

Spinal Congenital Anomalies (Spinal Dysraphism)

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*3rd week of gestation

The nervous system is **ectodermal** in origin (develops from neuro – ectoderm) except the **microglia** and **dura matter** which are **mesodermal** in origin

Spinal cord development :

1. **Gastrulation** (conversion of the embryonic disk from a **bilaminar** disk to a **trilaminar** disk composed of ectoderm , mesoderm and endoderm)



2. Primary neurulation (notochord and overlying ectoderm interact to form the neural plate)
Neural plate bends and folds to form the neural tube, which then closes bidirectionally in a zipperlike manner

3. Secondary neurulation (neural tube is formed by the caudal cell mass)

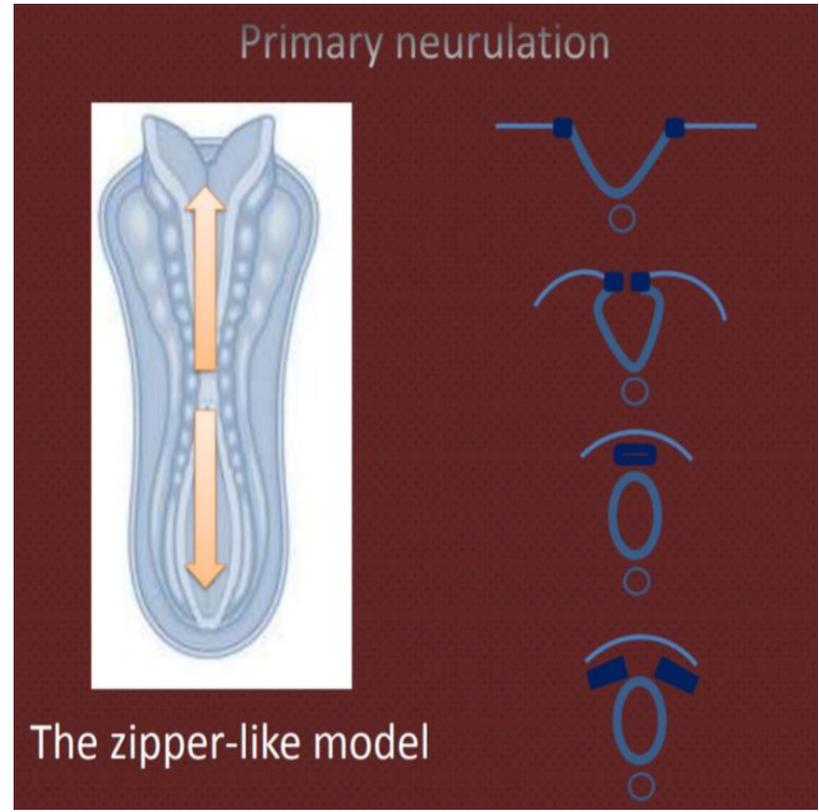
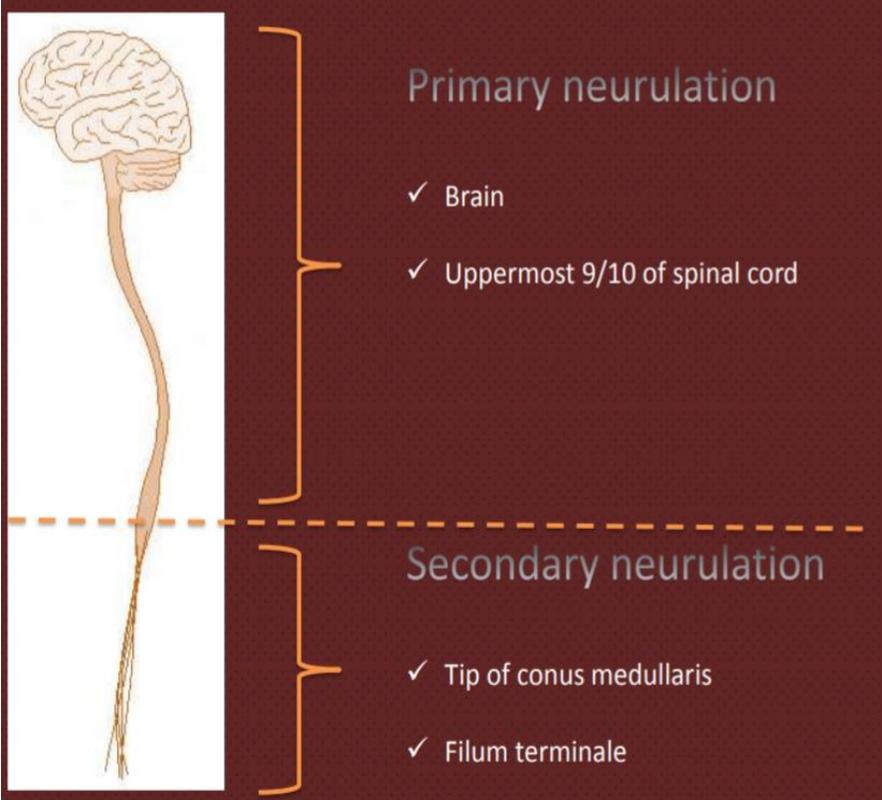
Abnormalities in any of these steps can lead to spine or spinal cord malformations.

