Y' is the anterior column, propped up with the aid of the shorter part, the posterior column.

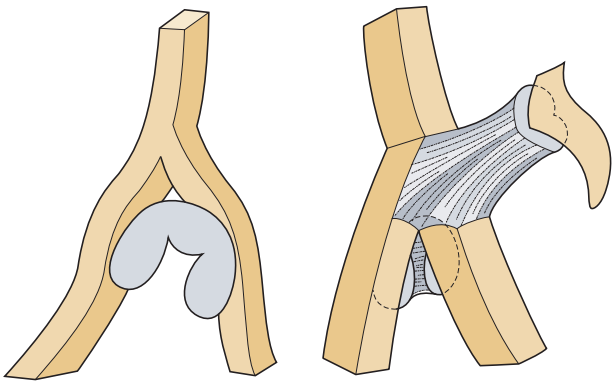


Figure 29.20 Inverted Y concept The two columns of the acetabulum.

Letournel classification

At first glance, this is one of the most daunting classifications in orthopaedics. The starting point to understanding the classification is to examine the lines on the AP pelvic X-ray, systematically looking at each line and seeing which is intact and which is not (see Figure 29.10).

There are two groups: *elemental fractures* and *associated fractures* (Figure 29.21). Four elemental fractures correspond to four lines: posterior wall, posterior column, anterior wall, and anterior column. The fifth elemental fracture is the transverse fracture, which is a complete fracture through the innominate bone.

The associated fractures are a combination of the elemental fractures.

POSTERIOR WALL FRACTURE

The posterior wall fracture is the most common fracture pattern seen and it is associated with a femoral head dislocation (Figure 29.22). It occurs when the leg is flexed, commonly in a road traffic accident where the knee hits the dashboard of a car at impact, forcing the femoral head posteriorly through the wall. These injuries can also be associated with posterior cruciate ligament injures of the ipsilateral knee. Emergent reduction of the femoral head should be performed. CT scanning is used to look for the presence of bone fragments in the joint and any marginal impaction of the articular surfaces.

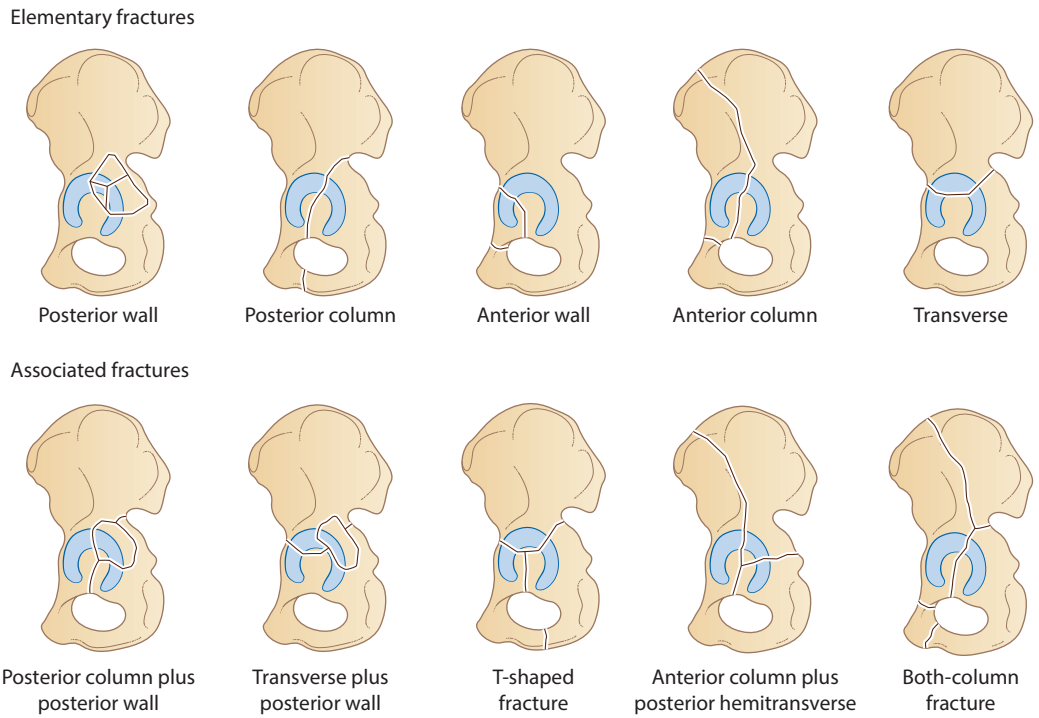


Figure 29.21 The Letournel classification of acetabular fractures

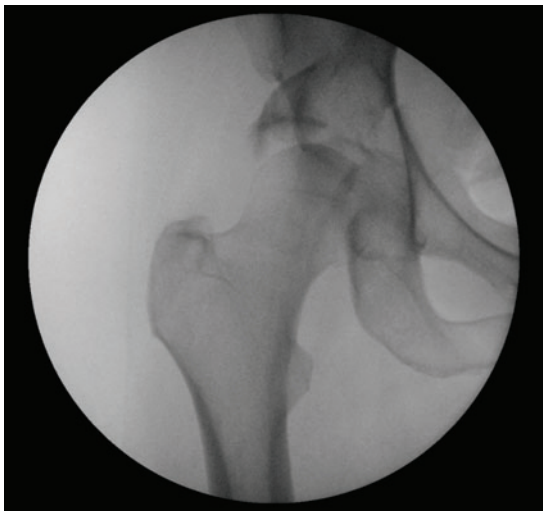


Figure 29.22 Intraoperative fluoroscopy image of a posterior acetabular wall fracture

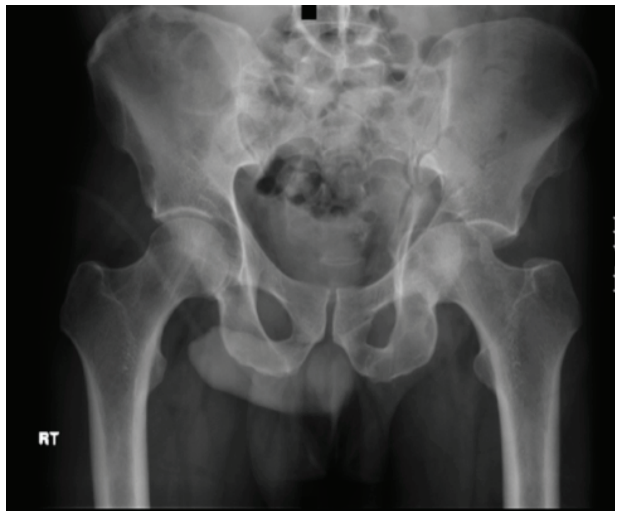


Figure 29.23 Posterior column acetabular fracture of the left hip

Treatment is examination under anaesthesia under fluoroscopic control and operative fixation (if unstable) via the Kocher-Langenbeck approach (see below).

POSTERIOR COLUMN FRACTURE

Posterior column fractures occur when the femoral head is directed medially. The fracture line may run from the obturator foramen through the posterior part of the dome of the acetabulum, and finally through the greater sciatic notch (Figure 29.23). There may be medial displacement of the femoral head and sciatic buttress.

Operative fixation is performed through a Kocher-Langenbeck approach. Care must be taken to expose and mobilize the superior gluteal neurovascular bundle; this is almost always interposed in the fracture at the level of the greater sciatic notch.

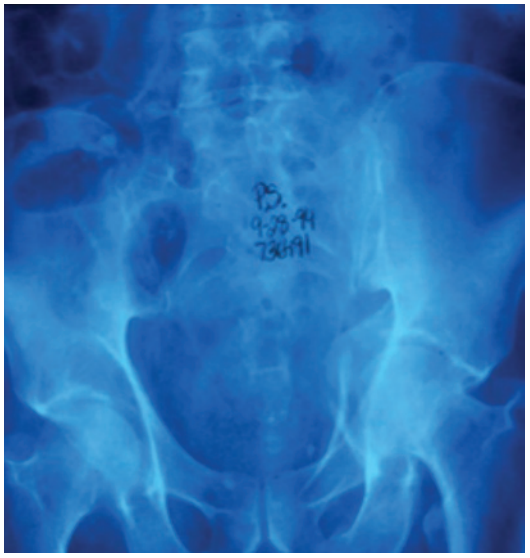
ANTERIOR WALL FRACTURE

This is the rarest fracture pattern seen. It is very commonly mistaken for a superior pubic ramus fracture that encroaches on the acetabular rim. A true anterior wall fracture is associated with anterior subluxation of the femoral head (Figure 29.24). The injury occurs with the leg in external rotation and radiographs will show the iliopectineal line broken in two places associated with a large trapezoid fragment.

If there is subluxation of the femoral head and the hip is unstable, operative fixation is performed. This is via either the Stoppa or the ilioinguinal approach.

ANTERIOR COLUMN FRACTURE

This injury occurs with the leg in external rotation. X-rays will show disruption of the iliopectineal line; look for the exiting fracture line through the



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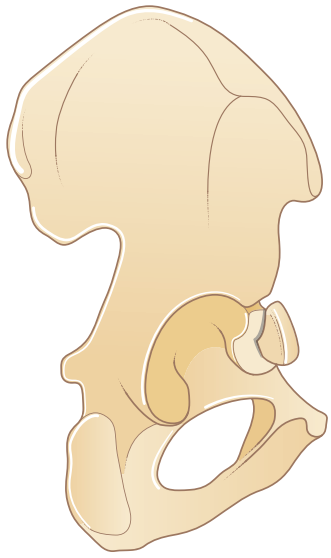


(b)

Figure 29.24 Anterior wall acetabular fracture

ischiopubic ramus. There are subtypes of fracture according to the fracture height.

Operative fixation is via the Stoppa or ilioinguinal approach. If a closed reduction is achievable, percutaneous anterior column screw fixation can be performed.

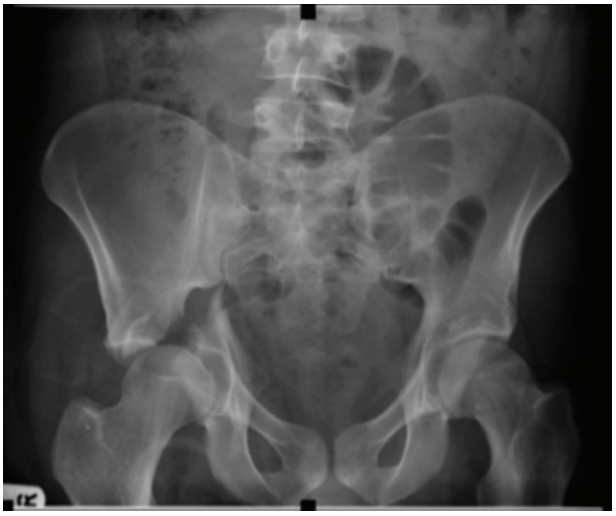
TRANSVERSE FRACTURE

This type of injury is relatively common and occurs with the leg in abduction and internal rotation. There is separation of the anterior and posterior columns into two parts. The innominate bone is divided into a proximal fragment containing the dome and intact ilium, and a distal segment. The femoral head usually follows the distal ischiopubic segment. Always exclude any associated posterior wall fracture (transverse posterior wall), and an exiting fracture line through the obturator foramen (T-shaped). Transverse fractures are associated with sacroiliac joint injuries. The height of the transverse fracture is key as it guides the surgical approach (Figure 29.25). Transverse fractures may be *transtectal*, *juxtatectal* or *infratectal*.

Usually, the transverse fracture is fixed via the Kocher-Langenbeck approach (juxtatectal and infratectal). However, if the fracture level is more superior, the decision must be made whether it is more appropriate to access the fracture via an anterior approach (ilioinguinal or iliofemoral approach).

T-SHAPED FRACTURE

This fracture is a transverse fracture with a second vertical fracture line across the acetabulum (Figure 29.26). In all transverse type fractures, look for a fracture line exiting through the obturator foramen as this indicates the presence of a vertical fracture as well. As in the transverse fracture, there may be an associated sacroiliac joint injury, and look at the height of the transverse fracture before deciding on the surgical approach.



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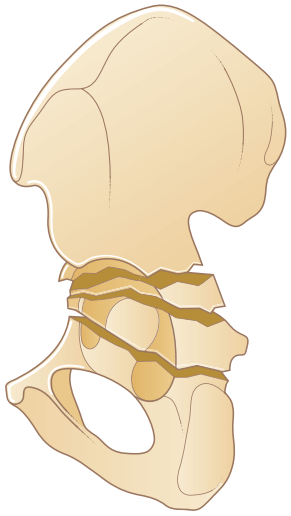
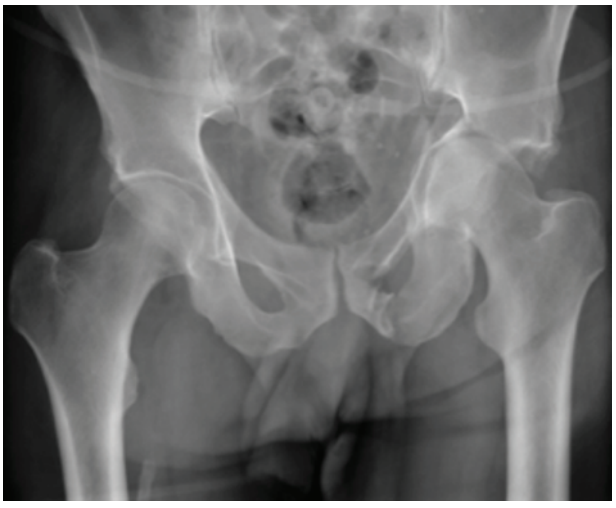


(b)

Figure 29.25 Transverse acetabular fracture (a) The femoral head goes with the ischiopubic segment. (b) I transtectal, II juxtatectal, III infratectal.



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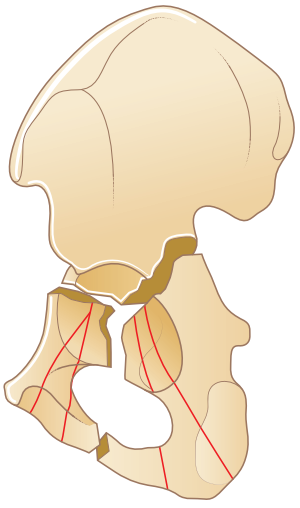


(b)

Figure 29.26 T-shaped acetabular fracture (a) The different configurations of the T-shaped fracture. (b) The height of the transverse element helps to guide the surgical approach.

TRANSVERSE POSTERIOR WALL FRACTURE

This is a transverse fracture with dislocation of the femoral head causing an associated posterior wall fracture (Figure 29.27). It occurs with the leg in flexion and abduction. As in the other transverse type fractures, there may be an associated sacroiliac joint injury. The incidence of sciatic nerve injury in this fracture pattern is very high (70%).

Immediate management is reduction of the femoral head. Operative fixation is via the Kocher-Langenbeck approach.

POSTERIOR WALL POSTERIOR COLUMN FRACTURE

This pattern occurs with the leg in internal rotation. The femoral head is dislocated in this fracture type and the fracture is at the level of the greater sciatic notch. There may be an associated sciatic nerve injury. Fixation is via the Kocher-Langenbeck approach.

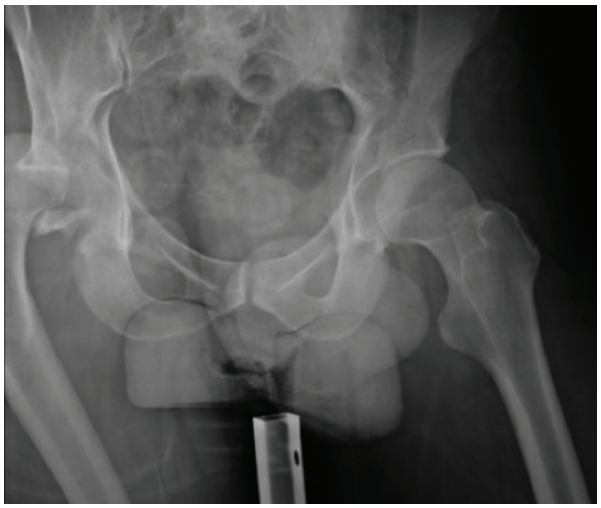


Figure 29.27 Transverse posterior acetabular wall fracture with dislocation of the femoral head

ANTERIOR COLUMN POSTERIOR HEMITRANSVERSE FRACTURE

This is a fracture of the anterior column, associated with a posterior column fracture at the posterior half of the innominate bone (Figure 29.28). This type of fracture is commonly seen in elderly patients. If operative fixation is required, it is performed via the Stoppa or the ilioinguinal approach.

ASSOCIATED BOTH COLUMN FRACTURE

There is complete dissociation of all parts of the acetabulum from the innominate bone (Figure 29.29). X-rays may show the 'spur' sign (intact ilium due to medial migration of the femoral head). These are challenging fractures and may require more than one surgical approach for fixation. Generally, the ilioinguinal approach is preferred. This allows fixation of the fracture fragments that have extended into the ilium.

Management

As with any other periarticular fracture in the body, the goal of treatment is to restore joint congruency, provide fracture stability to allow mobilization, and prevent osteoarthritis.

Undisplaced fractures are usually stable and can be managed conservatively. Bed rest and traction are rarely used as a definitive treatment nowadays (due to problems associated with prolonged immobility). Patients are mobilized with partial weight-bearing on the affected side for 6 weeks. If pain is severe, this indicates a lack of fracture stability. Repeat X-rays

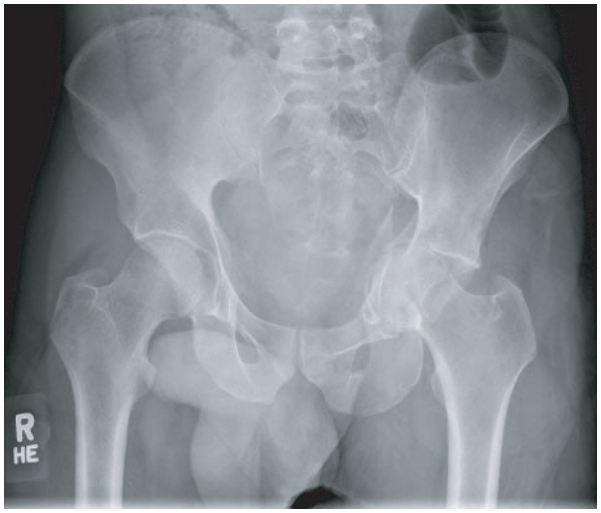


Figure 29.28 Anterior column posterior hemitransverse acetabular fracture

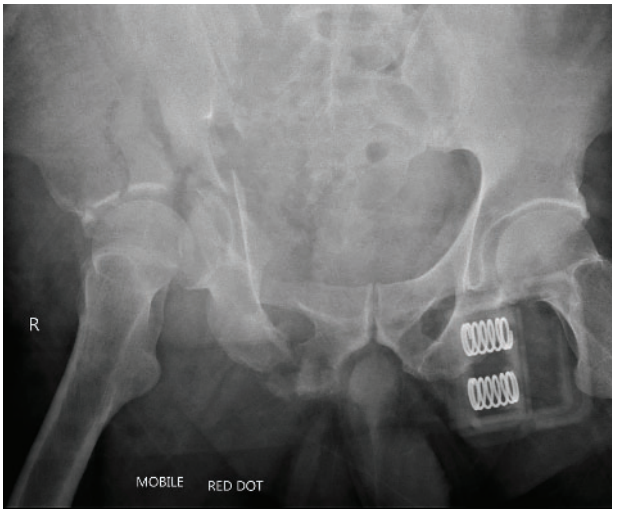


Figure 29.29 Associated both column acetabular fracture No part of the acetabulum is in continuity with the axial skeleton.

should be performed to look for displacement, and operative fixation needs to be considered.

If the hip is dislocated, reduction is urgent, followed by the application of skeletal traction until definitive surgery. This is best achieved via a distal femoral transfixion pin. Fractures with more than 2 mm of displacement of the articular surface should be anatomically reduced and stabilized. Patients with >3 mm of displacement have been shown to have a poor outcome, and patients with <1 mm displacement have been shown to have less progression to osteoarthritis. Articular surface impaction is a particularly poor prognostic factor, particularly if this is not anatomically reduced and supported.

Surgical approaches

The two 'work-horse' approaches in acetabular surgery are the *Kocher-Langenbeck approach* and the *ilioinguinal approach*. The iliofemoral approach is now very rarely used due to the high complication rate of heterotopic ossification, gluteal muscle dysfunction postoperatively and infection. However, in some fractures (high transverse patterns) it provides the best exposure and can still be useful.

ILIOINGUINAL APPROACH

This approach was devised by Letournel. It is performed with the patient supine, and provides access to the entire anterior column, symphysis pubis, quadrilateral plate, ilium and sacroiliac joint (Figure 29.30). There are three windows: the medial, middle and lateral windows. This approach is now used less commonly than previously. The Stoppa approach (medial window of the ilioinguinal approach via a rectus abdominis split) is now generally preferred and access through this approach can be gained right the way back to the sacroiliac joint if required. If there is iliac extension of a fracture, the lateral window of the ilioinguinal approach can also be opened up simultaneously. This less invasive approach has been made possible by specialist pelvic retractors and reduction clamps, specialized contoured pelvic plates which buttress the medial wall of the acetabulum, and specialist operating tables allowing lateral and longitudinal traction to distract the hip joint.

KOCHER-LANGENBECK APPROACH

This is an extension of the posterior approach used in hip arthroplasty. It can be performed in the lateral or prone positions. It provides access to the posterior wall and entire posterior column of the acetabulum (Figure 29.31). The sciatic nerve should be exposed from the greater sciatic notch proximally to beneath the gluteal insertion on the femur distally. Care must be taken not to cauterize branches of the posterior circumflex femoral artery when detaching the short external rotators. The hip joint capsule must be left intact. Lifting the obturator internus muscle away from the bone will expose the ischial tuberosity – the hamstring tendons provide a useful marker of the distal extension of the approach. Proximal exposure involves dissecting the gluteus medius and minimus muscles away from the bone. Care must also be taken not to place the superior gluteal neurovascular bundle under too much traction as this will lead to nerve palsy. Utilizing the trochanteric 'flip' where the abductors are detached with a sliver of bone is kinder to these muscles in well-built individuals rather than fighting them through the procedure with retractors. This technique also allows a surgical dislocation of the hip joint.

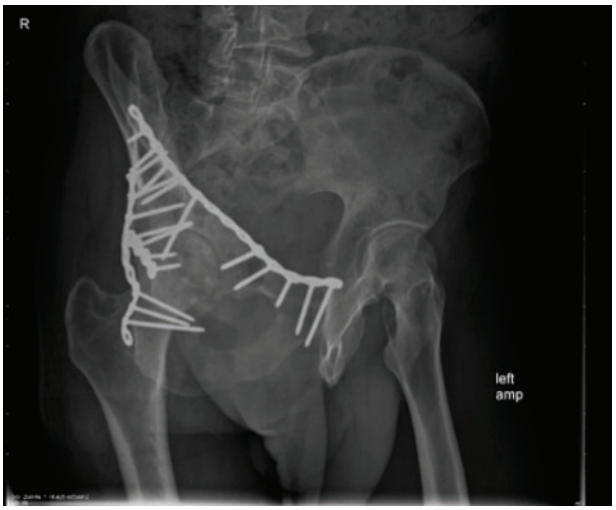


Figure 29.30 Two-column plating This obturator oblique view shows a plate inserted via the ilioinguinal approach, spanning the entire anterior column of the acetabulum.

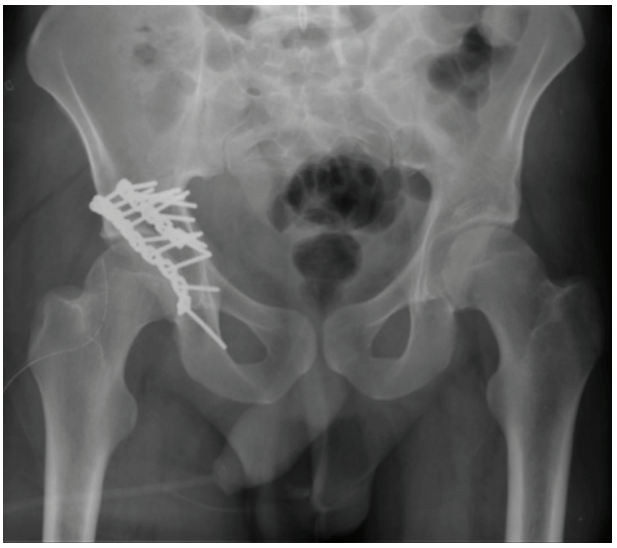


Figure 29.31 Posterior column plating Surgery performed via the Kocher-Langenbeck approach.

Special cases

ELDERLY PATIENTS

The incidence of acetabular fractures in elderly patients with osteoporosis is increasing. These patients present a challenge in the decision-making process. The bone quality is generally too poor to provide a hold for adequate fixation with standard techniques (Figure 29.32). The fracture pattern is frequently an associated pattern (most commonly an anterior column / posterior hemitransverse fracture). There is no clear evidence as to how best to manage these patients. They do not tolerate bed rest and traction well and are particularly vulnerable to venous thromboembolic disease, chest sepsis and pressure sores.

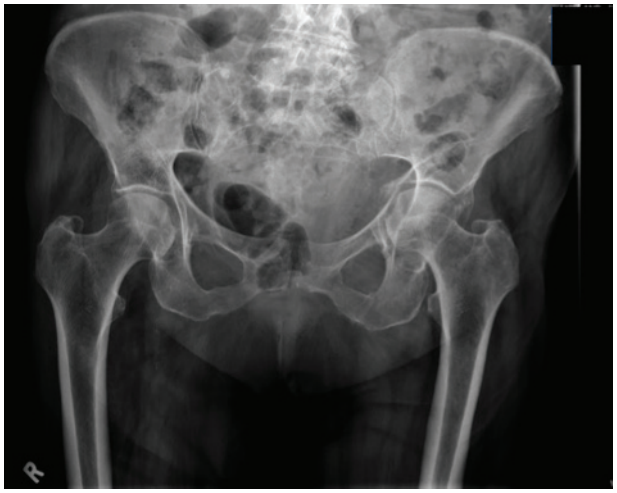


Figure 29.32 Anterior column posterior hemitransverse fracture

Conservative management in these patients is early mobilization, weight-bearing as tolerated. If the patients are in too much pain for conservative treatment, or the fracture is so displaced it is unlikely to heal, then surgery is required. In the very frail patient unlikely to tolerate the demands of major surgery, an excision arthroplasty for pain relief may be appropriate. Surgical reconstruction consists of combined fixation and arthroplasty, the so-called 'fix and replace' (Figure 29.33). Fixation is usually via a dual approach; depending on the fracture pattern, the Stoppa approach is used to stabilize the anterior column, and the Kocher-Langenbeck is used to stabilize the posterior column and perform hip arthroplasty (using a revision-type uncemented acetabular prosthesis). This surgery is a major undertaking; close liaison with the anaesthetist is required so they are aware of the extent of the surgery and are suitably prepared.

PELVIC DISCONTINUITY

The number of patients with a total hip arthroplasty *in situ* increases year on year. Pelvic discontinuity may occur where there is prosthetic loosening of the acetabular component with a fracture. This can occur either acutely due to trauma to an already loose acetabular component or as part of a chronic process due to progressive loosening and bone loss. Acute pelvic discontinuity is a challenging situation as there is frequently major bone loss in association with instability of the columns. Acute discontinuity may be seen following a fall (periprosthetic acetabular fracture), when there has been sudden bone failure (a fragility-type fracture), or where there has been an iatrogenic injury at the time of acetabular prosthesis implantation (Figure 29.34).

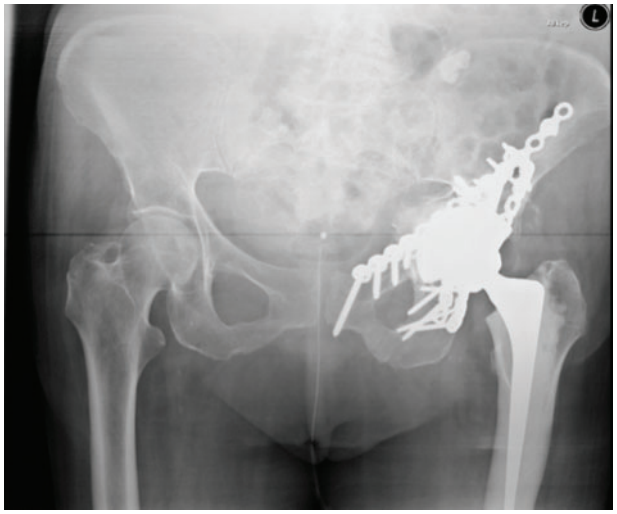


Figure 29.33 Fix and replace Acetabular reconstruction consisting of bi-column plating with combined hybrid total hip replacement.

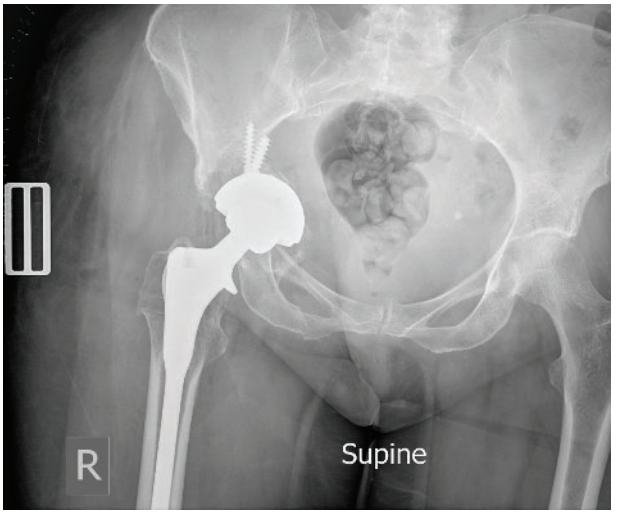


Figure 29.34 Pelvic discontinuity This example occurred at the time of acetabular component insertion during primary total hip replacement.

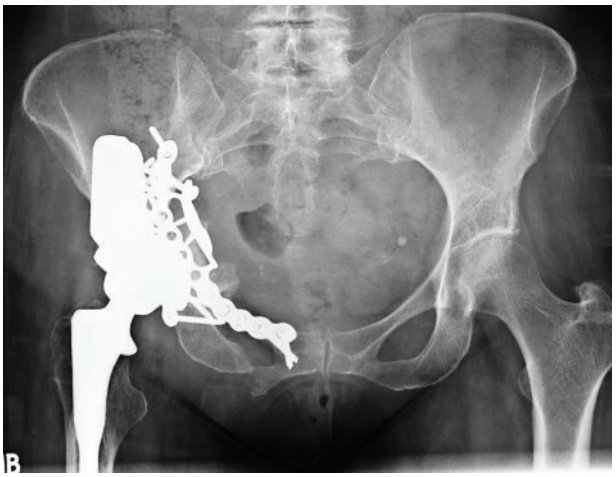


Figure 29.35 Reconstruction for pelvic discontinuity
Postoperative X-ray of case seen in Figure 29.34.

If there is displacement, and the prosthesis is loose (which is usually the case), surgical reconstruction is required; this consists of column stabilization with prosthetic revision. The principles of reconstruction are to decide whether one or both columns need stabilization. If the posterior column only is involved, then the reconstruction can be performed with posterior column plating and arthroplasty. If the anterior column is also involved, it needs plating first. Once column stabilization has been performed, deficiency needs to be assessed and can be managed with either bone grafting or augmentation, or both, before final insertion of the uncemented revision acetabular shell (Figure 29.35).

Complications

Possible complications are the same as in pelvic ring injuries with the following additions.

Heterotopic ossification This is common following the Kocher-Langenbeck approach. Any devitalized muscle should be excised, and patients are placed on prophylactic measures such as non-steroidal anti-inflammatory drugs to avoid this.

Avascular necrosis If the femoral head has been dislocated, the blood supply to it may have been damaged. Serial follow-up in these cases is necessary to monitor progression to avascular necrosis.

Hip abductor dysfunction This may occur following the Kocher-Langenbeck approach if there has been significant stretching of the superior gluteal nerve, or there may have been nerve damage at the time of injury.

Osteoarthritis This is related to the severity of the injury, the quality of reduction and the age of the patient. Patients with imperfect reductions have a higher risk of the development of arthritis, as do patients over the age of 50 years.

INJURIES TO THE SACRUM AND COCCYX

A blow from behind, or a fall onto the 'tail' may fracture the sacrum or coccyx, or sprain the joint between them. Women seem to be affected more commonly than men.

Bruising is considerable and tenderness is elicited when the sacrum or coccyx is palpated from behind or per rectum. Sensation may be lost over the distribution of sacral nerves.

X-rays may show: (1) a transverse fracture of the sacrum, in rare cases with the lower fragment pushed forwards; (2) a fractured coccyx, sometimes with the lower fragment angulated forwards; or (3) a normal appearance if the injury was merely a sprained sacrococcygeal joint.

Treatment If the fracture is displaced, reduction is worth attempting. The lower fragment may be pushed backwards by a finger in the rectum. The reduction is stable, which is fortunate. The patient is allowed to resume normal activity but is advised to use a rubber ring cushion when sitting. Occasionally, sacral fractures are associated with urinary problems, necessitating sacral laminectomy.

Persistent pain, especially on sitting, is common after coccygeal injuries. If the pain is not relieved by the use of a cushion or by the injection of local anaesthetic into the tender area, excision of the coccyx may be considered.

FURTHER READING

Dalal SA, Burgess AR, Siegel JH, *et al.* Pelvic fracture in multiple trauma. *J Trauma* 1989; **29**: 981–1000.

Giannoudis PV, Grotz MRW, Papakostidis C, Dinopoulos H. Operative treatment of displaced fractures of the acetabulum: a meta-analysis. *J Bone Joint Surg (Br)* 2005; **87B**: 2–9.

Hak DJ, Smith WR, Suzuki T. Management of haemorrhage in life-threatening pelvic fracture. *J Am Acad Orthop Surg* 2009; **17**: 447–57.

Letournel E. Acetabulum fractures: classification and management. *Clin Orthop* 1980; **151**: 81–106.

Letournel E, Judet R. *Fractures of the Acetabulum*, 2nd edn. Berlin: Springer Verlag, 1993, pp. 29–62.

Matta JM. Fractures of the acetabulum: accuracy of reduction and clinical results in patients managed operatively within three weeks after the injury. *J Bone Joint Surg (Am)* 1996; **78A**: 1632–45.

Matta JM, Merritt PO. Displaced acetabular fractures. *Clin Orthop Relat Res* 1988; **230**: 83–97.

Montgomery KD, Geerts WH, Potter HG, Helfet DL. Thromboembolic complications in patients with pelvic trauma. *Clin Orthop Relat Res* 1996; **329**: 68–87.

Tile M, Helfet DL, Kellam JF, Vrahas M. Fractures of the Pelvis and Acetabulum: principles and methods of management, 4th edn. Stuttgart: Thieme Publishing Group. 2015.

Young JWR, Burgess AR. *Radiologic Management of Pelvic Ring Fractures: Systematic Radiographic Diagnosis.* Baltimore: Urban and Schwarzenberg, 1987.

Young JWR, Burgess AR, Brumback RJ, Poka A. Lateral compression fractures of the pelvis: the importance of plain radiographs in the diagnosis and surgical management. *Skeletal Radiol* 1986; **15**: 103–9.

Injuries of the hip and femur

30

Richard Baker & Michael Whitehouse

DISLOCATION OF THE HIP

Hip dislocations are associated with high-energy trauma in the majority of cases. They often happen in conjunction with fractures of the femur or the polytraumatized patient (40–75%).

Usually great forces (>400 N, the force required simply to distract the femoral head from the acetabulum) are required to dislocate the hip joint, as it is well constrained by its bony and soft-tissue anatomy. The position of the femur at the time of trauma dictates the pattern of dislocation.

Hip dislocations are classified according to the direction of the femoral head displacement: *posterior* (the commonest type – 80% of cases), *anterior* and *central* (a comminuted or displaced fracture-dislocation through the floor of the acetabulum).

Posterior dislocation

Mechanism of injury

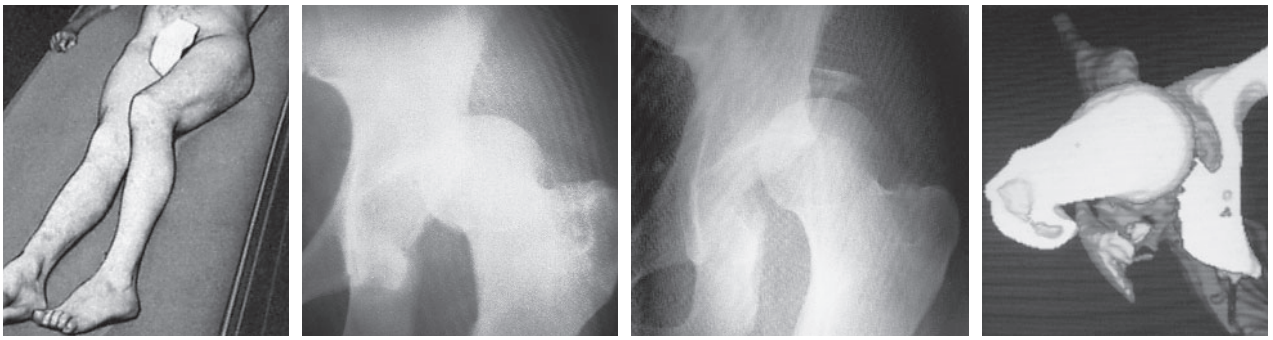
A posterior dislocation usually occurs when force is applied to the knee, commonly in a seated car passenger when the knee strikes the dashboard. The femur is thrust proximally and the femoral head is forced posteriorly; often a piece of bone from the acetabulum (usually the posterior wall) is sheared off, making a fracture-dislocation.

Clinical features

In an isolated posterior hip dislocation the leg is shortened and lies in an adducted, internally rotated and slightly flexed position (Figure 30.1). *In the presence of an ipsilateral femoral fracture the position may not be typical.* A high index of suspicion for this injury should be maintained and clinical and radiological screening performed in at-risk cases. The knee should be examined for bruising anteriorly and ligamentous injuries, and the neurovascular status of the limb determined. The sciatic nerve is particularly at risk.

Imaging

In the anteroposterior view *X-ray*, the femoral head is seen to be high-riding and smaller than expected when compared to the contralateral normal hip. Associated femoral head and posterior wall acetabular fractures may be evident.



(a) (b) (c) (d)

Figure 30.1 Posterior dislocation of the hip (a) This is the typical posture in a patient with a posterior hip dislocation: the left hip is slightly flexed and internally rotated. (b) The X-ray in this case demonstrates a simple dislocation, with the femoral head lying above and behind the acetabulum. (c) Dislocation and an associated acetabular rim fracture. CT scan helps delineates the acetabular fracture (d).

Impaction of the femoral head articular surface is best appreciated with a *CT scan*, as are acetabular fractures and loose bony fragments, but detailed imaging at this stage should be undertaken only if it does not delay reduction of the hip dislocation.

The *Thompson and Epstein classification* is helpful in planning treatment:

- *Type I* – dislocation with no more than minor chip fractures
- *Type II* – dislocation with single large fragment of posterior acetabular wall
- *Type III* – dislocation with comminuted fragments of posterior acetabular wall
- *Type IV* – dislocation with fracture through acetabular floor
- *Type V* – dislocation with fracture through acetabular floor and femoral head.

Types I and II are relatively simple dislocations; these are associated with either minor chip fractures (small fragments of the acetabular wall or fovea centralis) or a single large fragment from the posterior acetabular wall (type II). In type III, the posterior wall is comminuted. Type IV has an associated fracture of the acetabular floor, and type V an associated fracture of the femoral head, which can be further subdivided according to the Pipkin classification (see Figure 30.2).

Treatment

The dislocation must be reduced as soon as possible; rates of osteonecrosis and subsequent osteoarthritis are increased if the hip is dislocated for more than 6 hours. This requires sedation or general anaesthesia and is performed closed in the majority of cases. Closed reduction should not be attempted if there is an associated femoral neck fracture, to prevent further displacement of the femoral neck supply and disruption of the blood supply to the femoral head.

To achieve closed reduction, an assistant steadies the pelvis; the surgeon starts by applying traction in the line of the femur as it lies (usually in adduction and internal rotation) with the knee flexed at 90 degrees, and then gradually flexes the patient's hip, maintaining traction, followed by internal rotation and adduction to achieve reduction. In some cases hip flexion beyond 90 degrees may be required. Another assistant can help by applying direct medial and anterior pressure to the femoral head through the buttock. A satisfying 'clunk' indicates that reduction has been achieved.

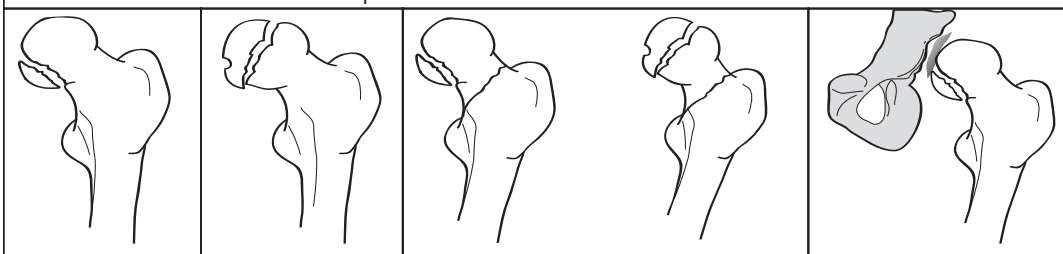
Stability is then tested of the reduced hip. This is performed by flexing the hip to 90 degrees and applying a longitudinal and posteriorly directed force while the hip is screened on an image-intensifier looking for signs of subluxation. Evidence of subluxation or associated significant fractures would prompt surgical repair; if definitive surgery is to be delayed, skeletal traction should be considered. When required, this is usually achieved through a distal femoral traction pin.

Regardless of the findings at this stage, post-reduction X-rays and/or CT scan should be performed to determine the quality of the reduction and the presence or absence of retained fragments in the joint.

POST-REDUCTION MANAGEMENT

In the hip with an isolated dislocation, following reduction, the patient can weight-bear as tolerated protected with the aid of crutches. This is also the case in stable hips that are congruent following reduction with fractures that are to be managed conservatively. It has been previously thought that non-weight-bearing should be considered to decrease the risk of osteonecrosis of the femoral head but there is little evidence to support

Pipkin classification of femoral head fractures



Type I	Type II	Type III	Type IV
The fracture line is inferior to the fovea	The fracture fragment includes the fovea	As with types I and II but with an associated femoral neck fracture	Any pattern of femoral head fracture and an acetabular fracture (coincides with Thompson and Epstein's type V)

Figure 30.2 Pipkin classification of femoral head fractures

this – unless protected weight-bearing is required for management of associated fractures.

If the post-reduction X-rays or CT scans show the presence of intra-articular bony fragments, unstable fractures, loss of congruity in the weight-bearing portion of the hip or articular surface impaction, an open procedure should be planned. If open reduction is required, the approach is planned on the basis of the fracture pattern and approaches that may be required for subsequent surgery; the operation is not an emergency and can be done once the patient's condition has stabilized or appropriate surgical expertise is available. Traction, which is usually achieved by means of skeletal traction applied via a distal femoral transfixion pin, can be applied until conditions are appropriate for surgery.

Thompson and Epstein type II or greater injuries usually require open reduction and anatomical fixation. For instance, fixation of the detached posterior wall fragment in a type II fracture will restore joint stability and joint congruency. Type III injuries are treated closed, but there are often retained fragments that require removal at operation. Fixation of comminuted posterior fragments may be achieved with spring plates or similar techniques; persistent instability may require delayed reconstruction.

Type IV and V hip dislocations are treated initially by closed reduction. The indications for surgery follow the principles outlined above, prompted by instability, retained fragments or joint incongruity. In type V injuries, a femoral head fragment may fall into place during hip reduction and this can be confirmed by post-reduction CT. If the fragment remains unreduced, operative treatment is indicated: a small fragment can simply be removed, but a large fragment should be replaced; the joint is opened, the femoral head dislocated and the fragment fixed in position with a countersunk screws. Postoperatively, weight-bearing is dictated by the fracture pattern and its stability – progression from non-weight-bearing to weight-bearing occurs over a course of 6–12 weeks.

Complications

EARLY

Sciatic nerve injury The sciatic nerve is damaged in 10–20% of cases. *Nerve function must be tested and documented before reduction of the hip is attempted.* If, after reducing the dislocation, a new onset sciatic nerve lesion is diagnosed, the nerve should be explored to ensure it has not been trapped by the reduction manoeuvre. If a foot drop occurs, the ankle is splinted in a neutral position to prevent an equinus deformity and in order to aid mobilization.

Vascular injury Occasionally the superior gluteal artery is torn and bleeding may be profuse. If this is suspected, an arteriogram should be performed. The torn vessel may need to be ligated or controlled by radiologically guided embolization.

Associated fractured femoral shaft When this occurs concurrently with a hip dislocation, the hip dislocation can be easily missed unless a high index of suspicion to look for one is employed. In the case of all femoral shaft fractures, proximal injury to the hip needs to be excluded clinically and radiographically. Closed reduction of the dislocation will be much more difficult in this situation. A prompt open reduction of the hip followed by internal fixation of the shaft fracture should be undertaken.

LATE

Osteonecrosis of the femoral head This has been reported in up to 20% of traumatic posterior hip dislocations; if reduction is delayed by more than 12 hours, the figure rises to over 40%. X-ray features, such as increased density of the femoral head, may not be seen for 6 weeks or more, and sometimes very much later (up to 2 years), depending on the rate of bone repair. Changes are seen earlier on MRI scans. Ischaemia occurs due to interruption of femoral head blood supply when the hip is dislocated. There is evidence to suggest that this results from compression, traction and arterial spasm rather than actual disruption of blood vessels, which explains why the consequences of ischaemia are proportional to the delay in reduction of the hip. Blood flow is restored on reduction of the hip, especially if this is performed early, which highlights the need for emergency treatment with a target of less than 12 hours (preferably less than 6 hours) from the time of injury.

Myositis ossificans This is an uncommon complication, probably related to the severity of the soft-tissue injury.

Unreduced dislocation After a few weeks an untreated dislocation can seldom be reduced by closed manipulation and open reduction is needed. The incidence of stiffness or osteonecrosis is considerably increased and the patient may later need reconstructive surgery.

Secondary osteoarthritis This is common and is due to: (1) cartilage damage at the time of the dislocation; (2) the presence of retained fragments in the joint (3rd body wear); and (3) osteonecrosis of the femoral head. In young patients treatment presents a difficult problem but this is now usually treated with arthroplasty when end-stage osteoarthritis occurs.

Recurrent instablility This is uncommon but it may require stabilization or delayed reconstruction when it does occur.

Femoral head fracture

Femoral head fractures are always seen in association with a dislocation or subluxation of the femoral head. In some cases the femoral head may have spontaneously reduced into the acetabulum, but this is unusual. The majority of femoral head fractures occur in posterior hip dislocations due to their greater frequency than anterior hip dislocations, although, if an anterior dislocation occurs, it is more likely to lead to a femoral head fracture.

Acetabular, femoral shaft and femoral neck fractures are often associated with femoral head fractures as are ligamentous injuries to the knee and sciatic nerve injuries.

Classification

The classic classification is by Pipkin, where the femoral head fracture patterns are described in four groups – all associated with a posterior hip dislocation (see Figure 30.2).

Treatment

A dislocated hip should be reduced regardless of the presence of a femoral head fracture. However, if a femoral neck fracture is present, it must be treated before any attempt at reduction of the hip.

Treatment decisions can be based on the *Pipkin classification* and other concomitant injuries.

- *Type 1* – The fragment should be excised if small or fixed if large.
- *Type 2* – This must be treated with open reduction and internal fixation.
- *Type 3* – The femoral neck fracture must be stabilized first, before any attempt to reduce and fix the dislocated femoral head.
- *Type 4* – The femoral head fracture should be fixed if large enough.

Complications

Complications following a femoral head fracture include: *osteoarthritis* in approximately 50%, *osteonecrosis* in approximately 20%, *sciatic nerve palsy*, fracture *malreduction*, *non-union* and *heterotropic ossification*.

Anterior dislocation

Anterior hip dislocations are rare, accounting for only approximately 10–20% of all hip dislocations. The usual mechanism is a high-energy accident. Anterior dislocations are sub-classified into type I – *pubic (superior)* or type II – *obturator (inferior)* injuries. Anterior dislocations are more commonly associated with femoral head fractures.

Clinical features

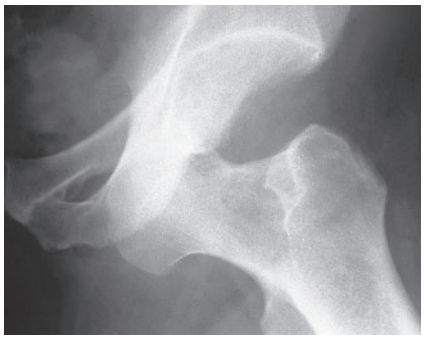
The leg lies externally rotated, abducted and slightly flexed. It is not typically short, because the attachment of rectus femoris prevents the head from migrating proximally by a significant amount. Occasionally the leg is abducted almost to a right angle. Seen from the side, the anterior bulge of the dislocated head is unmistakable when the patient's body habitus permits visualization, especially when the head has moved anteriorly and superiorly. The prominent head may be easy to feel, either anteriorly (superior type) or in the groin (inferior type). Hip movements are generally impossible (Figure 30.3).

X-rays

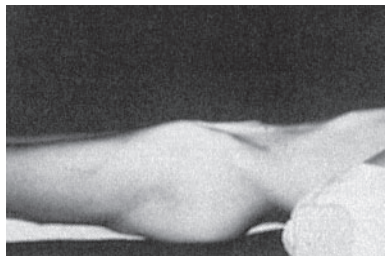
In the anteroposterior view the dislocation is usually obvious, but occasionally the head is almost directly in front of its normal position. The femoral will appear larger than the unaffected contralateral side. A lateral film helps confirm the diagnosis.

Treatment and complications

Reduction of the anteriorly dislocated hip is more difficult than a posteriorly dislocated hip. It should still be performed as soon as possible under sedation or general anaesthesia; if repeated attempts are required, open reduction should be considered. The closed reduction

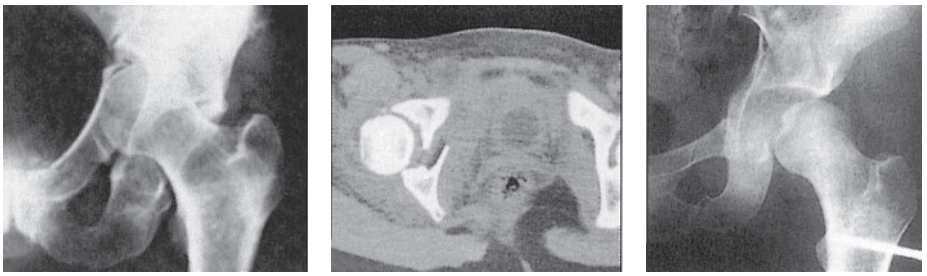


(a)



(b)

Figure 30.3 Anterior hip dislocation (a,b) The usual appearance of an anterior dislocation: the hip is only slightly abducted and the head shows clinically as a prominent lump.