gastrulation
*bilocameral
↓
trilaminar.

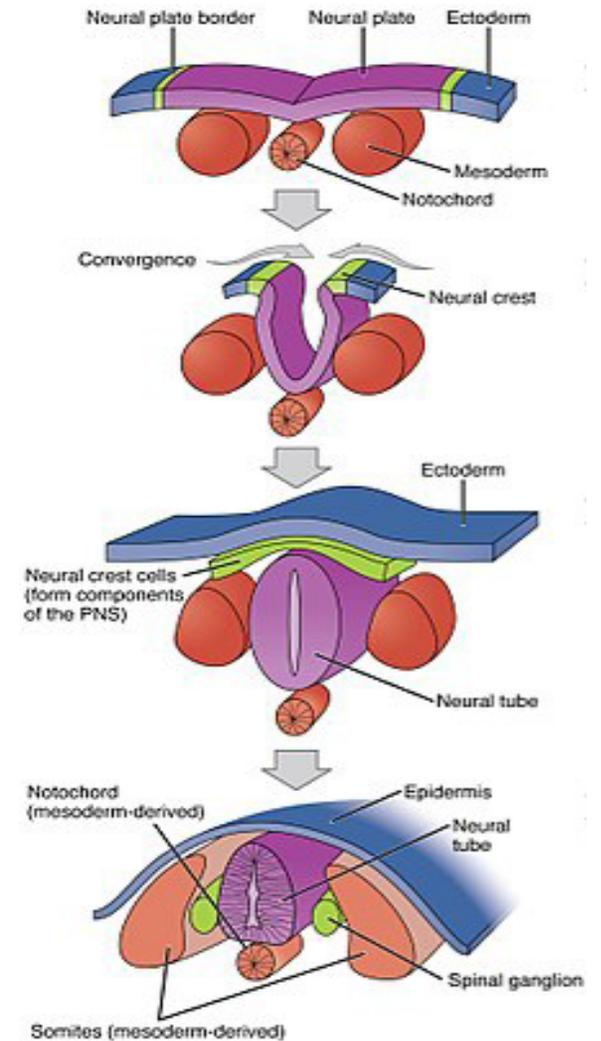
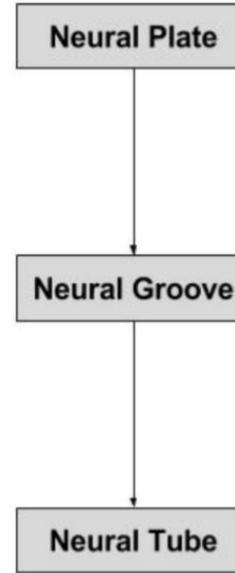
1ry neurulation

2ry neurulation

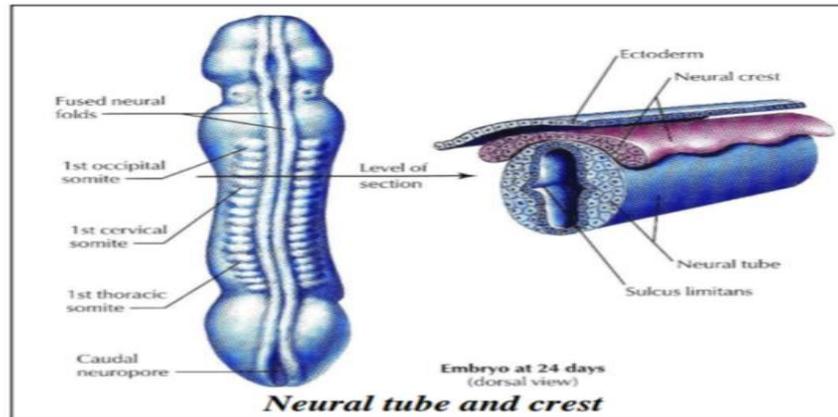
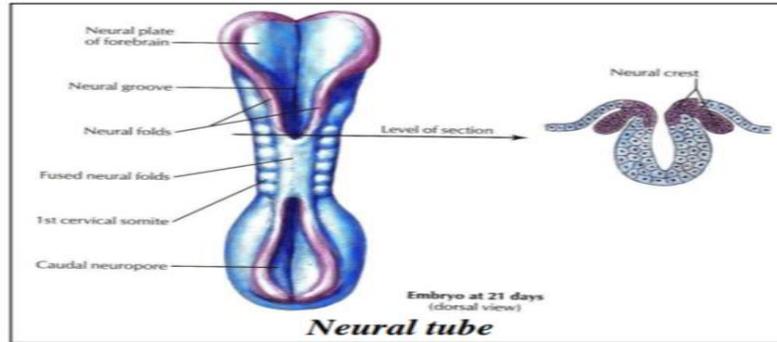
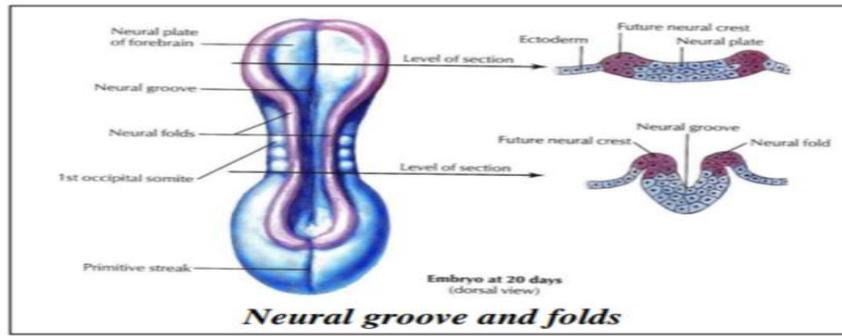




1. Formation of neural plate
2. Shaping of the neural plate
3. Bending of the neural plate
4. Fusion



Closure of the neural tube begins from the midline:



Fusion

Spinal Dysraphism :

Heterogeneous group of anomalies resulting from incomplete midline closure of osseous, mesenchymal and nervous tissue (Failure of normal fusion of the neural plate to form neural tube during the first 28 days following conception)

Prevalence :

Neural tube defects (NTDs) are one of the most common birth defects, occurring in approximately one in 1000 live births in the United States. *Common.*

Neural tube defects (NTDs) cause infant mortality (death) and serious disability

Categorization of Spinal Dysraphisms (neural tube defects) :

open
spinal
dysraphism

closed
spinal
dysraphism

no skin →
dysplastic tissue

1. Open spinal dysraphism

there is a defect in the overlying skin, and the neural tissue is exposed to the environment (caused by defective closure of the primary neural tube and are characterized clinically by exposure of the neural placode through a midline skin defect on the back)