(a) (b) (c)

Figure 30.4 Central dislocation (a) The plain X-ray of the central displacement; (b) a CT scan shows the pelvic injury more clearly. (c) Skeletal traction applied to the greater trochanter.

manoeuvre is as follows: the affected leg is held in external rotation, abduction and flexion, before longitudinal traction is applied. The leg is gently internally and externally rotated until the hip reduces – pressure anteriorly over the palpable femoral head may assist the reduction. The reduction is usually obvious and accompanied by a palpable or even audible clunk.

The subsequent treatment is similar to that employed for a posterior dislocation.

Central dislocation

A fall on the side, or a blow over the greater trochanter, may force the femoral head medially through the floor of the acetabulum. Although this is called a 'central dislocation', it is really a fracture of the acetabulum (Figure 30.4). The condition is dealt with in Chapter 29 'Injuries of the pelvis'. Distal femoral traction is applied first; if the reduction of the hip joint remains inadequate, a lateral trochanteric pin may also be required.

HIP FRACTURES

Hip fractures are defined as fractures that occur between the articular margin of the femoral head to 5 cm below the lesser trochanter. They are subdivided into *intracapsular* and *extracapsular* fractures. The blood supply to the femoral head is typically damaged in intracapsular fractures and rarely in extracapsular fractures. Extracapsular fractures are further subdivided into *pertrochanteric* (including the reverse oblique type) and *subtrochanteric* fractures.

Hip fractures typically occur in the elderly patient from low-energy falls from standing height and are secondary to osteoporosis. There is a significant risk of mortality and morbidity post injury; according to the National Hip Fracture Database England, Wales and Northern Ireland in 2015, 7.5% of patients will die within 1 month of hip fracture rising to 24% within 12 months. Around 4% of elderly patients sustain another fracture at the time of the index fall, most commonly of the wrist or proximal humerus. Younger patients suffer from femoral neck fractures secondary to high-energy injuries or underlying conditions which affect bone health and the risk of falls (e.g. alcoholism).

Hip fracture rates are highest in Northern and Central Europe, moderate in North America, Japan and Oceania and at the lowest in south Asia and Africa. For each decade after 50 the risk of hip fracture doubles and the worldwide incidence is predicted to rise from 1.7 million in 1990 to 6.3 million in 2050.

The very large economic impact of treating, rehabilitating and caring for this group of patients is increasingly being recognized, with many governments and healthcare administrations focusing on preventive strategies.

Mechanism of injury

In the elderly patient the fracture usually results from a simple fall from standing height. In severe osteoporosis a fracture may occur from simple twisting moments and it is the hip fracture itself which causes the reported fall. In typical fractures, patients rarely have preceding symptoms. Patients with prodromal symptoms may have a stress fracture through the femoral neck. In younger individuals there is usually a high-energy mechanism and the patient must be screened for associated injuries.

Pathological anatomy and classification

INTRACAPSULAR HIP FRACTURES

The most commonly used classification is the *Garden classification*, which is based on the amount of displacement apparent on X-rays of the hip (Figure 30.5). Once fractured, the head and neck become displaced in progressively severe stages:

- *Stage I* – an incomplete impacted fracture, including the so-called 'abduction fracture' in which the femoral head is tilted into valgus in relation to the neck
- *Stage II* – a complete but undisplaced fracture
- *Stage III* – a complete fracture with moderate displacement
- *Stage IV* – a severely displaced fracture.

Basicervical fractures occur at the very distal extent of the femoral neck and anatomically are intracapsular injuries although due to the relative stability of the fracture they behave and are treated like intertrochanteric fractures.

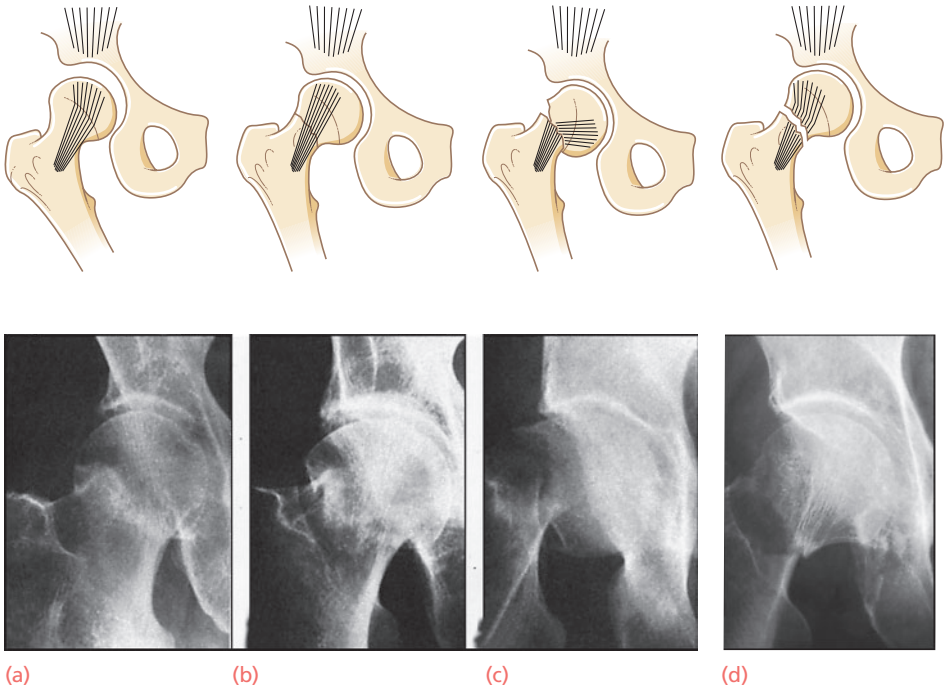


Figure 30.5 Garden classification of femoral neck fractures (a) *Stage I:* incomplete (so-called abducted or impacted hip fracture) – the femoral head in this case is in slight valgus. (b) *Stage II:* complete without displacement. (c) *Stage III:* complete with partial displacement – the fragments are still connected by the posterior retinacular attachment; the femoral head trabeculae are no longer in line with those of the innominate bone. (d) *Stage IV:* complete with full displacement – the proximal fragment is free and lies correctly in the acetabulum so that the trabeculae appear normally aligned with those of the innominate bone.

EXTRACAPSULAR HIP FRACTURES

Intertrochanteric fractures These are divided into stable and unstable varieties. In essence, *unstable fractures* are those where:

- poor contact exists between the fracture fragments, as in four-part intertrochanteric types (greater and lesser trochanter, proximal and distal femoral fragments), or if the posteromedial cortex is comminuted
- the fracture pattern is such that forces of weight-bearing continually displace the fragments further, as in those with a reverse oblique pattern or with a subtrochanteric extension.

The importance of the fracture pattern is detailed in the *Kyle classification*, which distinguishes four basic patterns that reflect increasing instability and increasing difficulty of reduction and fixation (Figure 30.6).

Reverse oblique intertrochanteric fractures These are a particularly unstable variant of this fracture pattern. As the plane of the fracture line is different from the usual pattern, the support offered by the medial column is lost. If a sliding hip screw is used, the screw is parallel to the fracture line rather than perpendicular to it, hence compression of the fracture is not achieved, shear forces across the fracture line are not addressed and the chance

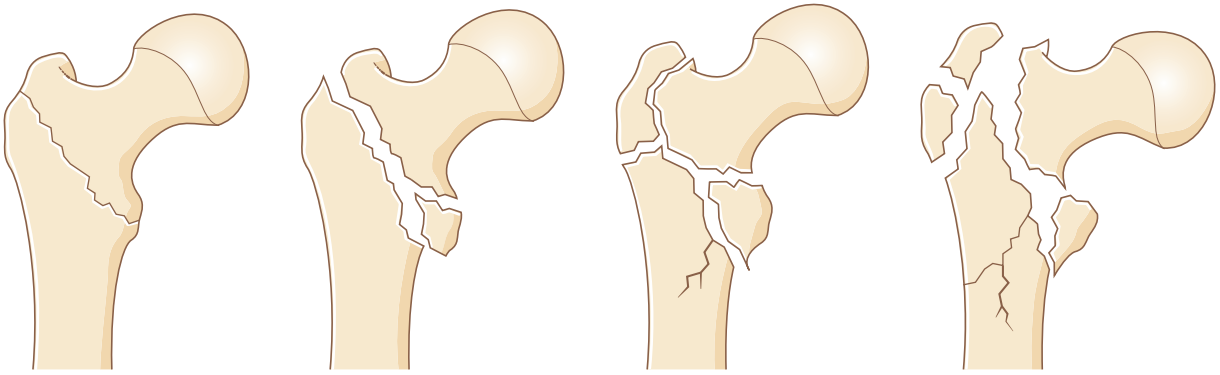


Figure 30.6 Intertrochanteric fractures – Kyle classification Types 1 to 4 are arranged in increasing degrees of instability and complexity. Types 1 and 2 account for the majority (nearly 60%). The reverse oblique type of intertrochanteric fracture represents a subgroup of type 4; it causes similar difficulties with fixation.

of failure is increased. Intramedullary devices are preferred for this fracture pattern.

Subtrochanteric hip fractures These fractures occur between the inferior margin of the lesser trochanter and 5 cm below this point. Fractures more distal than this are considered to be *femoral shaft fractures*. Fractures in this area are rare in young adults due to strength of the posteromedial calcar femorale, a thick area of cortical bone. The elderly patient is at higher risk of fracture in this area due to mechanisms that weaken the bone such as osteoporosis, osteomalacia, Paget's disease, metastatic deposits or the atypical changes seen in association with bisphosphonate use.

The fracture pattern is typically transverse or oblique and the proximal fragment sits in a flexed posture with posterior and external rotational displacement of the femoral shaft. A high index of suspicion should be maintained for such fractures being pathological. The pattern is becoming more frequent with the increased use of bisphosphonates. In this subgroup, there is often evidence of cortical thickening and beaking at the site of the fracture (Figure 30.7). In patients who present with a subtrochanteric fracture and this pattern, a similar pattern is often apparent on the contralateral side. In association with pain, this indicates an impending fracture and the surgeon should consider prophylactic fixation of the contralateral side. Due to the abnormal bone in the region of these atypical fractures, correction of the deformity and anatomical reduction are crucial in achieving satisfactory healing. This may require osteotomy to correct the plastic deformation and to excise the pathological bone, allowing direct compression of healthy bone at the time of fixation.

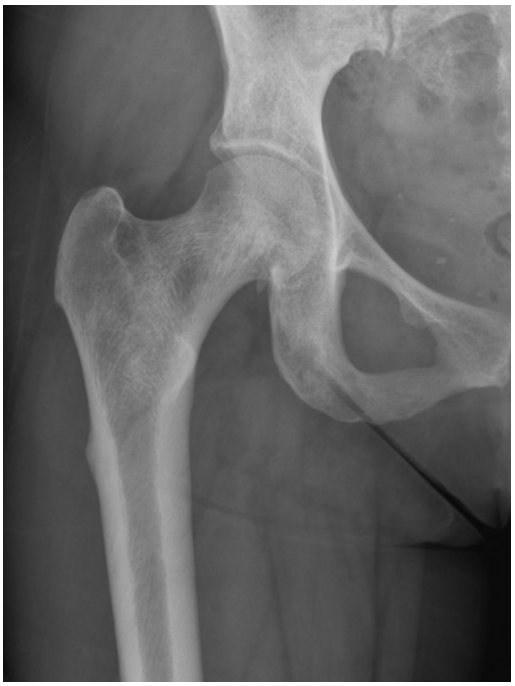


Figure 30.7 Subtrochanteric fracture The typical 'beaking' seen at the site of an impending bisphosphonate-mediated subtrochanteric fracture.

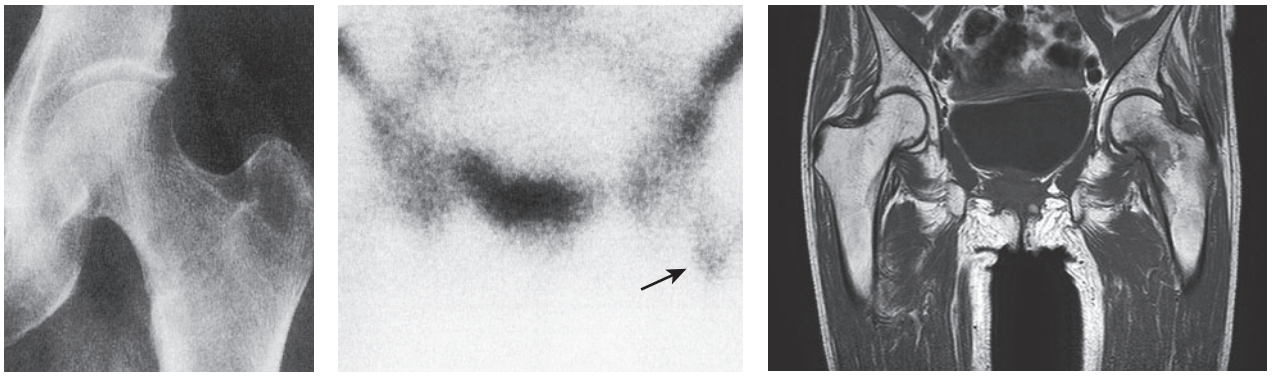
Clinical features

There is usually a history of a fall, followed by pain in the hip and an inability to walk. In any elderly patient who cannot mobilize with hip pain, a hip fracture should be ruled out. If the fracture is displaced, the leg is externally rotated and shortened. In subtrochanteric fractures, the leg lies in neutral or external rotation and looks short; the thigh is often markedly swollen. With an impacted intracapsular fracture the patient may still be able to walk, and patients who are debilitated or have dementia may not complain at all. In contrast, femoral neck fractures in young adults result from road traffic accidents or falls from heights and are often associated with multiple injuries. Young adults with severe injuries such as femoral shaft fractures, whether they complain of hip pain or not, should always be examined for an associated femoral neck fractures. Around 1–9% of femoral fractures have a concomitant ipsilateral femoral neck fracture and the injury is frequently missed at initial presentation.

Diagnosis

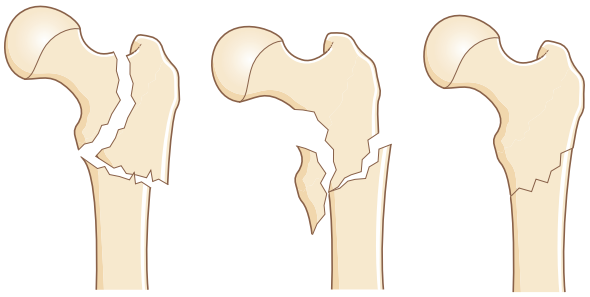
Simple *X-rays* (anteroposterior and lateral) of the hip diagnose fracture in the majority of cases. If there is suspicion of a pathological lesion, views of the whole femur must be obtained. In patients with pain in the hip and an inability to weight-bear with no fracture apparent on plain X-rays, further investigation is required (Figure 30.8). *MRI* is the most sensitive and specific to diagnose an occult hip fracture, although *CT* is often readily accessible and sufficient to detect most fractures of the femoral neck or elsewhere in the pelvic ring. If MRI is not available rapidly, CT is preferred. Occult fractures are detected in 50% of patients with normal X-rays and ongoing pain that prevents weight-bearing with additional imaging. Bone scans could be considered if MRI or CT scans are not available.

In subtrochanteric fractures, the fracture is through or below the lesser trochanter. It may be transverse, oblique or spiral, and it is frequently comminuted. The upper fragment is flexed and appears deceptively short; the shaft is adducted and is displaced proximally. Three important features should be looked for, as the presence of any one will influence treatment: (1) an unusually long fracture line extending proximally towards the greater trochanter and piriform fossa; (2) a large, displaced fragment which includes the lesser trochanter which may compromise stability if the wrong implant is selected; and (3) lytic lesions in the femur (Figure 30.9).



(a) (b) (c)

Figure 30.8 Fractures of the femoral neck – diagnosis (a) An elderly woman tripped on the pavement and complained of pain in the left hip. The plain X-ray showed no abnormality. Two weeks later she was still in pain and unable to fully weight-bear through the affected leg; (b) a bone scan showed a 'hot' area medially at the base of the femoral neck. (c) MRI of occult femoral neck fracture.



(a) (b) (c)

Figure 30.9 Subtrochanteric fractures of the femur – warning signs on the X-ray X-ray findings that should caution the surgeon: (a) comminution, with extension into the piriform fossa; (b) displacement of a medial fragment including the lesser trochanter; and (c) lytic lesions in the femur.

Treatment

Initial treatment consists of pain-relieving measures including analgesia and a femoral nerve block. Medical comorbidities that are amenable to stabilisation in the short term (e.g. cardiac ischaemia and tachycardias) are addressed and the patient optimized for theatre. Investigations such as echocardiography are rarely justified unless they will directly and immediately influence management. Most of the findings of such investigations are not amenable to preoperative optimization and simply lead to unacceptable delay to theatre with an attendant increase in the risk of mortality for the patient.

It is rare that injuries to the neck of the femur are open. Skin traction, or the application of a Thomas splint, can be useful in subtrochanteric fractures as an adjunct to pain relief and nursing care.

Non-operative treatment of hip fractures is limited to those patients who would not survive the surgical intervention and, in these cases, pain relief should be attempted by regional blockade. Even in patients considered high risk for surgery, surgical treatment of a hip fracture should be considered a useful palliative procedure to achieve pain relief and allow effective nursing care.

INTRACAPSULAR HIP FRACTURES

In the young patient every effort should be made to preserve the femoral head and fix the fracture; in contrast, reduction and fixation of fractures in the elderly patient is not recommended due to the associated high rates of subsequent re-operation (46%), non-union (30%) and osteonecrosis of the femoral head (14%). In the elderly, only in the truly undisplaced (on both anteroposterior and lateral X-rays) and stable fracture patterns without comminution should this be considered; this is a very rare occurrence.

In the young patient with a displaced femoral fracture neck fracture, surgery should be performed urgently to decrease the risk of subsequent osteonecrosis. Evidence for the use of a capsulotomy or aspiration of the hip to relieve the intracapsular pressure at the time of fixation is controversial. There is little evidence of benefit for the procedure but it is associated with a low risk of harm and therefore frequently performed at the time of surgery. Undisplaced fractures can be treated less urgently.

Internal fixation An anatomical reduction is mandatory as a fracture fixed in a displaced position is at high risk of failure (Figure 30.10). If screw fixation of an intracapsular hip fracture is performed, a minimum of three screws placed parallel to each other should be used (Figure 30.11). Cannulated screws are preferred due to the ease of the surgical technique, which can also be performed percutaneously under fluoroscopic guidance. Screws are placed from the lateral femoral cortex to the subchondral bone in the femoral

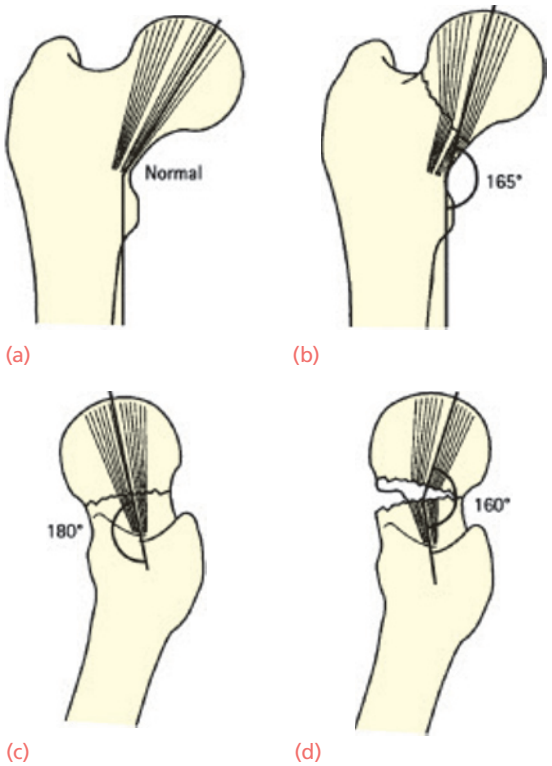


Figure 30.10 Assessing reduction On the antero-posterior X-ray (a,b) the medial femoral shaft and the axis of trabecular markings over the medial aspect of the femoral neck lie at an angle of 160°; an acceptable reduction is deemed to lie between 155° and 180°. On the lateral view (c,d) the trabecular markings would be in line (i.e. 180°) if the fracture was perfectly reduced; an acceptable reduction is within 20° of this ideal. Garden noted that there was a higher association with complications such as osteonecrosis, non-union and osteoarthritis if the quality of reduction was outside these acceptable limits.

head. At least one screw should be placed along the inferior neck and one along the posterior neck cortex to provide a buttress. Screw entry should not be below the lesser trochanter due to the risk of creating a stress riser and sustaining a subsequent fracture at this level. Alternatively, sliding hip screw devices can be used, often in conjunction with a derotation screw placed superiorly, which can either be retained or removed according to surgeon preference once the sliding hip screw is placed. If a derotation screw is used, it should be parallel to the sliding hip screw so that sliding can still occur. Patients are mobilized to touch weight-bearing for the first 6 weeks and weight-bearing is increased as X-ray progression towards union and the patient's symptoms allow.

Prosthetic replacement Both displaced and undisplaced femoral neck fractures in the elderly patient should be treated by prosthetic replacement due to the high failure rates of open reduction and internal fixation in this group. To fix an imperfectly reduced fracture is to risk failure.

If a stage III or IV fracture cannot be reduced closed, and the patient is under 60 years of age, open reduction through an anterior or anterolateral approach is advisable (Smith-Petersen or Watson-Jones being the most common examples). In older patients (and certainly in those over 70) this may not be justified. Prosthetic replacement may always be preferred for this older group as it carries a much lower risk of needing revision surgery.

In the frailer and less mobile patient, hemiarthroplasty should be performed. There is little evidence to support use of bipolar hemiarthroplasties over unipolar types in these patients; the mortality, morbidity and functional recovery following use of either are similar. If it is considered that the patient's functional

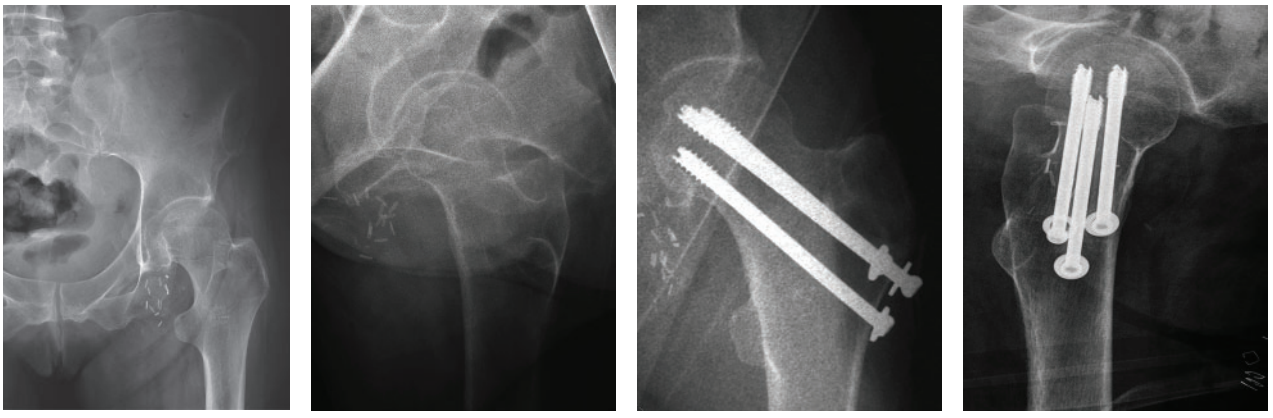
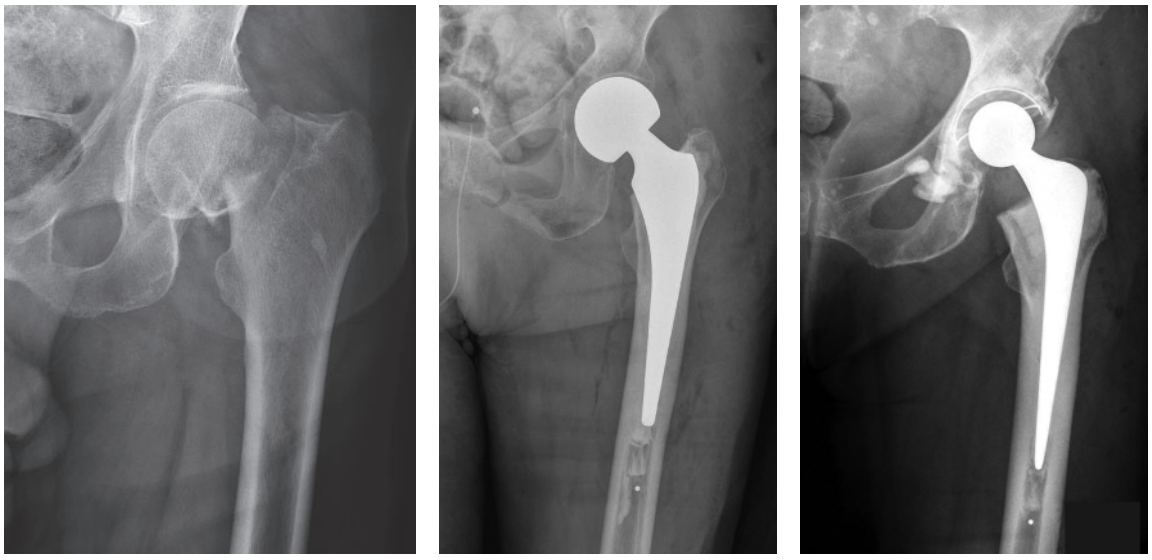


Figure 30.11 Femoral neck injuries – treatment (a,b) This Garden stage II fracture has been stabilized with three cannulated screws. (c,d) An optimum position for the screws is: one to support the inferior portion of the neck (centrally); and another two, central in level, skirting the anterior and posterior cortices of the femoral neck on the lateral X-ray. It is important that the most inferior screw enters the lateral cortex of the femur proximal to the level of the inferior margin of the lesser trochanter.



(a) (b) (c)

Figure 30.12 Fracture of the femoral neck – treatment (a) A fracture as severely displaced as this (stage IV), if treated by reduction and internal fixation, will probably end up needing revision surgery; instead, it could be treated by performing a hemiarthroplasty using a cemented femoral prostheses (b). A total hip replacement (c) provides a better outcome for active independent patients with this type of fracture.

demand may lead to acetabular erosion requiring revision surgery, total hip replacement should be considered as the first surgery (Figure 30.12). In the active independent mobile patient, total hip replacement is preferred. Other situations in which total hip replacement should be considered in preference to hemiarthroplasty are: (1) if treatment has been delayed for some weeks and acetabular damage is suspected; or (2) in patients with metastatic disease or Paget's disease. Cemented fixation is preferred due to lower postoperative pain scores and lower risk of periprosthetic fractures when compared to uncemented stems in osteoporotic bone. The risks of embolic complications associated with cement usage are addressed by adequate perioperative resuscitation of the patient, lavage, drying and venting of the femur prior to cementation and avoiding excessive pressurization. Although 'bone cement implantation syndrome' can occur, it is a very rare event, particularly if the precautions described are followed, and the use of cemented rather than uncemented has been shown to be associated with a survival benefit in properly adjusted analyses of national databases.

There is strong evidence for an increase in mortality in these frail patients if surgery is delayed for more than 24–36 hours following injury. The aim of surgery is to allow immediate full weight-bearing. Frail patients cannot effectively limit their weight-bearing and prolonged periods of immobility are associated with substantially increased risks of morbidity and hence mortality; always bear in mind the phrase 'weight-bear to survive' in this population. Speed of recovery depends largely on how active the patient was before the fracture; after 2–4 months, further improvement is unlikely.

EXTRACAPSULAR HIP FRACTURES

Intertrochanteric fractures Intertrochanteric fractures are almost always treated by early internal fixation – not because they fail to unite with conservative treatment (they unite quite readily), but (1) to obtain the best possible position and (2) to get the patient up and walking as soon as possible and thereby reduce the complications associated with prolonged immobility which are marked in this population. Non-operative treatment may be appropriate for a very small group who are too ill to undergo anaesthesia but this is limited to those that would not survive the procedure. Traction in bed until there is sufficient reduction of pain to allow mobilization can yield reasonable results but much depends on the quality of nursing care and physical therapy.

Fracture reduction at surgery is performed on a fracture table that provides traction against a post and the ability to internally rotate the leg; the position is checked by X-ray and the fracture is fixed with an angled device – preferably a sliding screw in conjunction with either a plate or intramedullary nail. Positioning the screw is important to prevent cut-out of the osteoporotic bone in the femoral head. The guide wire and screw should enter the bone above the inferior border of the lesser trochanter, pass up the middle of the femoral neck and end within the centre of the femoral head. A 'tip-apex' distance on the anteroposterior and lateral X-ray is described to identify a 'sweet-spot' for positioning

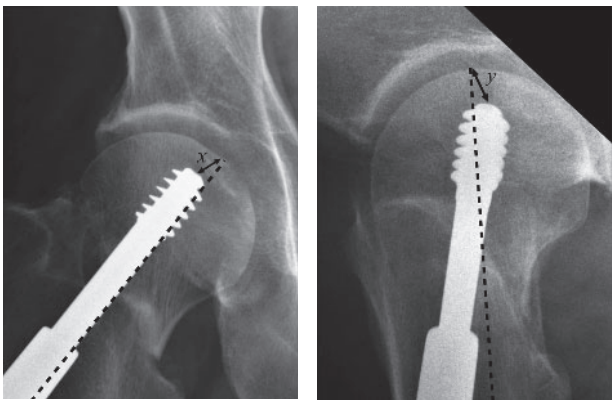
this sliding screw: if the tip is within 25 mm of the apex when the measurement on both views is combined, there is a lower risk of the screw cutting out of the femoral head (Figure 30.13).

Reverse oblique fractures With the less common 'reversed oblique' fracture pattern (where the fracture line runs downwards obliquely from the medial to the lateral cortex – Figure 30.14) there is a tendency for the distal fragment to shift medially under the proximal fragment as the hip screw slides in the barrel if a sliding hip screw is used. An intramedullary device is therefore recommended (Figure 30.15).

On the rare occasion that satisfactory reduction proves impossible, a valgus osteotomy may be needed to allow the proximal fragment to abut securely against the femoral shaft.

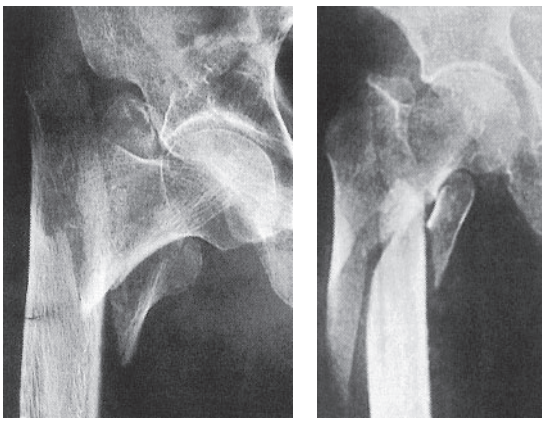
Subtrochanteric fractures Subtrochanteric fractures have several features which make them interesting (and challenging!) to treat.

- Blood loss is greater than with femoral neck or trochanteric fractures – the region is covered with anastomosing branches of the medial and lateral circumflex femoral arteries which come off the profunda femoris trunk.



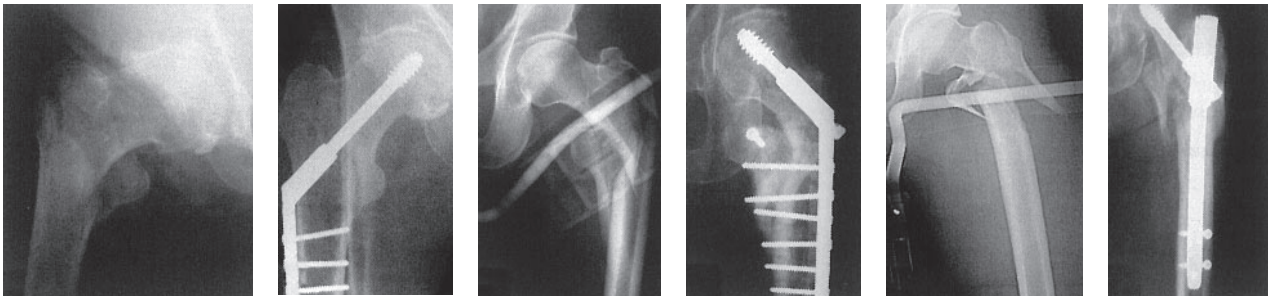
(a) (b)

Figure 30.13 Risk of screw cut-out The tip-apex distance is a measure that estimates the risk of screw cut-out from the femoral head. (a,b) It is the sum of the measured distances (after adjustment for magnification on the X-ray) from the tip of the screw to the apex of the femoral head – on both the AP (*x*) and lateral views (*y*). The risk of cut-out is low if the sum is less than 25 mm.



(a) (b)

Figure 30.14 Intertrochanteric fracture features Two contrasting types of intertrochanteric fracture. (a) Type 2 fracture: the fracture runs obliquely downwards from the lateral to medial cortex, in this case associated with a lesser trochanter fracture and resulting in a typical varus deformity. This is an unstable fracture. (b) Type 4 'reverse oblique' fracture: here the fracture line runs downwards from medial to lateral cortex, to give an even more unstable geometry.



(a) (b) (c) (d) (e) (f)

Figure 30.15 Intertrochanteric fractures – treatment Anatomical reduction is the ideal; but stable fixation is equally important. Type 1 and type 2 fractures (a,b) can usually be held in good position with a sliding screw and plate. If this is not possible, an osteotomy of the lateral cortex (c,d) will allow a screw to be inserted up to the femoral neck and into the head of the femur; this can be used as a lever to reduce the fracture so that the medial spike of the proximal fragment engages securely into the femoral canal; fixation is completed with a side plate. Reverse oblique fractures (e,f) are inherently unstable even after perfect reduction; here one can use an intramedullary device with an oblique screw that engages the femoral head. (Courtesy of Mr M. Manning and Mr J.S. Albert.)

- There may be subtle extensions of the fracture into the intertrochanteric region, which the surgeon should be aware of before attempting fixation in order to control this intraoperatively and ensure the selected device does not cause further displacement.
- The proximal part is abducted and externally rotated by the gluteal muscles, and flexed by the psoas. The shaft of the femur has to be brought into a position to match the proximal part or else a malunion is created by internal fixation. The surgeon should not rely on the fixation device to achieve reduction of the fracture for them during insertion – this will not work.

Traction may help to reduce blood loss and pain. It is an interim measure only until the patient is taken to theatre for fixation of the fracture. These fractures should be prioritized in the same manner as other hip fractures due to the increased mortality associated with delay to theatre.

Subtrochanteric hip fractures are most commonly treated with intramedullary devices with one or two screws into the femoral neck and head (Figure 30.16). Blade plates, 95-degree screw and plate devices and locking plates are other less commonly used alternatives, but the evidence to support their use is lacking. The selected device should achieve stabilization of the fracture and permit full weight-bearing postoperatively.

Due to the pattern of the fracture, subtrochanteric fractures are not biomechanically suitable for treatment with a sliding hip screw. Fixation with an intramedullary device that allows full protection of the femoral neck is the preferred treatment option due to a lower risk of non-union than with extramedullary devices. Long intramedullary devices are preferred to short ones due to the risk of fracture at the tip of the implant with the use of short devices. It is important to achieve anatomical reduction in these fractures to decrease the risk of non-union. In the case of atypical fractures, there may be plastic deformation of the femur at the site of the fracture requiring an osteotomy to correct.

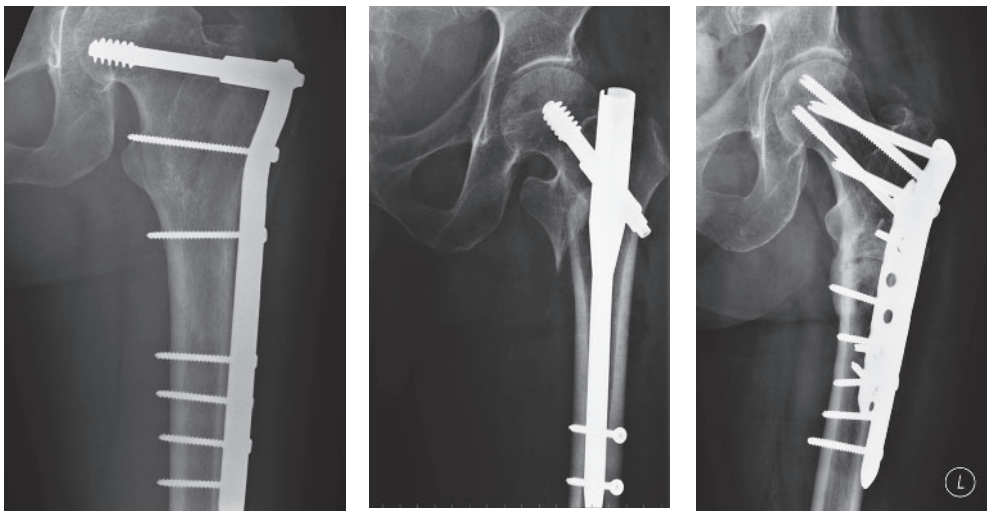
Key points to bear in mind when operating on these fractures are: (1) an anatomic reduction will provide the greatest surface area of contact between the fragments and reduce stresses on the implant; with intramedullary nails this has to be achieved *before* reaming is commenced – reaming and the insertion of the implant does not achieve this for you; (2) as little soft-tissue dissection as possible to accomplish reduction should be performed to reduce the risk of non-union but, if required, an open reduction is preferable to a malreduced and badly fixed fracture.

Postoperative rehabilitation Postoperatively, exercises are started on the day of surgery if possible and the patient is allowed up and weight-bearing as soon as they are able. The fixation achieved should aim to permit full weight-bearing immediately postoperatively due to the substantial risks and morbidity associated with prolonged bed rest in this population. It must be recognized that the majority of elderly, frail patients cannot achieve partial or limited weight-bearing and a postoperative instruction to limit weight-bearing often means that a patient is not mobilized, with the attendant risks associated with this.

Complications

GENERAL COMPLICATIONS OF HIP FRACTURES

These patients, most of whom are elderly, have a high rate of comorbidities prior to injury. A substantial proportion of these patients will fall due to these comorbidities and the reasons why they have fallen need to be considered as part of their treatment. They are prone to general complications such as deep vein thrombosis, pulmonary embolism, pneumonia and



(a) (b) (c)

Figure 30.16 Subtrochanteric fractures – internal fixation Several methods of fixation are in use: (a) a 95° screw and plate device; (b) an intramedullary nail with proximal interlocking screw into the femoral head; and (c) a proximal femoral plate with locking screws.

bed sores. Notwithstanding the advances in perioperative care, the mortality rates in elderly patients are as high as 7.5% at 30 days after injury and 24% at 1 year. Among the survivors over 80 years, about half fail to resume independent walking.

INTRACAPSULAR FRACTURES

Osteonecrosis Osteonecrosis of the femoral head occurs in about 30% of patients with displaced fractures and 10% of those with undisplaced fractures (Figure 30.17). There is no way of diagnosing this at the time of fracture. A few weeks later, an isotope bone scan may show diminished vascularity but, as this investigation is not performed routinely in those without problems, its utility is unproven and not justified. X-ray changes may not become apparent for 18 months following injury and surgery. Whether the fracture unites or not, collapse of the femoral head if it does occur will cause pain and progressive loss of function.

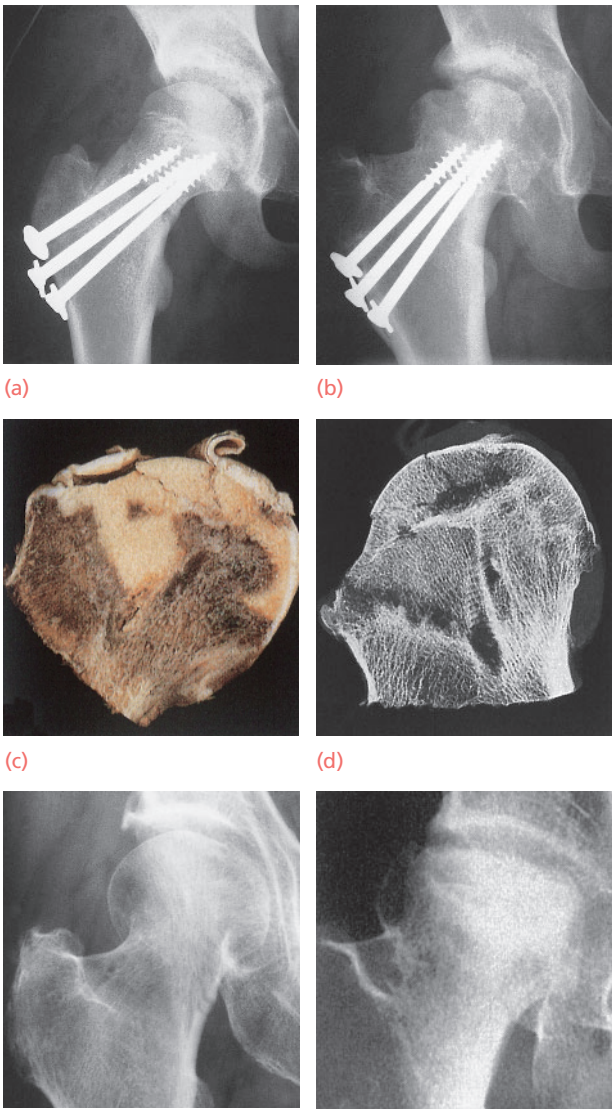


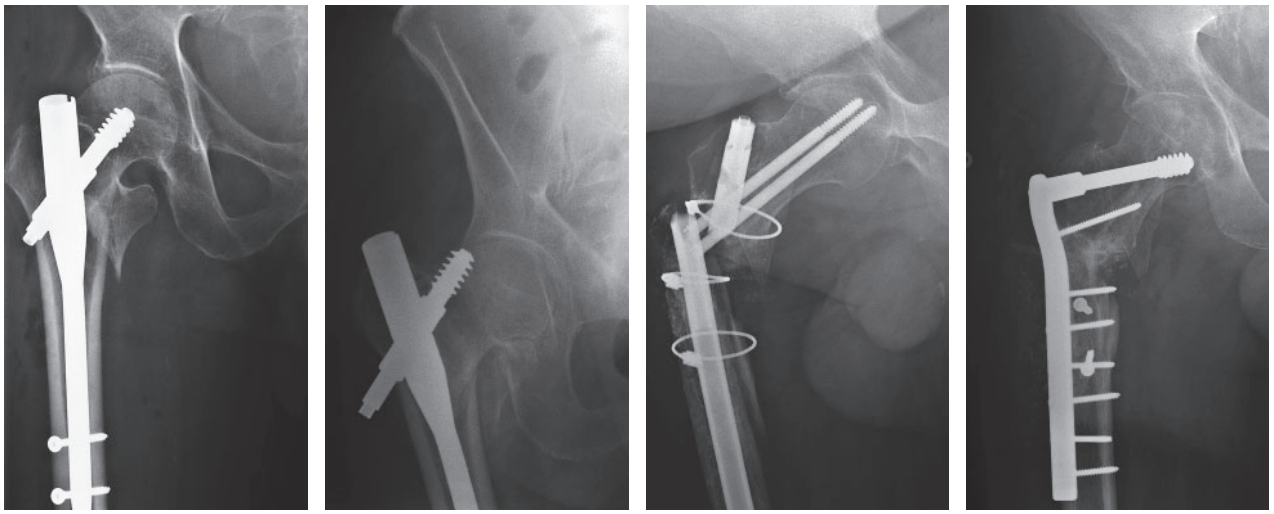
Figure 30.17 Fracture of the femoral neck – osteonecrosis (a) The post-reduction X-ray appearances may be adequate but the blood supply is compromised and 6 months later (b) there is obvious necrosis of the femoral head. (c) Section across the excised femoral head, showing the large necrotic segment and splitting of the articular cartilage. (d) Fine detail X-ray of the same. (e,f) Even an impacted fracture, if it is displaced in valgus, can lead to osteonecrosis.

In patients over 45 years, treatment of osteonecrosis is by total joint replacement. In younger patients, the choice of treatment is controversial. Core decompression (used in non-traumatic osteonecrosis) has no place in the management of traumatic osteonecrosis. Realignment or rotational osteotomy is suitable for those with a relatively small necrotic segment. Arthrodesis is often mentioned in armchair discussions, but in practice it is seldom carried out due to the difficulty of achieving union, the lack of contemporary expertise and the potential long-term consequences. Provided the risks are carefully explained, including the risk of revision surgery being required in the future, joint replacement is an effective treatment for relief of symptoms.

Non-union More than 30% of all femoral neck fractures fail to unite, and the risk is particularly high in fractures that are severely displaced. There are many causes of this: poor blood supply, imperfect reduction, inadequate fixation, and the tardy healing that is characteristic of intra-articular fractures. The bone at the fracture site is ground away, the fragments fall apart and the sliding screw cuts out of the bone or is extruded laterally. The patient complains of pain, shortening of the limb and difficulty with walking. The X-ray shows the evidence of the sorry outcome.

The method of *treatment* depends on the cause of the non-union and the age of the patient. In relatively *young patients*, three procedures are available:

1 *If the fracture is nearly vertical but the head is alive*, subtrochanteric osteotomy with internal fixation changes the fracture line to a more horizontal angle which is more biomechanically forgiving, compressed on weight-bearing and hence more likely to heal.
2 *If the reduction or fixation was faulty and there are no signs of osteonecrosis*, it is reasonable to remove the screws, reduce the fracture, insert further fixation that will achieve compression of the fracture and also apply a bone graft across the fracture (either a segment of fibula or a muscle pedicle graft).
3 *If the head is avascular but the acetabulum completely unaffected (rare)*, hemiarthroplasty may be suitable; if the joint is damaged or already arthritic, total hip replacement is indicated.



(a) (b) (c) (d)

Figure 30.18 Complications of treatment of intertrochanteric fractures (a,b) Failure to maintain reduction, which can be early – usually in osteoporotic bone or from poor implant seating (c,d). The implant may fracture if union is not timely. Revision surgery is complex and may involve bone grafts and a new implant.

In *elderly patients*, only two procedures should be considered:

1 *If pain is considerable*, the femoral head, no matter whether it is osteonecrotic or not, is best removed and (provided the patient is reasonably fit) total joint replacement is performed.
2 *If the patient is not fit for elective surgery and the pain is not unbearable*, a raised heel and a stout stick or elbow crutch may be sufficient.

Osteoarthritis Osteonecrosis or femoral head collapse may lead, after several years, to secondary osteoarthritis of the hip. If this is symptomatic, total joint replacement will be needed.

Complications of prosthetic replacement include dislocation (greater in the total hip replacement group (2–20%), infection, fracture, embolism, neurovascular injury and acetabular erosion in hemiarthroplasty (66% in active, independently mobile patients).

EXTRACAPSULAR FRACTURES

Failed fixation Screws may cut out of the osteoporotic bone if reduction is poor or if the fixation device is incorrectly positioned. If union is delayed, the implant itself may break. In either event, reduction and fixation may have to be re-done.

Malunion Varus and external rotation deformities are common. Fortunately, they are seldom severe and rarely interfere with function.

Non-union Intertrochanteric fractures seldom fail to unite. If healing is delayed (say beyond 6 months), the fracture probably will not unite and further operation is advisable; the fragments are repositioned as anatomically as is feasible, the fixation device is applied more securely and bone grafts are packed around the fracture (Figure 30.18).

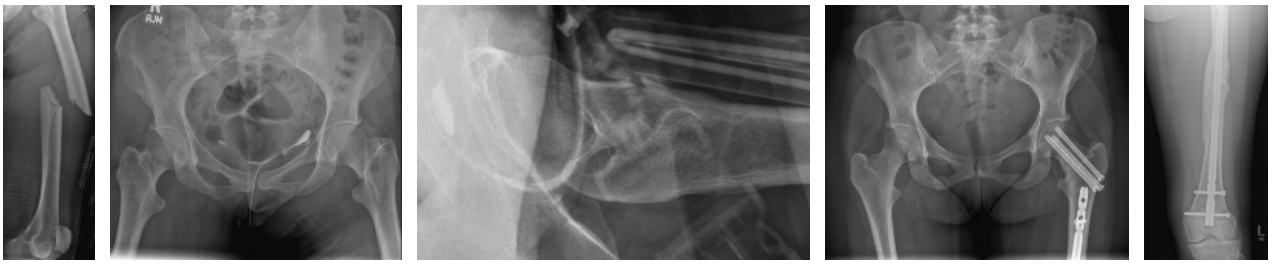
SUBTROCHANTERIC FRACTURES

Malunion Varus and rotational malunions are fairly common. This can be prevented by careful attention to accurate reduction before internal fixation is applied. If the degree of malunion produces symptoms, it may need operative correction, but this is rare.

Non-union The risk of delayed or non-union leading to implant failure is between 7% and 20%; the risk is higher in bisphosphonate-related fractures where the native bone is not normal and healing is slow. If symptomatic, it will require operative correction of any residual deformity, renewed fixation and bone grafting. Excision of the pathological deformity and osteotomy to correct the underlying plastic deformity is required.

Combined fractures of the neck and shaft

Patients with generally high-energy fractures of both the femoral neck and the ipsilateral femoral shaft present a special problem. The critical point is that the presence of the femoral neck fracture must be recognized at the time of first assessment or at least prior to surgery. Both fractures must be fixed, and there are several ways of doing this (Figure 30.19). The femoral neck fracture takes priority as complications following this fracture are more difficult to address than those of the shaft fracture. Anatomical reduction and stable fixation of the femoral neck fracture must not be compromised in order to accommodate fixation of the shaft fracture. The femoral neck fracture is reduced using closed or, if necessary, open methods and fixed.



(a) (b) (c) (d) (e)

Figure 30.19 Treatment of ipsilateral femoral neck and femoral shaft fracture (a) Femoral shaft fracture; (b,c) ipsilateral femoral neck fracture; (d) healed femoral neck fracture treated with cannulated screws; and (e) healing femoral shaft fracture treated with retrograde intramedullary nail.

The femoral shaft fracture can then be managed with a retrograde locked intramedullary nail (inserted through the knee), by a lateral plate or by an antegrade nail where the position of the neck fracture fixation allows.

Pathological fractures

Hip fractures, particularly intertrochanteric fractures may occur secondary to metastatic disease or myeloma. Solitary bone lesions must always be biopsied prior to definitive treatment (see Chapter 9). In the patient with known malignancy with multiple bone lesions a pragmatic approach may be taken. Surgery should be considered where life expectancy is long enough for the patient to gain benefit from the operation. Even if life expectancy is short, the benefits of pain relief and the facilitation of nursing with fracture fixation or replacement must be recognized. The reconstruction should allow immediate full weight-bearing and be robust enough to outlive the patient.