. All OSDs are anomalies of 1ry neurulation

2. Closed spinal dysraphism

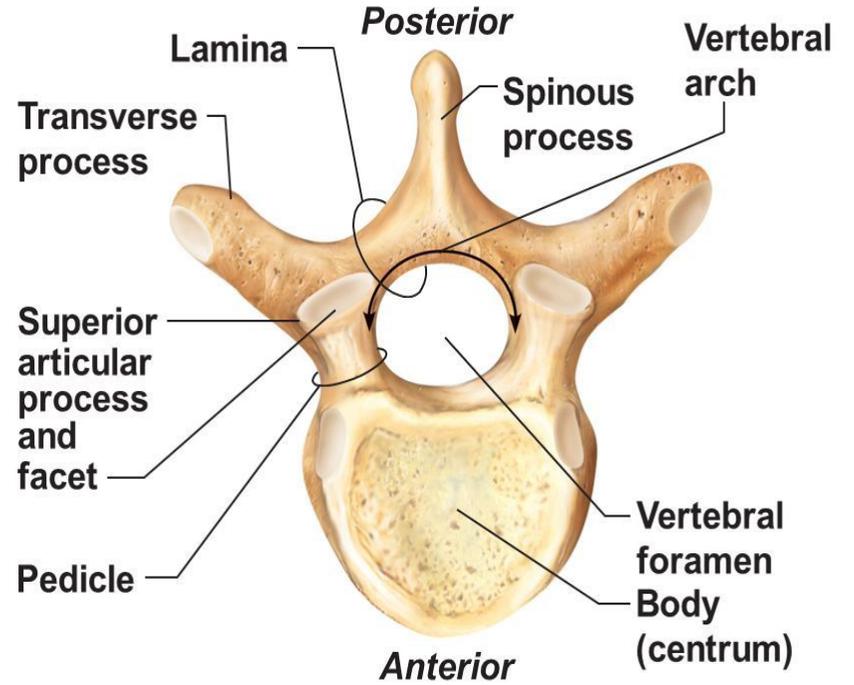
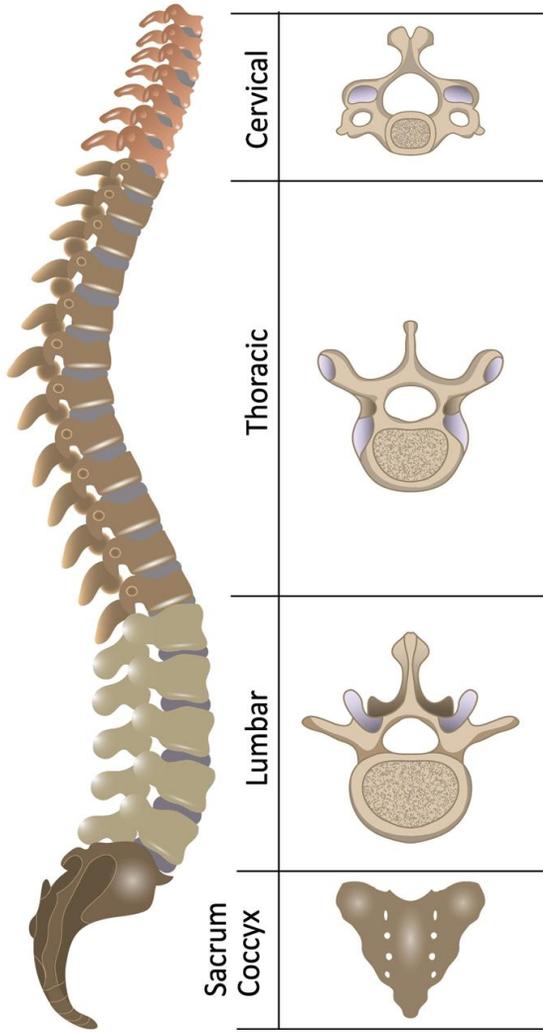
neural tissue is covered by skin (may be abnormal skin) , but with or without associated subcutaneous mass

Spina bifida Spine split

occulta cystica

Due to failure of fusion of the dorsal parts of vertebra “ absence of vertebral arch or part of them “

1. **Occulta** → no posterior arch
- . Most common and mildest form
 - . Mostly / typically affects the lumbosacral area
 - . May associated with overlying skin lesions as :
Skin dimples , Hemangioma or red spot ,
Abnormal tuft of hair , sinus tract , fatty mass or lipomas



Overlying skin lesions as :

1. Skin dimples
2. Heamangioma /red spot
3. Abnormal tuft of hair
4. Sinus tract
5. Lipomas





2. Cystica

1 → **Meningocele** (meningeal cyst)

. The meninges herniate through the spina bifida and forming subcutaneous sac / cyst filled with CSF

2 → **Myelomeningocele** *more common than meningocele*

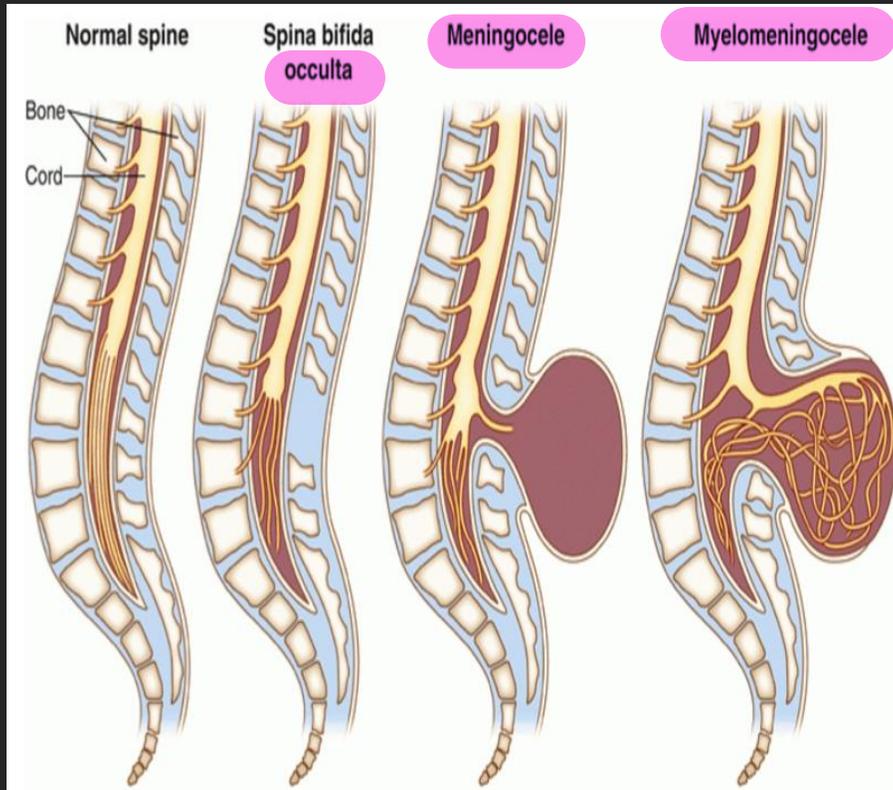
. The spinal cord herniates through the **Meningocele**

. May associated with other neurological anomalies as : **Chiari 2 malformation** , **hydrocephalus**

* **Myelocele (Rachischisis)** *open back deformity*

. Failure of obliteration of the neural tube

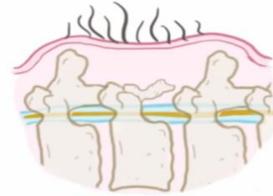
**all patient with myelomeningocele have chiari 2 [20-30%]*



SPINA BIFIDA

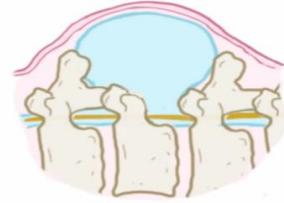
SPINA BIFIDA OCCULTA

- ↳ most common
- ↳ least severe



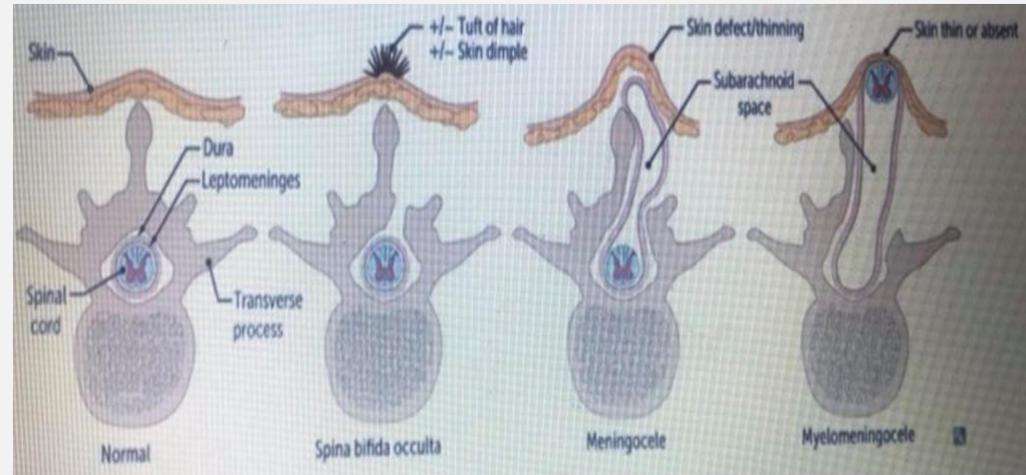
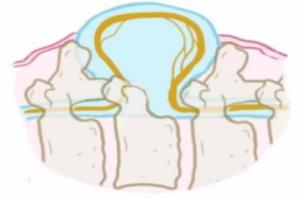
MENINGOCELE