In general, replacement is favoured as fixation has potentially higher failure rates due to disease progression. Where fixation is chosen, it can be augmented with polymethylmethacrylate bone cement. Lesions of the femoral head are adequately treated with hemiarthroplasty or total hip arthroplasty. If the lesion extensively involves the proximal femur to the subtrochanteric level, endoprosthetic replacement may be preferred. The surgeon should note the extent of any acetabular involvement when planning reconstruction and take this into account with their construct.

ISOLATED FRACTURES OF THE TROCHANTERS

In adolescents, the *lesser trochanter* apophysis may be avulsed by the pull of the psoas muscle; the injury nearly always occurs during hurdling. Treatment is rest, followed by return to activity when comfortable. In the elderly, separation of the lesser trochanter should arouse suspicions of metastatic malignant disease and be considered pathological until proven otherwise.

In the elderly, part of the *greater trochanter* can be fractured by a direct blow after a fall. The X-ray should be scrutinized for subtle associated intertrochanteric or intracapsular fractures. In the event this is absent and displacement is less than 1 cm, treatment is non-operative and functional recovery is usually good. Active abduction should be avoided and the patient mobilized with protected weight-bearing until signs of radiological union are present, usually at 6–8 weeks.

Occasionally, the greater trochanter is fractured and the fragment widely separated in a young individual. It can be fixed back in position with cancellous screws, tension-band wiring or a trochanteric plate. Removal of metal work is often required in these patients due to soft-tissue irritation once the fracture has united.

PROXIMAL FEMORAL FRACTURES IN CHILDREN

Hip fractures rarely occur in children but, when they do, they are potentially very serious. There is a high risk of complications, such as osteonecrosis, premature physeal closure and coxa vara. The fracture is usually due to high-energy trauma (e.g. falling from a height or an RTC). Pathological fractures sometimes occur through a bone cyst or benign tumour. In children, the possibility of non-accidental injury should be considered and investigated. Particular warning signs of non-accidental injury include external signs of other injuries particularly when these look to be of different ages, X-ray evidence of previous fractures of different ages, multiple attendances or attendances at a unit that is not local to the home address without good reason, a history that is inconsistent in terms of the events that occurred or injuries that are not consistent with the mechanism described (e.g. spiral femoral fractures in children who are not yet walking).

At birth the proximal end of the femur is entirely cartilaginous and for several years, as ossification proceeds, the area between the capital epiphysis and greater trochanter is vulnerable to trauma. Moreover, between the ages of 4 and 8 years the ligamentum teres contributes very little to the blood supply of the epiphysis, hence its susceptibility to post-traumatic ischaemia.

Classification

The most useful classification is that of Delbet, which is based on the location of the fracture.

- *Type I* – a fracture-separation of the epiphysis. The epiphyseal fragment may also be dislocated from the acetabulum.
- *Type II* – a transcervical fracture of the femoral neck. This is the commonest variety, accounting for almost 50% of paediatric hip fractures.
- *Type III* – a basal (cervicotrochanteric) fracture. This is the second most common injury.
- *Type IV* – an intertrochanteric fracture (Figure 30.20).

Clinical features

Diagnosis can be difficult, especially in infants where the epiphysis is not easily defined on X-ray. Type I fractures are easily misdiagnosed as hip dislocations. Ultrasonography, MRI and arthrography may be useful diagnostic adjuncts. In older children the diagnosis is usually obvious on plain X-ray examination. It is important to establish whether the fracture is displaced or unddisplaced; displaced fractures carry a much higher risk of complications. Type IV fractures are the least likely to give rise to complications.

Treatment

These fractures should be treated as a matter of urgency, and certainly within 24 hours. Initially the hip is supported or splinted to achieve pain relief and prevent secondary displacement while investigations are carried out. Early aspiration of the intracapsular haematoma is advocated by some authors as a means of reducing the risk of epiphyseal ischaemia; however, as in adults, the benefits are uncertain and the matter is controversial.

UNDISPLACED FRACTURES

Undisplaced fractures may be treated by immobilization in a plaster spica for 6–8 weeks. However, fracture position is not always maintained leading to a considerable risk of late displacement and malunion or non-union. Close X-ray follow-up is therefore mandated.

DISPLACED FRACTURES

Displaced *type IV fractures* may be treated non-operatively with closed reduction, traction and spica immobilization. Careful follow-up is essential; if position is lost, operative fixation will be required.

Type I, II and III fractures are treated by closed reduction and then internal fixation with smooth pins or cannulated screws. 'Closed reduction' means one gentle manipulation; if this fails, open reduction is performed. Repeated or forceful attempts at closed reduction risk further damage to the blood supply and increase the risk of subsequent complications and poor outcome. In small children, operative fixation is supplemented by a spica cast for 6–12 weeks.

Complications

Osteonecrosis of the femoral head This is the most common (and most feared) complication (Figure 30.21); it occurs in approximately 30% of all cases. Important risk factors are: (1) an age of more than 10; (2) a high-energy injury; (3) a type I or II fracture; and (4) fracture displacement.

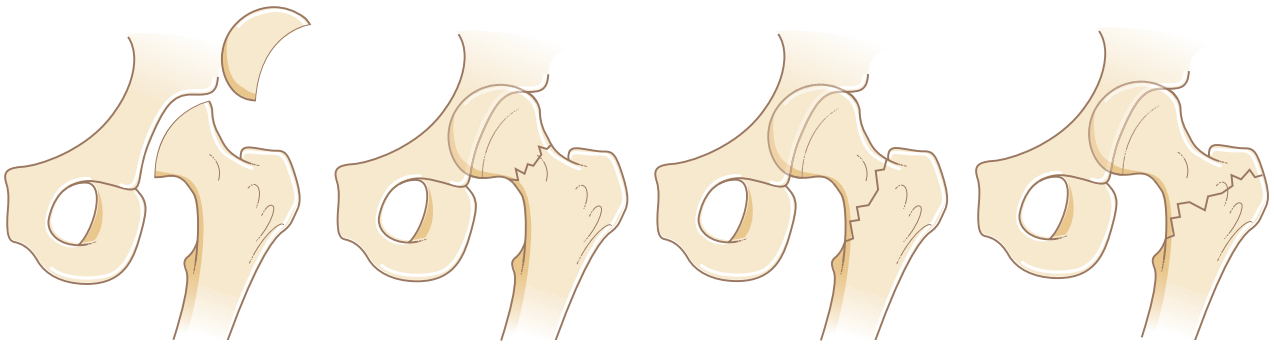
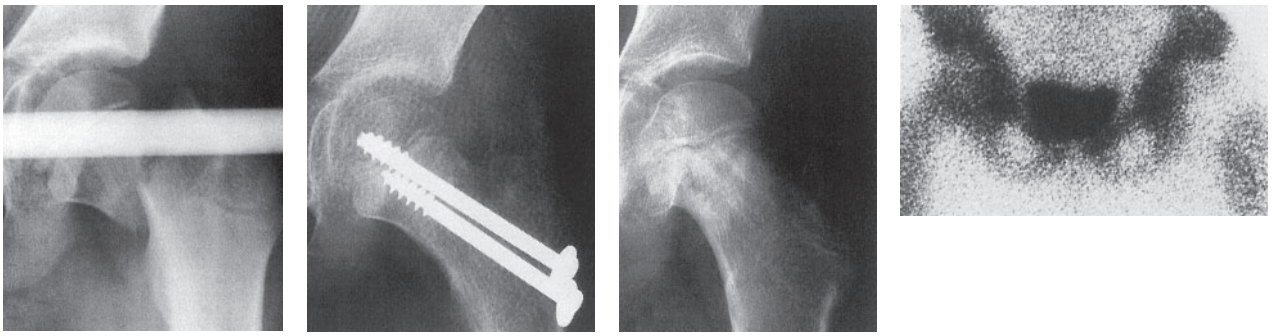


Figure 30.20 Proximal femoral fractures in children These are the result of strong forces or weak bone (e.g. through cysts). Delbet classification – depending on the level of the fracture: (a) type I at the physeal level; (b) type II through the middle of the neck; (c) type III at the base of the neck; and (d) type IV at the intertrochanteric level.



(a) (b) (c) (d)

Figure 30.21 Femoral neck fractures in children (a) Fracture of the femoral neck in a child is particularly worrying because, even with perfect fixation (b), there is often ischaemia of the femoral head. This fracture united and the screws were removed (c), but the radioisotope scan shows no activity in the left femoral head, i.e. osteonecrosis (d).

As osteonecrosis develops, the child complains of pain and loss of movement; X-ray changes usually appear within 3 months of injury. Treatment is problematic. Non-weight-bearing, or 'containment splintage' in abduction and internal rotation, is sometimes advocated but there is little evidence that this makes any difference to the outcome. The outcome depends largely on the size of the necrotic area; unfortunately, most end up with intrusive pain and marked restriction of movement. Arthrodesis may be advised as a late salvage procedure but it is now rarely performed. Arthroplasty may be considered in the young adult but careful counselling is required for the attendant risks including aseptic loosening, wear and the need for revision surgery in the future.

Coxa vara Femoral-neck deformity may result from malunion, osteonecrosis or premature physeal closure. If the deformity is mild, remodelling may take care of it. If the neck-shaft angle is less than 110 degrees, subtrochanteric valgus osteotomy will probably be needed.

Diminished growth Physeal damage may result in retarded femoral growth. Limb length equalization may be needed.

FEMORAL SHAFT FRACTURES

The femoral shaft is circumferentially padded with large muscles. Muscle contraction can displace femoral shaft fractures making reduction difficult. However, the muscle layers are advantageous, the healing potential is improved by having a well-vascularized sleeve and open fractures can be managed often with thorough debridement and primary closure; rarely, a simple split-thickness skin graft is required to obtain satisfactory cover. Soft-tissue flap coverage is very rarely required.

Mechanism of injury

High-energy trauma is the commonest mechanism of injury, with diaphyseal fractures occurring more frequently in younger adults. Non-accidental injury should always be considered in diaphyseal fractures in children. The fracture pattern is related to the type of force that produced the break. A *spiral fracture* is usually caused by a fall in which the foot is anchored while a twisting force is transmitted to the femur. *Transverse* and *oblique fractures* are more often due to angulation or direct violence and are therefore particularly common in road accidents. With severe violence (often a combination of direct and indirect forces) the fracture may be comminuted, or a segmental fracture pattern may be present.

Pathological anatomy

Most fractures of the femoral shaft have some degree of comminution, although it is not always apparent on X-ray. Small bone fragments, or a single large 'butterfly' fragment, may separate at the fracture line but usually remain attached to the adjacent soft tissue and retain their blood supply. With more extensive comminution, the fracture is completely unstable (Figure 30.22). This is the basis of a helpful classification.

Fracture displacement often follows a predictable pattern dictated by the pull of muscles attached to each fragment.

- In *proximal shaft fractures* the proximal fragment is flexed, abducted and externally rotated because of gluteus medius and iliopsoas muscle pull; the distal fragment is frequently adducted.
- In *mid-shaft fractures* the proximal fragment is again flexed and externally rotated but abduction is less marked.
- In *lower-third fractures* the proximal fragment is adducted and the distal fragment is tilted by gastrocnemius pull.

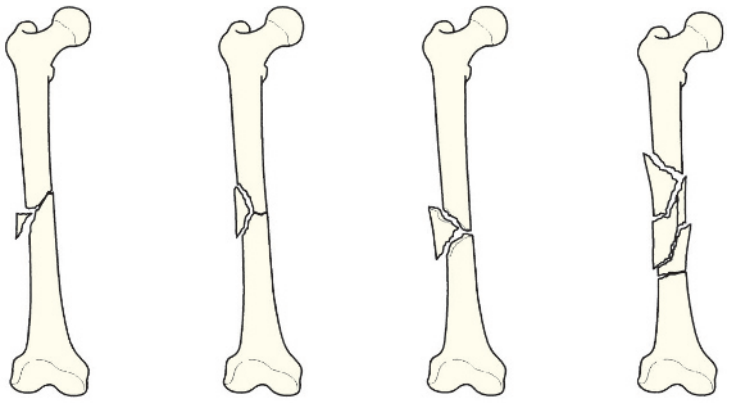


Figure 30.22 Femoral shaft fractures – classification Winquist's classification reflects the observation that the degree of soft-tissue damage and fracture instability increase with increasing grades of comminution. In *type 1* (a) there is only a tiny cortical fragment. In *type 2* (b) the 'butterfly fragment' is larger but there is still at least 50% cortical contact between the main fragments. In *type 3* (c) the butterfly fragment involves more than 50% of the bone width. *Type 4* (d) is essentially a segmental fracture.

AU: Not mentioned in text.

Extensive bleeding from femoral fractures may occur, and bleeding from the perforators of the profunda femoris may be severe. Over a litre of blood may be lost into the soft tissues and a patient can become hypotensive quickly if not adequately resuscitated. Fractures at the junction of the middle and distal thirds of the femoral shaft can directly damage the femoral artery in the adductor canal.

Clinical features

There is swelling and deformity of the limb, and movement of the limb is painful. Often there are associated injuries from the high-energy trauma. A careful secondary survey is necessary to exclude neurovascular problems, compartment syndrome and other lower-limb or pelvic fractures. An ipsilateral femoral neck fracture occurs in about 10% of cases; the treatment of the femoral neck fracture is to be prioritized over the shaft fracture. An ipsilateral tibia fracture produces a 'floating knee'; the femoral fracture should be treated first to help stabilize blood loss and makes nursing of the patient easier, especially if not all fractures can be treated at the same operation. Consideration should be given to using a retrograde femoral nailing and an antegrade tibial nailing through the same incision centred over the patella tendon. Multiple long-bone fractures have a high risk of multisystem injury in the patient. The effects of blood loss and other injuries, some of which can be life-threatening, may dominate the clinical picture.

X-rays

Adequate anteroposterior and lateral X-rays of the whole femur should be obtained. *The hip and knee joint must be included as well* (Figure 30.23). The fracture pattern should be noted; it will form a guide to treatment.

Emergency treatment

Traction with a splint is mandatory for a patient with a femoral shaft fracture. It is applied at the site of the accident, and before the patient is moved. A Thomas splint, or one of the modern derivations of this practical device, is ideal: the leg is pulled straight and threaded through the ring of the splint; the shod foot is tied to the cross-piece so as to maintain traction, and the limb and splint are firmly bandaged together. This temporary stabilization helps to control pain, reduces blood loss and makes transfer easier. Shock should be treated; blood volume is restored and maintained, and analgesia is commenced. The widespread adoption of such splints led to one of the largest reductions in mortality seen due to the introduction of an intervention in military conflicts.

Definitive treatment

The patient with multiple injuries The association of femoral shaft fractures with other injuries, including

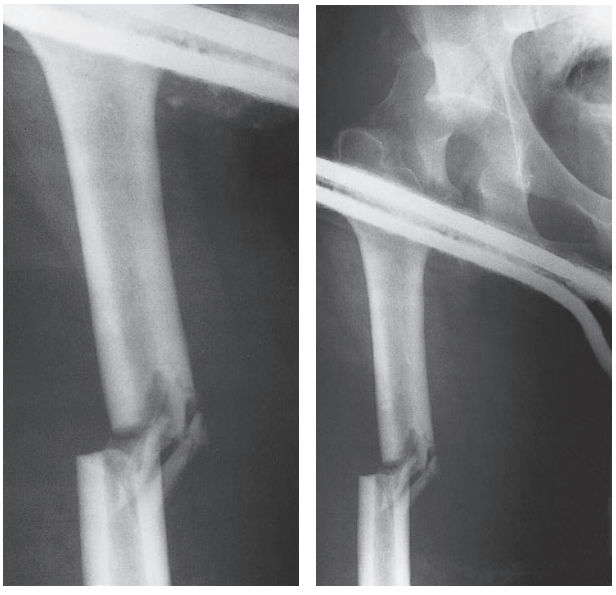


Figure 30.23 Femoral shaft fractures – diagnosis (a) The upper fragment of this femur is adducted, which should alert the surgeon to the possibility of (b) an associated hip dislocation. With this combination of injuries the dislocation is frequently missed; the safest plan is to X-ray the pelvis with every fracture of the femoral shaft.

head, chest, abdominal and pelvic trauma, increases the potential for developing fat embolism, ARDS and multi-organ failure. The polytraumatized patient and damage control surgery are discussed in Chapter 22.

THE ISOLATED FEMORAL SHAFT FRACTURE

Traction, bracing and spica casts Rarely is conservative treatment considered in contemporary practice. Traction can reduce and hold most fractures in reasonable alignment, except those of the upper third of the femur. Joint mobility can be ensured by active exercises. The chief drawback is the length of time spent in bed (10–14 weeks for adults) with the attendant problems of keeping the femur aligned until sufficient callus has formed plus reducing patient morbidity and frustration. Some of these difficulties are overcome by changing to a plaster spica or – in the case of lower-third fractures – functional bracing when the fracture is 'sticky', usually around 6–8 weeks.

The main indications for traction are: (1) fractures in children; (2) contraindications to anaesthesia or surgery; and (3) lack of suitable skill or facilities for internal fixation. Pathological fractures are highly unlikely to heal. The various methods of traction are described in Chapter 23. For young children, *skin traction* without a splint is usually all that is needed. Infants less than 12 kg in weight are most easily managed by suspending the lower limbs from overhead pulleys ('*gallows traction*'), but no more than 2 kg weight should be used and the feet must be checked frequently for circulatory problems. Older children are better suited to *Russell's traction* (Chapter 23) or use of a *Thomas splint*. Fracture union will have progressed sufficiently by 2–4 weeks (depending on the age of the child) to permit a *hip spica* to be applied.

Consolidation is usually complete by 6–12 weeks. Adults (and older adolescents) require *skeletal traction* through a pin or a tightly strung Kirschner wire behind the tibial tubercle. Traction (8–10 kg for an adult) is applied over pulleys at the foot of the bed. The limb is usually supported on a Thomas splint and a flexion piece allows movement at the knee (Figure 30.24). However, a splint is not essential; indeed, skeletal traction without a splint (*Perkins' traction*) has the advantages of producing less distortion of the fracture and allowing freer movement in bed (Figure 30.25). Exercises are begun as soon as possible. Once the fracture is starting to consolidate (at about 8 weeks in adults) traction can be discontinued and the patient allowed up and partial weight-bearing in a *cast* or *brace*. For fractures in the upper half of the femur, a *plaster spica* is the safest but it will almost certainly prolong the period of knee stiffness. For fractures in the lower half of the femur, *cast-bracing* is suitable. This type of protection is needed until the fracture has consolidated (16–24 weeks).

Plate and screw fixation This is a comparatively easy way of obtaining accurate reduction and strong fixation. High complication rates were seen with earlier plating techniques where often a wide open exposure of the fracture site and attempted anatomical reduction of all bone pieces was performed. Such extensive

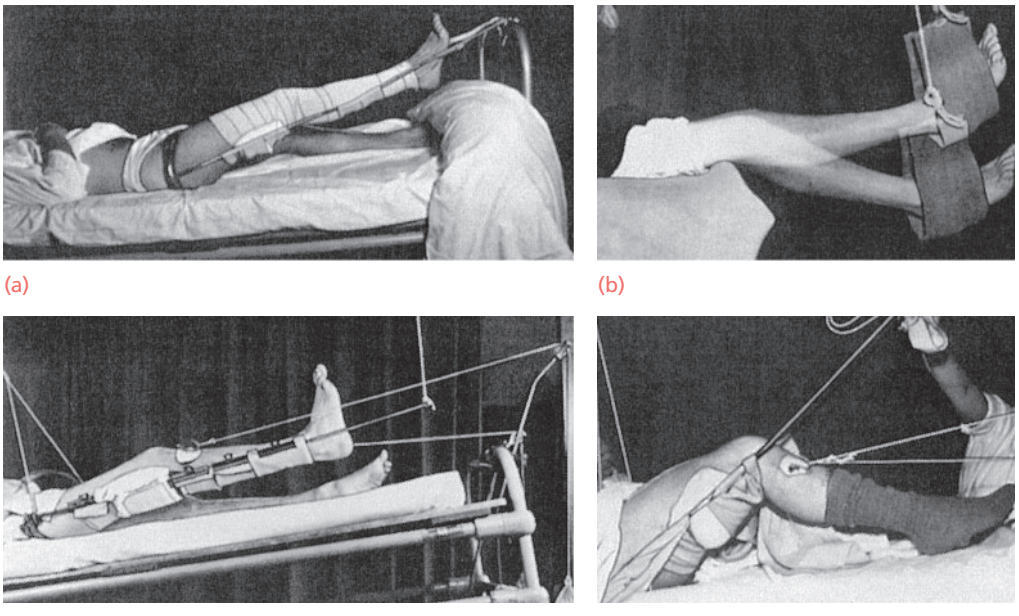


Figure 30.24 Femoral fractures – treatment by traction (a) *Fixed traction on a Thomas splint:* the splint is tied to the foot of the bed which is elevated. This method should be used only rarely because the knee may stiffen; (b) this was the range in such a case when the fracture had united. (c,d) *Balanced traction:* one way to minimize stiffness is to use skeletal balanced traction; the lower slings can be removed to permit knee flexion while traction is still maintained.

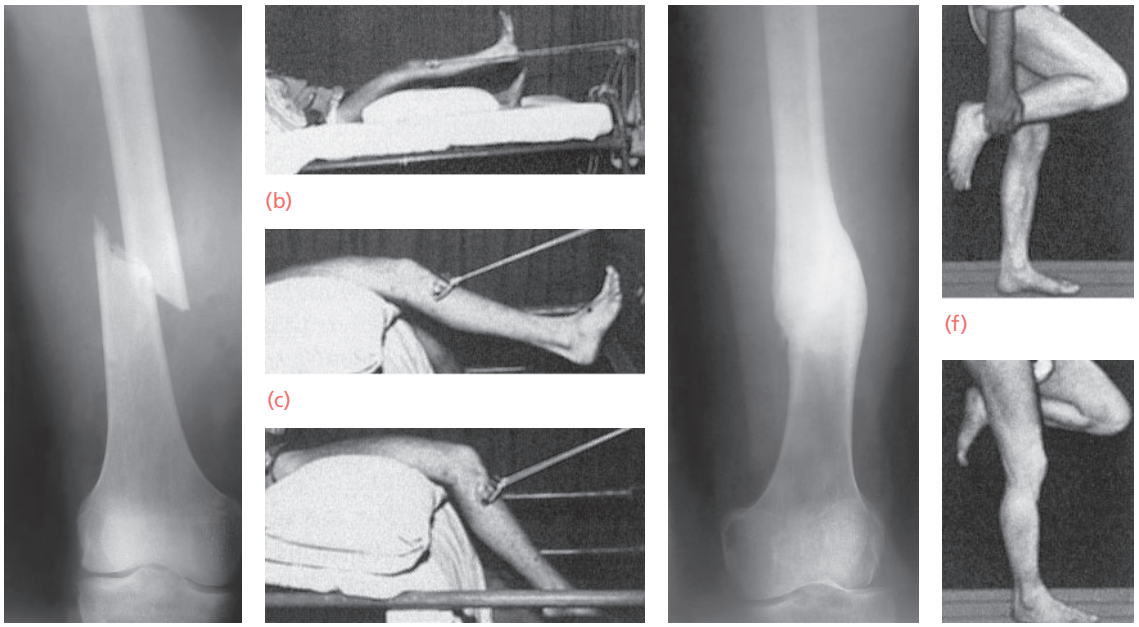


Figure 30.25 Femoral fractures – treatment by traction Even in the adult, traction without a splint can be satisfactory, but skeletal traction is essential. The patient with this rather unstable fracture (a) can lift his leg and exercise his knee (b,c,d). At no time was the leg splinted, but clearly the fracture has consolidated (e), and the knee range (f) is only slightly less than that of the uninjured left leg (g).

surgery damaged the soft tissues and healing potential and led to slow union and implant failure. However, modern techniques that insert plates through short incisions in submuscular planes (rather than deep to periosteum), with an indirect (closed) reduction of the fracture, using fewer screws which are usually placed at the ends of the plate, lead to a less rigid hold on the fracture. This technique of minimally invasive plate osteosynthesis (MIPO) has led to better union rates. However, postoperative weight-bearing may need to be modified as the implant is not as strong as an intramedullary nail, which is the main alternative. The main indications for plates are: (1) fractures at either end of the femoral shaft, especially those with extensions into the supracondylar or pertrochanteric areas; (2) a shaft fracture in a growing child; and (3) a fracture with a vascular injury which requires open repair (Figure 30.26).

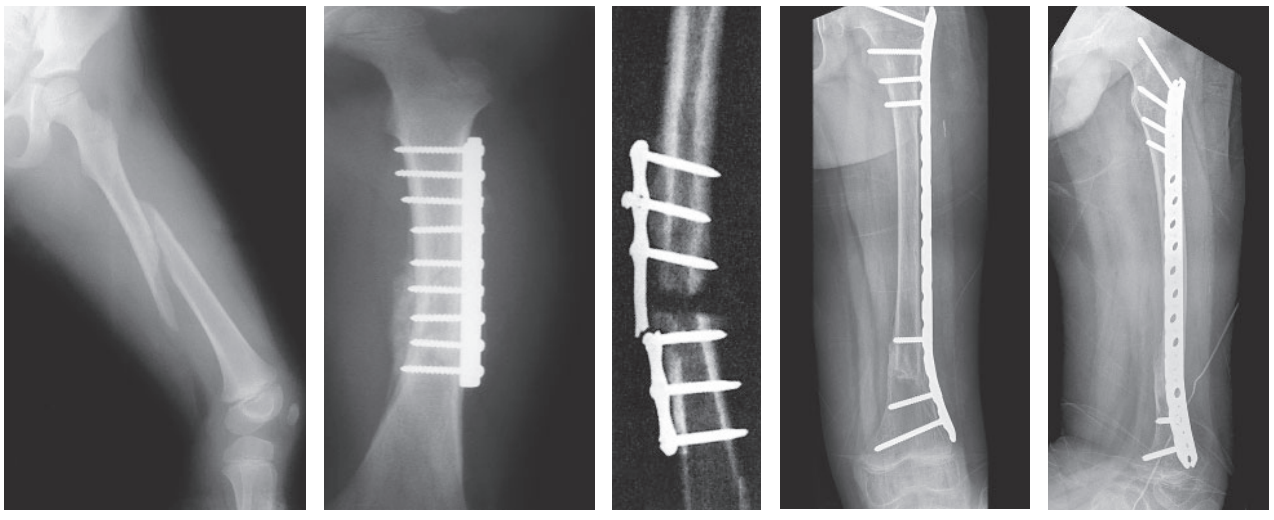


Figure 30.26 Plate fixation – past and present (a,b) Plate fixation was popular in the past, but it fell out of favour because of the high complication rate (c). Modern techniques of minimally invasive plate osteosynthesis (d,e) have shown that it still has a place in the treatment of certain types of femoral shaft fracture.

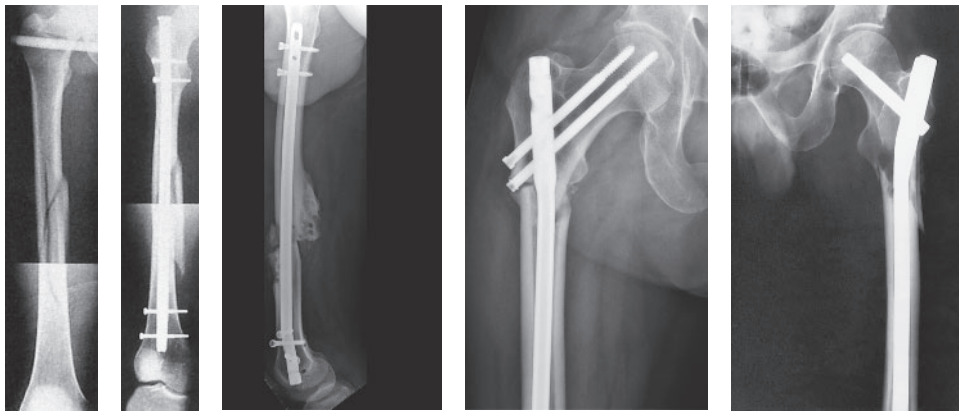


Figure 30.27 Intramedullary nailing This is the commonest way of treating femoral shaft fractures. Ideally a range of designs to suit different types of fracture should be available. (a,b) Antegrade nailing with insertion of the nail through the piriform fossa and transverse locking screws proximally and distally. (c) Retrograde nailing with insertion of the nail through the intercondylar notch at the knee. (d,e) Proximal locking can be achieved in other ways, e.g. by using parallel screws or a sliding hip screw.

Intramedullary nailing This is the method of choice for most femoral shaft fractures. The basic implant system consists of an intramedullary nail which is perforated near each end so that locking screws can be inserted transversely at the proximal and distal ends. This controls rotation and length of the femur, and ensures stability even for subtrochanteric and distal-third fractures (Figure 30.27).

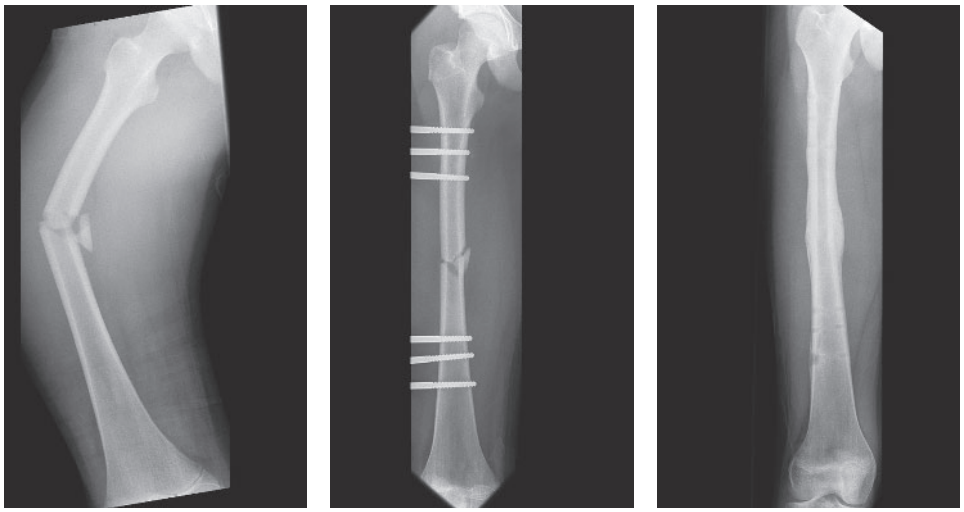
There are important considerations when using locked intramedullary nails:

- Reamed nails have a lower need for revision surgery when compared to unreamed nails.
- Select a nail that is approximately the size of the medullary isthmus so that it fills the canal reasonably well (after reaming) and adds to stability – small diameter nails are quicker to insert but are more likely to fail from fatigue fractures of the implant before union occurs.
- Consider alternative means of fracture fixation if the isthmus is so narrow that a large amount of canal reaming will have to be done in order to fit the smallest diameter nail available.
- Use a nail of sufficient length to span the canal fully.
- Antegrade insertion (through either the piriformis fossa or the tip of the greater trochanter, depending on the design of nail) or retrograde insertion (through the intercondylar notch distally) are equally suitable techniques to use; there is a small incidence of hip and thigh pain with antegrade nails, whereas there is a small incidence of knee pain with retrograde nails. If there is a total knee replacement *in situ*, compatibility of the knee prosthesis with retrograde femoral nailing should be checked. Retrograde insertion of intramedullary nails is particularly useful for: (1) obese patients; (2) when there are bilateral femoral shaft fractures (as the procedure can be performed without the need for a fracture table and the added time for setting up for each side); (3) when there is an ipsilateral tibial shaft fracture; and (4) if there is a femoral neck fracture more proximally, as screws can be inserted to hold the neck fracture without being impeded by the nail (see Figure 30.19).

Stability is improved by using interlocking screws. Often there is enough shared stability between the nail and fracture ends to allow weight-bearing early on. Full weight-bearing should be commenced as soon as the fixation allows. The fracture usually heals within 20 weeks and the complication rate is low; sometimes malunion (more likely malrotation) or delayed union (from overdistraction) occurs.

External fixation The main indications for external fixation are: (1) treatment of severe open injuries; (2) management of patients with multiple injuries where there is a need to reduce operating time and prevent the 'second hit'; and (3) the need to deal with severe bone loss by the technique of bone transport. External fixation is also useful for treating femoral fractures in adolescents where iatrogenic damage to growth plates can be avoided (Figure 30.28).

Similar to closed intramedullary nailing, it has the advantage of not exposing the fracture site and small amounts of axial movement can be applied to the bone by allowing a telescoping action in the fixator body (with some designs of external fixator). As the callus increases in volume and quality, the fixator can be adjusted to increase stress transfer to the fracture site, thus promoting quicker consolidation. However, there are still problems with pin-site infection, pin loosening and limitation of



(a) (b) (c)

Figure 30.28 External fixation for femoral shaft fractures in older children (a–c) External fixation is an option for treating femoral shaft fractures in adolescents. Elastic stable intramedullary nails shown in Figure 30.33 may not be strong enough for this heavier group of teenagers.

movement (if the pins are applied close to joints) due to interference with sliding structures. The patient is allowed up as soon as he or she is comfortable, and knee movement exercises are encouraged to prevent tethering by the half pins. Partial weight-bearing is usually possible immediately but this will depend on the X-ray appearance of callus – this may take some time (more than 6 weeks) if the fixator is a rigid device. Most femoral shaft fractures will unite in under 5 months but some take longer if the fracture is badly comminuted or contact between fracture ends is poor.

Treatment of open fractures

Open femoral fractures are high-energy injuries and should be carefully assessed for: (1) skin and soft-tissue loss; (2) wound contamination; (3) compartment syndrome; and (4) injury to vessels and nerves (see Chapter 23). The immediate treatment is similar to that of closed fractures; in addition, the patient is started on intravenous antibiotics, preferably as soon as possible after injury. Gross contamination is removed from the wound, photographs are taken if the facility is available, the wound is covered with moist swabs and an occlusive dressing, and plans are made for formal debridement in an operating theatre within 24 hours of injury but as soon as facilities and expertise allow. In theatre, the wound and zone of injury will need cleansing: the wound is extended to allow the bone ends to be delivered and adequately debrided, and contaminated and dead tissue to be excised, and the entire area should be washed thoroughly. Stabilization of open femoral shaft fractures is best achieved with locked intramedullary nails unless there is heavy contamination or bone loss – in which case external fixation (if necessary with the capacity to deal with bone loss through distraction osteogenesis) is preferable.

Complex injuries

FRACTURES ASSOCIATED WITH VASCULAR INJURY

Warning signs of an associated vascular injury are: (1) excessive bleeding and/or haematoma formation; and (2) paraesthesia, pallor or pulselessness in the leg and foot. Do not accept 'arterial spasm' as a cause of absent pulses; the fracture level on X-ray will indicate the region of arterial damage and arteriography may only delay surgery to re-establish perfusion. Most femoral fractures with vascular injuries will have had warm ischaemia times greater than 2–3 hours by the time the patient arrives in the operating theatre; when this exceeds 4–6 hours, salvage may not be possible and the risk of amputation rises. A prompt diagnosis and re-establishing perfusion is a priority; fracture stabilization is secondary.

A recommended *sequence for treatment*, particularly if the warm ischaemia time is approaching the salvage threshold, is as follows:

1 *Create a shunt* from the femoral vessels in the groin to beyond the point of injury using plastic catheters.
2 *Stabilize the fracture* (usually by plating or external fixation).
3 *Carry out definitive vascular repair.*

This sequence establishes blood flow quickly and permits fracture fixation and vascular repair to be carried out without pressure of time.

FRACTURES ASSOCIATED WITH KNEE INJURY

Femoral fractures are frequently accompanied by injury to the ligaments of the knee. Direct blows to the knee from the dashboard of a car in an accident may damage knee ligaments as well as break the femoral shaft and femoral neck – this triad of problems should be recognized. With attention focused on the femur, the knee injury is easily overlooked, only to

re-emerge as a persistent complaint weeks or months later. As soon as the fracture has been stabilized, the knee should be carefully examined and any associated abnormality investigated and treated.

COMBINED NECK AND SHAFT FRACTURES

This is dealt with in detail above. The most important thing is diagnosis: always examine the hip and obtain an X-ray of the pelvis. Both sites must be stabilized: first the femoral neck and then the femur.

PATHOLOGICAL FRACTURES

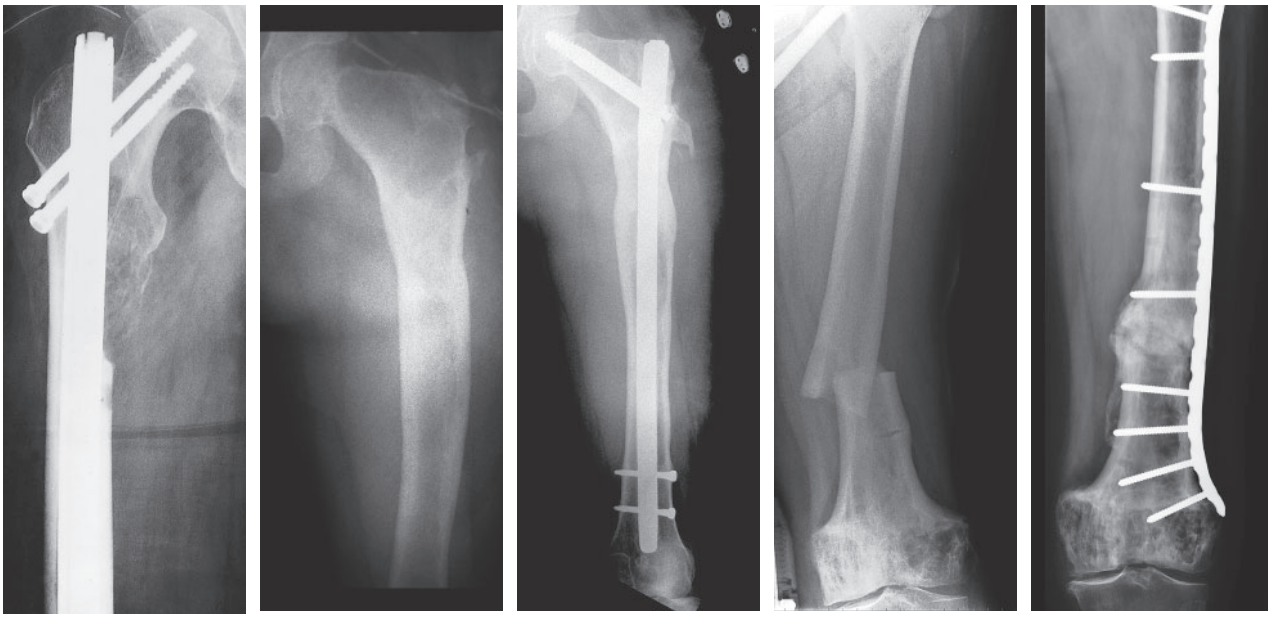
Patients with pathological fractures should be managed so that the reconstruction should outlive the patient and immediate full weight-bearing can occur postoperatively. Biopsy of solitary bone lesions should always be performed (see Chapter 9). Fractures through metastatic lesions should be fixed by intramedullary nailing. In patients with a good life expectancy an intercalary resection and spacer may be considered. Provided the patient is fit enough to tolerate the operation, a short life expectancy is not a contraindication. 'Prophylactic fixation' is also indicated if a lytic lesion is: (1) greater than half the diameter of the bone; (2) longer than 3 cm on any view; or (3) painful, irrespective of its size.

Paget's disease, fibrous dysplasia or rickets may present a problem. The femur is likely to be bowed and, in the case of Paget's disease, abnormally hard. An osteotomy to straighten the femur may be necessary to allow a nail to be inserted fully (Figure 30.29).

PERIPROSTHETIC FEMORAL FRACTURES

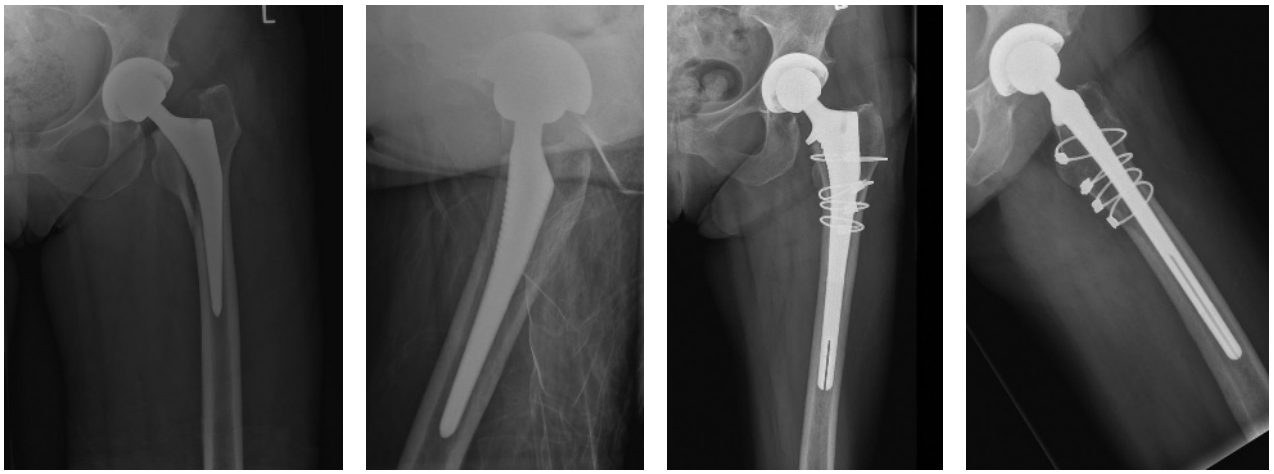
Femoral shaft fractures around a hip implant are relatively uncommon but the incidence is rising. They are most frequently classified using the Vancouver system, which uses the site of the fracture, whether the stem is loose and the available bone stock to categorize the fractures. Type A fractures occur in the trochanteric region (subdivided into A_G and A_L according to which trochanter is affected), type B occur in the diaphysis and the region up to two cortical diameters below the tip of the stem, and type C fractures occur well distal to the tip of the stem. In type B fractures, B1 fractures have a stable stem and adequate bone stock, B2 fractures have a loose stem and adequate bone stock, and B3 fractures have a loose stem and inadequate bone stock. Type B1 fractures are suitable for fixation rather than revision but should be carefully followed up in case of subsequent stem subsidence or loosening for those that have been misclassified. Type B2 and B3 fractures require revision surgery to address the loose stem. In B3 fractures, either long diaphyseal fitting stems with scaffold reconstruction of the remaining proximal bone or endoprosthetic replacement are required. Type C fractures occur distal enough to the construct that they can be treated as separate fractures. In all cases, the surgeon should be careful to avoid creating stress risers between implants that predispose to further fractures.

Femoral periprosthetic fractures may happen during primary or revision hip surgery during dislocation of the hip, when reaming or preparing the



(a) (b) (c) (d) (e)

Figure 30.29 Pathological fractures – internal fixation (a) Metastatic tumour, nailed before it actually causes a fracture. (b) Fibrous dysplasia with a stress fracture; (c) nailing provided the opportunity to correct the deformity. (d,e) Paget's disease, with a fracture; in this case (because of its site) treated by fixation with a plate and screws.



(a) (b) (c) (d)

Figure 30.30 Periprosthetic femoral fracture This patient sustained a Vancouver B2 periprosthetic fracture of the femur following a fall within the first few months of primary hip replacement (a,b). As the stem is loose, fixation is inappropriate and revision total hip replacement is required. The stem was revised to a longer stem with diaphyseal hold and the large posteromedial fragment that had separated and allowed the original stem to subside was cabled back on (c,d).

medullary canal, or when implanting trial or definitive implants. Periprosthetic fractures are more common with the use of uncemented implants. During revision surgery, they can also occur while extracting implants and cement. Sometimes the fracture occurs later, either as a consequence of trauma or in conjunction with osteolysis or implant loosening suggesting a reason for bone weakness. Osteolysis and loosening are usually apparent on X-ray.

Complications of femoral shaft fractures

EARLY

Shock Even with a closed fracture, 1–2 litres of blood can be lost and, if the injury is bilateral, shock may be severe. Prevention is better than cure; most patients will require a blood transfusion (see Chapter 22).

Fat embolism and ARDS Fracture through a large marrow-filled cavity almost inevitably results in small showers of fat emboli being swept to the lungs. This can usually be accommodated without serious consequences, but in some cases (and especially in those with multiple injuries and severe shock, or in patients with associated chest injuries) it results in progressive respiratory distress and multi-organ failure (adult respiratory distress syndrome). Blood gases should be measured if this is suspected and signs such as shortness of breath, restlessness or a rise in temperature or pulse rate should prompt a search for petechial haemorrhages over the upper body, axillae and conjunctivae. Treatment is supportive, with the emphasis on preventing hypoxia and maintaining blood volume.

Thromboembolism Prolonged traction in bed predisposes to thrombosis. Movement and exercise are important in preventing this, but high-risk patients should be given prophylactic anticoagulants as well. Vigilance is needed and full anticoagulant treatment is started immediately if thigh or pelvic vein thrombosis is diagnosed.

Infection In open injuries, and following internal fixation, there is always a risk of infection. Prophylactic antibiotics and careful attention to the principles of fracture surgery should keep the incidence below 2%. If the bone does become infected, the patient should be treated as for an acute osteomyelitis. Antibiotic treatment may suppress the infection until the fracture unites, at which time the metalwork can be removed and the canal reamed and washed out. However, if there is pus or a sequestrum, a more radical approach is called for: the wound is explored, all dead and infected tissue is removed and all metalwork as well; the canal is reamed and washed out and the fracture, if not united, is then stabilized by an external fixator. Replacement of the external fixator by another intramedullary nail can be risky, and much depends of the nature of the infecting organism (its sensitivity or resistance to antibiotics), the length of time during which the infection has been present and the quality of the surgical debridement. The long-term management of chronic osteomyelitis is discussed in Chapter 2.

LATE

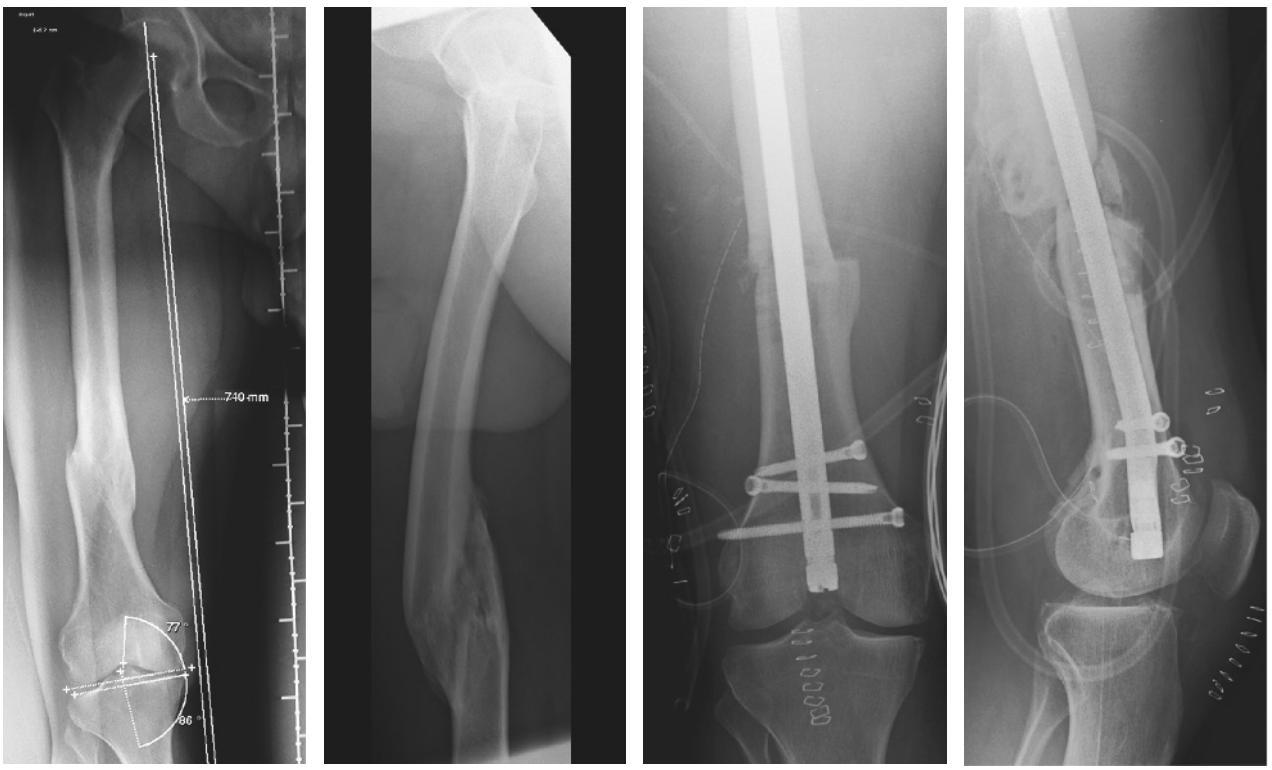
Delayed union and non-union The time-scale for declaring a delayed or non-union can vary with the type of injury and the method of treatment. If there is failure to progress by 6 months, as judged by serial X-rays, then intervention may be needed. A common

practice is to remove locking screws from the intramedullary nail to enable the fracture to 'collapse' ('*dynamize*' in modern orthopaedic parlance). This can be successful but may result in pain as rotational control of the fracture is lost (the femur is often subject to torsional forces in walking). A better course is to remove the nail, ream the medullary canal and introduce a larger diameter nail – *exchange nailing*. Bone grafts should be added to the fracture site if there are gaps not closed at the revision procedure.

Malunion Fractures treated by traction and bracing often develop some deformity; no more than 15 degrees of angulation should be accepted (Figure 30.31). Even if the initial reduction was satisfactory, until the X-ray shows solid union the fracture is too insecure to permit weight-bearing; the bone will bend and what previously seemed a satisfactory reduction may end up with lateral or anterior bowing. Malunion is much less likely in those treated with static interlocked nails; yet it does still occur – especially malrotation – and this can be prevented only by meticulous intraoperative and postoperative assessment followed, where necessary, by immediate correction. Shortening is seldom a major problem unless there was bone loss; if it does occur, treatment will depend on the amount and its clinical impact – sometimes all that is needed is a built-up shoe.