- ↳ Least common



MYELOMENINGOCELE

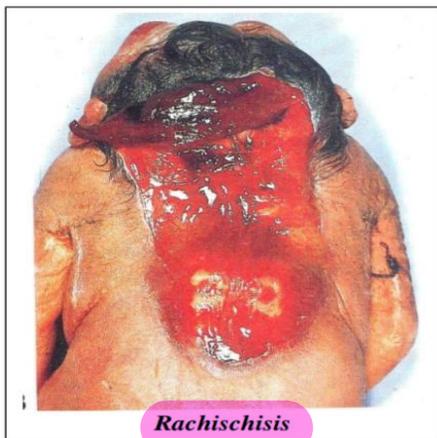
- ↳ most severe



	<i>Meningocele</i>	<i>Myelomeningocele</i>
<i>Incidence</i>	<i>Least common</i>	<i>More common</i>
<i>Prognosis</i>	<i>Less serious</i>	<i>Most serious</i>
<i>Sac</i>	<i>meninges and cerebro-spinal fluid</i>	<i>meninges and cerebro-spinal fluid & nerves and spinal cord.</i>
<i>Nerves</i>	<i>not badly damaged</i>	<i>badly damaged</i>
<i>Ability</i>	<i>❖ Limited disability is present.</i>	<i>bowel and bladder problems.</i>



Rachischisis + anencephaly



Rachischisis



قسطی بی پیدا



Spina bifida
* دالة مع Spina bifida في
- Rachisis.

Unknown cause , but the risk factors include :

1. Folate deficiency (Vitamin B9)
2. Obesity
3. Poorly controlled Diabetes
4. Medications that interfere with folate metabolism as :
Anti- Seizure medications
5. Genetic predisposition and Family history
6. Previous pregnancy with same condition

Diagnosed :

1. Prenatally *

. Screening (increased Alpha Fetoprotein " AFP " in mother's serum , Acetylcholinesterase)

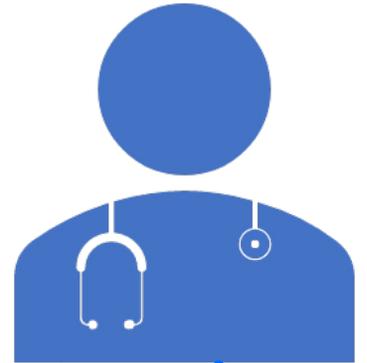
. US *most accurate* - *in amniotic fluid*
. Amniocentesis

2. Postnatally

- ✓ . Neonatal Examination " clinically " + imaging (MRI)
- ✓ . Get complications *→ weakness / paralysis.*
- ✓ . Incidentally by imaging

Fluid.
indicate → congenital anomaly
[confirm that]

this is CNS anomaly and open anomaly



Treatment :

1. Prenatal (with very high risk)

(↓ infection) * لا زرم نخود او repair جراحا و ع 5 ساعت (↓ infection)
48 hours.

2. Postnatal surgical repair shortly after birth
(starting initially by covering the lesion with Gauze soaked with normal saline or ringer solution to prevent dryness , sac excision , emptying of CSF , exposure of the contents , tissue repair)

avoid antiseptic (neurotoxic agents) ex: iodine .

avoid mechanical trauma

alcohol

General assessment

❖ Assess whether lesion is ruptured or unruptured

- Ruptured lesions start prophylactic antibiotic
- Urgent surgery

suture what of a كبر في line flap 2. عزى *
respiratory distress ← patient → RA is also causing

From scapula.
Erotationel Flap

❖ Measure size and site of defect for proper planing for Closure

❖ Evaluation by neonatologist if associated with other anomaly.