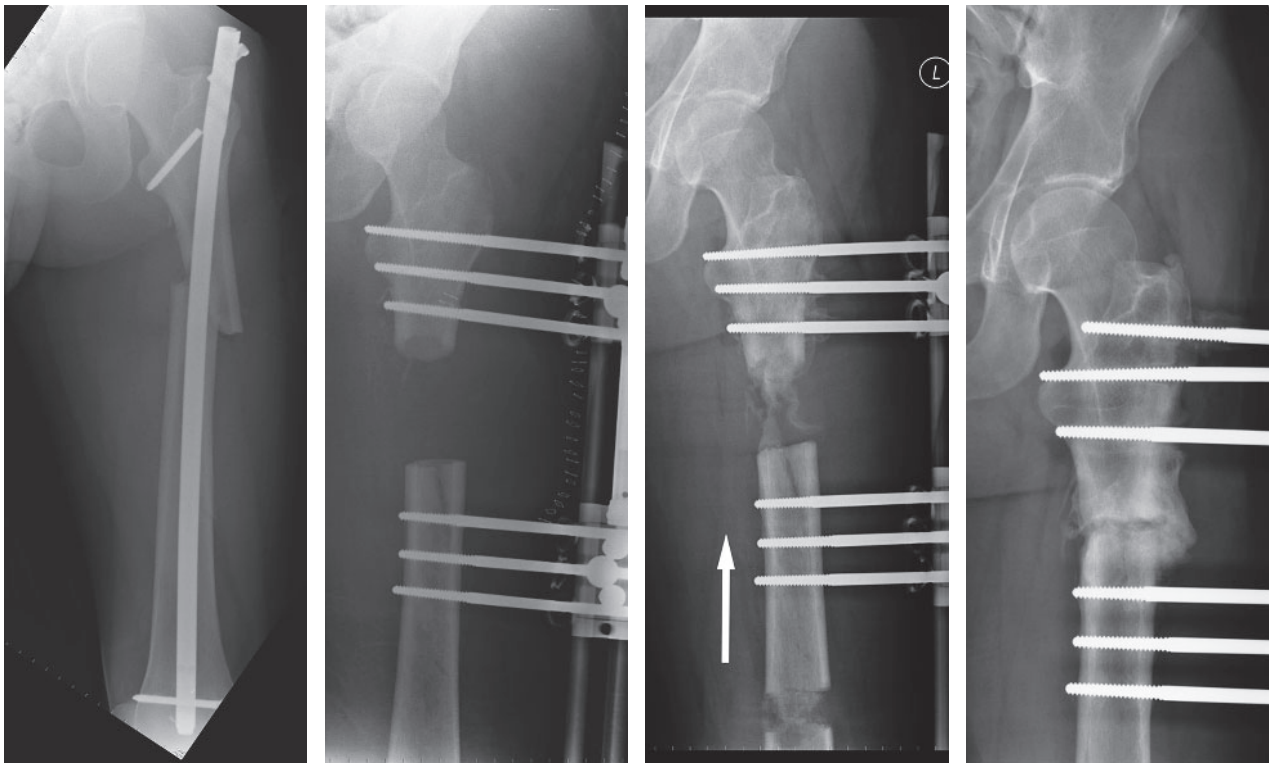
Joint stiffness The knee is often affected after a femoral shaft fracture. The joint may be injured at the same time, or it stiffens due to soft-tissue adhesions during treatment; hence the importance of early mobilization and physiotherapy.

Refracture and implant failure Fractures which heal with abundant callus are unlikely to recur. In those treated with absolute stability and internal fixation, bone healing is primary and there is no callus formation. If relative stability and internal fixation is used, bone healing is secondary and there will be callus formation as long as sufficient mechanical stability is achieved and an adequate biological environment maintained. With delayed union or non-union, the integrity of the femur may be almost wholly dependent on the implant, which, if union does not occur, will eventually fail by fatigue failure. If a comminuted fracture is plated, bone grafts should be added and weight-bearing delayed so as to protect the plate from reaching its fatigue limit too soon. Intramedullary nails are less prone to breakage. However, sometimes they do, especially with a slow-healing fracture of the



(a) (b) (c) (d)

Figure 30.31 Malalignment after treatment Treatment of femoral shaft fractures by traction can produce good results but, in some, a malunion can lead to symptoms. In this patient (a,b) the varus deformity produced knee symptoms from overloading of the medial compartment; this was relieved by corrective osteotomy and intramedullary nailing (c,d).



(a) (b) (c) (d)

Figure 30.32 Implant failure and non-union (a) This was an open injury with poor vascularity of the fracture ends. It was fixed with an intramedullary nail in the hope that it might unite. It didn't, and one of the proximal screws broke. The fracture ends were excised; an external fixator was applied (b); and an osteotomy was performed lower down (c); then the fracture ends were brought together with distraction osteogenesis at the osteotomy site. The fracture united (d).

distal third and a static locked nail; the break usually occurs through the screw hole closest to the fracture. Treatment consists of exchange nailing and bone grafting. In resistant cases, the fracture site may need excising (as viability of the bone ends is poor) followed by distraction osteogenesis which simultaneously stabilizes the limb and deals with the leg-length discrepancy (Figure 30.32).

FEMORAL SHAFT FRACTURES IN CHILDREN

Mechanism

Fractures of the femur are quite common in older children and are usually due to *direct violence* (e.g. an RTC) or a *fall* from a height. However, in children under 2 years of age the commonest cause is non-accidental injury; if there are several fractures in different stages of healing, this is very suspicious. *Pathological fractures* are common in generalized disorders such as spina bifida and osteogenesis imperfecta, and they may occur with local bone lesions (e.g. a benign cyst or tumour).

Treatment

The principles of treatment in children are the same as in adults but it should be emphasized that in young children open treatment is rarely necessary. The choice of closed method depends largely on the age and weight of the child. As children get older (and larger), fractures take longer to heal and conservative treatment is more likely to result in problems associated with long hospitalization and a greater risk of malunion. Coupled to this is the associated cost of protracted bed occupancy. Consequently, there has been a trend towards treating femoral shaft fractures in older children by operation, but the argument is flawed if this is based on cost alone – many of these children will have to return for implant removal. Perhaps it is the risk of malunion, particularly in unstable fracture patterns, that renders surgery a better option for older children and adolescents.

TRACTION AND CASTS

Infants need no more than a few days in balanced traction, followed by a spica cast for another 3–4 weeks. Angulation of up to 30 degrees can

be accepted, as the bone remodels quite remarkably with growth. Immediate spica casting has also found favour and this approach does not appear to increase the risk of complications. *Children between 2 and 10 years* of age can be treated either with balanced traction for 2–3 weeks followed by a spica cast for another 4 weeks, or by early reduction and a spica cast from the outset. Shortening of 1–2 cm and angulation of up to 20 degrees are acceptable.

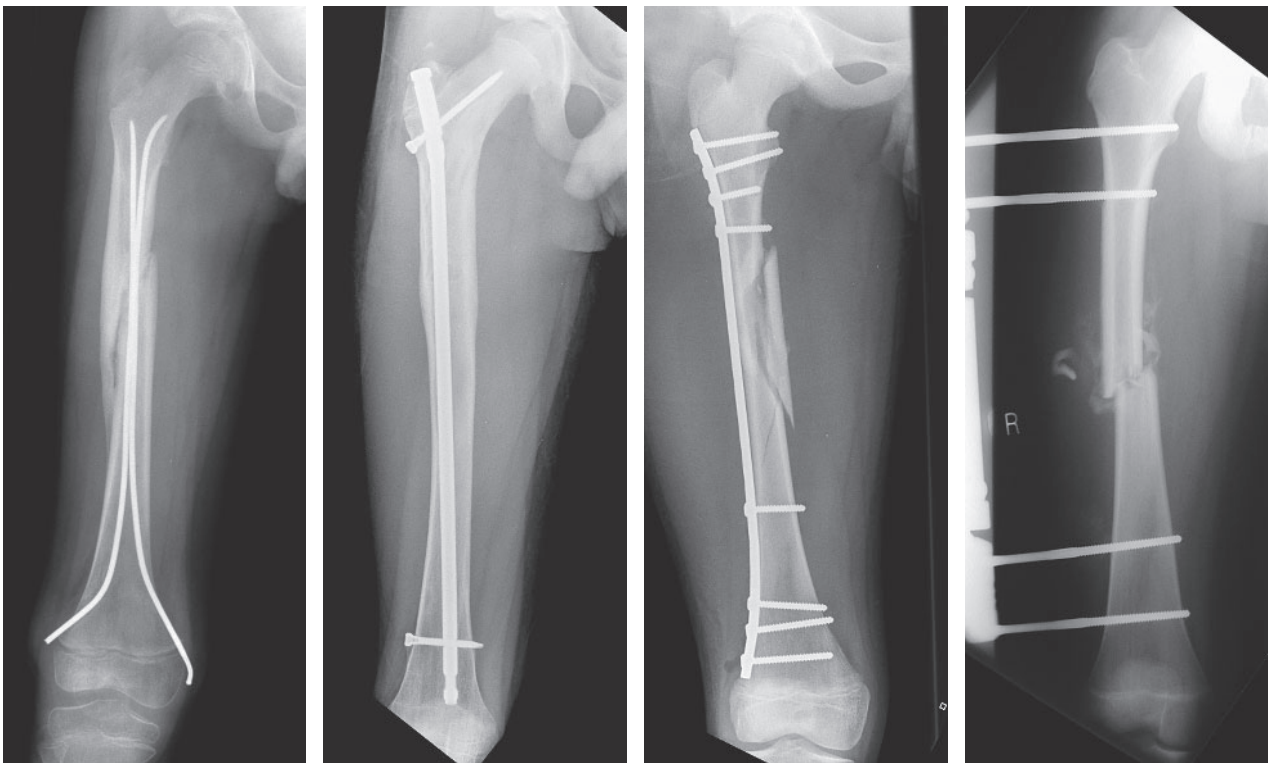
Teenagers require somewhat longer (4–6 weeks) in balanced traction, and those aged over 15 (or even younger adolescents if they are large and muscular) may need skeletal traction. Once the fracture feels firm, traction is exchanged for either a spica cast (in the case of upper-third and mid-shaft fractures) or a cast-brace (for lower-third fractures), which is retained for a further 6 weeks. The position should be checked every few weeks; the limit of acceptable angulation in this age group is 15 degrees on the anteroposterior X-ray and 25 degrees on the lateral X-ray. If a satisfactory reduction cannot be achieved by traction, internal fixation (plates or flexible intramedullary nails) or external fixation is justified. This applies to older children and those with multiple injuries.

OPERATIVE TREATMENT

This is growing in popularity as there is: (1) a shorter inpatient stay (and for the child, a quicker return home); (2) a lower incidence of malunion. Against this is the added risk of surgery, taking into account that many such fractures have good results when treated non-operatively. The tendency to adopt this approach in older children and adolescents may be justified. Surgical options include fixation with flexible intramedullary nails or trochanteric entry-point rigid nails with interlocking screws (neither of which damages the physes), plates inserted by the MIPO technique and external fixation (Figure 30.33).

Complications

Shortening Overlapping and comminution of the bone fragments may shorten the femur. However, anything up to 2 cm is quite acceptable in young children; indeed, some surgeons regard this as an advantage because there is a tendency for the fractured bone to grow faster for up to 2 years after the injury. This may be related to stimulation of the physes derived from the increased blood flow that accompanies fracture healing. Unfortunately, the effect on growth is unpredictable.



(a) (b) (c) (d)

Figure 30.33 Fixation techniques for femoral shaft fractures in children Non-operative treatment is safest for children. If surgery is indicated, options include: (a) flexible nailing; (b) trochanteric entry-point rigid nails; (c) plates and screws inserted by the minimally invasive percutaneous osteosynthesis (MIPO) technique; (d) external fixation.

Malunion Angulation can usually be tolerated within the limits mentioned above. However, the fact that bone remodelling is excellent in children is no excuse for casual management; bone may be forgiving but parents are not! Certainly rotational malunion is not corrected by growth or remodelling. It is probably wise to observe a malunited fracture for 2 years before offering a corrective osteotomy.

SUPRACONDYLAR FRACTURES OF THE FEMUR

Supracondylar fractures of the femur are encountered in young adults, usually as a result of high-energy trauma, and in elderly, osteoporotic individuals.

Mechanism and pathological anatomy

Direct violence is the usual cause. The fracture line is just above the condyles but may extend between them. In the worst cases the fracture is severely comminuted. A useful classification of these fractures is from the AO group:

- *Type A* – have no articular splits and are truly 'supracondylar'
- *Type B* – are simply shear fractures of one of the condyles
- *Type C* – have supracondylar and intercondylar fissures (Figure 30.34).

Gastrocnemius, arising from the posterior surface of the distal femur, will tend to pull the distal segment into extension, thus risking injury to the popliteal artery.

HOFFA FRACTURES

Hoffa fractures are an interesting variant of supracondylar femoral fractures. The defining characteristic is the presence of a fracture line in the coronal plane.

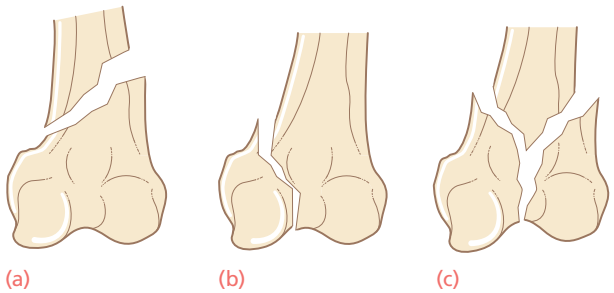


Figure 30.34 The AO classification of supracondylar fractures (a) Type A fractures do not involve the joint surface; (b) type B fractures involve the joint surface (one condyle) but leave the supracondylar region intact; (c) type C fractures have supracondylar and condylar components.

They are rare. Although they can occasionally be bicondylar, they are usually unicondylar and the lateral condyle is more commonly affected.

Clinical features

The knee is swollen because of a haemarthrosis – this can be severe enough to cause blistering later. Movement is too painful to be attempted. Distal pulses should always be checked to ensure the popliteal artery was not injured in the fracture.

Imaging

The entire femur should be X-rayed so as not to miss a proximal fracture or dislocated hip. The supracondylar fracture pattern will vary. Of importance are: (1) whether there is a fracture into the joint and if it is comminuted; (2) the size of the distal segment; and (3) whether the bone is osteoporotic. These factors influence the type of internal fixation required, if that is the chosen mode of treatment. Although most of these fractures can be diagnosed on X-ray, CT is often used as an adjunct to fully describe the pattern of the fracture and aid surgical decision making.

Treatment

NON-OPERATIVE TREATMENT

If the fracture is only slightly displaced and extra-articular, or if it reduces easily with the knee in flexion, it can be treated by traction through the proximal tibia; the limb is cradled on a Thomas splint with a knee flexion piece and movements are encouraged. If the distal fragment is displaced by gastrocnemius pull, a second pin above the knee, and vertical traction, will correct this. At 4–6 weeks, when the fracture is beginning to unite, traction can be replaced by a cast-brace and the patient allowed up and partially weight-bearing with crutches. Non-operative treatment should be considered as an option if the patient is young or the facilities and skill to treat by internal fixation are absent. Elderly patients tend not do as well with the 6 weeks of enforced recumbency.

SURGERY

Operative treatment with internal fixation can enable accurate fracture reduction, especially of the joint surface, and early movement of the joint to reduce the risk of stiffness. If the necessary facilities and skill are available, this is the treatment of choice. For the elderly, early mobilization is so important that internal fixation or replacement is almost obligatory. Sometimes the hold on osteoporotic bone is poor (despite modern implant designs)

or the patient may be old and frail, making early mobilization difficult or risky, but nursing in bed is made easier and knee movements can be started sooner. Several different devices are available (Figure 30.35):

- *Locked intramedullary nails* – These are introduced retrograde through the intercondylar notch – these are suitable for the type A and simpler type C fractures.
- *Plates* – These are applied to the lateral surface of the femur: traditional angled blade-plates or 95-degree condylar screw-plates. They are suitable for type A and the simpler type C fractures. For severely comminuted type C fractures, the newer plate designs with locking screws appear to offer an advantage over other implants; they provide adequate stability, even in the presence of osteoporotic bone. Care must be taken not to make the construct too stiff, as this inhibits union and predisposes to failure.
- *Simple lag screws* – These suffice for type B fractures and are inserted in parallel, with the screw heads buried within the articular cartilage to avoid abrading the opposing joint surface. They are also used to hold the femoral condyles together in type C fractures before intramedullary nails or lateral plates are used to hold the main supracondylar break and to lag on the condylar fragment in a Hoffa fracture (Figure 30.36).

In elderly patients with osteoporotic bone, there is a high rate of failure with attempted fixation of comminuted distal femoral fractures. Such patients may benefit from distal femoral replacement (Figure 30.37) in order to allow more rapid mobilization and to decrease the risk of failure. Knee movements are started soon after operation if wound healing allows. This limits adhesions forming within the knee joint.

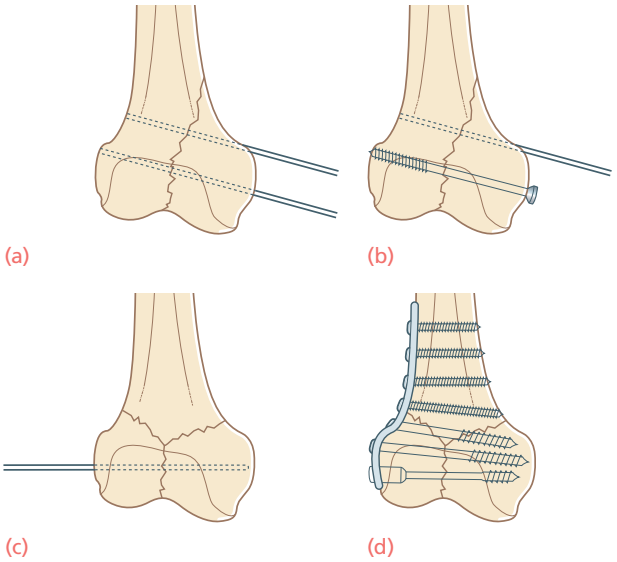


Figure 30.35 Femoral condyle fractures – treatment (a) A single condylar fracture can be reduced open and held with K-wires preparatory to (b) inserting compression screws. (c) T- or Y-shaped fractures are best fixed with a dynamic condylar screw and plate (d).

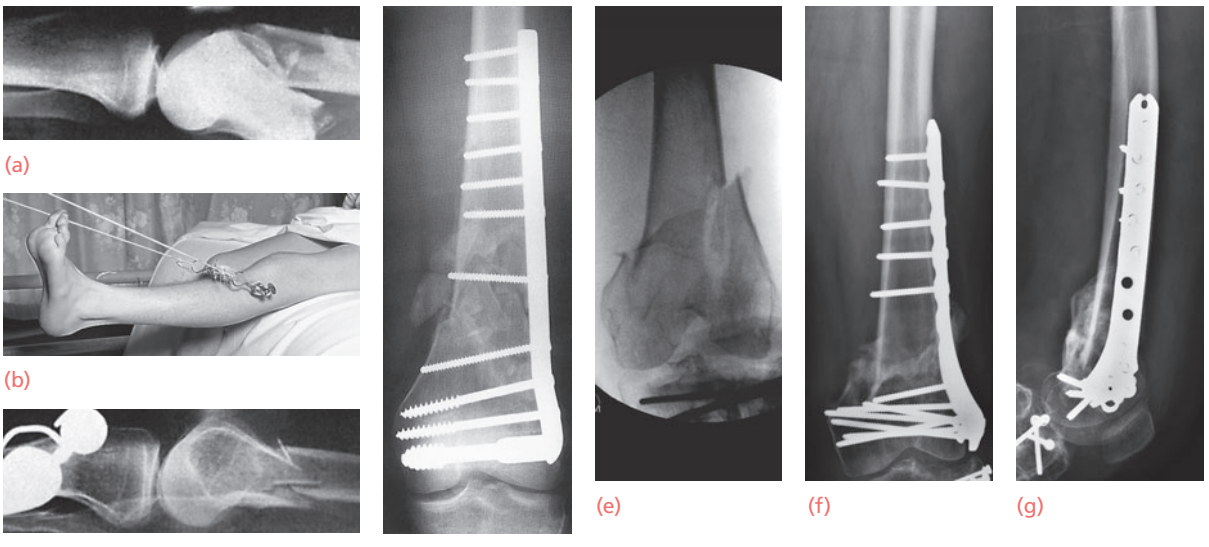
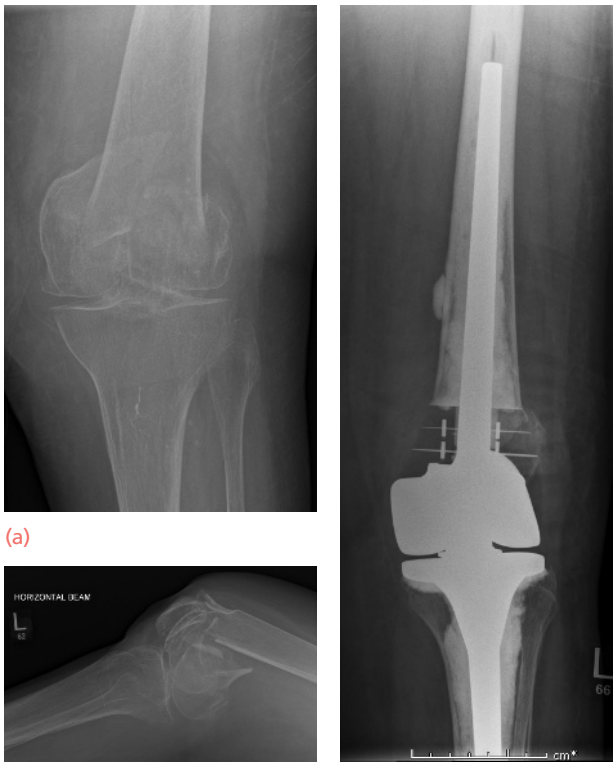


Figure 30.36 Supracondylar fractures (a–c) These fractures can sometimes be treated successfully by traction through the upper tibia. (d–g) If the bone is not too osteoporotic, internal fixation is often preferable and the patient can get out of bed sooner: a dynamic condylar screw and plate for a type A fracture (d) and a combination of lag screws and a lateral side plate for more complex fracture patterns (e,f,g).



(a) (b) (c) (d)

Figure 30.37 Supracondylar fracture of the femur (a,b) Highly comminuted distal femoral supracondylar fracture in a 92-year-old woman. (c,d) Postoperative images of a distal femoral endoprosthetic replacement.

Complications

EARLY

Arterial damage There is a small but definite risk of arterial damage and distal ischaemia. Careful assessment of the leg and peripheral pulses is essential, even if the X-ray shows only minimal displacement, as the degree of displacement at the time of injury would have been substantially greater.

LATE

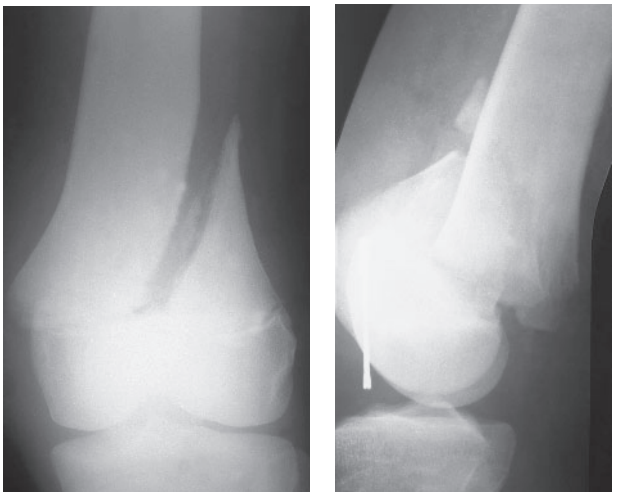
Joint stiffness Knee stiffness – probably due to scarring from the injury and the operation – is almost inevitable. A long period of rehabilitation is needed in all cases. For marked stiffness, arthroscopic division of adhesions in the joint or even a quadricepsplasty may be needed.

Malunion Internal fixation of these fractures is difficult and malunion – usually varus and recurvatum – is not uncommon. Corrective osteotomy may be needed for patients who are still physically active.

Non-union Modern surgical techniques of internal fixation recognize the importance of minimizing damage to the soft tissues around the fracture; where possible, only those parts that are essential for fracture reduction are exposed. The knee joint may need to be opened for reduction of articular fragments but the metaphyseal area is left untouched in order to preserve its vitality. If these precautions are taken, non-union is unlikely. If non-union does occur, autogenous bone grafts and a revision of internal fixation will be needed, particularly if there are signs that the fixation is working loose or has failed. Knee stiffness is another threat. Unless great care is exercised during mobilization, the ultimate range of movement at the knee may be less than that at the fracture!

FRACTURE-SEPARATION OF THE DISTAL FEMORAL EPIPHYSIS

In the child or adolescent, the lower femoral epiphysis may be displaced – either to one side (usually laterally) by forced angulation of the straight knee or forwards by a hyperextension injury. Although not nearly as common as physeal fractures at the elbow or ankle, this injury is important because of its potential for causing abnormal growth and deformity of the knee. The fracture is usually a Salter–Harris type 2 lesion – i.e. physeal separation with a large triangular metaphyseal bone fragment (Figure 30.38). Although this type of fracture



(a) (b)

Figure 30.38 Fracture-separation of the epiphysis

usually has a good prognosis, asymmetrical growth arrest is not uncommon and the child may end up with a valgus or varus deformity. All grades of injury, but especially Salter–Harris types 3 and 4, may result in femoral shortening. Nearly 70% of the femur's length is derived from the distal physis, so an early arrest can present a major problem.

Clinical features

The knee is swollen and perhaps deformed. The pulses in the foot should be palpated because, with forward displacement of the epiphysis, the popliteal artery may be obstructed by the lower femur.

Treatment

The fracture can usually be perfectly reduced manually, but further X-ray checks will be needed over the next few weeks to ensure that reduction is maintained. Occasionally open reduction is needed; a flap of periosteum may be trapped in the fracture line. Salter–Harris types 3 and 4 should be accurately reduced and fixed. If there is a tendency to redisplacement, the fragments may be stabilized with percutaneous K-wires or lag screws driven across the metaphyseal spike. The limb is immobilized in plaster and the patient is allowed partial weight-bearing on crutches. The cast can be removed after 6–8 weeks and physiotherapy started.

Complications

EARLY

Vascular injury There is danger of ischaemia unless the hyperextension injury is reduced without delay.

LATE

Physeal arrest Damage to the physis is not uncommon and residual deformity may require corrective osteotomy at the end of the growth period. Small areas of tethering across the growth plate can sometimes be successfully removed and normal growth restored. Shortening, if it is marked, can be treated by femoral lengthening.

FURTHER READING

Baker RP, Squires B, Gargan MF, *et al.* Total hip arthroplasty and hemiarthroplasty in mobile, independent patients with a displaced intracapsular fracture of the femoral neck: a randomized, controlled trial. *J Bone Joint Surg Am* 2006; **88**(12): 2583–9.

Bonnaire FA, Weber AT. The influence of haemarthrosis on the development of femoral head necrosis following intracapsular femoral neck fractures. *Injury* 2002; **33** Suppl 3: C33–40.

Boulton CL, Pollak AN. Special topic: Ipsilateral femoral neck and shaft fractures - does evidence give us the answer? *Injury* 2015; **46**(3): 478–83.

Bretherton CP, Parker MJ. Early surgery for patients with a fracture of the hip decreases 30-day mortality. *Bone Joint J* 2015; **97**(1): 104–8.

Costa ML, Griffin XL, Pendleton N, *et al.* Does cementing the femoral component increase the risk of peri-operative mortality for patients having replacement surgery for a fracture of the neck of femur? Data from the National Hip Fracture Database. *J Bone Joint Surg Br* 2011; **93**(10): 1405–10.

Dimon JH III, Hughston JC. Unstable intertrochanteric fractures of the hip. *J Bone Joint Surg Am* 1967; **49A**: 440–50.

Garbuz DS, Masri BA, Duncan CP. Periprosthetic fractures of the femur: principles of prevention and management. *Instr Course Lect* 1998; **47**: 237–42.

Garden RS. Low angle fixation in fractures of the femoral neck. *J Bone Joint Surg Br* 1961; **43B**: 647–63.

Hart GP, Kneisl JS, Springer BD, *et al.* Open reduction vs distal femoral replacement arthroplasty for comminuted distal femur fractures in the patients 70 years and older. *J Arthroplasty* 2017 Jan;**32(1)**:202–206.

Jones MD, Carpenter C, Mitchell SR, *et al.* Retrograde femoral nailing of periprosthetic fractures around total knee replacements. *Injury* 2016; **47**(2): 460–4.

Keating JF, Grant A, Masson M, *et al.* Randomized comparison of reduction and fixation, bipolar hemiarthroplasty, and total hip arthroplasty: treatment of displaced intracapsular hip fractures in healthy older patients. *J Bone Joint Surg Am* 2006; **88A**: 249–60.

Ohly NE, Whitehouse MR, Duncan CP. Periprosthetic femoral fractures in total hip arthroplasty. *Hip Int* 2014; **24**(6): 556–67.

Parker MJ, Gurusamy K. Internal fixation versus arthroplasty for intracapsular proximal femoral fractures in adults. *Cochrane Database Syst Rev* 2006; (4): CD001708.

Pipkin G. Treatment of grade IV fracture dislocation of the hip. *J Bone Joint Surg* 1957; **39**: 1027–42.

Robinson PM, O'Meara MJ. The Thomas splint: its origins and use in trauma. *J Bone Joint Surg Br* 2009; **91**(4): 540–4.

Thompson VP, Epstein VP. Traumatic dislocation of the hip. *J Bone Joint Surg Am* 1951; **33A**: 746–78.

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Injuries of the knee and leg

31

Nick Howells

The knee is a complex joint which is vulnerable to injury. The bony structure of the knee joint is inherently unstable; were it not for the strong capsule, intra- and extra-articular ligaments and controlling muscles, the knee would not be able to function effectively as a mechanism for support, balance and thrust (Figure 31.1, and see below). The tibia is also particularly vulnerable to injury: because of its subcutaneous position, it is more commonly fractured, and more often sustains an open fracture, than any other long bone.

KNEE LIGAMENT INJURIES

Mechanism of injury and pathological anatomy

Most ligament injuries occur while the knee is bent because the capsule and ligaments are relaxed and the femur is allowed to rotate on the tibia. The damaging force may be a straight thrust such as a dashboard injury forcing the tibia backwards or, more commonly, a combined rotation and thrust as in a football tackle.

Injuries of the knee ligaments are common, particularly in sporting pursuits but also in road accidents, where they may be associated with fractures or dislocations. They vary in severity from a simple sprain to complete rupture. The direction and nature of the force will directly influence the structure or structures injured. It is important to recognize that these injuries are seldom 'unidirectional'; they often involve more than one structure and it is therefore useful to refer to them in functional terms (such as 'anteromedial instability') as well as anatomical terms (such as 'torn medial collateral ligament (MCL)').

The cruciate ligaments provide both anteroposterior and rotary stability. They also help to resist

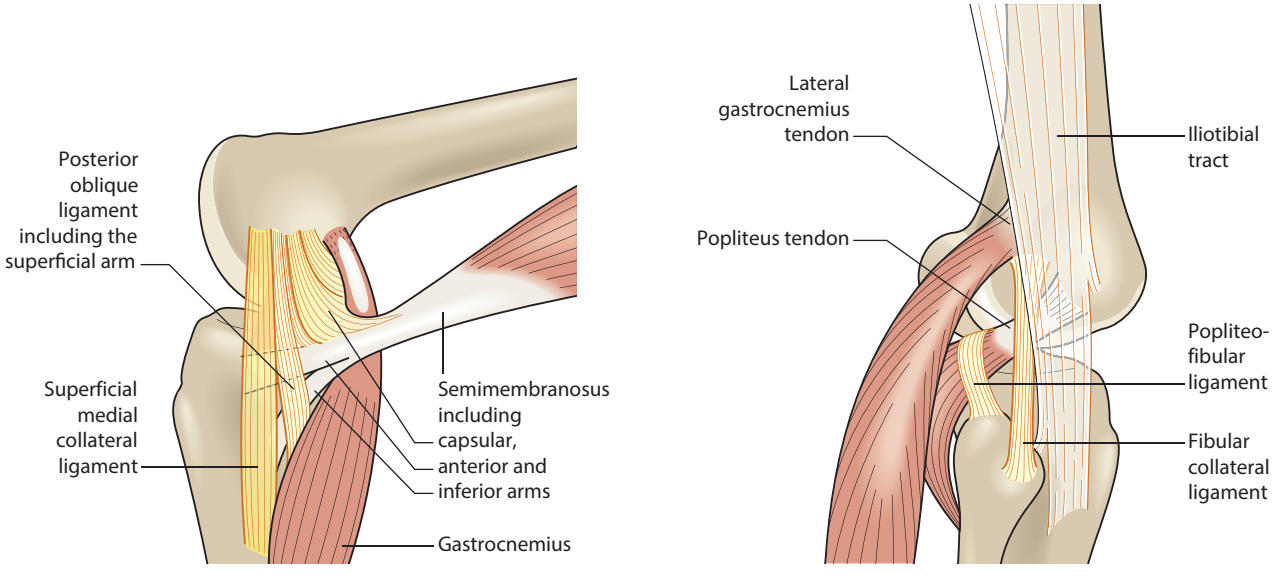


Figure 31.1 Extracapsular restraints to valgus and varus stresses on the knee (a) Restraints on valgus stresses: the deep and superficial parts of the medial collateral ligament, semimembranosus and the posterior oblique ligament. (b) Extracapsular restraints on varus stresses: lateral collateral ligament, popliteus tendon, popliteofibular ligament and the capsule.

excessive valgus and varus angulation. Both cruciate ligaments have a double bundle structure. The fibres within each bundle are arranged in such a way that at any position of knee flexion some of the fibres are taut. The anterior cruciate ligament (ACL) has anteromedial and posterolateral bundles, whereas the posterior cruciate has anterolateral and posteromedial bundles. Anterior displacement of the tibia (as in the anterior drawer test) is resisted by the anteromedial bundle of the ACL, while the posterolateral part tightens as the knee extends. Posterior displacement is prevented by the posterior cruciate ligament (PCL), specifically by the anterolateral bundle when the knee is in near 90 degrees of flexion and by the posteromedial bundle when the knee is straight (Figure 31.2).

Valgus stresses are resisted by the superficial and deep layers of the medial collateral ligament, semimembranosus tendon and its expansions, the tough posteromedial part of the capsule (referred to as the posterior oblique ligament) as well as the cruciate ligaments (Figure 31.1a). Depending on the position of the knee, some will act as primary and others as secondary stabilizers. At 30 degrees of flexion, the MCL is the primary stabilizer.

The main checks to varus angulation are the iliotibial band and the lateral (fibular) collateral ligament (LCL). Structures forming the posterolateral corner of the knee also make an important contribution to stability; they comprise the popliteus tendon and the popliteofibular ligament in addition to the LCL (Figure 31.1b). The iliotibial band and LCL are the primary stabilizers to a varus stress between full extension and 30 degrees of flexion; however, as flexion increases, the LCL relaxes and the posterolateral structures come into play to provide additional stability.

Clinical features

The patient gives a history of a twisting or wrenching injury and may even claim to have heard a 'pop' as the tissues snapped. The knee is painful and is usually swollen – and, in contrast to meniscal injury, the swelling appears almost immediately. Tenderness is most acute over the torn ligament, and stressing one or other side of the joint may produce excruciating pain. The knee may be too painful to permit deep palpation or much movement.

Abrasions suggest the site of impact, but bruising is more important and indicates the site of damage. The doughy feel of a haemarthrosis distinguishes ligament injuries from the fluctuant feel of the synovial effusion of a meniscus injury. The entire limb should be examined for other injuries and for vascular or nerve damage.

The most important aspect of the examination is to *test for joint stability*. Partial tears may permit no abnormal movement, but the attempt can cause pain. Complete tears tend to permit abnormal movement, which sometimes is almost painless.

Anteroposterior stability is assessed first by placing the knees at 90 degrees with the feet resting on the couch and looking from the side for posterior sag of the proximal tibia; when present, this is a reliable sign of posterior cruciate damage (Figure 31.3). Next, the *anterior and posterior drawer tests* are carried out; a positive drawer sign is diagnostic of a tear, but a negative test does not exclude one. The *Lachman test* is a more reliable test for AP instability; anteroposterior glide is tested with the knee flexed 15–20 degrees (Figure 31.4). Posterior cruciate ligament insufficiency can be confirmed with a positive *quadriceps active test* (Figure 31.5b). Rotational stability arising from acute injuries can usually be tested only under anaesthesia. After the

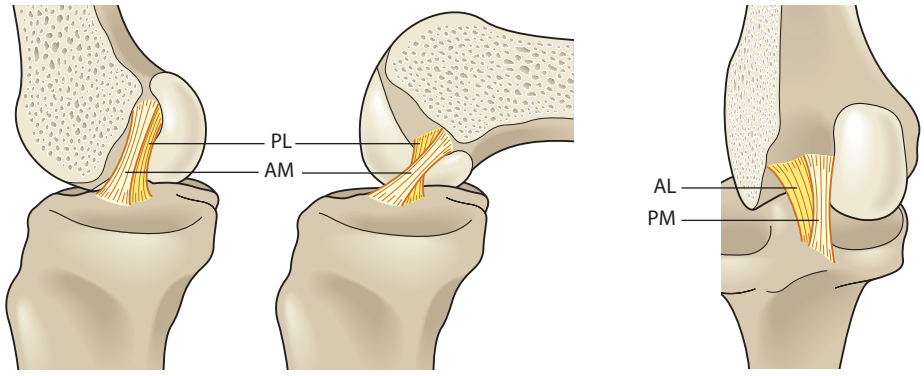
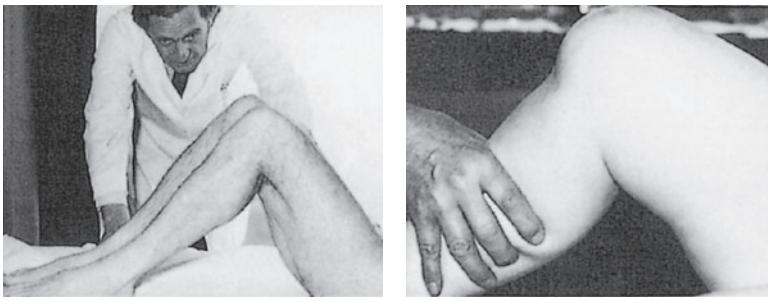
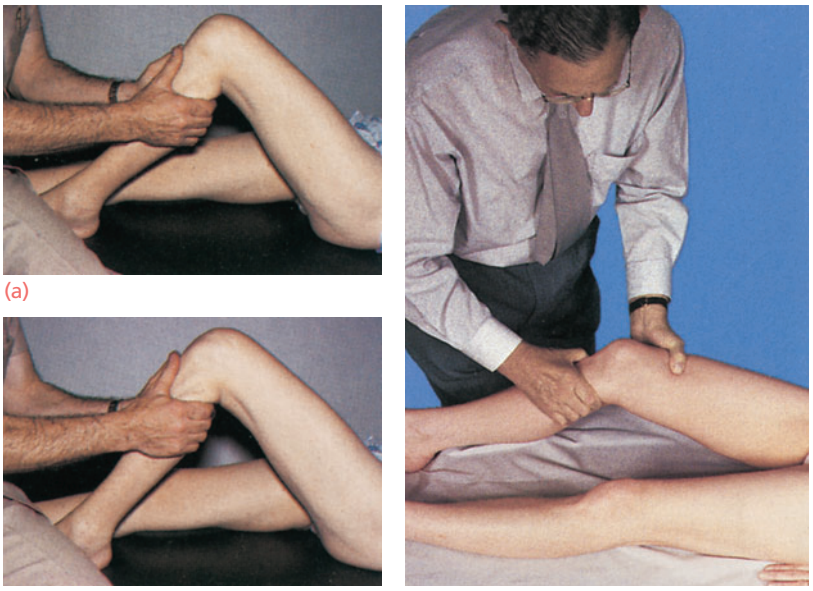


Figure 31.2 Dual-bundle structure of the anterior and posterior cruciate ligaments (a) The anteromedial (AM) bundle of an anterior cruciate ligament is taut in 90° of knee flexion whereas the posterolateral (PL) bundle tightens in extension. (b) In contrast, it is the anterolateral (AL) bundle of the posterior cruciate ligament that is tight in 90° flexion and the posteromedial (PM) bundle tightens in extension (and therefore resists hyperextension).



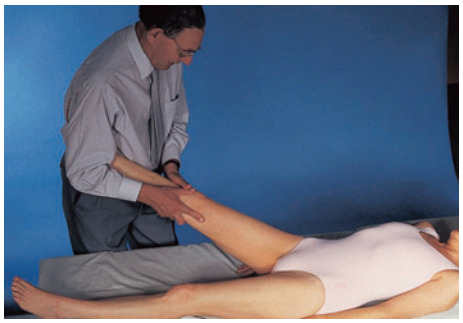
(a) (b)

Figure 31.3 Posterior cruciate ligament (a) Viewed from the side, any backwards displacement of the upper tibia is plainly visible and can be confirmed by (b) pushing the tibia backwards.



(a) (b) (c)

Figure 31.4 Tests for anterior cruciate ligament instability (a) *Drawer test:* With the knee at 90° and the hamstrings relaxed, grasp the top of the patient's leg and try to shift it forwards and backwards. (b) Note that there is some anterior shift in this patient when the tibia is pulled forwards - mild anterior cruciate laxity. (c) *Lachman test:* This is more sensitive than the drawer test. Note the position of the knee and the examiner's hands. There are modifications to this technique for more cumbersome knees.



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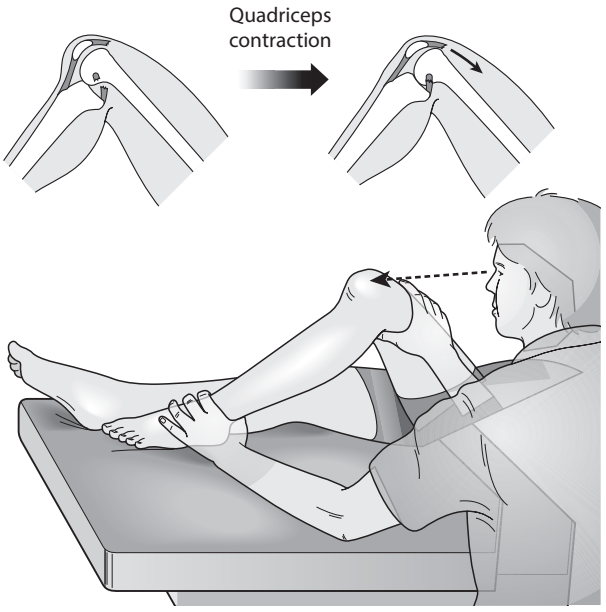


(b)

Figure 31.5 (a) *Testing collateral ligaments.* Side-to-side stability of the knee can be checked by holding the foot between the upper arm and body and moving the joint between supporting hands. This method is useful if the leg is large. (b) *The quadriceps active test.* Note the position of the examiner's hands in supporting the thigh and resisting knee extension by the ankle. At 90° of knee flexion, a posterior sag caused by a damaged posterior cruciate ligament is corrected when the quadriceps contracts.

acute injury has settled it is possible to perform a pivot shift test for ACL rotatory insufficiency whereby the lateral tibial condyle can be made to sublux forwards as the tibia rotates abnormally around an axis through the medial condyles.

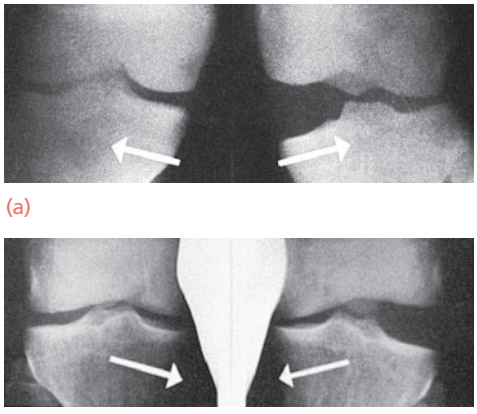
Sideways tilting (varus/valgus) is examined, first with the knee at 30 degrees of flexion and then with the knee straight (Figure 31.5a). Movement is compared with the normal side. If the knee angulates only in slight flexion, there is probably an isolated tear of

the collateral ligaments; if it angulates in full extension, there is almost certainly rupture of the capsule and cruciate ligaments as well. See Chapter 20 for more detailed descriptions of knee ligament stability tests.

Imaging

X-rays *Plain X-rays* may reveal associated fractures to the femoral condyles or tibial plateau in addition to small bony avulsions that can occur in conjunction with a ligament injury.
Bone may be avulsed from the:

- medial epicondyle of the femur at the origin of the MCL (*Pellegrini-Stieda lesion*)
- tip of the fibula, probably from the LCL or a posterolateral corner injury
- tibial spine at the insertion of the ACL
- central portion of the posterior tibia at the insertion of the PCL



(a)

(b)

Figure 31.6 Stress X-rays Stress films show: (a) complete tear of medial ligament, left knee; (b) complete tear of lateral ligament. In both, the anterior cruciate also was torn.

- near edge of the lateral tibial condyle by the iliotibial tract or capsule – this is called a *Segond fracture* and is associated with an ACL injury.

Stress X-rays are useful to document the degree of instability secondary to ligament injury. These can be performed on the table in theatre or in the radiology department. They should be compared to the contralateral limb and are particularly useful in assessing the multiligament-injured knee (Figure 31.6).

Computed tomography CT is useful for the delineation of subtle fractures associated with ligament injuries and to determine exact fracture patterns. This is useful in planning reconstruction. CT is not useful for assessment of ligamentous structures.

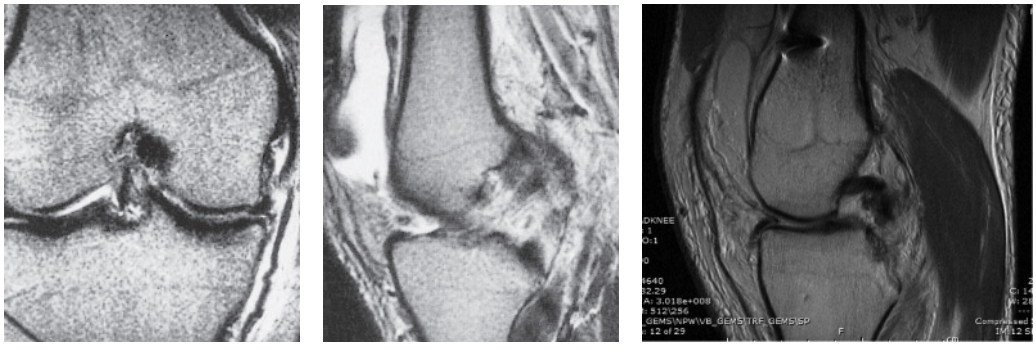
Magnetic resonance imaging MRI is the gold standard imaging tool for the assessment of knee ligament injuries. Increased availability has led to the widespread use of MRI to diagnose ligament, chondral and meniscal injuries, providing almost 100% sensitivity and over 90% accuracy (Figure 31.7).

Anterior cruciate ligament injury

Clinical features

ACL injuries are one of the most common knee injuries, particularly in women. The classic history is of an axial-loading twisting injury on a slightly flexed knee. This commonly occurs when suddenly changing direction or landing and twisting from a jump in pivoting sports such as football, netball and rugby. Risk factors include joint hypermobility, genetic predisposition, elevated BMI and increased tibial slope. The bleeding associated with ligament rupture will cause an acute haemarthrosis and the patient will describe swelling almost immediately.

It is very rare for the ACL to heal satisfactorily as the synovial fluid present around this intra-articular



(a) (b) (c)

Figure 31.7 Torn knee ligaments – MRI (a) Coronal T1-weighted image showing a medial collateral ligament tear with surrounding oedema and joint effusion. (b) Sagittal T1-weighted image showing an intra-substance tear of the anterior cruciate ligament with a large joint effusion. (c) Coronal T1-weighted image showing a PCL rupture.

ligament is thought to prevent formation of a consolidated clot that would promote a healing response in the tendon. In some individuals, however, the knee can stabilize symptomatically and clinically after a period of appropriate rehabilitation.

Treatment

ACL deficiency can be overcome with appropriate quadriceps rehabilitation and conscious control of the knee to minimize sagittal instability. This can allow individuals to return to sport. This does pose a risk, however, as unconscious quadriceps control can seldom be achieved. The potential therefore remains for future pivot instability in an ACL-deficient knee causing subsequent irreversible damage to the chondral and meniscal surfaces of the knee.

The majority of patients who sustain an ACL injury will continue to have demonstrable sagittal plane instability despite rehabilitation. In this situation treatment options consist of activity modification to avoid running, jumping, cutting and side-stepping activity, all of which put an ACL-deficient knee at risk, or surgical reconstruction.

ACL reconstruction is a widely performed operation for patients with persistent symptomatic instability in order to provide a functionally stable knee with reduced potential for secondary injury and long-term morbidity. The goals of ACL reconstruction (ACLR) are to stabilize the knee joint, restore normal kinematics and prevent early-onset degenerative arthrosis. Over the last 25–30 years ACLR techniques have evolved and yielded significant improvements in the clinical outcomes following ACL injury. The goal of modern surgical techniques is an intra-articular anatomical reconstruction in order to restore kinematics to as close to normal as possible. Graft options most commonly used are autologous hamstring tendons (semitendinosus + gracilis) or patella tendon. A number of other autologous, allogeneic and synthetic grafts have been trialled and are used with varying success. Graft should be of sufficient diameter to be strong enough to provide an adequate reconstruction. Four-stranded autologous hamstring grafts are the most commonly used worldwide but these sometimes produce too small a graft in smaller patients and therefore other options also need to be considered. Bony tunnels are drilled in the femur and the tibia within the footprint of the native ACL. The graft is pulled through these tunnels into position, tensioned then fixed to maintain this tension. Various fixation techniques are used but commonly include screws, staples and suspensory devices.

A stepwise structured quadriceps rehabilitation programme is important following surgery, focusing on range of movement, strengthening, proprioception and eccentric control. Patients should be counselled that return to pivoting sports is not advised for a minimum of 9–12 months. Active injury prevention programmes such as the PEP (Prevent injury and Enhance Performance) programme have been shown to dramatically reduce the risk of both primary ACL injury and re-injury following ACL reconstruction in soccer players. This is a focused series of stretches and muscle warm-up exercises that focus on strength, awareness and neuromuscular control. Well-performed, uncomplicated surgery can yield near-normal knee function at 18 months post surgery. At this stage postop rates of re-injury to the reconstructed knee reach equivalence with injury rates to the contralateral uninjured knee.

Posterior cruciate ligament injury

Clinical features

The PCL is most commonly injured by a direct anterior blow or a rapid deceleration injury such as the knee striking the dashboard in a motor vehicle collision. The initial clinical symptoms are very similar to an ACL injury. PCL rupture is much less common than ACL rupture. The PCL is a much stronger ligament with higher load to failure than the ACL.