- Other anomalies (average 2-2.5% additional anomalies)
- Condition oppose with surgery e.g lung immaturity

❖ Bladder

- Start with on regular urinary catheterization
- Urological consultation

❖ Orthopedic consultation for sever deformities

Clup Foot.

Neurological preop. assessment

- ❖ Watch for spontaneous movement of lower limbs which associated with better outcome.
- ❖ Assess lowest level of neurological function
 - Response to painful stimuli
 - Differ between voluntary movement from reflex movement which is stereotyped and not persist after stimulus
- ❖ Evaluate other neurological associations
 - A. Hydrocephalus
 - ✓ ▪ Anterior fontanel
 - ✓ ▪ Head circumference
 - B. Chiari II
 - Check for inspiratory stridor and apneic episodes

Lipomyelomeningocele : 2^{ry} neurulation

. Lipoma attached to dorsal surface of incompletely closed spinal cord

. Covered by skin



*normal conus medullaris at birth
at the level of L3-L4, during growth: ascend

↳ normal skin
*closed.

(L2-L3, L4-L5, L6-L7, L8-L9, L10-L11, L12-L13, L14-L15, L16-L17, L18-L19, L20-L21, L22-L23, L24-L25, L26-L27, L28-L29, L30-L31, L32-L33, L34-L35, L36-L37, L38-L39, L40-L41, L42-L43, L44-L45, L46-L47, L48-L49, L50-L51, L52-L53, L54-L55, L56-L57, L58-L59, L60-L61, L62-L63, L64-L65, L66-L67, L68-L69, L70-L71, L72-L73, L74-L75, L76-L77, L78-L79, L80-L81, L82-L83, L84-L85, L86-L87, L88-L89, L90-L91, L92-L93, L94-L95, L96-L97, L98-L99, L100-L101, L102-L103, L104-L105, L106-L107, L108-L109, L110-L111, L112-L113, L114-L115, L116-L117, L118-L119, L120-L121, L122-L123, L124-L125, L126-L127, L128-L129, L130-L131, L132-L133, L134-L135, L136-L137, L138-L139, L140-L141, L142-L143, L144-L145, L146-L147, L148-L149, L150-L151, L152-L153, L154-L155, L156-L157, L158-L159, L160-L161, L162-L163, L164-L165, L166-L167, L168-L169, L170-L171, L172-L173, L174-L175, L176-L177, L178-L179, L180-L181, L182-L183, L184-L185, L186-L187, L188-L189, L190-L191, L192-L193, L194-L195, L196-L197, L198-L199, L200-L201, 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Tethered cord syndrome

(tight filum terminale syndrome) :

The spinal cord could be caught against the vertebral by tight band , fibrosis or adhesions (abnormally attached to tissue around) resulting in abnormally low conus medullaris

Symptoms usually at age of puberty including (5 Ds) :

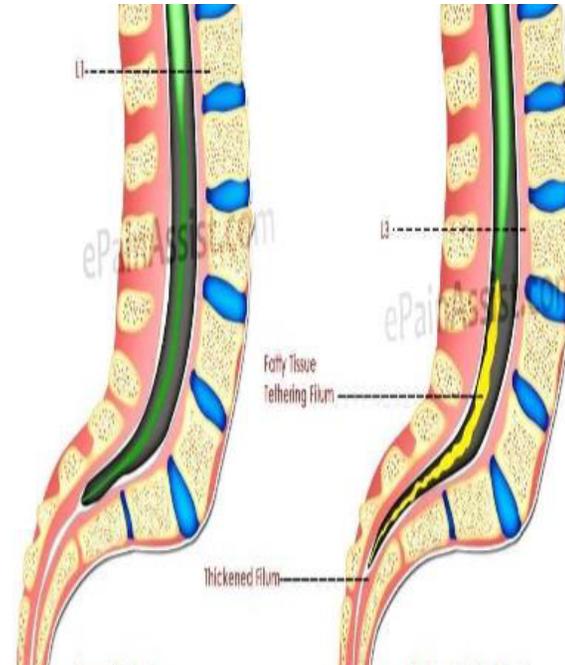
1. Discomfort , back and leg pain
2. Deformities of spine (scoliosis , kyphosis , lordosis) and feet (high arches , foot turned inward)
3. Dysfunctions (weakness , numbness , “ kidney , bladder and bowel dysfunctions “)
4. Decline of abilities
5. Dysraphisms of spine