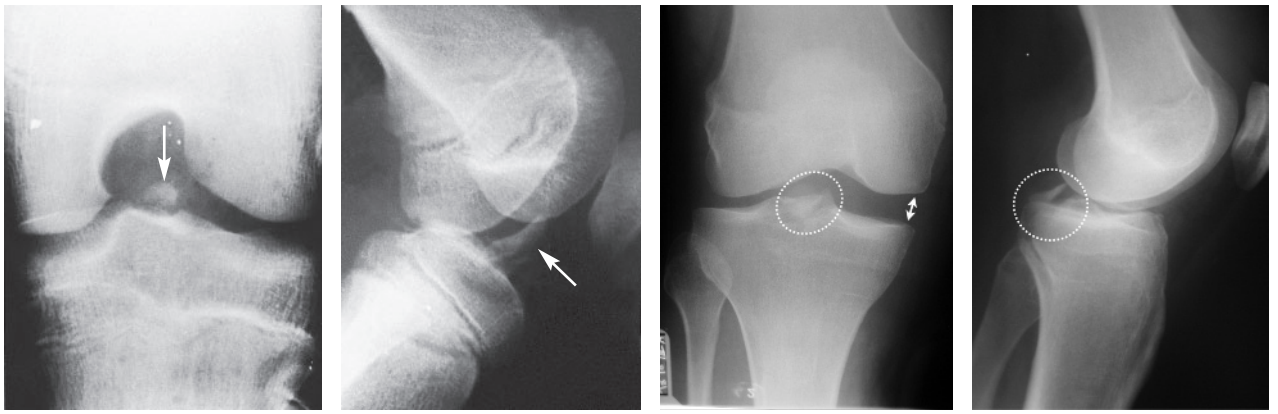
Treatment

With appropriate quadriceps rehabilitation, the majority of patients with an isolated PCL injury will return to a high level of function and avoid surgical intervention. Surgical reconstruction of the PCL is generally reserved for patients with marked and persistent instability despite rehabilitation.

Bony ACL/PCL ligament avulsions

A tibial spine fracture is a bony avulsion of the tibial insertion of the ACL. Often this is a traction type injury and it is seen most commonly in adolescents. The detached bone fragment may remain almost undisplaced, held in position by the soft tissues, it may be partially displaced, the anterior end lifted away on a posterior hinge, or it may be completely detached and displaced. Because the articular surface of the bony fragment is covered with cartilage, which is radiotranslucent, the image seen on radiographs is smaller than the actual fragment (Figure 31.8). CT gives a better impression of fragment displacement and position. MRI identifies any associated ligamentous, chondral or meniscal injuries. PCL bony avulsions are from the central posterior portion of the tibia.

Undisplaced avulsions can be managed non-operatively in a brace or cast. Displaced ACL and PCL avulsions are best managed with operative reduction and fixation. Both open and arthroscopic techniques



(a) (b) (c) (d)

Figure 31.8 Bony avulsion fractures (a,b) AP, lateral X-rays showing a large, displaced avulsion fracture of the tibial spine consistent with a bone ACL injury. This young man injured his knee while playing football. (c,d) AP and lateral X-rays showing a bony avulsion of the PCL.

have been described and are performed depending on fragment size, position and surgeon preference and experience. The fragments are held with screws, anchors or wires. Outcomes are usually good and patients regain good function. However, there is potential for some residual laxity if ligament fibres have been stretched when sustaining the injury.

Medial collateral ligament injury

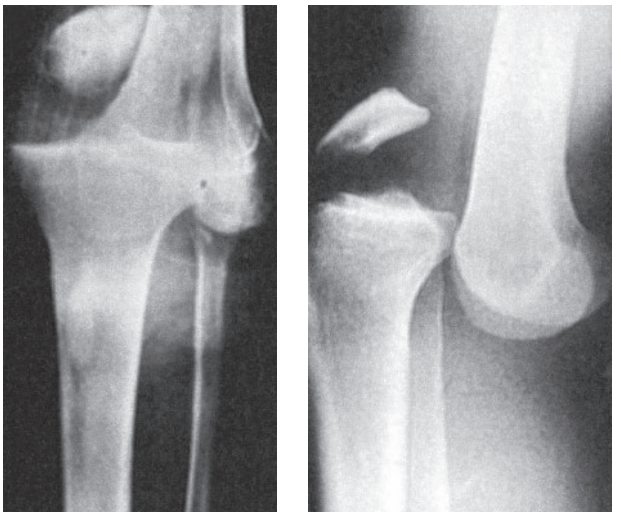
Isolated MCL tears are relatively rare. MCL injuries usually occur in conjunction with injuries to other structures, particularly the ACL and medial meniscus. An isolated MCL injury is caused by a purely valgus force. Partial ligament injuries and sprains will tend to stabilize in a brace as the intact fibres splint the torn ones and spontaneous healing will occur. Low-grade full-thickness injuries also have good healing potential with an appropriate bracing regime, which maintains range of motion and full flexion but limits full extension beyond 10 degrees of flexion, as this will prevent undue stress on the healing ligament. Rarely an isolated complete tear will require surgery, most commonly in the form of direct repair. Chronic instability from a missed or inappropriately braced MCL may require surgical reconstruction, although outcomes remain variable.

Lateral collateral ligament injury

Isolated LCL tears are also rare and result from a purely varus stress. Careful clinical examination is required to differentiate an isolated LCL from a more extensive posterolateral corner (PLC) injury. Isolated LCL injury can be managed non-operatively with bracing as for an MCL injury. If there is a bony avulsion, this warrants repair and often is an indication of a PLC injury. A PLC injury may benefit from acute repair +/– reconstruction.

Knee dislocation / multiligament injury

The dislocated knee is an under-diagnosed injury which relies on a high index of clinical suspicion on presentation of any knee injury. Traumatic knee dislocations are uncommon yet serious injuries that historically have had variable prognosis. A knee dislocation describes complete disruption of the integrity of the tibiofemoral articulation (Figure 31.9). Such disruption will result in a multiligament knee injury, defined most commonly as rupture to at least two of the four major knee ligament structures. Half of them are secondary to road traffic accidents (high-velocity dislocations), approximately a third are sports injuries (low-velocity dislocations) and nearly 10% are from simple falls (ultra-low-velocity dislocations).



(a) (b)

Figure 31.9 Knee dislocation X-rays showing an anterior dislocation of the knee.

Clinical features

For an acute dislocation, efficient patient assessment and management according to basic advanced trauma life support (ATLS) principles is the initial priority. A high index of suspicion is required, as a dislocated knee may have reduced spontaneously or may have been reduced before transfer to hospital. Rupture of the joint capsule produces a leak of the haemarthrosis, leading to severe bruising and swelling. The circulation in the foot must be examined because the popliteal artery may be torn or obstructed. Repeated examination is necessary as ischaemia may evolve and compartment syndrome is also a risk.

Acute ischaemia is a surgical emergency and requires in theatre angiography, vascular surgical involvement and revascularization. Subtle suspicion of vascular injury should be investigated with ankle brachial pressure index (ABPI) and vascular imaging such as angiography, CT angiography or MR angiography. ABPI of <0.9 should prompt vascular imaging. Common peroneal nerve injury occurs in nearly 20% of cases; distal sensation and movement should be tested and documented carefully.

Treatment

Should the knee remain dislocated at presentation, it should be reduced under conscious sedation or in the operating theatre as soon as possible and held with brace, plaster or external fixator. This is usually achieved by pulling directly in the line of the leg, but hyperextension must be avoided because of the danger to the popliteal vessels. For the definitive management of the multiligament-injured knee, a non-operative approach was formerly advocated in view of concerns over postoperative stiffness. More recent evidence has, in contrast, shown considerable benefit from surgical treatment in terms of improved functional outcomes, return to work and return to sport. There is increasing consensus that surgical intervention should be performed early (within 2–3 weeks). A recent systematic review of early versus delayed surgery found significantly better outcomes for early intervention.

A wide variety of surgical techniques have been described in the management of these injuries including primary repair, repair and augmentation, and reconstruction. The important principle is to define the components contributing to the instability and to repair or reconstruct the primary restraints as anatomically and isometrically as possible.

Complications

EARLY

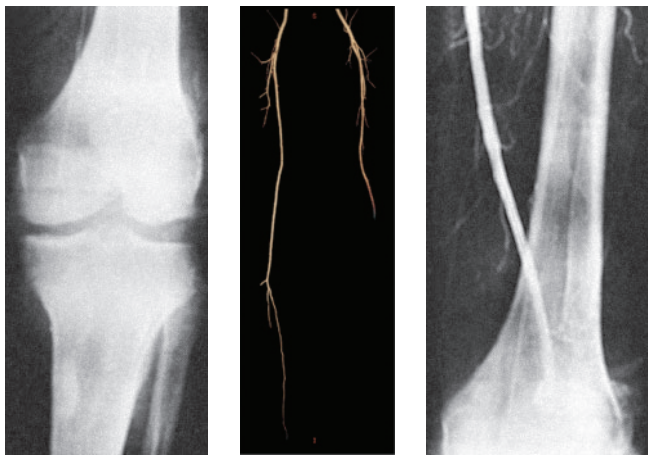
Arterial damage This occurs in 8–14% of patients. Popliteal artery injury causing acute ischaemia needs immediate repair. Delay resulting in an extended warm ischaemic period can result in amputation (Figure 31.10).

Common peroneal nerve injury Nerve injury can result in weak or absent ankle dorsiflexion. Spontaneous recovery is possible if the nerve is not completely disrupted and about 20% of patients with neuropraxia can be expected to improve. If nerve conduction studies or clinical examination show no sign of recovery, a transfer of tibialis posterior tendon through the interosseous membrane to the lateral cuneiform may help restore ankle dorsiflexion.

LATE

Joint instability Mild residual joint instability despite repair and/or reconstruction is common. Appropriately rehabilitated quadriceps muscle can compensate for mild instability and the functional disability is rarely severe.

Stiffness Stiffness due to prolonged immobilization and post-injury scarring is a common problem and it may be more troublesome than instability. Even with early surgical reconstruction, normal knee function is elusive after these severe injuries.



(a) (b) (c)

Figure 31.10 Knee dislocation and vascular trauma (a) This patient was admitted with a dislocated knee. After reduction the X-ray looked satisfactory, but the circulation did not. (b,c) An arteriogram showed vascular cut-off just above the knee; had this not been recognized and treated, amputation might have been necessary.

MENISCAL INJURIES

Meniscal injuries are common occurrences both in isolation and in conjunction with ligament injuries of the knee. The menisci have an important role in load distribution, stability and articular congruence. Menisci are relatively poorly vascularized structures that receive their blood supply from their periphery.

Meniscal injuries are discussed in detail in Chapter 20.

OSTEOCHONDRAL INJURIES

Patients who have suffered osteochondral injuries give a history of patellar dislocation or a blow to the front of the knee. The joint is swollen and aspiration yields blood-stained fluid mixed with fat globules.

Standard anteroposterior and lateral radiographs seldom show the bony part of the fragment and the deficit from which it arises. MRI imaging is required to delineate the lesion accurately.

Treatment

Small fragments should be removed because they may cause symptoms. Larger osteochondral fragments, and especially those from load-bearing areas, can be reattached with counter-sunk or 'headless' small fragment screws if the patient presents early.

Sometimes an area of purely chondral damage is identified on the articular surface that is not amenable to reattachment. For areas less than 15 mm in diameter the current gold standard treatment is arthroscopic trimming of any ragged parts and 'micro-fracturing'. Micro-fracturing consists of drilling through the crater to stimulate an inflammatory response and produce fibrocartilage. For larger lesions treatment remains more experimental. Techniques available that are associated with successful outcomes include cartilage transplantation techniques, collagen scaffold implantation, autologous chondral transfer, and resurfacing implants.

OSTEOCHONDRITIS DISSECANS

Teenagers and young adults who complain of intermittent pain in the knee are sometimes found to have developed a small segment of osteochondral necrosis, usually on the lateral aspect of the medial femoral condyle. This is probably a traumatic lesion, caused by repetitive contact with the overlying patella or an adjacent ridge on the tibial plateau. The condition is described in Chapter 20.

PATELLOFEMORAL INJURIES

Extensor mechanism injuries

Disruption of the extensor apparatus may occur in the quadriceps tendon, at the attachment of the quadriceps tendon to the proximal surface of the patella, through the patella and retinacular expansions, at the junction of the patella and the patellar tendon, in the patellar tendon, or at the insertion of the patellar tendon to the tibial tubercle.

In all but direct fractures of the patella, the mechanism of injury is the same: sudden resisted extension of the knee or (essentially the same thing) sudden passive flexion of the knee while the quadriceps is contracting. The patient gives a history of stumbling on a stair, catching the foot while running, or kicking hard at a muddy football.

The lesion tends to occur at progressively higher levels with increasing age: adolescents suffer avulsion fractures of the tibial tubercle; young adult sportspeople tear the patellar tendon, middle-aged adults fracture their patellae; and older people (as well as those whose tissues are weakened by chronic illness or steroid medication) suffer acute tears of the quadriceps tendon.

Quadriceps tendon rupture

The patient is usually elderly, may have a history of diabetes or rheumatoid disease, or may have been treated with corticosteroids. Occasionally acute rupture is seen in a young athlete. The typical injury is followed by tearing pain and giving way of the knee. There is bruising and local tenderness; sometimes a gap can be felt proximal to the patella. Active knee extension is either impossible (suggesting a complete rupture) or weak (partial rupture). Diagnosis can be confirmed by ultrasound or MRI.

Treatment

PARTIAL TEARS

Non-operative treatment will usually restore function: an extension brace or plaster cylinder is applied, followed by physiotherapy that concentrates on restoring knee flexion and quadriceps strength.

COMPLETE TEARS

Early operation is needed, or else the ruptured fibres will retract and repair will be more difficult. End-to-end suturing can be reinforced by proximal turndown techniques as required. If the tendon has been avulsed from the proximal pole of the patella, it should be reattached to a trough created at that site using longitudinal pull-through sutures through bone tunnels in

the patella. Postoperatively the knee is held in extension in a hinged brace. Early supervised movement through the brace is important to prevent adhesions; limits to the amount of flexion can be controlled through the brace and increased as the repair heals.

'Chronic' ruptures (usually the result of delayed presentations or missed diagnoses) are difficult to repair because the ends have retracted. The gap can be closed and residual defects covered with turndown techniques, or augmentation techniques can be used, most commonly using autologous hamstring tendons or synthetic meshes.

The results of acute repairs are good, with most patients regaining full power, a good range of movement and little or no extensor lag. Late repairs are less predictable.

Patella tendon rupture

This is an uncommon injury; it is usually seen in young athletes and the tear is almost always at the proximal or distal attachment of the ligament. There may be a previous history of 'tendinitis' and local injection of corticosteroid.

The patient gives a history of sudden pain on forced extension of the knee, followed by bruising, swelling and tenderness at the lower edge of the patella or more distally.

X-rays may show a high-riding patella and a telltale flake of bone torn from the proximal or distal attachment of the ligament.

Ultrasound or *MRI* will help to distinguish a partial from a complete tear.

Treatment

ACUTE TEARS

Partial tears can be treated by applying an extension brace or plaster cylinder. *Complete tears* need operative repair or reattachment to bone. Tension on the suture line can be lessened by inserting a temporary pull-out wire or a protective figure-of-eight strong suture to keep the distance between the inferior pole and attachment to the tibial tuberosity constant. Immobilization in full extension may precipitate stiffness – it is, after all, a joint injury – and it may be better to support the knee in a hinged brace with limits to the amount of flexion permitted. This range can be gradually increased incrementally with time.

LATE CASES

Late cases are difficult to manage because of proximal retraction of the patella. A two-stage operation may be needed: first to release the contracted tissues and apply traction directly to the patella, then at a later stage to repair the patellar tendon and augment it with autologous hamstrings.

Early repair of acute ruptures gives excellent results. Late repairs are less successful and the patient may be left with a permanent extension lag.

Patella fractures

The patella is a sesamoid bone in continuity with the quadriceps tendon and the patellar tendon. There are additional insertions from the vastus medialis and lateralis into the medial and lateral edges of the patella. The extensor 'strap' is completed by the medial and lateral extensor retinacula (or quadriceps expansions), which bypass the patella and insert into the proximal tibia.

The mechanical function of the patella is to hold the entire extensor 'strap' away from the centre of rotation of the knee, thereby lengthening the anterior lever arm and increasing the efficiency of the quadriceps muscles.

The key to the management of patellar fractures is assessment of the state of the entire extensor mechanism. It is important to remember that if the extensor retinacula are intact, active knee extension is still possible, even if the patella itself is fractured.

Mechanism of injury and pathological anatomy

The patella may be fractured, either by a direct force or by an indirect traction force that pulls the bone apart (and often tears the extensor expansions as well).

Direct injury – usually a fall onto the knee or a blow against the dashboard of a car – causes either an undisplaced crack or a comminuted ('stellate') fracture without severe damage to the extensor expansions.

Indirect injury occurs, typically, when someone catches the foot against a solid obstacle and, to avoid falling, contracts the quadriceps muscle forcefully. This is a transverse fracture with a gap between the fragments.

Clinical features

Following one of the typical injuries, the knee becomes swollen and painful. There may be an abrasion or bruising over the front of the joint. The patella is tender and sometimes a gap can be felt. There is an associated haemarthrosis.

X-rays

The X-rays may show one or more fine fracture lines without displacement, multiple fracture lines with irregular displacement or a transverse fracture with a gap between the fragments (Figure 31.11).

Patella fractures are classified as transverse, longitudinal, polar or comminuted (stellate). Any of these may be either undisplaced or displaced. Separation of the fragments is significant if it is sufficient to create a step on the articular surface of the patella or, in the case of a transverse fracture, if the gap is more than 3 mm wide.

A fracture line running obliquely across the superolateral corner of the patella should not be confused with the smooth, regular line of a (normal) bipartite patella. Check the opposite knee; bipartite patella is often bilateral.

Treatment

UNDISPLACED OR MINIMALLY DISPLACED FRACTURES

The extensor mechanism is generally intact and treatment is mainly protective. A plaster cylinder or an extension brace holding the knee straight should be worn for 3–4 weeks, and during this time quadriceps exercises are to be practised every day.

LONGITUDINAL FRACTURES

The extensor mechanism is nearly always intact and the fracture is inherently more stable to early movement. Extension brace or cylinder plaster can be converted to a range of movement brace with incremental increases in movement range usually after 2 weeks.

COMMINUTED (STELLATE) FRACTURES

The extensor expansions are intact and the patient may be able to lift the leg. However, the undersurface of the patella is irregular and there is a serious risk of damage to the patellofemoral joint. Open reduction and fixation is generally advocated. Some small fragments that obviously distort the articular surface may be removed but the principle is to restore and hold the articular surface as much as possible. A combination of K-wires, mini fragment screws, cerclage wires or sutures are most commonly employed. A hinged brace is used in extension but can be unlocked for exercises to mould the fragments into position and to maintain mobility (see Figure 31.11).

DISPLACED TRANSVERSE FRACTURES

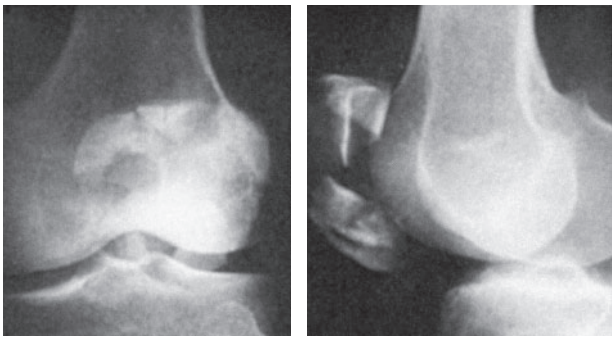
The lateral expansions are torn and the entire extensor mechanism is disrupted. Operative treatment is essential. Through a longitudinal incision the fracture is exposed and the patella repaired by the tension-band principle. The fragments are reduced and transfixed with two stiff K-wires; flexible wire is then looped tightly around the protruding K-wires and over the front of the patella (Figure 31.12). The tears in the extensor expansions are then repaired. A plaster backslab or hinged brace is worn until active extension of the knee is regained; either may be removed every day to permit active knee-flexion exercises.

Outcome

Patients usually regain good function but, depending on the severity of the injury, there is a significant incidence of late patellofemoral osteoarthritis.

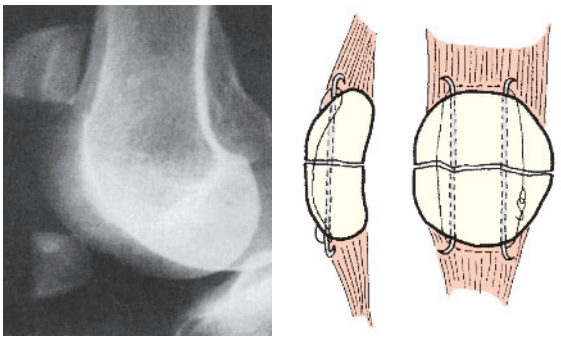
Patella dislocation

Because of the offset created by the femoral neck, the knee is normally angled in slight valgus that creates a natural tendency for the patella to pull towards the lateral side when the quadriceps muscle contracts. Lateral deviation of the patella during knee extension is prevented by a number of factors: the patella is seated in the intercondylar groove or trochlea, which has a high lateral 'embankment'; the force of



(a) (b)

Figure 31.11 Displaced comminuted patella fracture Displacement to the joint surface and disruption of the extensor mechanism.



(a) (b)

Figure 31.12 Fractured patella – transverse The separated fragments (a) are transfixed by K-wires; (b) malleable wire is then looped around the protruding ends of the K-wires and tightened over the front of the patella.

extensor muscle contraction pulls it firmly into the groove; and the extensor retinacula and patellofemoral ligaments guide it centrally as it tracks along the trochlea groove. The most important static checkrein on the medial side is the medial patellofemoral ligament, a more or less distinct structure extending from the superomedial border of the patella towards the medial femoral condyle deep to vastus medialis. Additional restraint is provided by the medial patellomeniscal and patellotibial ligaments and the associated medial retinacular fibres. In a typical knee, considerable force is required to wrench the patella out of its track. However, if the intercondylar groove is unusually shallow (trochlea dysplasia), or the patella is seated higher than usual (patella alta), or the ligaments are abnormally lax (hypermobility), dislocation occurs more easily.

Mechanism of injury

Patella dislocation may be traumatic or atraumatic. The patella dislocates laterally and the medial patellofemoral ligament and retinacular fibres may be torn. Atraumatic dislocations occur in patients with predisposing factors as discussed, such as trochlea dysplasia, patella alta or hypermobility. Traumatic dislocations are rarely caused by direct violence while the knee is flexed and the quadriceps muscle relaxed, although the patella may be forced laterally by direct violence. More often, traumatic dislocation is due to indirect force: sudden, severe contraction of the quadriceps muscle while the knee is stretched in valgus and external rotation. Typically this occurs in field sports when a runner dodges to one side.

The rate of first-time dislocation is highest among female adolescents. Following a first event, 17% of patients will experience subsequent instability. The younger a patient is at the time of first dislocation and the more severe the dislocation, the greater the risk of subsequent dislocation. If the patient has an established history of subluxation or dislocation, the risk of subsequent episodes rises to 50%.

Clinical features

In a 'first-time' dislocation the patient may experience a tearing sensation and a feeling that the knee has gone 'out of joint.' Often the patella springs back into position spontaneously; however, if it remains unreduced, there is an obvious (if somewhat misleading) deformity: the displaced patella, seated on the lateral side of the knee, is not easily noticed but the uncovered medial femoral condyle is unduly prominent and may be mistaken for the patella. Neither active nor passive movement is possible (Figure 31.13). If the dislocation has reduced spontaneously, the knee may be swollen and there may be bruising and tenderness on the medial side.

With recurrent dislocation the symptoms and signs are much less marked, though still unpleasant. After spontaneous reduction the knee looks normal, but the apprehension test is positive, i.e. the patient experiences pain and apprehension of attempts to push the patella laterally.

Imaging

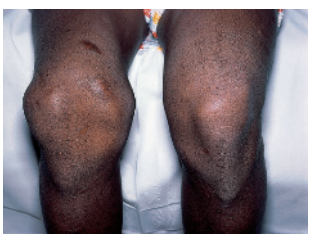
Anteroposterior (AP), lateral and tangential ('patella skyline') *X-rays* are needed. In an unreduced dislocation, the patella is seen to be laterally displaced and tilted or rotated. In 5% of cases there is an associated osteochondral fracture. The lateral radiograph enables assessment for patella alta and trochlear dysplasia. A shallow trochlea and a crossing sign are features of dysplasia.

MRI provides invaluable information on the multifactorial aspects of patellofemoral instability and is the investigation of choice for patients being considered for surgical intervention. The cartilage contour in the trochlea typically varies from that of the underlying bone and accentuates the dysplasia. This is not appreciable on conventional radiographs or CT.

Treatment

If unreduced, dislocations can in most cases be pushed back into place without much difficulty. There is no need for immobilization or bracing and it is safe to weight-bear on the knee as soon as it is comfortable to do so.

First-time dislocations are generally managed non-operatively in the first instance. *Non-operative therapy* initially aims to reducing swelling and increase the range of motion of the knee. This is followed by muscle-strengthening exercises. Physiotherapy is directed towards closed chain exercises and vastus medialis oblique (VMO) strengthening (the main dynamic stabilizer of the patella).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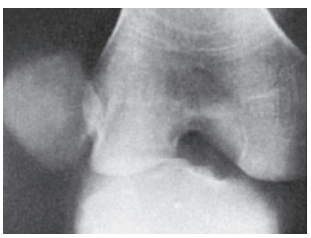
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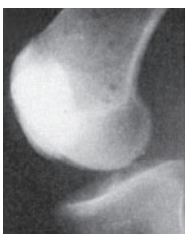

(c)

Figure 31.13 Dislocation of the patella (a) The right patella has dislocated laterally; the flattened appearance is typical. (b,c) AP and lateral X-rays of lateral patella dislocation.

Surgery should not be considered until non-operative treatments have failed and the recurrent nature of the disease has resulted in functional impairment. Surgical strategies involve addressing the underlying factors predisposing to recurrent dislocation. For patients with minimal or moderate trochlea dysplasia this may involve medial patellofemoral ligament reconstruction +/– tibial tubercle osteotomy if there is associated patella alta. For patients with severe dysplasia this may involve trochleoplasty. (The subjects of recurrent dislocation, subluxation, chronic patellar instability and patellar maltracking are dealt with in more detail in Chapter 20).

KNEE INJURIES IN CHILDREN

Proximal tibial epiphyseal injuries

This uncommon injury is usually caused by a severe hyperextension and valgus strain. It is a rare injury due to the insertion of knee ligaments being distal to the tibial epiphysis, thereby protecting the growth plate and transmitting stresses to the metaphysis. The epiphysis displaces forwards and laterally, often taking a small fragment of the metaphysis with it (a Salter–Harris type 2 injury). There is a risk of popliteal artery damage where the vessel is stretched across the step at the back of the tibia.

Imaging

Salter–Harris type 1 and 2 injuries may be undisplaced and difficult to define on *X-ray*; a few small bone fragments near the epiphysis may be the only clue. *CT* or *MRI* will more accurately delineate the injury. In the more serious injuries the entire upper tibial epiphysis may be tilted forwards or sideways. The fracture is categorized by the direction of displacement, so there are hyperextension, flexion, varus and valgus types.

Treatment

Under anaesthesia, closed reduction can usually be achieved. The direction of tilt may suggest the mechanism of injury; the fragment can be reduced by gentle traction and manipulation in a direction opposite to that of the fracturing force. Fixation using smooth K-wires or screws may be needed if the fracture is unstable. The rare Salter–Harris type 3 or 4 fractures also may need open reduction and fixation.

Complications

Epiphyseal fractures in young children sometimes result in angular *deformity* of the proximal tibia. This may later require operative correction.

With the higher grades of injury there is a risk of complete *growth arrest* at the proximal tibia. If the predicted leg length discrepancy is greater than 2.5 cm, tibial lengthening (or epiphysiodesis of the opposite limb) may be needed. Epiphysiodesis is surgical damage to the physis leading to growth arrest.

Proximal tibial metaphyseal fractures

This rare injury (Cozen's fracture) usually occurs between the ages of 4 and 8 years, resulting in fracture of the medial cortex with valgus angulation at the fracture site. Most fractures can be managed non-operatively with reduction and immobilization. It is vital to ensure that any valgus angulation is corrected with careful varus moulding of the cast and this must be monitored while the fracture heals. Open reduction is rarely required, usually due to failed reduction because of interposed soft tissues.

A unique complication seen with this fracture type is a progressive valgus deformity after bony healing and possible tibial overgrowth resulting in leg lengthening.

Tibial tuberosity injuries

Fracture or avulsion of the tibial tubercle usually occurs as a sports injury in young people. The knee is suddenly forced into flexion while the quadriceps is contracting, and a fragment of the tubercle – or sometimes the entire apophysis – may be wrenched from the bone. The diagnosis is suggested by the history. The area over the tubercle is swollen and tender; active extension causes pain.

The lateral radiograph shows the fracture, although this can be subtle. Patella alta may be present (the patella is abnormally high), having lost part of its distal attachment. CT/MRI may help delineate anatomy of the injury.

An incomplete fracture can be treated by applying a long-leg cast or extension brace with the knee in extension. Complete separation requires open reduction and fixation.

Patella sleeve fractures

Sleeve fractures occur between the cartilage 'sleeve' and main part of the patella and ossific nucleus. The causative mechanism is similar to that of tibial tubercle avulsions, i.e. a forced quadriceps contraction on a flexed knee. These fractures occur most commonly in children aged 8–12 years when patella ossification is nearly complete.

Lateral radiographs may reveal small flecks of bone adjacent to the distal pole of the patella and patella alta. CT/MRI may help delineate anatomy of the injury.

Undisplaced fractures with intact extensor mechanisms may be treated in long-leg casts or extension braces. Displaced fractures require open reduction and internal fixation.

FRACTURES OF THE TIBIA AND FIBULA

TIBIAL PLATEAU FRACTURES

Mechanism of injury

Fractures of the tibial plateau are caused by a varus or valgus force combined with axial loading (a pure valgus force is more likely to rupture the ligaments). This is sometimes the result of a car striking a pedestrian (hence the term 'bumper fracture'); more often it is due to a fall from a height in which the knee is forced into valgus or varus. The tibial condyle is crushed or split by the opposing femoral condyle, which remains intact.

Pathological anatomy

The fracture pattern and degree of displacement depend on the type and direction of force as well as the quality of the bone at the upper end of the tibia. A useful classification is that of Schatzker (Figure 31.14):

- *Type 1 – a vertical split of the lateral condyle*
  This is a fracture through dense bone, usually in younger people. It may be virtually undisplaced, or the condylar fragment may be pushed inferiorly and tilted; the damaged lateral meniscus may be trapped in the crevice.
- *Type 2 – a vertical split of the lateral condyle combined with depression of an adjacent load-bearing part of the condyle*
  The wedge fragment, which varies in size from a portion of the rim to a sizeable part of the lateral condyle, is displaced laterally; the joint is widened and, if the fracture is not reduced, may later develop a valgus deformity.
- *Type 3 – depression of the articular surface with an intact condylar rim*
  Unlike type 2, the split to the edge of the plateau is absent. The depressed fragments may be wedged firmly into the subchondral bone. The joint is usually stable and may tolerate early movement.
- *Type 4 – fracture of the medial tibial condyle*
  Two types of fracture are seen: (1) a depressed, crush fracture of osteoporotic bone in an elderly person (a low-energy lesion); and (2) a high-energy fracture resulting in a condylar split that runs obliquely from the intercondylar eminence to the medial cortex. The momentary varus angulation may be severe enough to cause a rupture of the lateral collateral ligament and a traction injury of the peroneal nerve. The severity of these injuries should not be underestimated.
- *Type 5 – fracture of both condyles*
  Both condyles are split but there is a column of the metaphysis wedged in between that remains in continuity with the tibial shaft.
- *Type 6 – combined condylar and subcondylar fractures*
  This is a high-energy injury that may result in severe comminution. Unlike type 5 fractures, the tibial shaft is effectively disconnected from the tibial condyles.

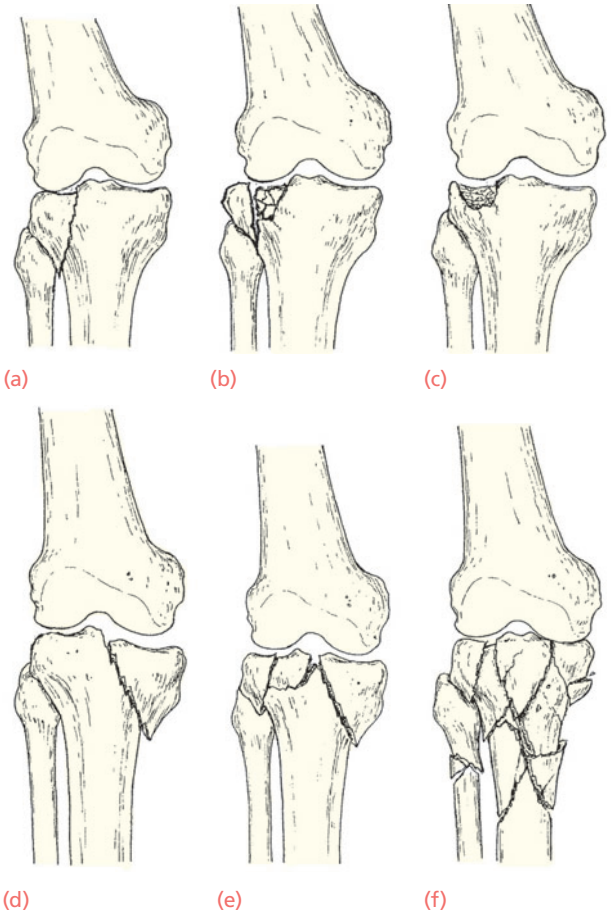


Figure 31.14 Tibial plateau fractures - Schatzker classification (a) *Type 1:* simple split of the lateral condyle. (b) *Type 2:* a split of the lateral condyle with a more central area of depression. (c) *Type 3:* depression of the lateral condyle with an intact rim. (d) *Type 4:* a fracture of the medial condyle. (e) *Type 5:* fractures of both condyles, but with the central portion of the metaphysis still connected to the tibial shaft. (f) *Type 6:* combined condylar and subcondylar fractures; effectively a disconnection of the shaft from the metaphysis.

Clinical features

The knee is swollen and may be deformed. Bruising is usually extensive and the tissues feel 'doughy' because of haemarthrosis. Examining the knee may suggest medial or lateral instability but this is usually painful and adds little to the radiographic diagnosis. More importantly, the leg and foot should be carefully examined for signs of vascular or neurological injury. Traction injury of the peroneal or tibial nerves is not uncommon and it is important to establish the extent of any neurological injury at the time of admission and before surgery.

Imaging

Anteroposterior, lateral and oblique *X-rays* will usually show the fracture, but the amount of comminution or plateau depression may not be appreciated without *CT*. This provides information on the location of the main fracture lines, the site and size of the portion of condyle that is depressed and the position of major parts of articular surface that have been displaced. Software-generated reassembly of the axial images can provide sagittal and coronal views that aid in surgical planning (Figure 31.15). It is important not to miss a posterior condylar component in high-energy fractures because this may require a separate posteromedial or posterolateral exposure for internal fixation.

Treatment

TYPE 1 FRACTURES

Undisplaced type 1 fractures can be treated conservatively. As soon as the acute pain and swelling have subsided (usually within 1 week), a hinged cast-brace or ROM brace is fitted to allow early mobilization. Weight-bearing is restricted initially.

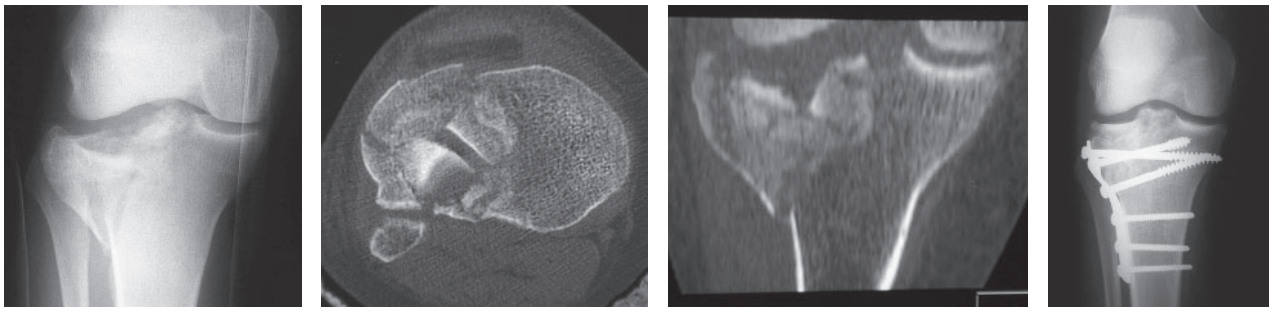
Displaced fractures should be treated by open reduction and internal fixation. The condylar surface is examined (open or arthroscopically) and trapped fragments are released or removed. The aim is for an accurate reduction and fixation; lag screws alone or in combination with a buttress plate are usually sufficient for fixation.

TYPE 2 FRACTURES

If depression is slight (less than 5 mm) and the knee is not unstable, or if the patient is old and frail or osteoporotic, the fracture can be treated non-operatively with the aim of regaining mobility and function rather than anatomical restitution. In younger patients, and more so in those with a central depression of more than 5 mm, open reduction with elevation of the plateau and internal fixation is required. The joint is seen to allow a check on the quality of reduction (either with a submeniscal arthrotomy or arthroscopically). After elevation and restoration of the joint line bone graft may be needed to support the elevated fragments. Screws can be placed in parallel just beneath the subchondral bone to hold up the elevated fragments well. These are sometimes referred to as 'raft' screws, describing the arrangement of parallel screws (Figure 31.16). The wedge of lateral condyle is then fixed with a buttress plate (Figure 31.17). Newer designs of anatomically contoured and angle-stable locking plates (using screws that lock into the plate) are available and increasingly used but are not always necessary. Early knee movement is encouraged after surgery to minimize joint stiffness.

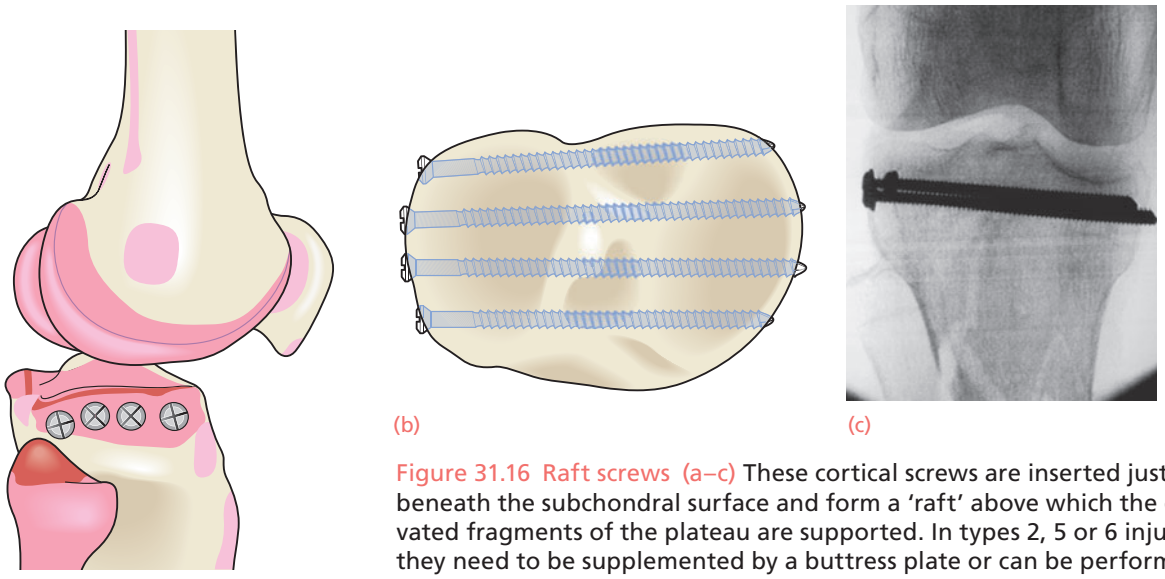
TYPE 3 FRACTURES

The principles of treatment are similar to those applying to type 2 fractures. However, the fact that the lateral rim of the condyle is intact means that the knee is usually stable and a satisfactory outcome is more predictable. The depressed fragments may need to be elevated through a window in the metaphysis; reduction should be checked by X-ray or arthroscopy. The elevated fragments can be supported with



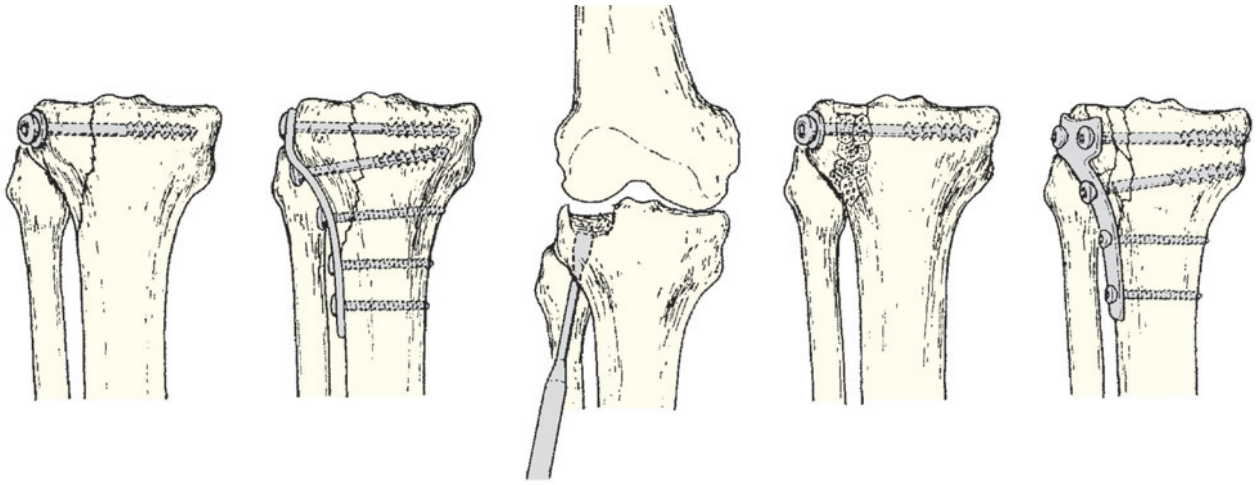
(a) (b) (c) (d)

Figure 31.15 Tibial plateau fractures – imaging (a) X-rays provide information about the position of the main fracture lines and areas of articular surface depression. (b,c) CT reconstructions reveal the extent and direction of displacements, vital information for planning the operation. (d) The postoperative X-ray shows that reduction has been achieved.



(a) (b) (c)

Figure 31.16 Raft screws (a–c) These cortical screws are inserted just beneath the subchondral surface and form a 'raft' above which the elevated fragments of the plateau are supported. In types 2, 5 or 6 injuries, they need to be supplemented by a buttress plate or can be performed through holes in a plate.



(a) (b) (c) (d) (e)

Figure 31.17 Tibial plateau fractures – fixation (a) Two or three lag screws may be sufficient for simple split fractures (type 1), though (b) a buttress plate and screws may be more secure. (c) Depression of more than 5 mm in a type 3 fracture can be treated by elevation from below and (d) supported by bone grafts and fixation. (e) Type 2 fractures require a combination of both techniques – direct reduction, elevation of depressed areas, bone grafting and buttress plate fixation.

bone grafts and the whole segment is held with 'raft' screws.

TYPE 4 FRACTURES

Osteoporotic *crush fractures* of the medial plateau are difficult to reduce; in the long term the patient is likely to be left with some degree of varus deformity. The principles of treatment are the same as for type 2 fractures of the lateral plateau.

Medial condylar *split fractures* usually occur in younger people and are caused by high-energy trauma. The fracture itself is often more complex than is appreciated at first sight; there may be a second, posterior split in the coronal plane that cannot be fixed through the standard anterior approach. Good lateral radiographs or CT scans are needed to define the fracture pattern. There may be an associated underlying ligament injury on the lateral side. Stable fixation of the medial side, along the lines described for the type 2 fracture, will then allow an assessment of the ligament injury. If the joint is unstable after fracture fixation, the torn structures on the lateral side may need repair.

TYPE 5 AND 6 FRACTURES

These are severe injuries that carry the added risk of a compartment syndrome. Surgical intervention with the aim of achieving stable internal fixation and early

joint movement for these injuries is the goal, but surgery is not without significant risk. The danger is that the wide exposure necessary to gain access to both condyles may strip the supporting soft tissues, thus increasing the risk of wound breakdown and delayed union or non-union.

The current approach to management is provisional stabilization with splint, plaster or external fixator, and waiting for the soft-tissue conditions to improve – sometimes as long as 2–3 weeks. Then a double incision approach (anterior and posteromedial usually) is made, which provides access to the main fracture fragments and limits the amount of subperiosteal elevation carried out if both condyles are approached through a single anterior incision only.

The three-column theory for the management of these injuries based on CT fracture pattern has been recently proposed (lateral column, medial column and posterior column). This provides an excellent strategy for planning surgical approach and fixation methods according to the columns involved and the fragments to be fixed (Figure 31.18). Fixation is most commonly then done using a combination of screws with locking plates and/or buttress plates with bone graft as required.

An alternative method is to perform the articular reduction through a limited surgical exposure (this can often be done percutaneously) and to stabilize the metaphysis to the diaphysis using a circular external fixator (Figure 31.19).

NOTE: Stability is all-important; no matter which method is used, fixation must be secure enough to permit early joint movement. There is little point in ending up with a pleasing X-ray and a stiff knee.

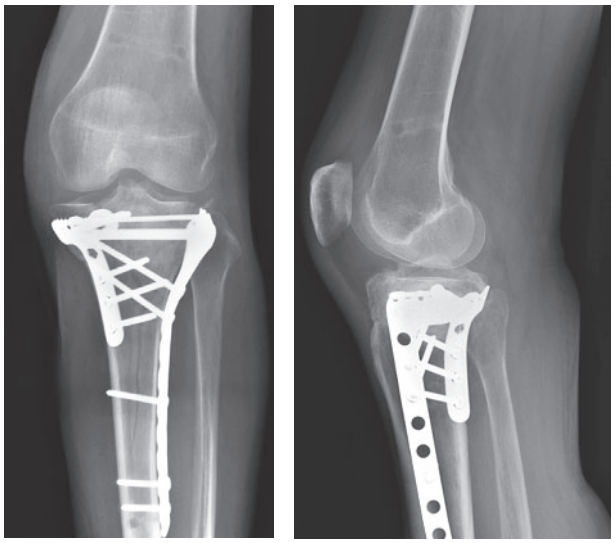


Figure 31.18 Complex tibial plateau fractures – internal fixation AP and lateral X-rays showing use of a lateral locking plate and a postero-medial buttress plate to address all three (anterolateral, anteromedial and posterior) columns involved in this injury. This requires two separate approaches through a lateral and a posteromedial skin incision.

Complications

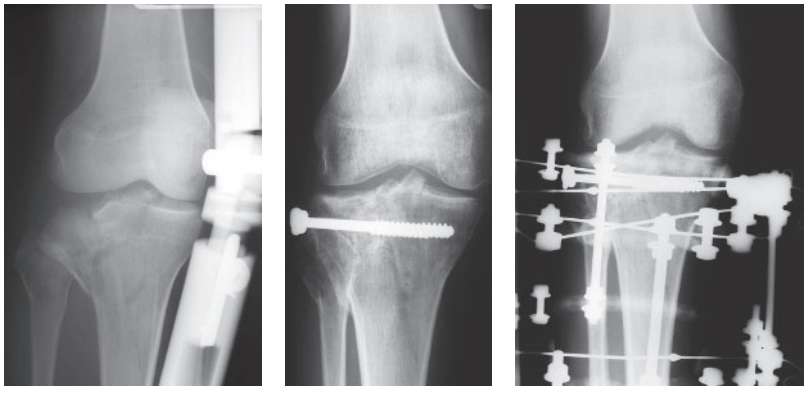
EARLY

Compartment syndrome With closed type 4 and 5 fractures there is considerable bleeding and swelling of the leg – and a risk of developing a compartment syndrome. The leg and foot should be examined repeatedly for signs such as disproportionate pain on passive stretching of muscles which run through the compartment and distal neurological or vascular compromise.

LATE

Joint stiffness With severely comminuted fractures, and after complex operations, there is a considerable risk of developing a stiff knee. This is prevented by avoiding prolonged immobilization and encouraging movement as early as possible.

Deformity Some residual valgus or varus deformity is quite common – either because the fracture was incompletely reduced or because, although adequately reduced, the fracture became redisplaced during treatment. Fortunately, moderate deformity



(a) (b) (c)

Figure 31.19 Complex tibial plateau fractures – external fixation An alternative method. Rather than expose the joint formally in order to reduce the fracture, this can be done percutaneously, albeit with X-ray control, and the articular fragments held with screws and wires (a,b). The tibial metaphysis is then held to the shaft using a circular external fixator (c).

is compatible with good function, although constant overloading of one compartment may predispose to osteoarthritis in later life.

Osteoarthritis If, at the end of treatment, there is marked depression of the plateau, or deformity of the knee or ligamentous instability, secondary osteoarthritis is likely to develop. This may subsequently require reconstructive/arthroplasty surgery.

Tibia and fibula shaft fractures

Because of its subcutaneous position, the tibia is more commonly fractured, and more often sustains an open fracture (23.5% in one large study), than any other long bone. The commonest mechanisms are falls, sporting and transport accidents, with higher-energy mechanisms seen more commonly in younger patients.

Mechanism of injury

A twisting force causes a spiral fracture of both leg bones at different levels; an angulatory force produces transverse or short oblique fractures, usually at the same level.

Indirect injury is usually low-energy; with a spiral or long oblique fracture one of the bone fragments may pierce the skin from within.

Direct injury crushes or splits the skin over the fracture; this is usually a high-energy injury and the most common cause is a motorcycle accident.

Pathological anatomy

The behaviour of these injuries – and therefore the choice of treatment – depends on the following factors:

- *The state of the soft tissues* – The risk of complications and the progress to fracture healing are directly related to the amount and type of soft-tissue damage. Closed fractures are best described using Tscherne's method (Table 31.1); for open injuries, Gustilo's grading is more useful (Table 31.2). The incidence of tissue breakdown and/or infection ranges from 1% for Gustilo type I to 30% for type IIIC.
- *The severity of the bone injury* – High-energy fractures are more damaging and take longer to heal than low-energy fractures; this is regardless of whether the fracture is open or closed. Low- energy breaks are typically closed or Gustilo I or II, and spiral. High-energy fractures are usually caused by direct trauma and tend to be open (Gustilo III A–C), transverse or comminuted.
- *The stability of the fracture* – Consider whether it will displace if weight-bearing is allowed. Long oblique fractures tend to shorten; those with a butterfly fragment tend to angulate towards the butterfly. Severely comminuted fractures are the least stable of all.
- *The degree of contamination* – In open fractures this is an important additional variable.
- *Patient factors* – Patient comorbidities, frailty, BMI, pre-morbid level of function, smoking status and psychosocial circumstances are also important in prognosis and hence decision making in these injuries.

Clinical features

The limb should be carefully examined for signs of soft-tissue damage: bruising, severe swelling, crushing or tenting of the skin, an open wound, circulatory changes, weak or absent pulses, diminution or loss of sensation and inability to move the toes. Any deformity should be noted before splinting the limb. *Always be on the alert for signs of an impending compartment syndrome.*

X-rays

The entire length of the tibia and fibula, as well as the knee and ankle joints, must be seen. The type of fracture, its level and the degree of angulation and displacement are recorded. Rotational deformity can

Table 31.1 Tscherne's classification of skin lesions in closed fractures

Grade	Skin lesion
IC1	No skin lesion
IC2	No skin laceration but contusion
IC3	Circumscribed degloving
IC4	Extensive, closed degloving
IC5	Necrosis from contusion

Table 31.2 Gustilo's classification of open fractures

Grade	Wound	Soft-tissue injury	Bone injury
I	<1 cm long	Minimal	Simple low-energy fractures
II	>1 cm long	Moderate, some muscle damage	Moderate comminution
IIIA	Usually >1 cm long	Severe deep contusion; + compartment syndrome	High-energy fracture patterns; comminuted but soft-tissue cover possible
IIIB	Usually >10 cm long	Severe loss of soft-tissue cover	Requires soft-tissue reconstruction for cover
IIIC	Usually >10 cm long	As IIIB, with need for vascular repair	Requires soft-tissue reconstruction for cover

be gauged by comparing the width of the tibiofibular interspace above and below the fracture.

Management

The main objectives are:

1 to limit soft-tissue damage and preserve (or restore, in the case of open fractures) skin cover
2 to prevent – or at least recognize – a compartment syndrome
3 to obtain and hold fracture alignment
4 to start early weight-bearing (loading promotes healing)
5 to start joint movements as soon as possible.

LOW-ENERGY FRACTURES

Low-energy fractures can in certain circumstances be treated by non-operative methods. If the fracture is *undisplaced or minimally displaced*, a full-length cast from upper thigh to metatarsal necks is applied with the knee slightly flexed and the ankle at a right angle (Figure 31.20). Displacement of the fibular fracture, unless it involves the ankle joint, is unimportant and can be ignored. Apposition need not be complete but alignment must be near-perfect and rotation absolutely perfect. The position is checked by X-ray. Minor degrees of angulation can still be corrected by making a transverse cut in the plaster and wedging it into a better position. If there is excessive swelling, the cast is split. After 2 weeks the position is checked by X-ray. A change from an above- to a below-the-knee cast is possible around 4–6 weeks, when the fracture becomes 'sticky'. An alternative is a 'Sarmiento' cast which allows knee flexion but confers some additional stability. The cast is retained (or renewed if it becomes loose) until the fracture unites, which is around 8 weeks in children but seldom under 12 weeks in adults.

SURGICAL FIXATION METHODS

Closed intramedullary nailing This is the method of choice for internal fixation in most tibial shaft fractures. The fracture is reduced under X-ray control and image intensification. The proximal end of the tibia is exposed; a guide-wire is passed down the medullary canal and the canal is reamed. A nail of appropriate size and shape is then introduced from the proximal end across the fracture site. Transverse locking screws are inserted at the proximal and distal ends (Figure 31.21). Postoperatively, partial weight-bearing is started as soon as possible, progressing to full weight-bearing when this is comfortable.

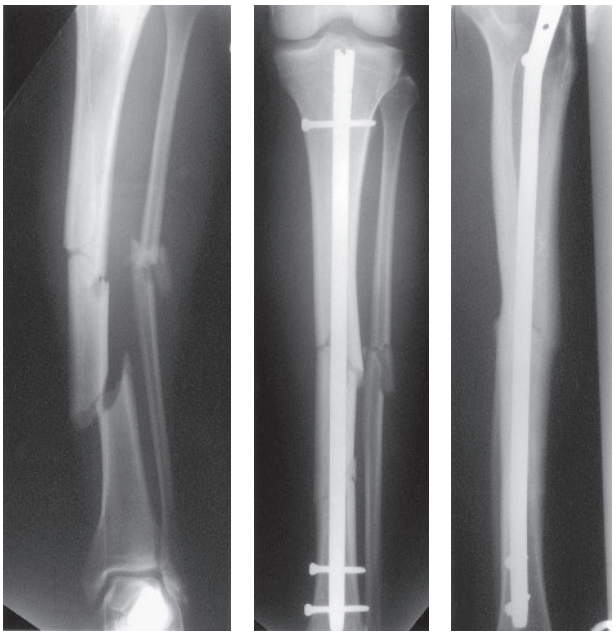
For diaphyseal fractures, union can be expected in over 95% of cases. For metaphyseal fractures near the bone ends, the method is more technically demanding but, as techniques have evolved, it has become commonly performed and is an excellent way of managing these injuries. Care must be taken: there may be existing intra-articular extensions of metaphyseal fracture lines and there is the potential for propagation of a fracture into the joint when the nail is passed.

(a)

(b)

Figure 31.20 Fractured tibia and fibula – closed treatment (a) The position is held while an assistant applies plaster from the knee downwards. When the plaster has set, the leg is lifted and the above-knee plaster completed; note that the foot is plantigrade, the knee slightly bent and the plaster moulded round the patella. A rockered boot can be fitted for walking (b).

Plate fixation Plating can be used for metaphyseal fractures deemed unsuitable for nailing. It is also sometimes used for unstable tibial shaft fractures in children as it avoids the potential damage to the growth plate from passing an intramedullary nail through an open physis. Previously, the disadvantages of plate fixation included the need to expose the fracture site and, in so doing, stripping the soft tissues around the fracture, which could increase the risk of introducing infection and delaying union. Newer techniques of plating overcome these disadvantages. The plate is slid across the fracture through proximal and distal 'access incisions' on the anterolateral aspect



(a) (b) (c)

Figure 31.21 Fractured tibia and fibula – intramedullary nailing Closed intramedullary nailing is now the preferred treatment for unstable tibial fractures. This series of X-rays shows the fracture before (a) and after (b,c) nailing. Active movements and partial weight-bearing were started soon after operation.

of the tibia and then fixed to the bone *only at these levels*. This method of 'submuscular' plating preserves the soft tissues around the fracture site better than conventional open plating, and it provides a relative stability that appears to hasten union. The soft tissues are still at risk using this technique, however, and considerable care is required in tissue handling to minimize the risk of wound complications.

External fixation This is a more rarely used alternative to closed nailing; it avoids exposure of the fracture site and allows further adjustments to be made if this should be needed. It has a particular role in long, segmental, multifragmentary fractures. Monolateral external fixation is most commonly used as a temporizing method of fixation in the context of an open injury in adults but can be used as a definitive method. It is more commonly used in children. Disadvantages include the need to span joints for sufficient stability, which can cause stiffness and the potential for pin-site infection. Circular external fixators confer greater stability and often negate the need to span joints (Figure 31.22). In both cases the tissues around the fracture are left undisturbed.

HIGH-ENERGY FRACTURES

Initially, the most important consideration is the viability of the damaged soft tissues and underlying bone. Tissues around the fracture should be disturbed as little as possible.

Comminuted and segmental fractures, those associated with bone loss, and indeed any high-energy fracture that is inherently unstable, require early surgical stabilization. For closed fractures, most commonly this is done by intramedullary nailing hence the tissues around the fracture are left undisturbed.

In cases of bone loss, small defects can be treated by delayed bone grafting as required; limb salvage options for larger defects include either bone transport or compression-distraction (acute shortening to close the defect, with subsequent lengthening at a different level) with a circular external fixator (see Chapter 12).

OPEN FRACTURES

For open fractures, the use of internal fixation has to be accompanied by judicious and expert debridement and prompt cover of the exposed bone and implant; alternatively, initial temporary external (or internal) fixation can be safer if these prerequisites cannot be met and definitive fixation can be delayed until coverage can be achieved. In the UK all high-energy open fractures are managed in Major Trauma Centres where orthopaedic and plastic surgeons can jointly manage these injuries to provide the highest standards of care according to BOA Standards for Trauma (BOAST 4) guidelines.

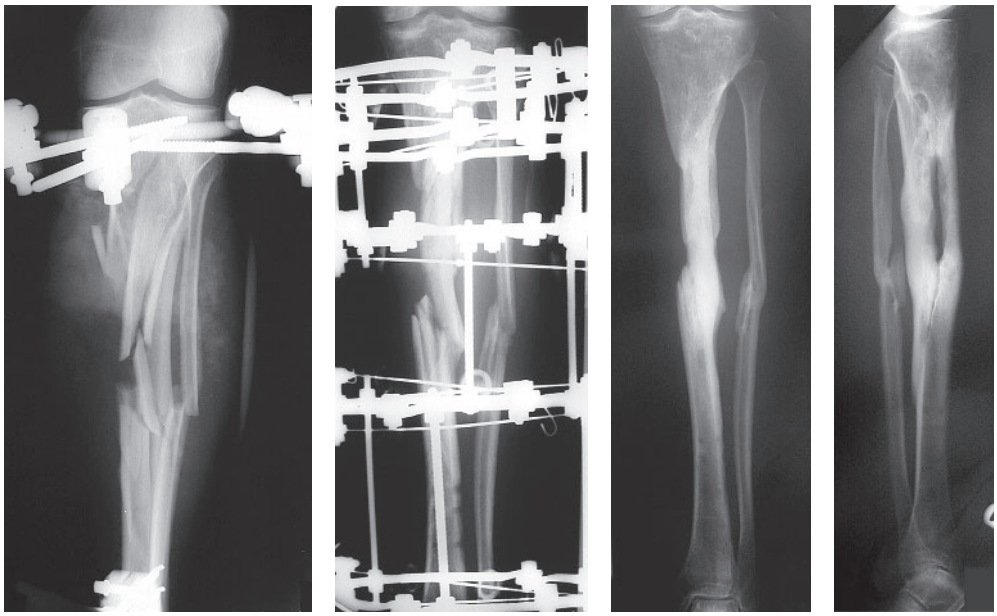
A suitable mantra for the treatment of open tibial fractures is:

- antibiotics
- debridement
- stabilization
- prompt soft-tissue cover
- rehabilitation.

Antibiotics are started immediately. A first- or second-generation cephalosporin or co-amoxiclav is suitable in most cases and should be continued until soft-tissue closure or for a *maximum* of 72 hours, whichever is sooner. The evidence for prolonged antibiotic use is lacking. Good debridement of the fracture and prompt cover remain the strongest defence against infection.

The wound should be photographed on first inspection in the Emergency Department by medical photography or using a hospital-registered camera, and then covered with a sterile dressing. The photograph can then be printed for inclusion in the patient's case notes to serve as a record and prevent further disturbance to the wound.

Adequate debridement is possible only if the original wound is extended. Wounds should be reviewed and discussed with plastic surgeons, especially if there appears to be a need for local or free skin or muscle flaps. Ideally the debridement should be performed



(a) (b) (c) (d)

Figure 31.22 Circular external fixation (a–d) This method of fixation offers the benefit of multilevel stability and can be carried out with little additional damage to the soft tissues around the injury.

jointly with the plastic surgeon. All dead and foreign material is removed; this includes bone without significant soft-tissue attachments. The wound and fracture site are then washed out with large quantities of normal saline. Gustilo grade I injuries can be closed primarily – being a low-energy injury with a small wound, closure should be possible without skin tension – and the fracture is then treated as for closed injuries.

More severe wounds should, ideally, be closed or covered at primary surgery as long as the debridement has been thorough and the skills of a plastic surgeon are at hand. If there is tissue of doubtful viability that requires another look, or a local flap cover deemed to be inappropriate, a second planned operation is needed. This allows further debridement and, hopefully, sufficient time to plan cover by free tissue transfer. Temporary cover of the exposed bone is often done using vacuum closure devices that can reduce oedema, improve local blood flow and encourage granulation tissue formation.

It is important to stabilize the fracture. For Gustilo I, II and IIIA injuries, locked intramedullary nailing is permissible where definitive wound cover is possible at the time of debridement. For more severe grades of open tibial fracture, definitive fixation should be performed only at the time of definitive soft-tissue cover. If this is not feasible at the time of primary debridement, the fracture should be stabilized temporarily with a spanning external fixator or a temporary plate through the open defect. Exchange of the temporary fixation for an intramedullary nail can be done at the point when definitive soft-tissue cover is carried out – ideally within 5 days of the injury.

POSTOPERATIVE MANAGEMENT

Swelling is common after tibial fractures; even after skeletal fixation the soft tissues continue to swell for several days. The limb should be elevated and frequent checks made for signs of compartment syndrome.

After intramedullary nailing of a transverse or short oblique fracture, weight-bearing can be started within a few days and increased to full weight when this is comfortable. If the fracture is comminuted or segmental, meaning that almost the entire load will be taken by the nail initially, only partial weight-bearing is permitted until some callus is seen on X-ray.

Early complications

VASCULAR INJURY

Fractures of the proximal half of the tibia may damage the popliteal artery. This is an emergency of the first order, requiring exploration and repair. Damage to one of the two major tibial vessels may also occur and go unnoticed if there is no critical ischaemia.

COMPARTMENT SYNDROME

Tibial fractures – both open and closed – are among the commonest causes of compartment syndrome in the leg. The combination of tissue oedema and bleeding (oozing) causes swelling in the muscle compartments and this may precipitate ischaemia. Additional risk factors are proximal tibial fractures, severe crush injury, a long ischaemic period before revascularization (in type IIIC open fractures), a long delay to treatment, haemorrhagic shock, difficult and prolonged operation and a fracture fixed in distraction.

The diagnosis in most instances is a clinical one. Severe unremitting pain is the most important symptom. Additional warning symptoms are increasing pain, a feeling of tightness or 'bursting' in the leg and numbness in the leg or foot. These complaints should always be taken seriously and followed by careful and repeated examination for pain provoked by passive muscle stretching and loss of sensibility and/or muscle strength. Heightened awareness is all! Compartment pressure measurements can aid in diagnosis. This is particularly useful in patients who are unconscious or uncooperative, and those with multiple injuries. A split-tip 20-gauge catheter is introduced into the anterior compartment of the leg and the pressure is measured close to the level of the fracture. A differential pressure (ΔP) – the difference between diastolic pressure and compartment pressure – of less than 30 mmHg (4.00 kPA) is regarded as critical and an indication for compartment decompression. Ideally the pressure should be measured in all four compartments.

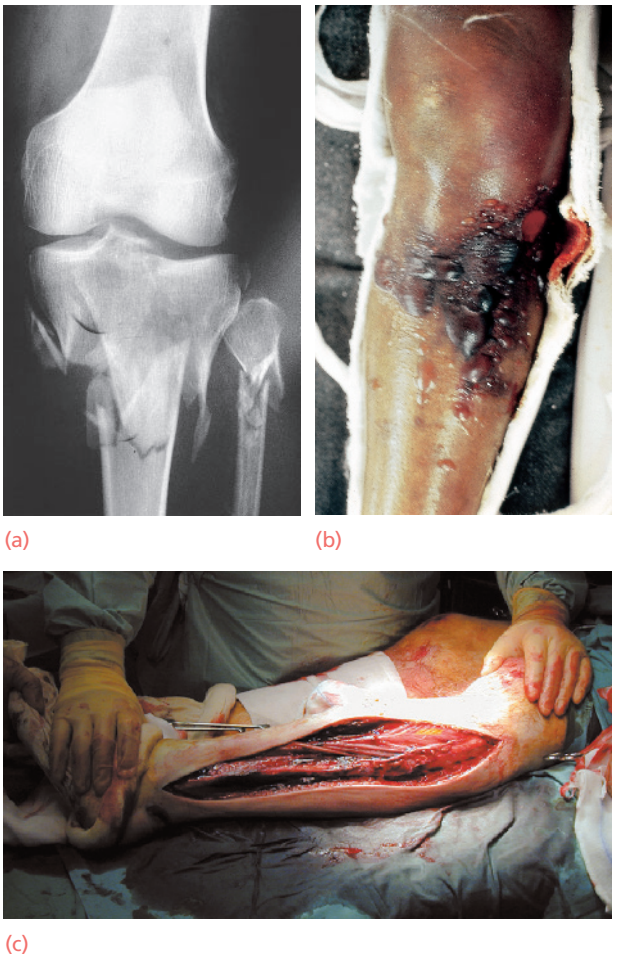
Fasciotomy and decompression Once the diagnosis is made, decompression of all four leg compartments should be carried out with the minimum delay. This is best and most safely accomplished through two incisions, one anterolateral and one posteromedial. The anterolateral incision is made about 2–3 cm lateral to the crest of the tibia and extends from the level of the tibial tuberosity to just above the ankle (Figure 31.23). The fascia is split along the length of the anterior and lateral compartments, taking care not to damage the superficial peroneal nerve. A second, similar incision is made just posterior to the posteromedial border of the tibia; the fascial covering of the superficial posterior compartment is split. The deep posterior compartment is identified just above the ankle and traced proximally; the muscle bulk of the superficial compartment needs to be retracted posteriorly, exposing the fascial envelope of the deep posterior compartment, which is likewise split down its entire length. Segmental arteries that perforate the fascia from the posterior tibial artery should be preserved for possible use in local skin flaps (Figure 31.24). The incisions are left open, a well-padded dressing is applied and the leg is splinted with the ankle in the neutral position. The fracture is treated as a grade III open injury requiring a spanning external fixator and prompt return for wound closure or skin grafting.

Outcome Compartment decompression within 6 hours of the onset of symptoms (or critical pressure measurement) should result in full recovery. Delayed decompression carries the risk of permanent dysfunction, the extent of which varies from mild sensory and motor loss to severe muscle and nerve damage, joint contractures and trophic changes in the foot.

Infection Open fractures are always at risk of infection; even a small perforation should be treated with respect and debridement carried out before the wound is closed.

If the diagnosis is suspected, wound swabs and blood samples should be taken. If debridement is required, deep tissue samples will aid microbiological diagnosis and antibiotic treatment should be withheld until samples have been taken. If not, then antibiotics should be started forthwith, using a 'best guess' intravenous preparation. Once the laboratory results are obtained, a more suitable antibiotic may be substituted.

With established infection, skeletal fixation should not be abandoned if the system is stable; infection control and fracture union are more likely if fixation is



(a) (b) (c)

Figure 31.23 Compartment syndrome (a) X-ray showing a complex proximal tibial fracture. All tibial fractures should be monitored for signs of compartment syndrome. Severe pain is the most important clinical feature. Regular clinical evaluation is important. (b) Swelling, blistering and skin necrosis associated with late, severe compartment syndrome that progressed quickly, unchecked underneath a complete plaster cast. (c) 4 compartment fasciotomies required in the context of lower limb compartment syndrome.

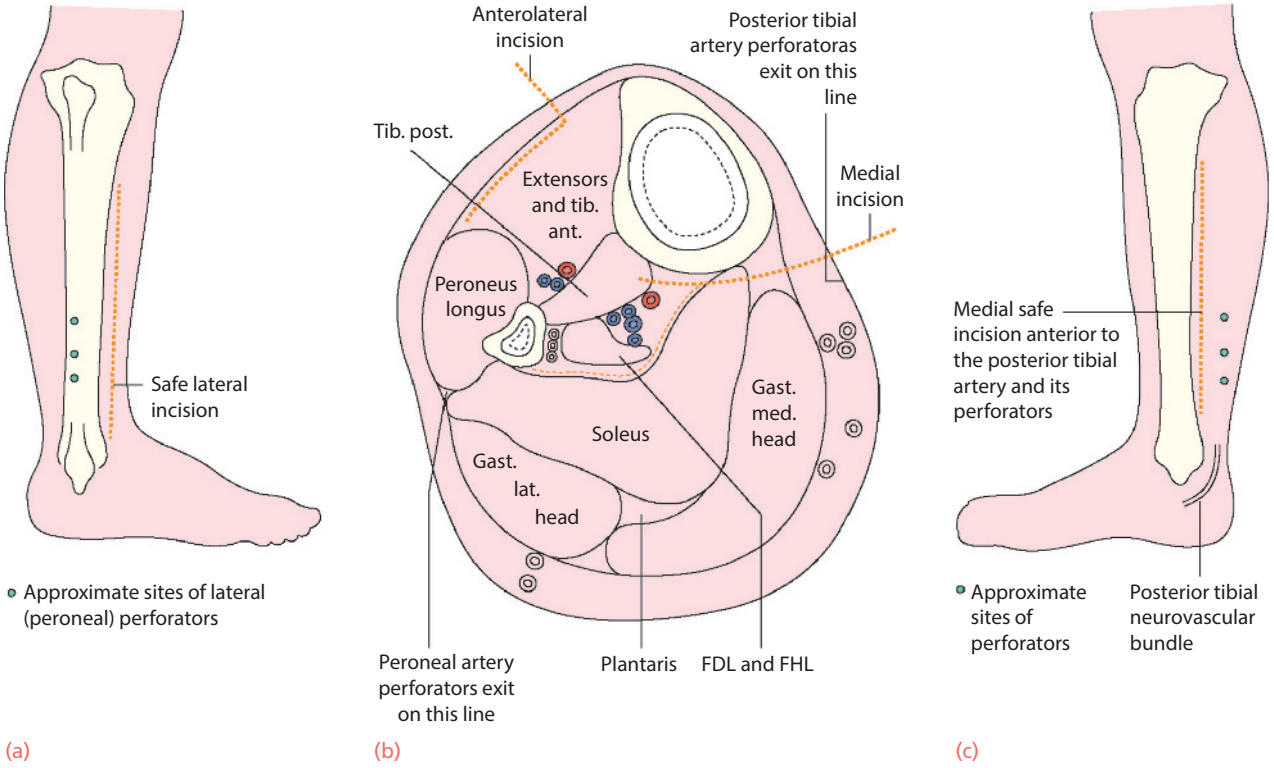


Figure 31.24 Fasciotomies for compartment decompression (a) The first incision is usually anterolateral, giving access to the anterior and lateral compartments. But this is not enough. The superficial and deep posterior compartments also must be opened; their position is shown in (b), a cross-section of the leg. This requires a second incision (b,c), which is made a finger's breadth behind the posteromedial border of the tibia; care must be taken not to damage the deep perforators of the posterior tibial artery. Note that the two incisions should be placed at least 7 cm apart so as to ensure a sufficient skin bridge without risk of sloughing.

secure. However, if there is a loose implant, it should be removed and replaced by external fixation.

Late complications

Malunion Slight shortening (up to 1.5 cm) is usually of little consequence, but rotation and angulation deformity, apart from being unsightly, can be disabling because the knee and ankle no longer move in the same plane.

Angulation should be prevented at all stages; anything more than 7 degrees in either plane is unacceptable. Angulation in the sagittal plane, especially if accompanied by a stiff equinus ankle, produces a marked increase in sheer forces at the fracture site during walking; this may result in either refracture or non-union.

Varus or valgus angulation will alter the axis of loading through the knee or ankle, causing increased stress in some part of the joint. This is often cited as a cause of secondary osteoarthritis; however, while this may be true for angular deformities close to the joint, long-term studies have failed to show that it applies to moderate deformities in the middle third of the bone.

Rotational alignment should be near-perfect (as compared with the opposite leg). This may be difficult to achieve with closed methods, but it should be possible with locked intramedullary nailing.

Late deformity, if marked, can be corrected by tibial osteotomy.

Delayed union High-energy fractures are slow to unite. If there is insufficient contact at the fracture site, either through bone loss or comminution, early 'prophylactic' bone grafting can be considered in high-risk patients. If there is a failure of union to progress on X-ray by 6 months, secondary intervention should be considered. The first nail is removed, the canal reamed and a larger nail inserted. If the fibula has united before the tibia, it should be osteotomized to allow better apposition and compression of the tibial fragments.

Non-union This may follow bone loss or deep infection, but a common cause is faulty treatment. Either the risks and consequences of delayed union have not been recognized, or splintage has been discontinued too soon, or the patient with a recently united fracture has walked with a stiff equinus ankle.

Hypertrophic non-union can be treated by intramedullary nailing (or exchange nailing) or compression plating. Atrophic non-union needs bone grafting in addition. If the fibula has united, a small segment should be excised so as to permit compression of the tibial fragments. Intractable cases may require radical Ilizarov techniques (Figure 31.25).

Joint stiffness Prolonged cast immobilization is liable to cause stiffness of the ankle and foot, which may persist for 12 months or longer in spite of active exercises. This can be avoided by changing to a functional brace as soon as it is safe to do so, usually by 4–6 weeks.

Osteoporosis Osteoporosis of the distal fragment is so common with all forms of treatment as to be regarded as a 'normal' consequence of tibial fractures. Axial loading of the tibia is important and weight-bearing should be re-established as soon as possible. After prolonged external fixation, special care should be taken to prevent a distal stress fracture.

Complex regional pain syndrome This is not uncommon in fractures of the distal third of the tibia. Exercises should be encouraged throughout the period of treatment. The management of the established condition is discussed in Chapter 10.

Isolated tibial fractures

A direct injury, such as a kick or blow with a club, may cause a transverse or slightly oblique fracture of the tibia alone at the site of impact. In children, the fracture is usually caused by an indirect injury; the fibula is intact or may show plastic deformation.

A fracture of the tibia alone takes just as long to unite as if both bones were broken, so at least 12 weeks is needed for consolidation and sometimes much longer.

Delayed union Isolated tibial fractures, especially in the lower third, may be slow to join and the temptation is to discard splintage too soon. Even slight displacement and loss of contact at the fracture level may delay union, so internal fixation is often preferred as primary treatment. This fracture also has a tendency to drift into varus in the later stages of healing; sometimes a fibular osteotomy is needed to allow correction of the deformity at surgery.

Tibial stress fractures

Repetitive stress may cause a fatigue fracture of the tibia (usually in the upper half of the bone) or the fibula (most often in the lower third). This injury is seen in army recruits, mountaineers, runners and ballet dancers, who complain of pain in the leg. There is local tenderness and slight swelling. The condition may be mistaken for a chronic compartment syndrome.

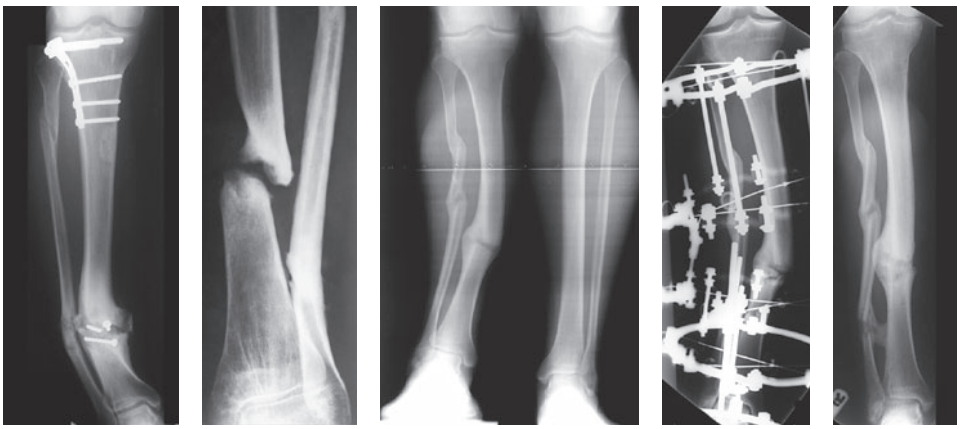
X-rays

For the first 4 weeks there may be no visible radiographic signs, but a bone scan shows increased activity at the fracture site. After some weeks periosteal new bone may be seen, with a small transverse defect in the cortex.

There is a danger that these appearances may be mistaken for erosive bone lesion. If the diagnosis of stress fracture is kept in mind, such mistakes are unlikely.

Treatment

The patient is told to avoid the stressful activity. Usually after 8–10 weeks the symptoms settle down. A short leg gaiter can be applied for comfort during weight-bearing.



(a) (b) (c) (d) (e)

Figure 31.25 Fractured tibia and tibula – late complications (a) *Hypertrophic non-union:* the exuberant callus formation and frustrated healing process are typical. (b) *Atrophic non-union:* there is very little sign of biological activity at the fracture site. (c) *Malunion:* treated, in this case, by gradual correction in an Ilizarov fixator (d,e).

FURTHER READING

Alford JW, Cole BJ. Cartilage restoration: techniques, outcomes and future directions. *Am J Sports Med* 2005; **33**: 443–60.

Bourke HE, Gordon DJ, Salmon LJ, *et al.* The outcome at 15 years of endoscopic anterior cruciate ligament reconstruction using hamstring tendon autograft for 'isolated' anterior cruciate ligament rupture. *J Bone Joint Surg Br* 2012 May; **94**(5): 630–7.

British Orthopaedic Association, British Association of Plastic, Reconstructive and Aesthetic Surgeons Standards for Trauma 2009. The management of severe open lower limb fractures. *J Bone Joint Surg Am* 2012 Jun 20; **94**: 1057e64. http://dx.doi.org/10.2106/JBJS.K. 00582.

Canadian Orthopaedic Trauma Society. Open reduction and internal fixation compared with circular fixator application for bicondylar tibial plateau fractures: results of a multicenter, prospective, randomized clinical trial. *J Bone Joint Surg* 2006; **88A**: 2613–23.

Cramer KE, Moed BR. Patellar fractures: contemporary approach to treatment. *J Am Acad Orthop Surg* 1997; **5**: 323–31.

Gustilo RB, Anderson JT. Prevention of infection in the treatment of one thousand and twenty-five open fractures of long bones: retrospective and prospective analyses. *J Bone Joint Surg Am* 1976; **58**: 453e8.

Howells NR, Barnett A, Ahearn A, *et al.* Medial patellofemoral ligament reconstruction: prospective outcome assessment of a large single centre series. *J Bone Joint Surg Br* 2012 Sep; **94**(9).

Howells NR, Brunton LR, Robinson J, *et al.* Acute knee dislocation: an evidence based approach to the management of the multiligament-injured knee. *Injury* 2011; **42**(2011): 1198–204.

Koval KJ, Helfet DL. Tibial plateau fractures: evaluation and treatment. *J Am Acad Orthop Surg* 1995; **3**: 86–94.

LaPrade RF, Wentorf F. Diagnosis and treatment of posterolateral knee injuries. *Clin Orthop Relat Res* 2002; **402**: 110–21.

Larson E, Lund PM. Ruptures of the extensor mechanism of the knee joint. *Clin Orthop Relat Res* 1986; **213**: 150.

Levy BA, Dajani KA, Whelan DB, *et al.* Decision making in the multiligament-injured knee: an evidence-based systematic review. *Arthroscopy* 2009; **2**: 430–8.

Luo CF, Sun H, Zhang B, Zeng BF. Three-column fixation for complex tibial plateau fractures. *J Orthop Trauma* 2010 Nov; **24**(11): 683–92.

Lyman S, Koulouvaris P, Sherman S, *et al.* Epidemiology of anterior cruciate ligament reconstruction: trends, readmissions and subsequent knee surgery. *J Bone Joint Surg Am* 2009; **91**: 2321.

Oestern H, Tscherne H. Pathophysiology and classification of soft tissue injuries associated with fractures. In: Tscherne H, Gotzen L (eds), *Fractures with Soft Tissue Injuries.* Berlin: Springer Verlag, 1984.

Upadhyay N, Wakeley C, Eldridge JD. Patellofemoral instability. *Orthopaedics and Trauma Journal* 2010; **24**(2): 139–48.

Injuries of the ankle and foot

32

Gavin Bowyer

INTRODUCTION

The foot and ankle act to both support and propel the body and are well adapted for these roles. During running and jumping, loads well in excess of 10 times the body weight are transmitted through the ankle and foot. If this loading is excessive, or excessively repeated, it can lead to foot and ankle injuries.

The ankle is a close-fitting hinge-like joint in which the two parts interlock like a mortise (the box formed by the distal ends of the tibia and fibula) and tenon (the upward projecting talus). The mortise bones are held together as a syndesmosis by the distal (inferior) tibiofibular and interosseous ligaments, and the talus is prevented from slipping out of the mortise by the medial and lateral collateral ligaments and joint capsule. The peroneal tendons provide additional stability, as active resistors of inversion.

The ankle rotates only in one plane (flexion/extension), but with a complex axis of rotation, actually rolling forward as the talus goes into plantar flexion; sideways movement is prevented by the malleolar buttresses and the collateral ligaments, but the bony constraint lessens as the ankle flexes. If the talus is forced to tilt or rotate, something must give: the ligaments, the malleoli or both. Movements of the talus into internal or external rotation come about from a rotatory force upon the foot, or more commonly inversion/supination of the foot, which, through the orientation of the subtalar joint, causes external rotation of the talus. Whenever a fracture of the malleolus is seen, it is important to assess the associated ligament injury.

ANKLE LIGAMENT INJURIES

Ankle sprains are the most common of all sports-related injuries, accounting for over 25% of cases. They are probably even more common in pedestrians and country walkers who stumble on stairways, pavements and potholes.

In more than 75% of cases it is the lateral ligament complex that is injured, in particular the anterior talofibular and calcaneofibular ligaments. Medial ligament injuries are usually associated with a fracture or joint injury.

A sudden twist of the ankle momentarily tenses the structures around the joint. This may amount to no more than a painful wrenching of the soft tissues – what is commonly called a *sprained ankle*. If more severe force is applied, the ligaments may be strained to the point of rupture. With a *partial tear*, some of the ligament remains intact and, once it has healed, it is able to stabilize the joint. With a *complete tear*, the ligament may still heal but it might not regain its original form and length; the joint will potentially be unstable (Figure 32.1).

Functional anatomy

The *lateral collateral ligaments* consist of the anterior talofibular, the posterior talofibular and (between them) the calcaneofibular ligaments. The anterior talofibular ligament (ATFL) runs almost horizontally from the anterior edge of the lateral malleolus to the neck of the talus; it is relaxed in dorsiflexion and tense in plantarflexion. In plantarflexion the ligament essentially changes its orientation from horizontal with respect to the floor to almost vertical. Thus the ligament at greatest stretch, and most vulnerable, with the foot plantar-flexed is the ATFL – hence the propensity for ATFL injury with the plantar-flexed, inverting, foot (e.g. down a pothole, off a kerb). The calcaneofibular ligament runs from the tip of the lateral malleolus to the posterolateral part of the calcaneum, thus it helps also to stabilize the subtalar joint. Maximum tension is produced by inversion and dorsiflexion of the ankle. The posterior talofibular ligament runs from the posterior border of the lateral malleolus to the posterior part of the talus.

The *medial collateral (deltoid) ligament* consists of superficial and deep portions. The superficial fibres spread like a fan from the medial malleolus as far anteriorly as

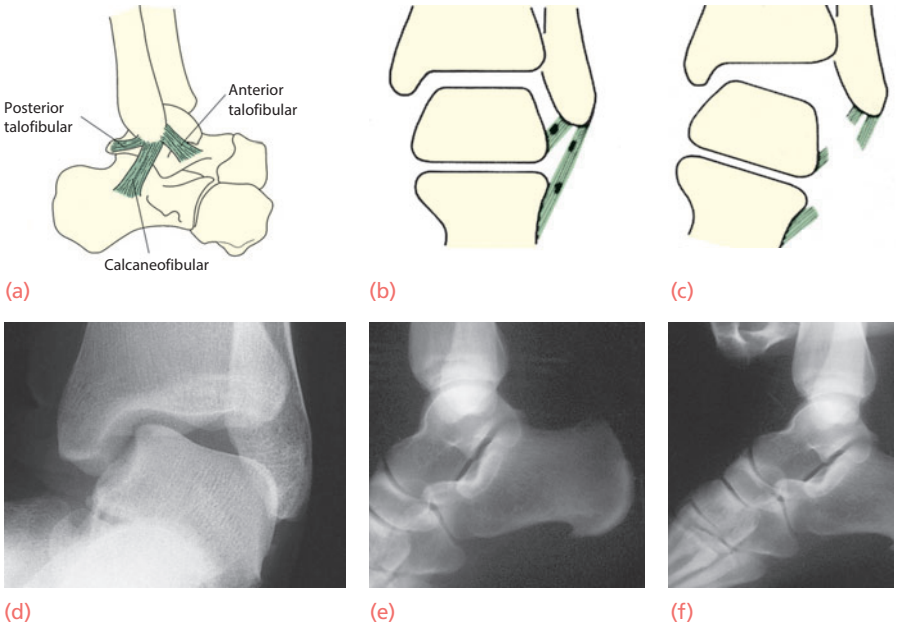


Figure 32.1 Ankle ligament injuries (a) Schematic diagram showing the mortise-and-tenon articulation and main ligaments of the ankle. (b) The three components of the lateral collateral ligament. (c) The commonest injury is a partial tear of one or other component of the lateral ligament. Following a complete tear, the talus may be displaced in the ankle mortise; the tibiofibular ligament may have ruptured as well, shown here in somewhat exaggerated form. (d) Stress X-ray showing talar tilt. (e,f) X-rays demonstrating anteroposterior instability. Pulling the foot forward under the tibia causes the talus to shift appreciably at the ankle joint; this is usually seen after recurrent sprains.

the navicular and inferiorly to the calcaneum and talus. Its chief function is to resist eversion of the hindfoot. The deep portion is intra-articular, running directly from the medial malleolus to the medial surface of the talus. Its principal effect is to prevent external rotation of the talus. The combined action of restraining eversion and external rotation makes the deltoid ligament a major stabilizer of the ankle.

The *distal tibiofibular joint* is held by four ligaments: the anterior, posterior, inferior transverse and interosseous 'ligament', which is really a thickened part of the interosseous membrane. This strong ligament complex still permits some movement at the tibiofibular joint during flexion and extension of the ankle.

Pathology

The common 'twisted ankle' is due to unbalanced loading with the ankle inverted and plantarflexed. First the ATFL and then the calcaneofibular ligament is strained; sometimes the talocalcaneal ligaments are also injured. If fibres are torn, there is bleeding into the soft tissues. The tip of the malleolus may be avulsed and in some cases the peroneal tendons are injured. There may be a small fracture of an adjacent tarsal bone or (on the lateral side) the base of the fifth metatarsal.

Acute injury of lateral ligaments

Clinical features

A history of a twisting injury followed by pain and swelling could suggest anything from a minor sprain to a fracture. If the patient is able to walk and bruising is only faint and slow to appear, it is probably a sprain; if bruising is marked and the patient is unable to put any weight on the foot, this suggests a more severe injury. In an ATFL sprain, tenderness is maximal just distal and slightly anterior to the lateral malleolus. The slightest attempt at passive inversion of the ankle is extremely painful. It is impossible to test for abnormal mobility in the acute phase without using local or general anaesthesia.

With all ankle injuries it is essential to examine the entire leg and foot; undisplaced fractures of the fibula or the tarsal bones, or even the fifth metatarsal bone are easily missed and injuries of the distal tibiofibular joint and the peroneal tendon sheath cause features that mimic those of a lateral ligament strain.

Imaging

About 15% of ankle sprains reaching the Emergency Department are associated with an ankle fracture. This complication can be excluded by obtaining an X-ray, but there are doubts as to whether all patients with ankle injuries should be subjected to X-ray examination. More than 20 years ago The Ottawa Ankle Rules were developed to assist in making this decision. X-ray examination is called for if there is: (1) pain around the malleolus; (2) inability to take weight on the ankle immediately after the injury; (3) inability to take four steps in the Emergency Department; (4) bone tenderness at the posterior edge or tip of the medial or lateral malleolus or the base of the fifth metatarsal bone.

If X-ray examination is considered necessary, anteroposterior, lateral and 'mortise' (30-degree oblique) views of the ankle should be obtained.

Localized soft-tissue swelling and, in some cases, a small avulsion fracture of the tip of the lateral malleolus or the anterolateral surface of the talus may be the only corroborative signs of a lateral ligament injury. However, it is important to exclude other injuries, such as an undisplaced fibular fracture or diastasis of the tibiofibular syndesmosis. If tenderness extends onto the foot, or if swelling is so severe that the area cannot be properly examined, additional X-rays of the foot are essential.

Persistent inability to weight-bear over 1 week should call for re-examination and review of all the initial 'negative' X-rays. For patients who have had persistent pain, swelling, instability and impaired function over 6 weeks or longer – despite appropriate early treatment – magnetic resonance imaging (MRI) or computed tomography (CT) will be required to assess the extent of soft-tissue injury or subtle bony changes.

Treatment

NON-OPERATIVE TREATEMENT

Initial treatment consists of **r**est, **i**ce, **c**ompression and **e**levation (RICE), which is continued for 1–3 weeks depending on the severity of the injury and the response to treatment. Cold compresses should be applied for about 20 minutes every 2 hours, and after any activity that exacerbates the symptoms.

More recently the acronym has been extended to 'PRICE' by adding **p**rotection (crutches, splint or brace) and still further to 'PRICER', adding **r**ehabilitation (supported return to function). The principles remain the same – a phased approach, to support the injured part during the first few weeks and then allow early mobilization and a supported return to function. An advice leaflet for patients is probably helpful.

The use of non-steroidal anti-inflammatory drugs (NSAIDs) in the acute phase can be helpful, with the usual contraindications and caveats. There is evidence that in acute injuries topical non-steroidal anti-inflammatory gels or creams might be as beneficial as oral preparations, probably with a better risk profile.

Functional treatment, i.e. 'protected mobilization', leads to earlier recovery of all grades of injury – without jeopardizing stability – than either rigid immobilization or early operative treatment.

OPERATIVE TREATMENT

If the ankle does not start to settle within 1–2 weeks of starting RICE, further review and investigation are called for. Persistent problems at 12 weeks after injury, despite physiotherapy, may signal the need for operative treatment. Residual complaints of ankle pain and stiffness, a sensation of instability or giving way and intermittent swelling are suggestive of cartilage damage or impinging scar tissue within the ankle. Arthroscopic repair or ligament substitution is now effective in many cases, allowing a return to full function and sports.

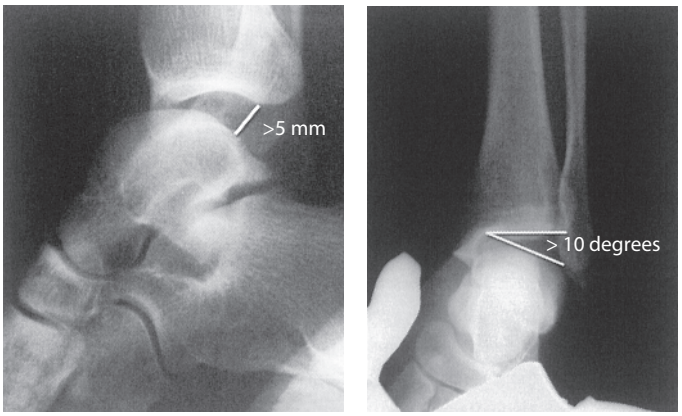
Recurrent lateral instability

Recurrent sprains are potentially associated with cartilage damage, and warrant careful investigation by MRI, arthroscopy and examination under anaesthesia.

Clinical features

The patient gives a history of a 'sprained ankle' that never quite seems to recover and is followed by recurrent 'giving way' or a feeling of instability when walking on uneven surfaces. This is said to occur in about 20% of cases after acute lateral collateral ligament tears.

The ankle looks normal and passive movements are full, but stress tests for abnormal lateral ligament laxity may show either excessive talar tilting in the sagittal plane or anterior displacement (an anterior drawer sign) in the coronal plane (Figure 32.2). In the chronic phase these tests are painless and can be performed either manually or with the use of special mechanical stress devices. Both ankles are tested, so as to allow comparison of the abnormal with the normal side.



(a) (b)

Figure 32.2 Recurrent lateral instability – special tests (a) *Anterior drawer test:* When the heel is drawn forwards under the tibia, the abnormally lax ligaments allow the talus to displace anteriorly. (b) *Talar tilt test:* Forcibly inverting the ankle causes the talus to tilt abnormally in the mortise. For both tests comparison with the normal side is important.

Talar tilt test With the ankle held in the neutral position, the examiner stabilizes the tibia by grasping the leg with one hand above the ankle; the other hand is then used to force the heel into maximum inversion. The range of movement can be estimated clinically and compared with that of the normal ankle. The exact degree of talar tilt can also be measured by X-rays, which should be taken with the ankles in 30 degrees of internal rotation (mortise views); 15 degrees of talar tilt (or 5 degrees more than in the normal ankle) is regarded as abnormal. Inversion laxity suggests injury to both the calcaneofibular and anterior talofibular ligaments.

Anterior drawer test The patient should be sitting with the knee flexed to 90 degrees and the ankle in 10 degrees of plantarflexion. The lower leg is stabilized with one hand while the other hand forces the patient's heel forward under the tibia. In a positive test the talus can be felt sliding forwards and backwards. The position of the talus is verified by lateral X-rays; anterior displacement of 10 mm (or 5 mm more than on the normal side) indicates abnormal laxity of the ATFL. With an isolated tear of the ATFL, the anterior drawer test may be positive in the absence of abnormal talar tilt. (Note: A positive anterior drawer test can sometimes be obtained in normal, asymptomatic individuals; the finding should always be considered in conjunction with other symptoms and signs).

Treatment

Recurrent 'giving way' can sometimes be prevented by modifying footwear, raising the outer side of the heel and extending it laterally. More effectively, the secondary dynamic ankle stabilizers, the peronei, can be strengthened and brought into play by specific physiotherapy regimes. Ankle exercises to strengthen the peroneal muscles are helpful, and a light brace can be worn during vigorous activities that stress the ankle.

If, in spite of these measures, the patient continues to experience mechanical instability (true giving way) during everyday activities, reconstruction of the lateral ligament should be considered. More commonly the persisting problem will be functional instability, in which the patient does not trust the ankle, and there are recurrent episodes in which the patient has rapidly or suddenly to unload the ankle, probably because of inhibitory feedback from the injured ankle.

Most patients with functional instability can be improved and returned to sport by arthroscopic debridement of the impinging tissue within the ankle joint, followed by physiotherapy.

Various operations for mechanical stabilization are described; they fall mainly into two groups: (1) those that aim to repair or tighten the ligaments; and (2) those that are designed to construct a 'check-rein' against the unstable movement. The *Broström–Karlsson* or *Gould operation* is an example of the first type: the anterior talofibular and calcaneofibular ligaments are exposed and repaired, usually by an overlapping – or 'double-breasting' – technique (Figure 32.3a). In the second type of operation a substitute ligament is constructed by using peroneus brevis to act as a tenodesis and prevent sudden movements into varus (the Chrisman operation) (Figure 32.3b). The disadvantages of the non-anatomical reconstructions are that they sacrifice or partially sacrifice the secondary stabilizers, the peroneal tendons.

There are various post-operative regimes that aim to protect the repair but allow an early, supported, return to activity. These might involve a plaster cast

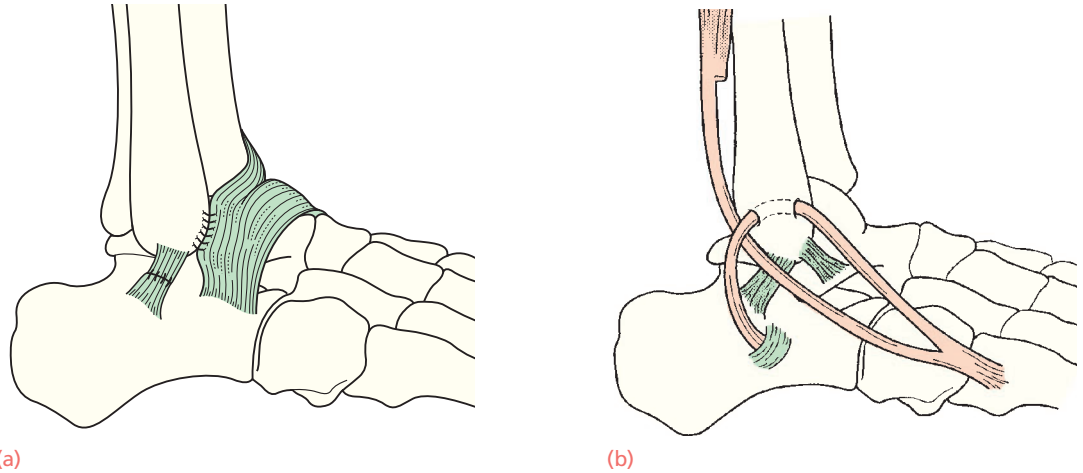


Figure 32.3 Recurrent lateral instability – operative treatment (a) The lax anterior talofibular and calcaneofibular ligaments can be reinforced by a double-breasting technique (the Boström–Karlsson operation). (b) Another way of augmenting the lateral ligament is to re-route part of the peroneus brevis tendon so that is acts as a check-rein (tenodesis) (the Chrisman operation).

but more commonly now use a special supportive boot (the 'moon boot'). Sometimes a removable brace is worn as the patient returns to active exercise and sports. The brace can usually be discarded after 3 months but it may need to be used from time to time for sports activities.

Deltoid ligament tears

Rupture of the deltoid ligament is usually associated with either a fracture of the distal end of the fibula or tearing of the distal tibiofibular ligaments (or both). The effect is to destabilize the talus and allow it to move into eversion and external rotation. The diagnosis is made by imaging: there is widening of the medial joint space in the mortise view; sometimes the talus is tilted, and diastasis of the tibiofibular joint may be obvious.

When there is a deltoid ligament or medial malleolar injury but no apparent lateral disruption at the ankle, it is important to look for a fracture or dislocation of the proximal fibula – the highly unstable *Maisoneuve injury*.

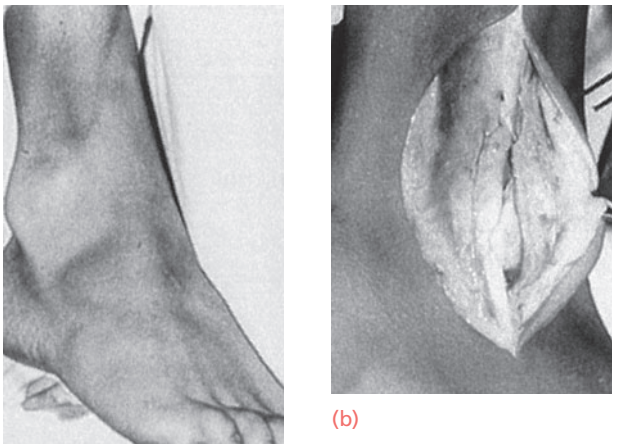
Treatment

Provided the medial joint space is completely reduced, the ligament will heal. The fibular fracture or diastasis must be accurately reduced, if necessary by open reduction and internal fixation. Occasionally the medial joint space cannot be reduced; it should then be explored in order to free any soft tissue trapped in the joint. A below-knee cast or support boot is applied with the foot plantigrade and is retained for 8 weeks.

Dislocation of peroneal tendons

Acute dislocation of the peroneal tendons may accompany – or may be mistaken for – a lateral ligament strain. Telltale signs on X-ray are an oblique fracture of the lateral malleolus (the so-called 'rim fracture') or a small flake of bone lying lateral to the lateral malleolus (avulsion of the retinaculum). Treatment in a below-knee cast, or similar support/immobilization in a brace or boot, for 6 weeks will help in a proportion of cases; the majority will complain of residual symptoms.