neck pain, back pain & sciatica before age 4? red flag.

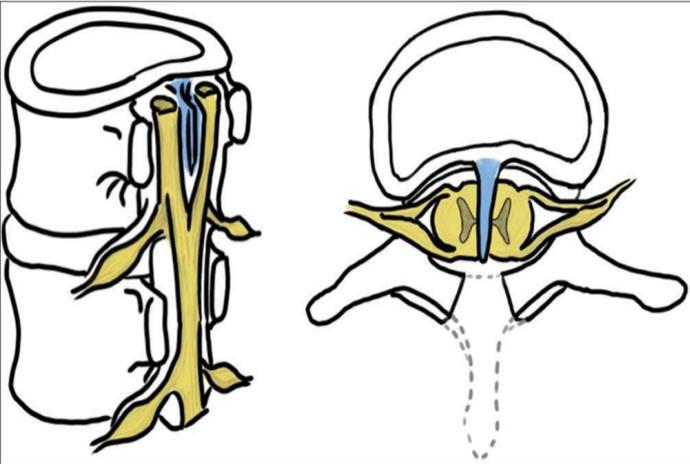
Diagnosed by MRI to confirm the diagnosis and rule out Neoplasia

↳ abnormal diameter > 2mm

Treated surgically by Un-tethering surgery by cutting the filum terminale



Split Cord Malformation (SCM) :



Rare anomaly characterized by a split along the midline of the spinal cord by fibrous or bony septum , which divides it into 2 symmetric or nonsymmetric entities

* تقسيم جزء معين

It NOT involving the whole length of spinal cord , but segment of it mostly at level of lumber or thoracic area

Mostly associated with Tethered cord , so share it's symptoms

Diagnosed mainly by MRI

Treated by Un- tethering surgery (but after removal of the bony septum and reconstitution of dura into single tube of affected segments until reaching the normal segments)

* اولا نبي البنية او bony septum ، بعدئذ نزيل
• untethering • عشان ممكن نغير confusion

Type 1



hemicord ٥ *
بجیر کان
قالب cord

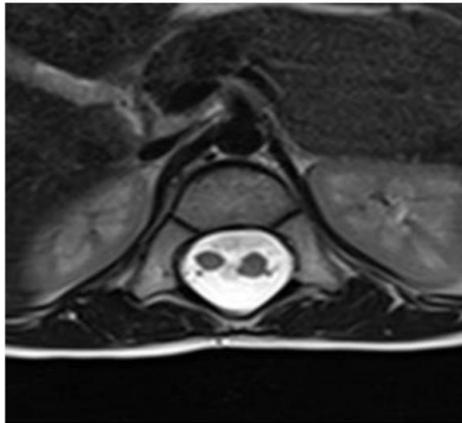
Two types of SCM :

1. Type 1 (**Diastematomyelia**)

by bone

- . Two hemicords , each with it's **own central canal** , surrounding pia and **dural tube** , separated by **rigid osseocartilaginous** (bony spur) median septum
- . With abnormalities of spine at the level of split (absent disc)
- . $\frac{2}{3}$ associated with overlying skin abnormalities

Type 2



2. Type 2 (**Diplomyelia**)

by Fibrous

- . Two hemicords within a **single dural tube** , separated by a **nonrigid fibrous median septum**
- . Without abnormalities of spine at the level of split , but usually with **spina bifida occulta**

Impact of life :

1. Financial (cost)
2. Physical (paralysis , hydrocephalus , learning disabilities , bowel and bladder control problems , other congenital anomalies)
3. Emotional (miscarriage , stillbirth , infant mortality , disability , feelings)



Thank you