Recurrent subluxation or *dislocation* is unmistakable; the patient can demonstrate that the peroneal tendons dislocate forwards over the fibula during dorsiflexion and eversion (Figure 32.4). Treatment is operative and is based on the observation that the attachment of the retinaculum to the periosteum on the front of the fibula has come adrift, creating a pouch into which the tendons displace. Using non-absorbable sutures through drill holes in the bone, the normal anatomy is recreated. An alternative approach is to modify the morphology of the distal fibula, posteriorly translating a shelf of bone to constrain the tendons mechanically in a deepened posterior channel. Whichever method of stabilization is used, it is important also to assess the state of the tendons themselves, as an associated longitudinal split tear is commonly found, and this will lead to continuing pain and dysfunction around the lateral border of the ankle if it is not repaired.



(a) (b)

Figure 32.4 Dislocation of peroneal tendons (a) On movement of the ankle, the peroneal tendons slip forwards over the lateral malleolus. (b) The anterior part of the retinaculum is being reconstructed.

Tears of inferior tibiofibular ligaments

The inferior tibiofibular ligaments may be torn, allowing partial or complete separation of the tibiofibular joint (diastasis). *Complete diastasis*, with tearing of both the anterior and posterior fibres, follows a severe abduction strain. *Partial diastasis*, with tearing of only the anterior fibres, is due to an external rotation force. These injuries may occur in isolation, but they are usually associated with fractures of the malleoli or rupture of the collateral ligaments.

Clinical features

Following a twisting injury, the patient complains of pain in the front of the ankle. There is swelling and marked tenderness directly over the inferior tibiofibular joint. A 'squeeze test' has been described by Hopkinson and colleagues: when the leg is firmly compressed some way above the ankle, the patient experiences pain over the syndesmosis. Be sure, though, to exclude a fracture before carrying out the test.

X-rays

With a partial tear the fibula usually lies in its normal position and the X-ray looks normal. With a complete tear the tibiofibular joint is separated and the ankle mortise is widened; sometimes this becomes apparent only when the ankle is stressed in abduction. There may be associated fractures of the distal tibia or fibula, or an isolated fracture more proximally in the fibula.

Treatment

Partial tears can be treated by strapping the ankle firmly or bracing for 2–3 weeks. Thereafter exercises are encouraged.

Complete tears are best managed by internal fixation with a transverse screw just above the joint. This must be done as soon as possible so that the tibiofibular space does not become clogged with organizing haematoma and fibrous tissue. If the patient is seen late and the ankle is painful and unstable, open clearance of the syndesmosis and transverse screw fixation may be warranted. The ankle is immobilized in plaster for 8 weeks or similar support and off-loading is used, after which the screw is removed. However, some degree of instability usually persists.

MALLEOLAR FRACTURES OF THE ANKLE

Fractures and fracture – dislocations of the ankle are common. Most are low-energy fractures of one or both malleoli, usually caused by a twisting mechanism. Less common are the more severe fractures involving the tibial plafond, the pilon fractures, which are high-energy injuries, often caused by a fall from a height.

In malleolar injuries, the patient usually presents with a history of a twisting injury, usually with the ankle going into inversion, followed by immediate pain, swelling and difficulty weight-bearing. Bruising often comes out soon after injury.

One such injury was described by Percival Pott in 1768, and the group as a whole was for a long time referred to as Pott's fracture – although, as with many eponyms, he was not the first to notice or describe it, and what became known by this eponym was not what he described anyway!

The most obvious injury is a fracture of one or both malleoli; often, though, the 'invisible' part of the injury – rupture of one or more ligaments – is just as serious.

Mechanism of injury

The patient stumbles and falls. Usually the foot is anchored to the ground while the body lunges forward. The ankle is twisted and the talus tilts and/or rotates forcibly in the mortise, causing a low-energy fracture of one or both malleoli, with or without associated injuries of the ligaments. If a malleolus is pushed off, it usually fractures obliquely; if it is pulled off, it fractures transversely. The precise fracture pattern is determined by: (1) the position of the foot; (2) the direction of force at the moment of injury. The foot may be either pronated or supinated and the force upon the talus is towards adduction, abduction or external rotation, or a combination of these.

Pathological anatomy

There is no completely satisfactory classification of ankle fractures. Lauge-Hansen in the 1950s grouped these injuries according to the likely position of the foot and the direction of force at the moment of fracture. This is useful as a guide to the method of reduction (reverse the pathological force); it also gives a pointer to the associated ligament injuries. However, some people find this classification overly complicated. (For a detailed description the reader is referred to the original 1950 paper by Lauge-Hansen).

A simpler classification is that of *Danis* and *Weber*, which focuses on the fibular fracture.

- *Type A* – is a transverse fracture of the fibula below the tibiofibular syndesmosis, perhaps associated with an oblique or vertical fracture of the medial malleolus. This is almost certainly an adduction (or adduction and internal rotation) injury.
- *Type B* – is an oblique fracture of the fibula in the sagittal plane (and therefore better seen in the lateral X-ray) at the level of the syndesmosis; often there is also an avulsion injury on the medial side (a torn deltoid ligament or fracture of the medial malleolus). This is probably an external rotation injury and it may be associated with a tear of the ATFL.
- *Type C* – is a more severe injury, above the level of the syndesmosis, which means that the tibiofibular ligament and part of the interosseous membrane must have been torn. This is due to severe abduction or a combination of abduction and external rotation. Associated injuries are an avulsion fracture of the medial malleolus (or rupture of the medial collateral ligament), a posterior malleolar fracture and diastasis of the tibiofibular joint.

Clinical features

Ankle fractures are seen in sportsmen and sportswomen, as well as those who catch their foot, twist and fall in day-to-day activities; an older group includes women with postmenopausal osteoporosis who trip and fall.

A history of a twisting injury, followed by intense pain and inability to stand on the leg suggests

something more serious than a simple sprain. The ankle is swollen and deformity may be obvious. The site of tenderness is important; if both the medial and lateral sides are tender, a double injury (bony or ligamentous) must be suspected.

X-rays

At least three views are needed: anteroposterior, lateral and a 30-degree oblique 'mortise' view. The level of the fibular fracture is often best seen in the lateral view; diastasis may not be appreciated without the mortise view. Further X-rays may be needed to exclude a proximal fibular fracture, up as high as the knee.

From a careful study of the X-rays it should be possible to reconstruct the mechanism of injury. The four most common patterns are shown in Figure 32.5.

Treatment

Swelling is usually rapid and severe, particularly in the higher-energy injuries. If the injury is not dealt with within a few hours, definitive treatment may have to be deferred for several days while the leg is elevated so that the swelling can subside; this can be hastened by using a foot pump (which also reduces the risk of deep-vein thrombosis) and cold compression.

Fractures are visible on X-ray; ligament injuries are not. Always look for clues to the invisible ligament injury – widening of the tibiofibular space, asymmetry of the talotibial space, widening of the medial joint space, or tilting of the talus – before deciding on a course of action.

Like other intra-articular injuries, ankle fractures must be accurately reduced and held if later mechanical dysfunction is to be prevented. Persistent displacement of the talus, or a step in the articular surface, leads to increased stress and predisposes to secondary osteoarthritis.

In assessing the accuracy of reduction, four objectives must be met:

- the fibula must be restored to its full length
- the talus must sit squarely in the mortise, with the talar and tibial articular surfaces parallel

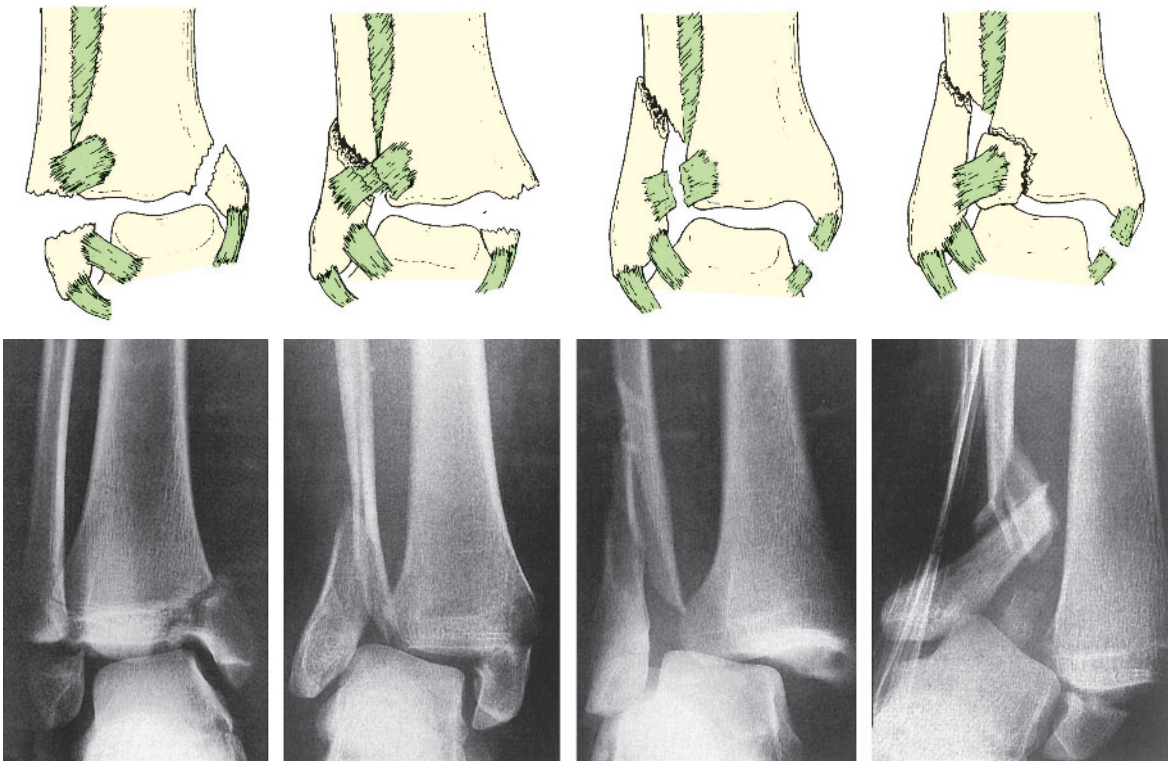


Figure 32.5 Ankle fractures – classification The Danis–Weber classification is based on the level of the fibular fracture. (a) *Type A:* a fibular fracture below the syndesmosis and an oblique fracture of the medial malleolus (caused by forced supination and adduction of the foot). (b) *Type B:* a fracture at the syndesmosis often associated with disruption of the anterior fibres of the tibiofibular ligament and fracture of the posterior and/or medial malleolus, or disruption of the medial ligament (caused by forced supination and external rotation). (c) *Type C:* a fibular fracture above the syndesmosis; the tibiofibular ligament must be torn, or else (d) the ligament avulses a small piece of the tibia. Here, again, there must also be disruption on the medial side of the joint – either a medial malleolar fracture or rupture of the deltoid ligament.

- the medial joint space must be restored to its normal width, i.e. the same width as the tibiotalar space (about 4 mm)
- oblique X-rays must show that there is no tibiofibular diastasis.

Ankle fractures are often unstable. Whatever the method of reduction and fixation, the position must be checked by X-ray during the period of healing.

UNDISPLACED FRACTURES

The first step is to decide whether the injury is stable or unstable (Figure 32.6).

Isolated undisplaced Danis–Weber type A fractures These fractures are stable and will need minimal splintage: a firm bandage or stirrup brace is applied mainly for comfort until the fracture heals.

Undisplaced type B fractures These are potentially unstable only if the tibiofibular ligament is torn or avulsed, or if there is a significant medial-sided injury. X-rays will show if the syndesmosis or mortise is intact; if it is, a below-knee cast is applied with the ankle in the neutral (anatomical) position. The plaster may need to be split and, if so, it must be completed or replaced when swelling has subsided. A check X-ray is taken at 2 weeks to confirm that the fracture remains undisplaced. An overboot is fitted and the patient is taught to walk correctly as soon as possible. The cast can usually be discarded after 6–8 weeks. Ankle and foot movements are regained by active exercises when the plaster is removed. As with any lower limb fracture, the leg must not be allowed to dangle idly – it must be exercised and elevated.

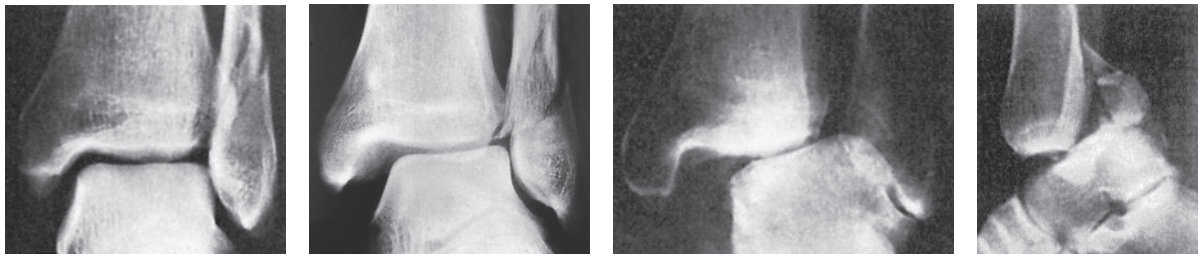
Undisplaced type C fractures These fractures are deceivingly innocent-looking but are often accompanied by disruption of the medial joint structures as well as the tibiofibular syndesmosis and interosseous membrane. These defects may become apparent only when the fracture displaces in a cast; arguably, therefore, type C fractures are better fixed from the outset.

DISPLACED FRACTURES

Reduction of these joint disruptions is a prerequisite to all further treatment; knowledge of the causal mechanism (and this is where the Lauge-Hansen classification is useful) helps to guide the method of closed reduction. Although internal fixation is usually performed to stabilize the reduction, not all such fractures require surgery.

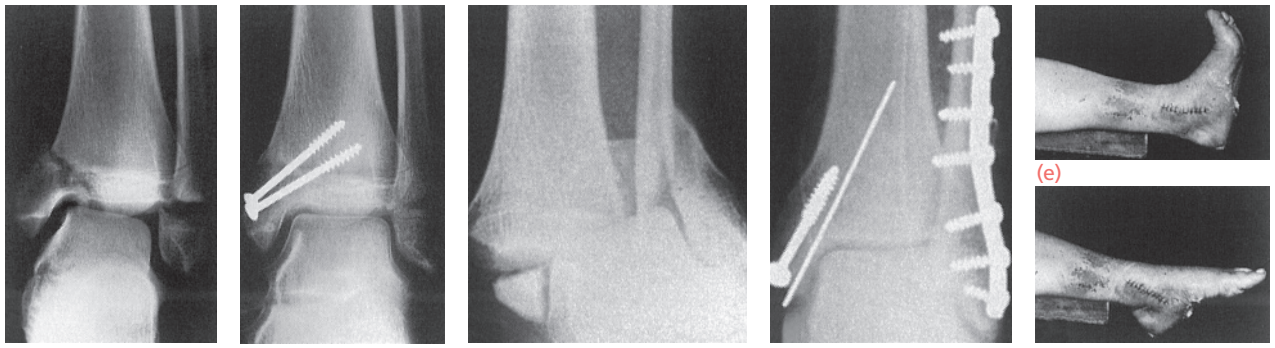
Displaced Danis-Weber type A fractures The medial malleolar fracture is nearly vertical and after closed reduction it often remains unstable; internal fixation of the malleolar fragment with one or two screws directed almost parallel to the ankle joint is advisable. A perfect reduction should be aimed for, with accurate restoration of the tibial articular surface. Loose bone fragments are removed. The lateral malleolar fracture, unless it is already perfectly reduced and stable, should be fixed with a plate and screws or tension-band wiring (Figure 32.7). Postoperatively a 'walking cast' or removable splintage boot is applied for 6 weeks; the advantage of removable splintage is that early physiotherapy can be commenced.

Displaced Danis-Weber type B fractures The most common fracture pattern is a spiral fracture of the fibula and an oblique fracture of the medial malleolus. The causal mechanism is external rotation of the ankle when the foot is caught in a supinated position.



(a) (b) (c) (d)

Figure 32.6 Ankle fractures – stable or unstable? (a) *Stable fracture:* In this Danis–Weber type B fracture the tibiofibular syndesmosis has held; the surfaces of the tibia and talus are precisely parallel and the width of the joint space is regular both superiorly and medially. (b) *Slight subluxation:* The syndesmosis is intact but the talus has moved laterally with the distal fibular fragment; the medial joint space is too wide, signifying a deltoid ligament rupture. It is vital, after reduction of the fibular fracture, to check that the medial joint space is normal; if it is not, the ligament has probably been trapped in the joint and it must be freed so as to allow perfect repositioning of the talus. (c) *Fracture dislocation:* In this high fibular fracture the syndesmosis has given way, the medial collateral ligament has been torn and the talus is displaced and tilted. The fibula must be fixed to full length and the tibiofibular joint secured before the ankle can be stabilized. (d) *Posterior fracture-dislocation:* If the posterior margin of the tibia is fractured, the talus may be displaced upwards. The fragment must be replaced and fixed securely.



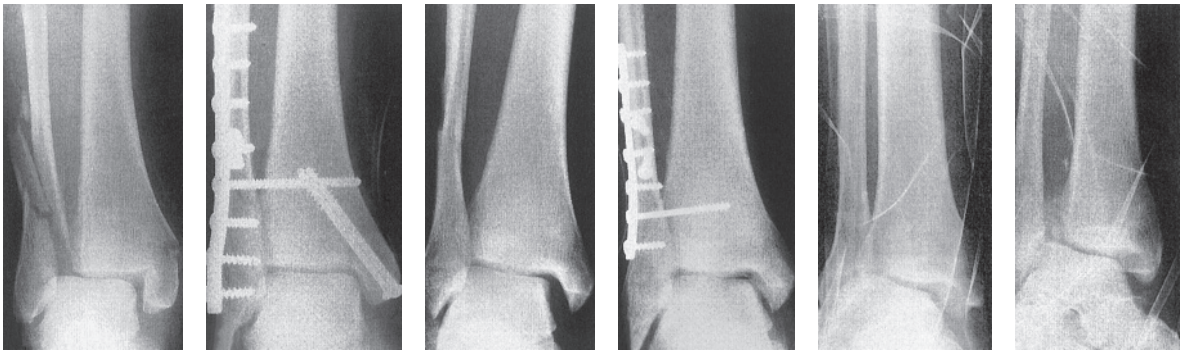
(a) (b) (c) (d) (e) (f)

Figure 32.7 Ankle fractures – open treatment (1) (a,b) Danis–Weber type A fractures can often be treated conservatively but, if the medial malleolar fragment involves a large segment of the articular surface, it is best treated by accurate open reduction and internal fixation with one or two screws. (c,d) An unstable fracture-dislocation such as this almost always needs open reduction and internal fixation. The fibula should be restored to full length and fixed securely; in this case the medial malleolus also needed internal fixation; (e) and (f) show the range of ankle movement a few days after operation and before a 'walking plaster' was applied.

Closed reduction therefore needs traction (to disimpact the fracture) and then internal rotation of the foot. If closed reduction succeeds, a cast is applied, following the same routine as for undisplaced fractures. Failure of closed reduction (sometimes a torn medial ligament is caught in between the talus and medial malleolus) or late redisplacement calls for operative treatment.

Type B fractures may also be caused by abduction; often the lateral aspect of the fibula is comminuted and the fracture line more horizontal. Despite accurate reduction (the ankle is adducted and the foot supinated), these injuries are unstable and often poorly controlled in a cast; internal fixation is therefore preferred.

Displaced Danis-Weber type C fractures The fibular fracture is well above the syndesmosis and frequently there are associated medial and posterior malleolar fragments. An isolated type C fibular fracture should raise strong suspicions of major ligament damage to the syndesmosis and medial side of the joint. Almost all type C fractures are unstable and will need open reduction and internal fixation (Figure 32.8). The first step is to reduce the fibula, restoring its length and alignment; the fracture is then stabilized using a plate and screws. If there is a medial fracture, this also is fixed. The syndesmosis is then checked, using a hook to pull the fibula laterally. If the joint opens out, it means that the ligaments are torn; the syndesmosis is stabilized by inserting a



(a) (b) (c) (d) (e) (f)

Figure 32.8 Ankle fractures with diastasis – open treatment (2) (a) In this type B fracture there is partial disruption of the distal tibiofibular syndesmosis. Treatment (b) required medial and lateral fixation as well a tibiofibular screw. (c) A type C fracture must, inevitably, disrupt the tibiofibular ligament; in this case the medial malleolus was intact but the deltoid ligament was torn (look at the wider than normal medial joint space). (d) By fixing the fibular fracture and using a tibiofibular screw, the ankle was completely reduced and it was therefore unnecessary to explore the deltoid ligament. (e) This patient presented 5 days after his injury; he, too, had a diastasis with disruption of the deltoid ligament (f). In this case the tibiofibular joint as well as the deltoid ligament had to be explored before the ankle could be reduced.

transverse screw across from the fibula into the tibia (the ankle should be held in 10 degrees of dorsiflexion when the screw is inserted).

Fracture subluxations more than 1–2 weeks old may prove difficult to reduce because of clot organization in the syndesmosis. Granulation tissue should be removed from the syndesmosis and transverse tibiofibular fixation secured.

Postoperative management After open reduction and fixation of ankle fractures, movements should be regained before applying a below-knee plaster cast or removable support boot. The patient is then allowed partial weight-bearing with crutches; the support is retained until the fractures have consolidated (6–12 weeks).

Management of the syndesmosis- or diastasis-screw remains controversial. Some advocate removal of the screw when the syndesmosis has healed, and before weight-bearing has commenced (6 weeks is too early; 10 weeks is probably more appropriate). Others are happy to allow early weight-bearing with the screw still in place, accepting that the screw may break (especially if four cortices are engaged).

OPEN FRACTURES

Open fractures of the ankle pose special problems. If the fracture is not reduced and stabilized at an early stage, it may prove impossible to restore the anatomy. For this reason unstable injuries should be treated by internal fixation even in the presence of an open wound, provided the soft tissues are not too severely damaged and the wound is not contaminated. As with all open fractures, attention must be paid to the extent of damage to the soft-tissue envelope and the involvement of other structures, particularly neurovascular or tendinous injuries. If internal fixation seems inappropriate, an external fixator can be used, often as a temporary spanning option. Treatment in other respects follows the principles outlined in Chapter 23.

Complications

EARLY

Vascular injury With a severe fracture-subluxation the pulses may be obliterated. The ankle should be reduced immediately and held in a splint until prompt definitive treatment has been initiated.

Wound breakdown and infection Diabetic patients are at greater than usual risk of developing wound-edge necrosis and deep infection. In dealing with displaced fractures, these risks should be carefully weighed against the disadvantages of conservative treatment; casts may also cause skin problems if not well padded and they are less effective in preventing malunion.

LATE

Incomplete reduction Incomplete reduction is common and, unless the talus fits the mortise accurately, degenerative changes may occur. This can sometimes be prevented by a corrective osteotomy.

Non-union The medial malleolus occasionally fails to unite because a flap of periosteum or other tissue is interposed between it and the tibia. It should be prevented by operative reduction and screw fixation.

Joint stiffness Swelling and stiffness of the ankle are usually the result of the soft-tissue injury. Optimal treatment can help to mitigate these potential problems. The patient must walk correctly in plaster and, when the plaster is removed, he or she must, until circulatory control is regained, wear a support bandage and elevate the leg whenever it is not being used actively. Physiotherapy is always helpful.

Complex regional pain syndrome (CRPS) This often follows fractures of the ankle. The patient complains of pain in the foot; there may be swelling and diffuse tenderness, with gradual development of trophic changes and severe osteoporosis. Management is discussed in Chapter 10.

Osteoarthritis Malunion and/or incomplete reduction may lead to secondary osteoarthritis of the ankle in later years. Unless the ankle is unstable, symptoms can often be managed by judicious analgesic treatment and the use of firm, comfortable footwear. However, in the longer term, if symptoms become severe, arthrodesis may be necessary.

PILON FRACTURES

Unlike the twisting injuries that cause the common ankle fractures, pilon injury to the ankle joint occurs when a large force drives the talus upwards against the tibial plafond, like a pestle (pilon) being struck into a mortar. There is considerable damage to the articular cartilage and the subchondral bone may be broken into several pieces; in severe cases, the comminution extends some way up the shaft of the tibia.

Clinical features

There may be little swelling initially but this rapidly changes and fracture blisters are common. The ankle may be deformed or even dislocated; prompt approximate reduction is mandatory.

Imaging

This is a comminuted fracture of the distal end of the tibia, extending into the ankle joint. The fracture may be classified according to the amount of displacement and comminution, but in all cases assessment is far better

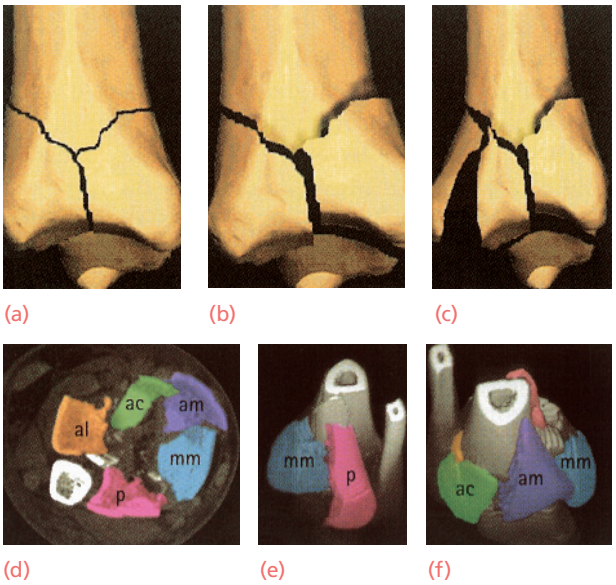


Figure 32.9 Pilon fractures – imaging These may be (a) undisplaced (type 1), (b) minimally displaced (type 2); (c) markedly displaced (type 3). CT (d) shows that there are usually five major tibial fragments: anterolateral (al), anterocentral (ac), anteromedial (am), the medial malleolus (mm) and the posterior fragment (p). These elements are better defined by 3D CT reconstruction.

with *CT scanning* (preferably including 3D reconstruction) than with plain X-ray examination (Figure 32.9).

Treatment

The three points of early management of these injuries are: *span*, *scan*, *plan*. Staged treatment has reduced the complication rate in these injuries.

Control of soft-tissue swelling is a priority; this is best achieved by elevation and applying an external fixator across the ankle joint (the spanning external fixator). It may take 2–3 weeks before the soft tissues improve, and fracture blisters can be actively managed rather than hidden under plaster (Figure 32.10). Surgery can be planned, based on the CT scan.

Once the skin has recovered, or using an approach that respects the zone of injury and pattern of vascularity, an open reduction and fixation with plates and screws (usually with bone grafting) may be possible (Figure 32.11). However, the more severe injuries do not readily tolerate large surgical exposures for plating, and significant wound breakdown and infection rates have been reported. Better results have followed wider use of indirect reduction techniques (e.g. applying a bone distractor or utilizing the spanning fixator across the joint to obtain as much reduction as possible through ligamentotaxis) and plating through limited exposures. Recently, these injuries have been successfully treated by using a combination of indirect reduction methods and small screws to hold the articular fragments, coupled with axially stable locking plates. Circular frame and fine-wire fixation has also been successful.

The soft-tissue swelling following these injuries is substantial. After fixation, elevation and early movement help to reduce the oedema; arteriovenous impulse devices applied to the sole of the foot are also helpful.

Outcome

Pilon fractures usually take several months to heal. Postoperatively, physiotherapy is focused on joint movement and reduction of swelling. There remains,

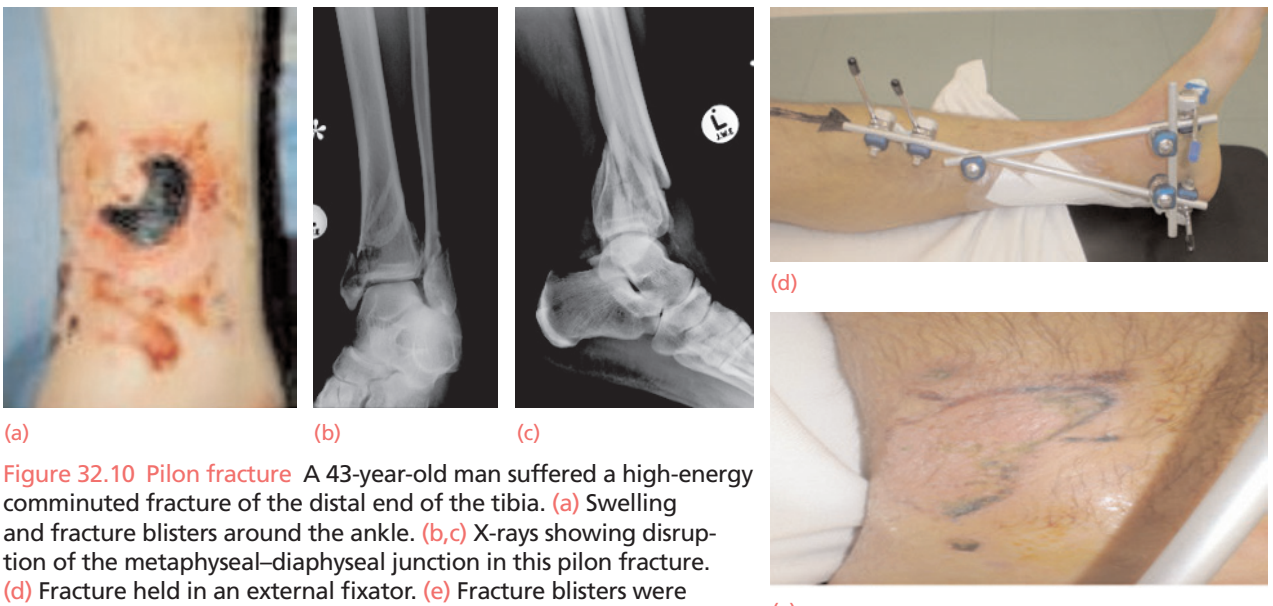
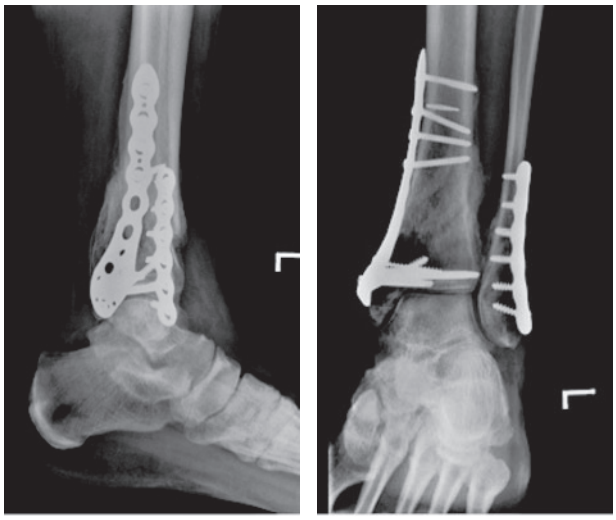


Figure 32.10 Pilon fracture A 43-year-old man suffered a high-energy comminuted fracture of the distal end of the tibia. (a) Swelling and fracture blisters around the ankle. (b,c) X-rays showing disruption of the metaphyseal–diaphyseal junction in this pilon fracture. (d) Fracture held in an external fixator. (e) Fracture blisters were de-roofed and treated with Flamazine (silver sulphadiazine); the skin has re-epithelialized and is free of infection 5 days after injury.



(a) (b)

Figure 32.11 Same case as Figure 32.10 – outcome
At 3 months after minimal approach reduction and fixation with distal locking plates the fractures have healed and the joint is congruent and normally aligned.

however, a challenging problem with poor functional results in these complex fractures, which represent a significant soft tissue injury as well as bony jigsaw. Although bony union may be achieved, the fate of the joint is decided by the degree of cartilage injury – the 'invisible' factor on X-rays. Secondary osteoarthritis, stiffness and pain are still frequent late complications in these injuries.

ANKLE FRACTURES IN CHILDREN

Physeal injuries are quite common in children and almost a third of these occur around the ankle (Figure 32.12).

Mechanism of injury

The foot is fixed to the ground or trapped in a crevice and the leg twists to one or the other side. The tibial (or fibular) physis is wrenched apart, usually resulting in a *Salter–Harris type 1 or 2 fracture*. With severe external rotation or abduction the fibula may also fracture more proximally. The tibial metaphyseal spike may come off posteriorly, laterally or posteromedially; its position is determined by the mechanism of injury and suggests the method of reduction. With adduction injuries the tip of the fibula may be avulsed.

Type 3 and 4 fractures are uncommon. They are due to a supination–adduction force. The epiphysis is split vertically and one piece of the epiphysis (usually the medial part) may be displaced.

Two unusual injuries of the growing ankle are the *Tillaux fracture* and the notorious *triplane fracture*. The *Tillaux fracture* is an avulsion of a fragment of tibia by the anterior tibiofibular ligament (Figure 32.13); in the child or adolescent this fragment is the lateral part of the epiphysis and the injury is therefore a Salter–Harris type 3 fracture.

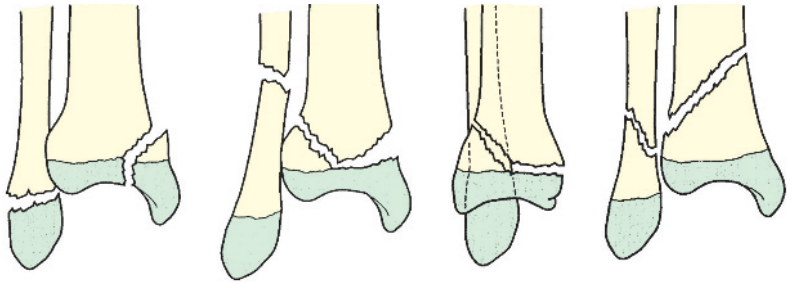
The *triplane fracture* occurs on the medial side of the tibia and is a combination of Salter–Harris type 2 and 3 injuries. Fracture lines appear in the coronal, sagittal and transverse planes. Injury to the physis may result in either asymmetrical growth or arrested growth.

Clinical features

Following a sprain the ankle is painful, swollen, bruised and acutely tender. There may be an obvious deformity, but sometimes the injury looks deceptively mild.

Imaging

Undisplaced physeal fractures – especially those in the distal fibula – are easily missed. Even a hint of physeal widening should be regarded with great suspicion and



(a) (b) (c) (d)

Figure 32.12 Physeal injuries of the distal tibia The classification suggested by Dias and Tachdjian has the merit of pointing to the required reduction manoeuvre – the reverse of the causal mechanism. (a) *Supination–inversion*: the fibular fracture is usually an avulsion (Salter–Harris type 1) whereas the medial malleolar fracture can be variable. (b) *Pronation–eversion–external rotation:* the fibular fracture is often high and transverse. (c) *Supination–plantarflexion:* a fracture of the distal tibia only (Salter–Harris type 1 or 2) with posterior displacement. (d) *Supination–external rotation:* an oblique fibular fracture coupled with a fracture of the distal tibia.

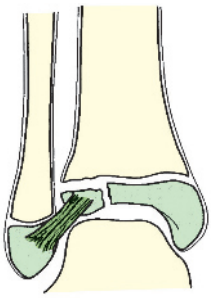
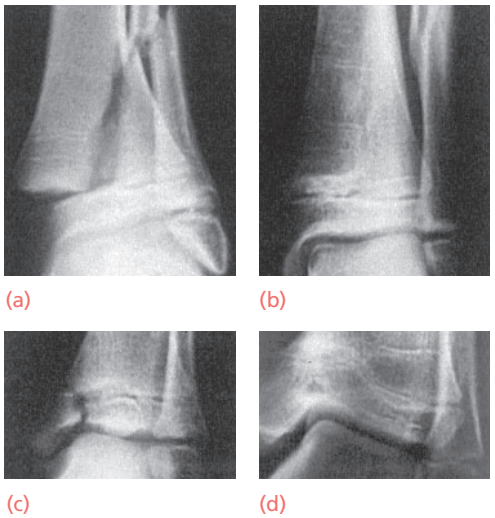


Figure 32.13 Tillaux fracture Diagram illustrating the elements of this unusual injury.

the child *X-rayed* again after 1 week. In an infant the state of the physis can sometimes only be guessed at, but a few weeks after injury there may be extensive periosteal new bone formation.

In triplane fractures the tibial epiphysis may be split in one plane and the metaphysis in another, thus making it difficult to see both fractures in the same X-ray. *CT scans* are particularly helpful in these and other type 3 injuries.



(a) (b) (c) (d)

Figure 32.14 Ankle fractures in children (a) Salter–Harris type 2 injury; after reduction (b) growth has proceeded normally. (c) Salter–Harris type 3 injury; (d) the medial side of the physis has fused prematurely, resulting in distorted growth.

Treatment

Salter–Harris types 1 and 2 injuries are treated closed. If it is displaced, the fracture is gently reduced under general anaesthesia; the limb is immobilized in a full length cast for 3 weeks and then in a below-knee walking cast for a further 3 weeks. Occasionally, surgery is needed to extract a periosteal flap, which prevents an adequate reduction (Figure 32.14).

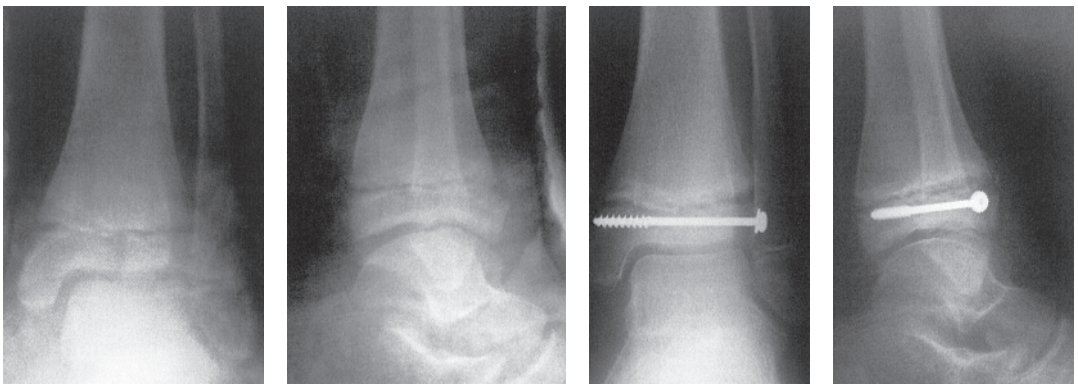
Type 3 or 4 fractures, if undisplaced, can be treated in the same manner, but *the ankle must be re-X-rayed after 5 days to ensure that the fragments have not slipped*. Displaced fractures can sometimes be reduced closed by reversing the forces that produced the injury. However, unless reduction is near-perfect, the fracture should be reduced open and fixed with interfragmentary screws, which are inserted parallel to the physis. Postoperatively the leg is immobilized in a below-knee cast for 6 weeks.

Tillaux fractures are treated in the same way as type 3 fractures (Figure 32.15). Triplane fractures, if undisplaced, can be managed closed but require vigilant monitoring for late displacement (Figure 32.16). Displaced fractures must be reduced and fixed.

Complications

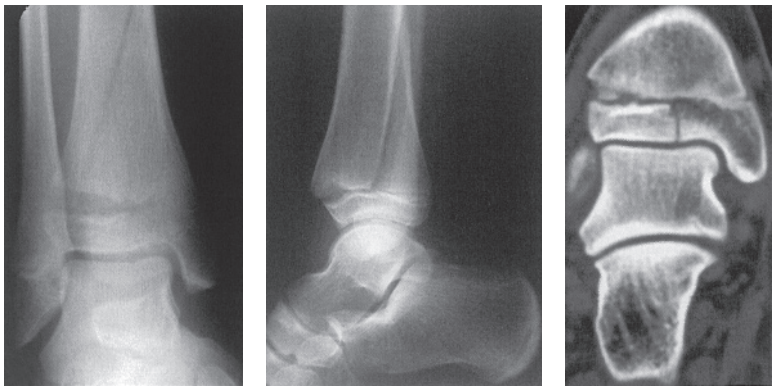
Malunion Imperfect reduction may result in angular deformity of the ankle – usually valgus. In children under 10 years old, mild deformities may be accommodated by further growth and modelling. In older children the deformity should be corrected by a supramalleolar closing-wedge osteotomy.

Asymmetrical growth Fractures through the epiphysis (Salter–Harris type 3 or 4) may result in localized fusion of the physis. The bony bridge is usually in the



(a) (b) (c) (d)

Figure 32.15 Tillaux fracture (a,b) This avulsion fracture of the lateral part of the physis was reduced and fixed percutaneously (c,d).



(a) (b) (c)

Figure 32.16 Triplane fracture The three fracture planes may not be seen in a single X-ray, but can be visualized from a combination of images (a,b). In this case the epiphyseal fracture is clearly seen only in the coronal plane CT scan (c).

medial half of the growth plate; the lateral half goes on growing and the distal tibia gradually veers into varus. MRI and CT are helpful in showing precisely where physeal arrest has occurred. If the bony bridge is small (less than 30% of the physeal width), it can be excised and replaced by a pad of fat in the hope that physeal growth may be restored. If more than half of the physis is involved, or the child is near the end of the growth period, a supramalleolar closing-wedge osteotomy is indicated.

Shortening Early physeal closure occurs in about 2% of children with distal tibial injuries. Fortunately, the resulting limb length discrepancy is usually mild. If it promises to be more than 2 cm and the child is young enough, proximal tibial epiphysiodesis in the opposite limb may restore equality. If the discrepancy is marked, or the child near the end of the growth period, leg lengthening is indicated.

PRINCIPLES IN MANAGING INJURIES OF THE FOOT

Injuries of the foot are apt to be followed by residual symptoms and loss of function that seem out of proportion to the initial trauma. Severe injuries affect the foot as a whole, whatever the particular bone that might be fractured. A global approach is therefore essential in dealing with these injuries, the objective being a return to full weight-bearing without pain, with an appropriate propulsive gait.

Identification of these injuries is particularly challenging in the patient with multiple trauma, where the more subtle foot injuries might be missed as the life-threatening truncal injuries and limb-threatening long bone injuries distract attention from the more subtle injuries to the foot, which may nonetheless impair eventual function.

Clinical assessment

The entire foot should be examined systematically, no matter that the injury may appear to be localized to one spot. Multiple fractures, or combinations of fractures and dislocations, are easily missed. The circulation and nerve supply must be carefully assessed; a well-reduced fracture is a useless achievement if the foot becomes ischaemic or insensate. Similarly, attention must be paid to the soft tissues and functional movement of the foot; the stiff, painful foot is impaired for propulsion, and maybe even for stance.

Fractures and dislocations may cause tenting of the skin; this is always a bad sign because there is a risk of skin necrosis if reduction is delayed.

Imaging

Imaging routinely begins with anteroposterior, lateral and oblique *X-rays* of the foot. If a fracture of the talus or calcaneum or fracture-dislocation of the midtarsal joints is suspected, a *CT scan* of the foot should be obtained.

CT is especially useful for evaluating fractures of the calcaneum, and *MRI* is helpful in diagnosing osteochondral fractures of the talus. *Familiarity with the talocalcaneal anatomy is essential if fractures of the hindfoot are to be diagnosed properly* (Figure 32.17).

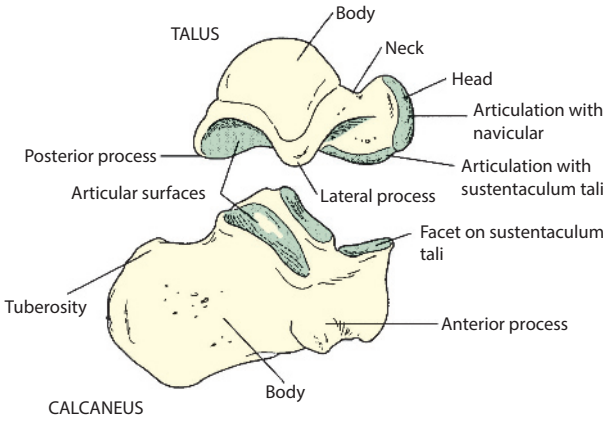


Figure 32.17 Talus and calcaneum The main features of these two bones, and their relationship to each other, are shown here.

Treatment

Swelling is always a problem. Not only does it make clinical examination difficult, but more importantly it may lead to definitive treatment being delayed; fractures and dislocations are more difficult to reduce in a swollen foot. The principles are:

- realign and splint the foot, keep it elevated and apply Cryo-Cuff or ice-packs and intermittent pneumatic compression foot pumps
- make the diagnosis, defining the extent of injury
- start definitive treatment as soon as the fracture pattern is properly defined and swelling permits.

In the rehabilitation phase, if the foot has to be immobilized, exercise those joints that can be left free. Start weight-bearing as soon as the patient will tolerate it, provided this will not jeopardize the reduction. If a removable splint will fit the purpose, use it so that non-weight-bearing exercises can be started as soon as possible. Prolonged immobilization predisposes to stiffness, impaired function, localized osteoporosis and complex regional pain syndrome.

INJURIES OF THE TALUS

Talar fractures and dislocations are relatively uncommon. They usually involve considerable violence, such as car accidents in which the occupants are thrown against the resistant frame of the vehicle, falls from a height, or severe wrenching of the ankle. The injuries include fractures of the neck, body, head or bony processes of the talus, dislocations of the talus or the joints around the talus, osteochondral fractures of the superior articular surface, and a variety of chip or avulsion fractures.

The significance of the more serious injuries is enhanced by two important facts: (1) the talus is a major weight-bearing structure (the superior articular surface carries a greater load per unit area than any other bone in the body); (2) it has a vulnerable blood supply and is a relatively common site for post-traumatic ischaemic necrosis.

Blood vessels enter the bone from the anterior tibial, posterior tibial and peroneal arteries, as well as anastomotic vessels from the surrounding capsule and ligaments. The head of the talus is richly supplied by intraosseous vessels. However, the body of the talus is supplied mainly by vessels that enter the talar neck from the tarsal canal and then run retrograde from distal to proximal. In fractures of the talar neck these vessels are disrupted; if the fracture is displaced, the extraosseous plexus too may be damaged and the body of the talus is at risk of ischaemia.

Mechanism of injury

Fracture of the talar neck is produced by violent hyperextension of the ankle. The neck of the talus is forced against the anterior edge of the tibia, which acts like a cleaver. If the force continues, the fracture is displaced and the surrounding joints may sublux or dislocate.

Fracture of the body is usually a compression injury due to a fall from a height, or an everting force across the body, fracturing the lateral process (the snowboarder's fracture). Avulsion fractures are associated with ligament strains around the ankle and hindfoot.

Clinical features

The patient has most commonly been involved in a motor vehicle accident or has fallen from a height. The foot and ankle are painful and swollen; if the fracture is displaced, there may be an obvious deformity, or the skin may be tented or split. Tenting is a dangerous sign; if the fracture or dislocation is not promptly reduced, the skin may slough and become infected. The pulses should be checked and compared with those in the opposite foot.

Imaging

Anteroposterior, lateral and oblique *X-ray* views are essential (Figure 32.18); *CT scanning* helps to identify associated injuries of the ankle and foot. Both malleoli, the ankle mortise, the talus and all the adjacent tarsal bones should be carefully assessed. Undisplaced fractures are not always easy to see, and sometimes even severely displaced fractures are missed in the initial assessment because of unfamiliarity with the normal appearance – sad but true.

Classification

Fractures of the neck of the talus These fractures are classified according to the system devised by Hawkins and modified by Canale:

- *Type I* – undisplaced
- *Type II* – displaced (however little) and associated with subluxation or dislocation of the subtalar joint
- *Type III* – displaced, with dislocation of the body of the talus from the ankle joint
- *Type IV* – displaced vertical talar neck fracture with associated talonavicular joint disruption.

Fractures of the head of the talus This is a rare injury; the fracture usually involves the talonavicular joint.

Fractures of the body of the talus These are also uncommon. The fracture is often displaced and may cause distortion of the talocalcaneal joint. Rotational malalignment of the fragments is difficult

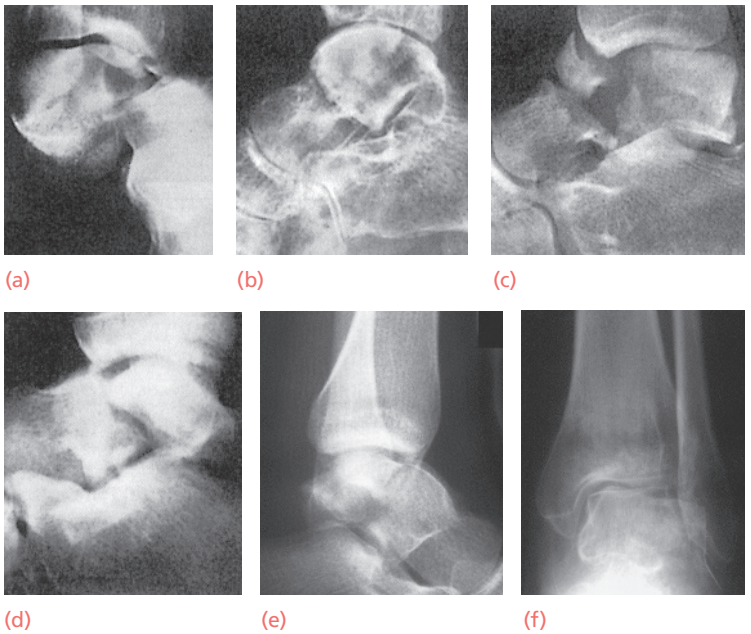


Figure 32.18 Injuries of the talus – X-rays (a) Talocalcaneal fracture-dislocation; (b) undisplaced fracture of the talar neck; (c) type III fracture of the neck; (d) displaced fracture of the body of the talus. (e) This fracture of the body was thought to be well reduced; however, in the AP view (f) it is possible to see two overlapping outlines, indicating that the fragments are malrotated.

to diagnose on plain X-ray examination; the deformity is best visualized by 3D CT reconstruction.

Fractures of the lateral and posterior processes These are usually associated with ankle ligament strains. It is sometimes difficult to distinguish between a fracture of the posterior process and a normal os trigonum. A simple rule is, 'If it's not causing symptoms, it doesn't really matter.'

Osteochondral fractures Osteochondral fractures following acute trauma usually occur on the lateral part of the dome of the talus. The diagnosis is often missed when the patient is first seen and may come to light only after CT or MRI scan.

Treatment

The general principles in managing injuries of the foot set out above should be observed.

UNDISPLACED FRACTURES

A split below-knee plaster, or a supportive boot, is applied and, when the swelling has subsided, is replaced by a complete cast with the foot plantarflexed. Weight-bearing is not permitted for the first 4 weeks; thereafter, the plaster is removed, the fracture position is checked by X-ray, a new cast is applied and weight-bearing is gradually introduced. Further plaster changes or use of an adjustable splintage boot will allow the foot to be brought up, slowly, to plantigrade; physiotherapy is commenced. At 8–12 weeks the splintage is discarded and function is regained by normal use.

DISPLACED FRACTURES OF THE NECK

Even the slightest displacement makes it a type II fracture, which needs to be reduced. If the skin is tight, reduction becomes urgent because of the risk of skin necrosis (Figure 32.19). Reduction must be *perfect*: (1) in order to ensure that the subtalar joint is mechanically sound; and (2) to lessen the chance – or at any rate lessen the effects – of avascular necrosis.

With *type II fractures*, closed manipulation under general anaesthesia can be tried first. Traction is

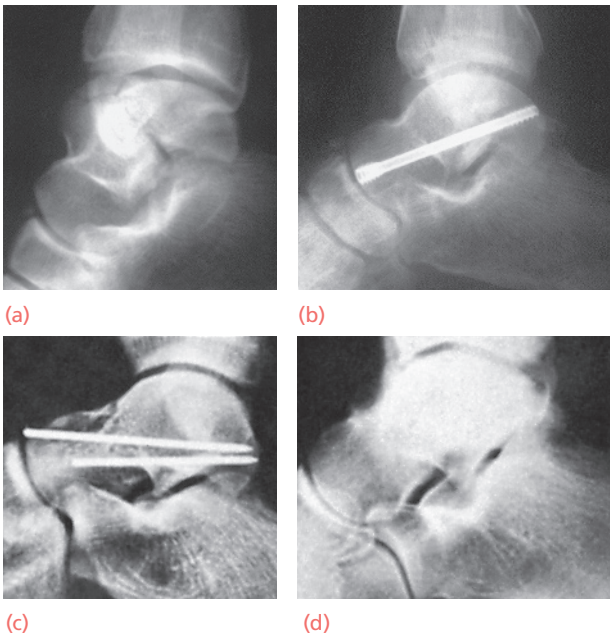


Figure 32.19 Fractures of the talus – treatment (a) This displaced fracture of the body was reduced and fixed with a countersunk screw (b), giving a perfect radiological result. Fractures of the neck, even if well reduced (c) are still at risk of developing ischaemic necrosis (d). The stabilization with K-wires would probably now be improved upon by using definitive screw fixation, but the risk of avascular necrosis would be the same.

applied with the ankle in plantarflexion; the foot is then steered into inversion or eversion to correct the displacement shown on the X-ray. The reduction is checked by X-ray; nothing short of 'anatomic' is acceptable. A below-knee cast is applied (with the foot still in equinus) and this is retained, non-weight-bearing, for 4 weeks. Cast changes after that will allow the foot to be gradually brought up to plantigrade; however, weight-bearing is not permitted until there is evidence of union (8–12 weeks).

If closed reduction fails (which it often does), open reduction is essential; indeed, some would say that *all* type II fractures should be managed by open reduction and internal fixation without attempting closed treatment. Through an anteromedial incision the fracture is exposed and manipulated into position. Wider access can be obtained by pre-drilling and then osteotomizing the medial malleolus. The fracture reduction is checked by X-ray and the fracture is then fixed with two screws. The osteotomized malleolar fragment is fixed back in position with screws. Postoperatively a below-knee cast is applied; weight-bearing is not permitted until there are signs of union (8–12 weeks).

Type III fracture-dislocations need urgent open reduction and internal fixation. The approach will depend on the fracture pattern and position of displaced fragments. Osteotomy of the medial malleolus might help; the malleolus is pre-drilled for screw fixation and osteotomized and retracted distally without injuring the deltoid ligament. This wide exposure is essential to permit removal of small fragments from the ankle joint and perfect reduction of the displaced talar body under direct vision; even then, it is difficult! The position is checked by X-ray and the fracture is then fixed securely with screws. If there is the slightest doubt about the condition of the skin, the wound is left open and delayed primary closure carried out 5 days later. Postoperatively the foot is splinted and elevated until the swelling subsides; a below-knee cast or splintage boot is then applied, following the same routine as for type II injuries.

DISPLACED FRACTURES OF THE BODY

Fractures through the body of the talus are usually displaced or comminuted and involve the ankle and/or the talocalcaneal joint; occasionally the fragments are completely dislocated.

Minimal displacement can be accepted; a below-knee, non-weight-bearing cast is applied for 6–8 weeks; this is then replaced by a weight-bearing cast for another 4 weeks.

The small number of horizontal fractures that do not involve the ankle or subtalar joint are treated by closed reduction and cast immobilization (as earlier).

Displaced fractures with dislocation of the adjacent joints should be accurately reduced. In almost all cases open reduction and internal fixation will be needed (Figure 32.19). An osteotomy of the medial malleolus is useful for adequate exposure of the talus; the malleolus is predrilled before the osteotomy and fixed back into position after the talar fracture has been dealt with. The prognosis for these fractures is poor: there is a considerable incidence of malunion, joint incongruity, avascular necrosis and secondary osteoarthritis of the ankle or talocalcaneal joint.

DISPLACED FRACTURES OF THE HEAD

The main problem is injury to the talonavicular joint. If the fragments are large enough, open reduction and internal fixation with screws is the recommended treatment. If there is much comminution, it may be better simply to excise the smaller fragments. Postoperative immobilization is the same as for other talar fractures.

FRACTURES OF THE TALAR PROCESSES

If the fragment is large enough, open reduction and fixation with K-wires or small screws is advisable. Tiny fragments are left but can be removed later if they become symptomatic.

OSTEOCHONDRAL FRACTURES

These small surface fractures of the dome of the talus usually occur with severe ankle sprains or subtalar dislocations. Most acute lesions can be treated by cast immobilization for 4–6 weeks. Occasionally a displaced fragment is large enough to warrant operative replacement and internal fixation – easier said than done! More often it is separated from its bed and is excised: the exposed bone is then drilled to encourage repair by fibrocartilage.

OPEN FRACTURES

Fractures of the talus are often associated with burst skin wounds. In some cases the fracture becomes 'open' when stretched or tented skin starts sloughing. There is a high risk of infection in these wounds and prophylactic antibiotics are advisable.

The injury is treated as an emergency. Under general anaesthesia, the wound is cleaned and debrided and all necrotic tissue is removed. The fracture is then dealt with as for closed injuries, except that the wound is left open and closed by delayed primary suture or skin grafting 5–7 days later, when swelling has subsided and it is certain that there is no infection. The plastic surgeons may have a role to play in providing early cover and closure.

Sometimes, in open injuries, the talus is completely detached and lying in the wound. After adequate debridement and cleansing, the talus should be replaced in the mortise and stabilized, with fixaton. Later definitive fixation is then performed.

Complications

Malunion The importance of accurate reduction has been stressed. Malunion will lead to distortion of the joint surface, limitation of movement and pain on weight-bearing. If early follow-up X-rays show redisplacement of the fragments, a further attempt at reduction is justified. Persistent malunion predisposes to osteoarthritis.

Avascular necrosis Avascular necrosis of the body of the talus occurs in displaced fractures of the talar neck. The incidence varies with the severity of displacement: in type I fractures it is less than 10%; in type II about 30–40%; and in type III more than 90%. The earliest X-ray sign (often present by the sixth week) is apparent increased density of the avascular segment; in reality it is the rest of the tarsus that has become slightly porotic with disuse, but the avascular portion remains unaffected and therefore looks more 'dense'. The opposite is also true: if the dome of the talus becomes osteoporotic, this means that it has a blood supply and it will not develop osteonecrosis. This is the basis of Hawkins' sign, which should be looked for 6–8 weeks after injury.

If osteonecrosis does occur, the body of the talus will eventually appear on X-ray to be more dense than the surrounding bones. Despite necrosis, the fracture may heal, so treatment should not be interrupted by this event; if anything, weight-bearing should be delayed in the hope that the bone is not unduly flattened. Function may yet be reasonable. However, if the talus becomes flattened or fragmented, or pain and disability are marked, the ankle may need to be arthrodesed.

Secondary osteoarthritis Osteoarthritis of the ankle and/or subtalar joints occurs some years after injury in over 50% of patients with talar neck fractures. There are a number of causes: (1) articular damage due to the initial trauma; (2) malunion and distortion of the articular surface; (3) avascular necrosis of the talus. Pain and stiffness may be managed by judicious analgesic medication and orthotic adjustments, but in some cases the painful hindfoot will simply not allow a return to function; arthrodesis of the affected joints can help to relieve symptoms. Operative fusion of one joint may predispose to overload of the associated foot joints, and hence to later arthritis, but this should be accepted, and is usually many years later.

FRACTURES OF THE CALCANEUM

The calcaneum is the most commonly fractured tarsal bone, and in 5–10% of cases both heels are injured simultaneously. Crush injuries, although they always heal in the biological sense, are likely to be followed by long-term disability. The general attitude to these injuries at the beginning of the twentieth century (at least from an industrial point of view) was that 'the man who breaks his heel-bone is finished'. This was followed by attempts, throughout the latter part of that century, to modify the outcome through open reduction and internal fixation of these fractures. The efficacy of fixation and the results of operative intervention have been studied and questioned in the past few years. There is still a role for fixation, but the decision-making and operative techniques require considerable expertise.

Mechanism of injury

In most cases the patient falls from a height, often from a ladder, onto one or both heels. The calcaneum is driven up against the talus and is split or crushed. Over 20% of these patients suffer associated injuries of the spine, pelvis or hip.

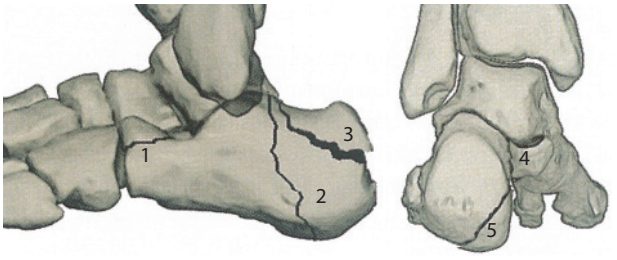
Avulsion fractures sometimes follow traction injuries of the tendo Achillis or the ankle ligaments. Occasionally the bone is shattered by a direct blow.

Pathological anatomy

Based largely on the work of Palmer and Essex-Lopresti in the 1940s and 1950s, it has been customary to divide calcaneal fractures into *extra-articular fractures* (those involving the various calcaneal processes or the body posterior to the talocalcaneal joint – Figure 32.20) and *intra-articular fractures* (those that split the talocalcaneal articular facet).

EXTRA-ARTICULAR FRACTURES

These account for 25% of calcaneal injuries. They usually follow fairly simple patterns, with shearing or avulsion of the anterior process, the sustentaculum tali, the tuberosity or the inferomedial process. Fractures of the posterior (extra-articular) part of



(a) (b)

Figure 32.20 Extra-articular fractures of the calcaneum
Fractures may occur through: 1 the anterior process, 2 the body, 3 the tuberosity, 4 the sustentaculum tali, or 5 the medial tubercle. Treatment is closed unless the fragment is large and badly displaced, in which case it will need to be fixed back in position.

the body are caused by compression. Extra-articular fractures are usually easy to manage and have a good prognosis.

INTRA-ARTICULAR FRACTURES

These injuries are much more complex and unpredictable in their outcome. They are best understood by imagining the impact of the talus cleaving the bone from above to produce a *primary fracture line* that runs obliquely across the posterior articular facet and the body from posteromedial to anterolateral (Figure 32.21). Where it splits, the posterior articular facet depends upon the position of the foot at impact: if the heel is in valgus (abducted), the fracture is in the lateral part of the facet; if the heel is in varus (adducted), the fracture is more medial.

The upward displacement of the body of the calcaneum produces one of the classic X-ray signs of a 'depressed' fracture: flattening of the angle subtended by the posterior articular surface and the upper surface of the body posterior to the joint (Böhler's angle).

The advent of CT, and the trend towards operative reduction and fixation of displaced calcaneal fractures, have sharpened our understanding of these complex injuries (Figure 32.22). There are two important ways of assessing or classifying these injuries that are of relevance to the treating surgeon (and the patient). The work of Sanders and Gregory has helped to define the intra-articular fracture pattern and the associated outcome and prognosis. Knowledge of the variations in fracture pattern, particularly in relation to the lateral wall of the calcaneum, has improved our understanding of the anatomy that is likely to be encountered at operation, approaching from an extended L-shaped incision; the lateral joint fragment may sometimes be trapped within the body of the calcaneum and can only be reduced if the lateral wall of the body is osteotomized so as to gain access to it.

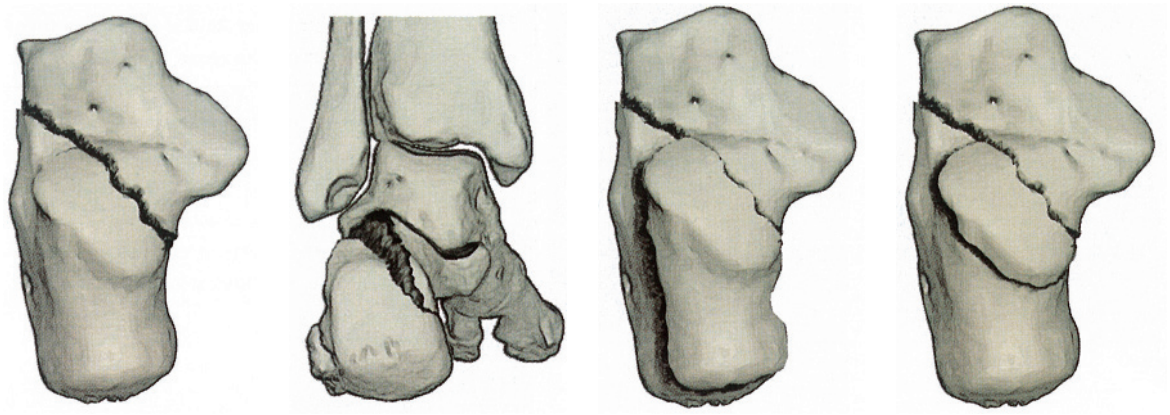


Figure 32.21 Intra-articular fractures of the calcaneum 1 The primary fracture line (a,b) is created by the impact of the talus on the calcaneum – it runs from posteromedial to anterolateral. Secondary fracture lines may create 'tongue' (c) or 'joint depression' (d) variants to the fracture pattern.

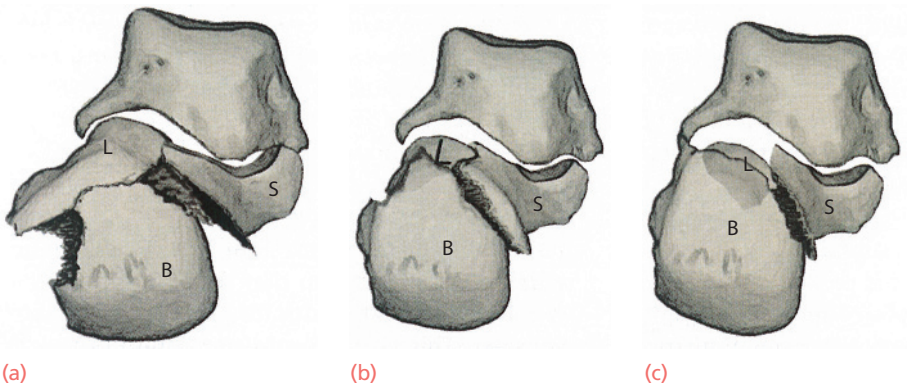


Figure 32.22 Intra-articular fractures of the calcaneum 2 CT scans have allowed a better understanding of the fracture anatomy. A coronal CT scan enables the identification of three major fragments in most intra-articular fractures: the lateral joint fragment (L), the sustentaculum tali (S) and the body fragment (B). In type 1 fractures (a) the lateral joint fragment is in valgus whereas the body is in varus. In type 2 fractures (b), the sustentaculum tali is in varus and the lateral joint is elevated in relation to it. In type 3 fractures (c) the lateral joint fragment is impacted and buried within the body fragment.

Clinical features

There is usually a history of a fall from a height or a road traffic accident; in elderly osteoporotic people even a comparatively minor injury may fracture the calcaneum.

The foot is painful and swollen and a large bruise appears on the lateral aspect of the heel. The heel may look broad and squat. The surrounding tissues are thick and tender, and the normal concavity below the lateral malleolus is lacking. The subtalar joint cannot be moved but ankle movement is possible.

Always check for signs of a compartment syndrome of the foot (intense pain, very extensive bruising and swelling, diminished sensation, with pain on passive toe movement).

Imaging

Plain X-rays should include lateral views, but once a fracture has been identified then cross-sectional imaging (*CT scan*) is the standard of care. Extra-articular fractures are usually fairly obvious. Intra-articular fractures can also often be identified in the plain films and, if there is displacement of the fragments, the lateral view may show flattening of the tuber-joint angle (Böhler's angle) (Figure 32.23).

For accurate definition of intra-articular fractures, however, CT is essential and 3D reconstruction views are even better. Coronal sections will show the fracture 'geometry' clearly enough to permit accurate diagnosis of most intra-articular fractures.

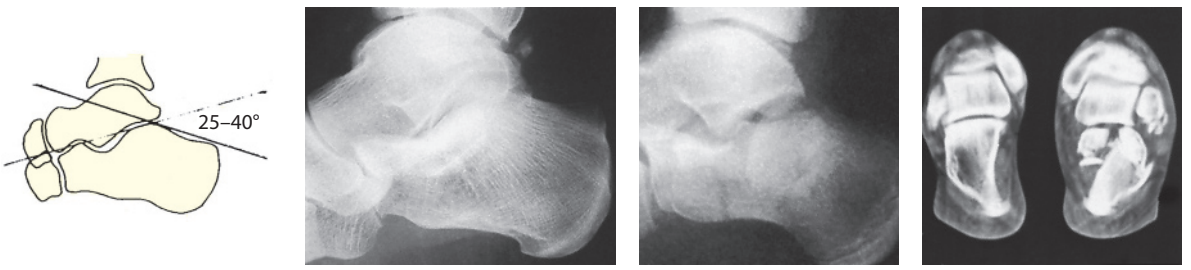
With severe injuries – and especially with bilateral fractures (Figure 32.24) or in the unconscious patient – *it is essential to assess the knees, spine and pelvis as well.*

Treatment

For all except the most minor injuries, the patient is admitted to hospital so that the leg and foot can be elevated and treated with cold (ice or Cryo-Cuff) and compression until swelling subsides. This also gives time to obtain the necessary CT scans.

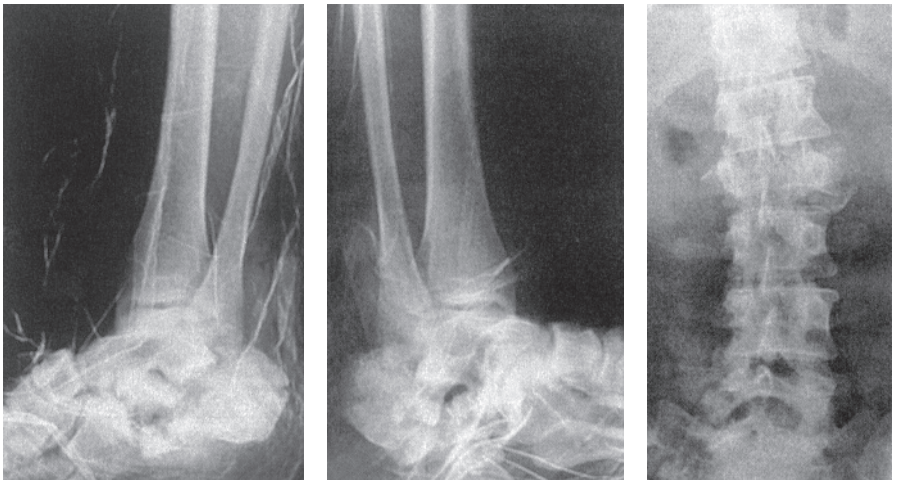
EXTRA-ARTICULAR FRACTURES

The essence of management of extra-articular fractures is 'mobility and function are more important than anatomical repositioning'. The vast majority are treated closed: (1) compression bandaging, ice packs and elevation until the swelling subsides; (2) exercises as soon as pain permits; (3) no weight-bearing for 4 weeks and partial weight-bearing for another 4 weeks. Variations from this routine relate to specific injuries.



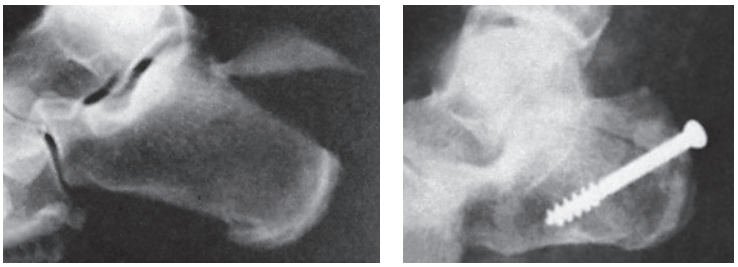
(a) (b) (c) (d)

Figure 32.23 Fracture of the calcaneum – imaging (a,b) Measurement of Böhler's angle and the X-ray appearance in a normal foot. (c) Flattening of Böhler's angle in a fractured calcaneum. (d) The CT scan in this case shows how the articular fragments have been split apart.



(a) (b) (c)

Figure 32.24 Calcaneal fractures – imaging Bilateral calcaneal fractures (a,b) are caused by a fall on the heels from a height or by an explosion from below. In either case the spine also may be fractured, as it was in this patient (c). With bilateral heel fractures, always assess the spine.



(a) (b)

Figure 32.25 Extra-articular calcaneal fractures – treatment (a) Avulsion fracture of posterosuperior corner (b) fixed by a screw.

Fractures of the anterior process Most of these are avulsion fractures and many are mistaken for an ankle sprain. Oblique X-rays will show the fracture, which almost always involves the calcaneocuboid joint. If there is a large displaced fragment, internal fixation may be needed; this is followed by the usual 'closed' routine.

Fractures of the tuberosity These are usually due to avulsion by the tendo Achillis; clinical signs are similar to those of a torn Achilles tendon. If the fragment is displaced, it should be reduced and fixed with cancellous screws (Figure 32.25); the foot is then immobilized in slight equinus to relieve tension on the tendo Achillis. Weight-bearing can be permitted after 4 weeks.

Fractures of the body If it is certain that the subtalar joint is not involved, the prognosis is good and the fracture can be treated by the usual 'closed' routine. However, if there is much sideways displacement and widening of the heel, closed reduction by manual compression should be attempted. Weight-bearing is avoided for 6–8 weeks; however, cast immobilization is unnecessary except if both heels are fractured or if the patient simply cannot or will not manage a one-legged gait with crutches (e.g. those who are elderly, frail or poorly compliant).

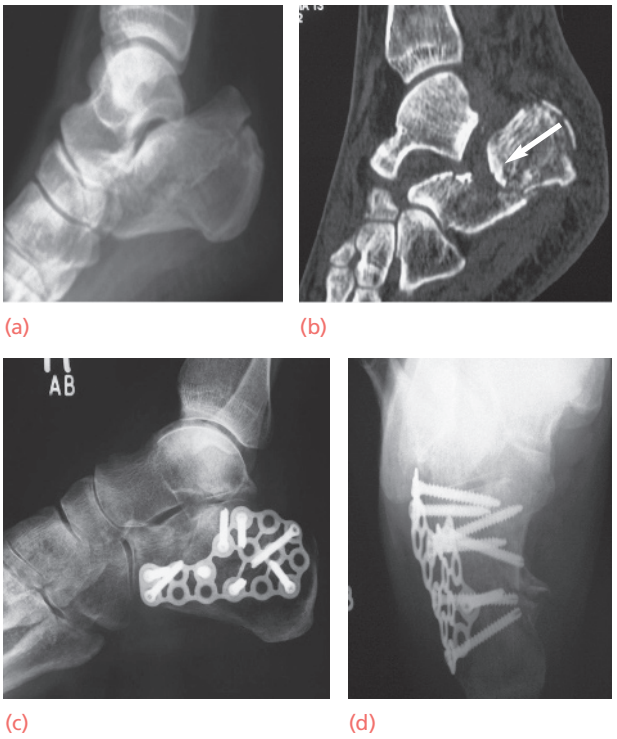
INTRA-ARTICULAR FRACTURES

Undisplaced fractures These are treated in much the same way as extra-articular fractures: compression bandaging, ice-packs and elevation followed by exercises and non-weight-bearing for 6–8 weeks. As long as vertical stress is avoided, the fracture will not become displaced; cast immobilization is therefore unnecessary and it may even be harmful in that it increases the risk of stiffness and algodystrophy. Good or excellent results can be expected in most patients with undisplaced intra-articular fractures.

Displaced intra-articular fractures Open reduction and internal fixation as soon as the swelling subsides is the best treatment for these fractures. CT has greatly facilitated this approach; the medial and lateral fragments can be clearly defined and, with suitable drawings or models, the surgical procedure can be carefully planned and rehearsed.

The operation is usually performed through a single, wide lateral approach; access to the posterior facet and medial fragment is achieved by taking down the lateral aspect of the calcaneum, performing the reduction, and then rebuilding this wall. The various fragments are held with interfragmentary screws – bone grafts are sometimes added to fill in defects. The anterior part of the calcaneum and the calcaneocuboid joint also need attention; the fragments are similarly reduced and fixed. Finally a contoured plate is placed on the lateral aspect of the calcaneum to buttress the entire assembly (Figure 32.26). The wound is then closed and may be drained.

Postoperatively the foot is lightly splinted and elevated. Exercises are begun as soon as pain subsides and after about 2 weeks the patient can be allowed



(a) (b) (c) (d)

Figure 32.26 Intra-articular calcaneal fracture – treatment (a) X-ray gives limited information, but the CT (b) shows the severe depression of the posterior calcaneal facet. This was treated operatively with a calcaneal locking plate, to reconstitute the posterior facet (arrow) and restore the height of the calcaneum (c,d).

up non-weight-bearing on crutches. Partial weight-bearing is permitted only when the fracture has healed (seldom before 8 weeks) and full weight-bearing about 4 weeks after that. Restoration of function may take 6–12 months.

Outcome

Extra-articular fractures and *undisplaced intra-articular fractures*, if properly treated, usually have a good result. However, the patient should be warned that it may take 6–12 months before full function is regained, and in about 10% of cases there will be residual symptoms that might preclude a return to their previous job if this involved walking on uneven surfaces or balancing on ladders.

The outcome for *displaced intra-articular fractures* is much less predictable. The results of operative treatment are heavily dependent on the severity of the fracture and the experience of the surgeon. A Canadian multicentre study showed a shorter time off work and lower requirement for subtalar arthrodesis in those managed operatively. Results were particularly favourable with internal fixation in younger men and those not working with heavy loads or receiving workmen's compensation. In experienced hands, for selected fractures, this is a rational treatment. A large multicentre UK trial cast doubt on the need for fixation, but scrutiny of this revealed significant selection bias. However, calcaneal surgery is not an enterprise for the occasional foot and ankle/trauma surgeon and, unless the appropriate skills and facilities are available, the patient should be referred to a specialist centre.

The fact remains that the heel fracture is a serious and disabling injury in many patients with heavy or physically demanding jobs; mechanical reconstruction of the bony anatomy does not necessarily improve the functional outcome.

Complications

EARLY

Swelling and blistering Intense swelling and blistering may jeopardize operative treatment. The limb should be elevated with the minimum of delay.

Compartment syndrome About 10% of patients develop intense pressure symptoms. The risk of a full-blown compartment syndrome can be minimized by starting treatment early. If operative decompression is carried out, this will delay any definitive procedure for the fracture.

LATE

Malunion Closed treatment of displaced fractures, or injudicious weight-bearing after open reduction, may result in malunion. *The heel is broad and squat*, and the patient has a problem fitting shoes. Usually the foot is in valgus and walking may be impaired.

Peroneal tendon impingement Lateral displacement of the body of the calcaneum may cause painful compression of the peroneal tendons against the lateral malleolus. Treatment consists of operative paring down of protuberant bone on the lateral wall of the calcaneum.

Insufficiency of the tendo Achillis The loss of heel height may result in diminished tendo Achillis action. If this interferes markedly with walking, subtalar arthrodesis with insertion of a bone block may alleviate the problem.

Talocalcaneal stiffness and osteoarthritis Displaced intra-articular fractures may lead to joint stiffness and, eventually, osteoarthritis. This can usually be managed conservatively but persistent or severe pain may necessitate subtalar arthrodesis. If the calcaneocuboid joint is also involved, a triple arthrodesis is better.

MIDTARSAL INJURIES

Injuries in this area vary from minor sprains, often incorrectly labelled as 'ankle' sprains, to severe fracture-dislocations that can threaten the survival of the foot. The mechanism differs accordingly, from benign twisting injuries to crushing forces that produce severe soft-tissue damage; bleeding into the fascial compartments of the foot may cause a typical compartment syndrome.

Isolated injuries of the navicular, cuneiform or cuboid bones are rare. Fractures in this region should be assumed to be 'combination' fractures or fracture-subluxations, until proved otherwise.

Remember that small flakes of bone on X-ray often have large ligaments attached to them, and that 'midfoot sprain' (like 'partial Achilles tendon rupture') is a dangerous diagnosis to make.

Pathological anatomy

The most useful classification is that of Main and Jowett, which is based on the mechanism of injury.

- *Medial stress injuries* are caused by violent inversion of the foot and vary in severity from sprains of the midtarsal joint to subluxation or fracture-subluxation of the talonavicular or midtarsal joints.
- *Longitudinal stress injuries* are the most common. They are caused by a severe longitudinal force with the foot in plantarflexion. The navicular is compressed between the cuneiforms and the talus,

resulting in fracture of the navicular and subluxation of the midtarsal joint.

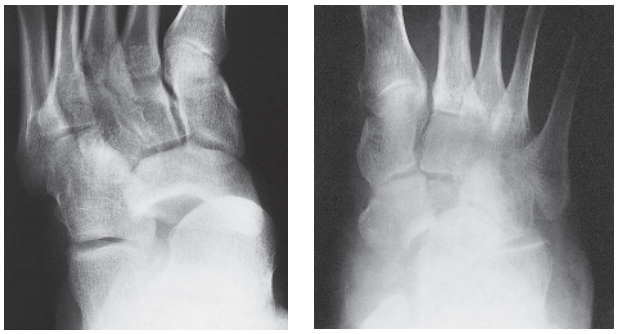
- *Lateral stress injuries* are usually due to falls in which the foot is forced into valgus. Injuries include fractures and fracture-subluxations of the cuboid and the anterior end of the calcaneum as well as avulsion injuries on the medial side of the foot.
- *Plantar stress injuries* result from falls in which the foot is twisted and trapped under the body; they usually present as dorsal avulsion injuries or fracture-subluxation of the calcaneocuboid joint.
- *Crush injuries* usually cause open comminuted fractures of the midtarsal region.

Clinical features

The foot is bruised and swollen. Tenderness is usually diffuse across the midfoot. A medial midtarsal dislocation looks like an 'acute club foot' and a lateral dislocation produces a valgus deformity; with longitudinal stress injuries there is often no obvious deformity. Any attempt at movement is painful. It is important to exclude distal ischaemia or a compartment syndrome.

Imaging

Multiple X-ray views are necessary to determine the extent of the injury; be sure that *all* the tarsal bones are clearly shown. Tarsometatarsal dislocation may be missed if the forefoot falls back into place; fractures of the tarsal bones or bases of the metatarsals should alert the surgeon to this possibility (Figure 32.27). Abnormality of alignment, or fracture, on any view should lead to CT scanning to better assess the extent of injury (Figure 32.28).



(a) (b)

Figure 32.27 Midtarsal injuries (a) X-ray showing dislocation of the talonaviclar joint. (b) X-ray on another patient showing longitudinal compression fracture of the navicular bone and subluxation of the head of the talus. This injury is often difficult to demonstrate accurately on plain X-ray.

Treatment

Ligamentous strains The foot may be bandaged until acute pain subsides. Thereafter, movement is encouraged. Be prepared to re-examine and re-X-ray the foot that does not settle within a few weeks.

Undisplaced fractures The foot is elevated to counteract swelling. After 3–4 days a below-knee cast or removable splintage boot is applied and the patient is allowed up on crutches with limited weight-bearing. The plaster is retained for 4–6 weeks.

Displaced fractures An isolated navicular or cuboid fracture is sometimes displaced and, if so, may need open reduction and screw fixation.

Fracture-dislocation These are severe injuries. Under general anaesthesia, the dislocation can usually be reduced by closed manipulation but holding it is a problem. If there is the least tendency to redisplacement, percutaneous K-wires are run across the joints to fix them in position.

The foot is immobilized in a below-knee cast for 6–8 weeks. Exercises are then begun and should be practised assiduously; it may be 6–8 months before function is regained.

If accurate reduction cannot be achieved by closed manipulation, open reduction and screw fixation is necessary; the importance of anatomical reduction cannot be overemphasized. However, missed fractures are a lost cause and open reduction will seldom improve the situation in those who present late (more than a few weeks after injury). In these cases an arthrodesis of the involved joints might be appropriate.

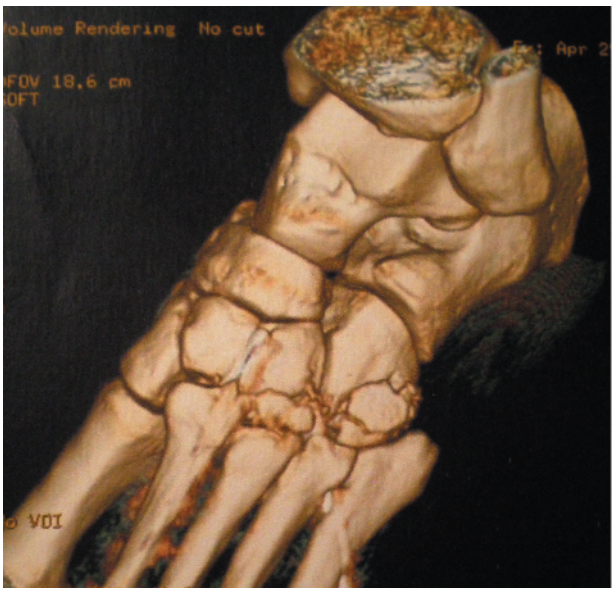


Figure 32.28 Midtarsal injuries Reconstructed CT after reduction of a severe tarsometatarsal injury reveals associated injuries of the cuboid and the lateral cuneiform.

Comminuted fractures Severely comminuted fractures defy accurate reduction. Attention should be paid to the soft tissues; there is a risk of ischaemia. The foot is splinted in the best possible position and elevated until swelling subsides. Early arthrodesis, with restoration of the longitudinal arch, is advisable, with stable fixation and interpositional bone graft block.

Outcome

A major problem with midtarsal injuries is the frequency with which fractures and dislocations are missed at the first examination, resulting in undertreatment and a poor outcome. Even with accurate reduction of midtarsal fracture-dislocations, post-traumatic osteoarthritis may develop and about 50% of patients fail to regain normal function. If symptoms are persistent and intrusive, arthrodesis may be indicated.

TARSOMETATARSAL INJURIES

The five tarsometatarsal (TMT) joints form a structural complex that is held intact partly by the interdigitating joints and partly by the strong ligaments that bind the metatarsal bones to each other and to the tarsal bones of the midfoot.

An appreciation of the anatomy across the TMT joints is important in understanding these injuries. The second metatarsal base is set into a recess formed by the medial, intermediate and lateral cuneiforms. There is no ligament between the first and second metatarsal bases, but the plantar ligament between second metatarsal base and medial cuneiform is short and thick. In the coronal plane, the second metatarsal base forms the apex or keystone in the arch.

Dislocation is rare, but important not to miss; twisting and crushing injuries are the usual causes, with the foot buckling or twisting at the midfoot–forefoot junction. The term *Lisfranc injury* is often used for the disruptions that occur at the midfoot–forefoot junction. Classifying these by direction of forefoot dislocation is, however, pointless – it is neither a guide to treatment nor an indication of outcome. These are often high-energy injuries with extensive damage to the whole region of the foot, and simply to assess the direction of metatarsal displacement is to miss the complexity of the injury pattern.

Clinical features

TMT dislocation or fracture-dislocation should always be suspected in patients with pain and swelling of the foot after high-velocity car accidents and falls. Unfortunately, about 20–30% of these injuries are initially missed. Only with severe injury is there an obvious deformity.

Imaging

X-rays may be difficult to interpret; something looks wrong but it is often difficult to tell what. A systematic method for examining the foot X-rays can help to improve the pick-up rate for these injuries. Concentrate on the second and fourth metatarsals in the oblique views: the medial edge of the second should be in line with the medial edge of the second cuneiform, and the medial edge of the fourth should line up with the medial side of the cuboid. A true lateral may show the dorsal displacement of the second metatarsal base. If a fracture-dislocation is suspected (the displacement may reduce spontaneously and not be immediately detectable), stress views may reveal the abnormality, but a *CT scan* is a more efficient way of showing the extent of injury.

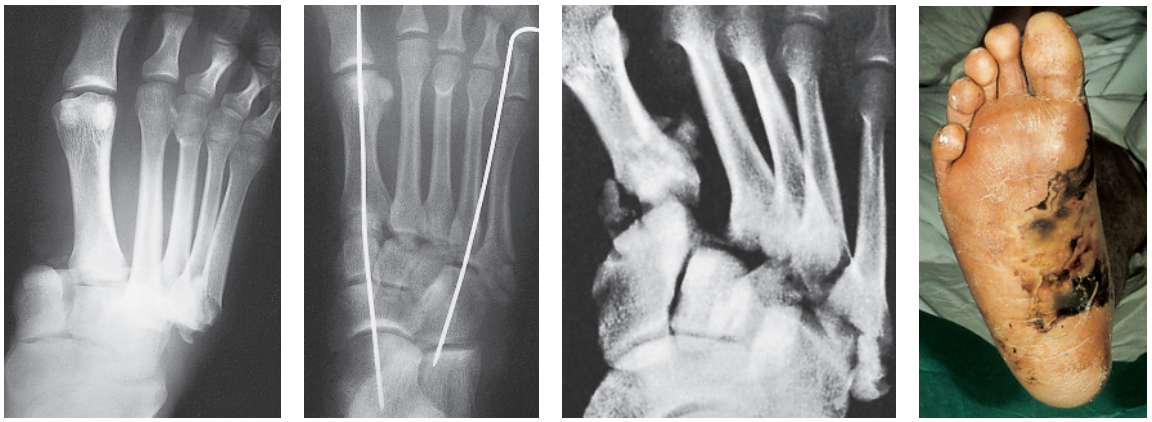
Treatment

The method of treatment depends on the severity of the injury. Undisplaced sprains require cast or boot immobilization for 4–6 weeks. Subluxation or dislocation calls for accurate reduction. This can often be achieved by traction and manipulation under anaesthesia; the position is then held with percutaneous K-wires or screws and cast immobilization (Figure 32.29). The cast is changed after a few days when swelling has subsided; the new cast is retained, non-weight-bearing, for 6–8 weeks. The K-wires are then removed and rehabilitation exercises begun.

If closed reduction fails, open reduction is essential. The key to success is the second TMT joint. Through a longitudinal incision, the base of the second metatarsal is exposed and the joint manipulated into position. Reduction of the remaining parts of the tarsometatarsal articulation will not be too difficult. The bones are fixed with percutaneous K-wires or screws and the foot is immobilized as described earlier.

Complications

Compartment syndrome A tensely swollen foot may hide a serious compartment syndrome that could result in ischaemic contractures. If this is suspected, intracompartmental pressures should be measured (see Chapter 23). Treatment should be prompt and effective in decompressing the affected area. Through a medial longitudinal incision, or two well-spaced dorsal incisions, all the compartments can be decompressed; the wound is left open until swelling subsides and the skin can be closed without tension or grafted with a meshed graft that will later contract.



(a) (b) (c) (d)

Figure 32.29 TMT injuries (a) Dislocation of the TMT joints. (b) X-ray after reduction and temporary stabilization with K-wires – later stabilization with screw fixation was performed. (c) X-ray showing a high-energy fracture-dislocation involving the TMT joints. These are serious injuries that may be complicated by (d) compartment syndrome of the foot.

INJURIES OF METATARSAL BONES

Metatarsal fractures are relatively common and are of four types: (1) crush fractures due to a direct blow; (2) a spiral fracture of the shaft due to a twisting injury; (3) avulsion fractures due to ligament strains; (4) insufficiency fractures due to repetitive stress.

Clinical features

In acute injuries pain, swelling and bruising of the foot are usually quite marked; with stress fractures, the symptoms and signs are more insidious.

X-rays

X-rays should include routine AP, lateral and oblique views of the entire foot; multiple injuries are not uncommon. Undisplaced fractures may be difficult to detect and stress fractures usually show nothing at all until several weeks later.

Treatment

Treatment will depend on the type of fracture, the site of injury and the degree of displacement.

UNDISPLACED AND MINIMALLY DISPLACED FRACTURES

These can be treated by support in a below-knee cast or removable boot splint; the foot is elevated and active movements are started immediately, partial weight-bearing for about 4–6 weeks. At the end of that period, exercise is very important and the patient is encouraged to resume normal activity. Slight malunion rarely results in disability once mobility has been regained.

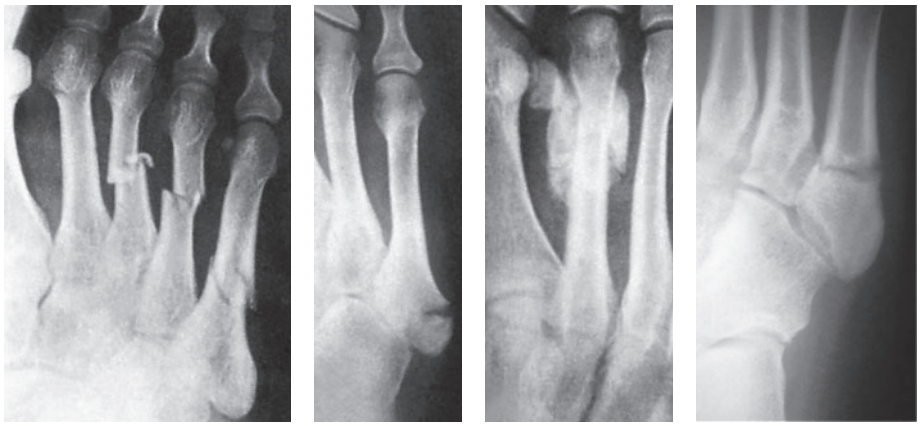
DISPLACED FRACTURES

Displaced fractures can usually be treated closed. The foot is elevated until swelling subsides. The fracture may be reduced by traction under anaesthesia and the leg immobilized in a cast – non-weight-bearing – for 4 weeks. Alternatively the fracture position might be accepted, depending on the degree of displacement. For the second to fifth metatarsals, displacement in the coronal plane can be accepted and closed treatment, as above, is satisfactory. However, for the first metatarsal and for all fractures with significant displacement in the sagittal plane (i.e. depression or elevation of the displaced fragment) open reduction and internal fixation with K-wires, or better with stable fixation using a plate and small screws, is advisable. A below-knee cast or boot is applied and weight-bearing is avoided for 4 weeks; this is then replaced by a weight-bearing cast or boot for another 4 weeks.

Fractures of the metatarsal neck have a tendency to displace, or re-displace, with closed immobilization. It is therefore important to check the position repeatedly if closed treatment is used. If the fracture is unstable, it may be possible to maintain the position by percutaneous K-wire or screw fixation. The wire is removed after 4 weeks; cast immobilization is retained for 4–6 weeks.

FRACTURES OF THE FIFTH METATARSAL BASE

Forced inversion of the foot (the 'pothole injury') may cause avulsion of the base of the fifth metatarsal, with pull-off by the peroneus brevis tendon or the lateral band of the plantar fascia (Figure 32.30). Pain due to a sprained ankle may overshadow pain in the foot. Examination will disclose a point of tenderness



(a) (b) (c) (d)

Figure 32.30 Metatarsal injuries (a) Transverse fractures of three metatarsal shafts. (b) Avulsion fracture of the base of the fifth metatarsal – the pothole injury, or Robert Jones fracture. (c) Florid callus in a stress fracture of the second metatarsal. (d) Jones fracture of the fifth metatarsal.

directly over the prominence at the base of the fifth metatarsal bone.

A careful assessment of the fracture pattern will provide a guide to prognosis and treatment. Again, an appreciation of the pathoanatomy explains these factors.

The fifth metatarsal base extends much more proximal into the midfoot region, compared to the other metatarsal bases. It articulates with the cuboid and with the fourth metatarsal. The peroneus brevis tendon and lateral band of the plantar fascia insert onto the base of the fifth metatarsal. There is a relative watershed in the blood supply to the fifth metatarsal at the junction between the diaphysis and metaphysis.

Robert Jones, a founding father and doyen of orthopaedics, described his own fracture (sustained while dancing), as a fracture of the fifth metatarsal about three-fourths of an inch from its base. Unfortunately, as observed above with Pott's fractures, what has passed into history as this eponymous fracture is often not what was actually described, and the term 'Jones fracture' is now sometimes used for any fracture of the proximal fifth metatarsal. A more useful classification system takes account of the fracture line, and whether it is proximal, affecting the tuberosity, in the region of articulation with the fourth metatarsal, or at the metaphyseal/diaphyseal junction – the latter has a higher rate of non-union, probably as a consequence of the relatively poor blood supply in that region.

Occasionally a normal peroneal ossicle in this area may be mistaken for a fracture; there is also an apophyseal ossification centre in the tuberosity.

Treatment

The proximal avulsion fractures can usually be treated symptomatically, with initial rest and support, but with early mobilization and return to function.

The intra-articular injuries and those at the metaphyseal–diaphyseal junction may also be treated non-operatively, but there is a greater risk of non-union and slower return to function. The role of fixation with an interfragmentary screw or screws and plate is therefore an issue for discussion between the surgeon and the patient, depending to a large extent on the patient's functional demands and expectations with respect to sport and activity, and time away from these.

Stress injury (march fracture)

In a young adult (often a military recruit or a runner building up training) the foot may become painful and slightly swollen after overuse. A tender lump is palpable just distal to the midshaft of a metatarsal bone. Usually the second metatarsal is affected, especially if it is much longer than an 'atavistic' first metatarsal. The X-ray appearance may at first be normal but a radioisotope scan will show an area of intense activity in the bone. Later a hairline crack may be visible and later still (4–6 weeks) a mass of callus is seen.

Unaccountable pain in elderly osteoporotic people may be due to the same lesion; X-ray diagnosis is more difficult because callus is minimal and there may be no more than a fine linear periosteal reaction along the metatarsal. If osteoporosis has not already been diagnosed, then this should be considered and assessed with bone densitometry.

Metatarsal pain after forefoot surgery may also be due to stress fractures of the adjacent metatarsals, a consequence of redistributed stresses in the foot.

No displacement occurs and neither reduction nor splintage is necessary. The forefoot may be supported with an elastic bandage and normal walking is encouraged.

INJURIES OF METATARSOPHALANGEAL JOINTS

Sprains and dislocations of the metatarsophalangeal (MTP) joints are common in dancers and athletes. A simple sprain requires no more than light splinting; strapping a lesser toe (second to fifth) to its neighbour

for a week or two is the easiest way. If the toe is dislocated, it should be reduced by traction and manipulation; the foot is then protected in a short walking cast for a few weeks.

FRACTURED TOES

A heavy object falling on the toes may fracture phalanges. If the skin is broken, it must be covered with a sterile dressing, and antibiotics are given; a contaminated wound will require formal surgical washout and exploration. The fracture is disregarded and the patient encouraged to walk in a supportive boot or shoe. If pain is marked, the toe may be splinted by strapping it to its neighbour for 2–3 weeks.

FRACTURED SESAMOIDS

One of the sesamoids (usually the medial) may fracture from either a direct injury (landing from a height on the ball of the foot) or sudden traction; chronic, repetitive stress is more often seen in dancers and runners.

The patient complains of pain directly over the sesamoid. There is a tender spot in the same area and sometimes pain can be exacerbated by passively hyperextending the big toe. X-rays will usually show the fracture (which must be distinguished from a smooth-edged bipartite sesamoid).

Treatment

Treatment is often unnecessary, though a local injection of lignocaine helps for pain. If discomfort is marked, the foot can be supported in a removable boot/splint for 2–3 weeks. An insole with differential padding or cut-out under the sesamoid might also speed a return to sporting activities. Occasionally, intractable symptoms call for excision of the offending ossicle; care should be taken not to disrupt the flexor attachment to the proximal phalanx as this may result in great toe deformity.

FURTHER READING

Bajammal S, Tornetta P 3rd, Sanders D, Bhandari M. Displaced intra-articular calcaneal fractures. *J Orthop Trauma* 2005; 19: 360–4.

Bosse MJ, McCarthy ML, Jones AL, *et al.* Lower Extremity Assessment Project (LEAP) Study Group. The insensate foot following severe lower extremity trauma: an indication for amputation? *J Bone Joint Surg* 2005; **87A**: 2601–8.

Broström L. Sprained ankles. Surgical treatment of 'chronic' ligament ruptures. *Acta Chir Scand* 1966; **132**: 551–65.

Buckley R, Tough S, McCormack R, *et al.* Operative compared with non-operative treatment of displaced intraarticular calcaneal fractures: a prospective, randomized, controlled multicenter trial. *J Bone Joint Surg* 2002; **84A**: 1733–44.

Coetzee JC. Making sense of Lisfranc injuries. *Foot Ankle Clin* 2008; **13**: 695–704.

Coetzee JC, Ly TV. Treatment of primarily ligamentous Lisfranc joint injuries: primary arthrodesis compared with open reduction and internal fixation. Surgical technique. *J Bone Joint* Surg 2007; **89A**(Suppl 2 Pt 1): 122–7.

Eastwood DM, Gregg PJ, Atkins RM. Intra-articular fractures of the calcaneum. Part 1: Pathological anatomy and classification. *J Bone Joint Surg* 1993; **75B**: 183–8.

Eastwood DM, Langkamer VG, Atkins RM. Intra-articular fractures of the calcaneum. Part 2: Open reduction and internal fixation by the extended lateral transcalcaneal approach. *J Bone Joint Surg* 1993; **75B**: 189–95.

Griffin D, Parsons N, Shaw E, *et al.* Operative versus non-operative treatment for closed, displaced, intra-articular fractures of the calcaneus: randomised controlled trial. *BMJ* 2014; **349**: 4483.

Lauge-Hansen N. Fractures of the ankle. II. Combined experimental-surgical and experimental-roentgenologic investigations. *Arch Surg* 1950; **60**: 957–85.

Muller ME, Allgöwer M, Schneider R, Willeneger H. *Manual of Internal Fixation* (3rd edn). Berlin: Springer, 1991, pp. 598–600.

Pearce CJ, Wong KL, Calder JD. Calcaneal fractures: selection bias is key. *Bone Joint J* 2015; **97-B**: 880–2.

Pearse EO, Klass B, Bendall SP. The 'ABC' of examining foot radiographs. *Ann R Coll Surg Engl* 2005; **87**: 449–51.

Tarkin IS, Clare MP, Marcantonio A, Pape HC. An update on the management of high-energy pilon fractures. *Injury* 2008; **39**: 142–54.

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