Pupil and optic nerve

Khaled albayaydah
abdullah albayaydah
abdulrahman abu-feddah
Obada Alayed
Hamzah altamimi

The pupil

The pupil is a hole located in the center of the iris of the eye that allows light to strike the retina.

Introduction

Movements of the pupil are controlled by the parasympathetic and sympathetic nervous systems. The pupils constrict (miosis) when the eye is illuminated (parasympathetic activation, sympathetic relaxation) and dilate (mydriasis) in the dark (sympathetic activation, parasympathetic relaxation). When the eyes focus on a near object, they convergeandthepupilsconstrict(thenearresponse).Thepupilsarenormallyequalinsize

but some 20% of people may have noticeably unequal pupils (anisocoria) with no associated disease. The key to diagnosis of pupillary disorders is to:

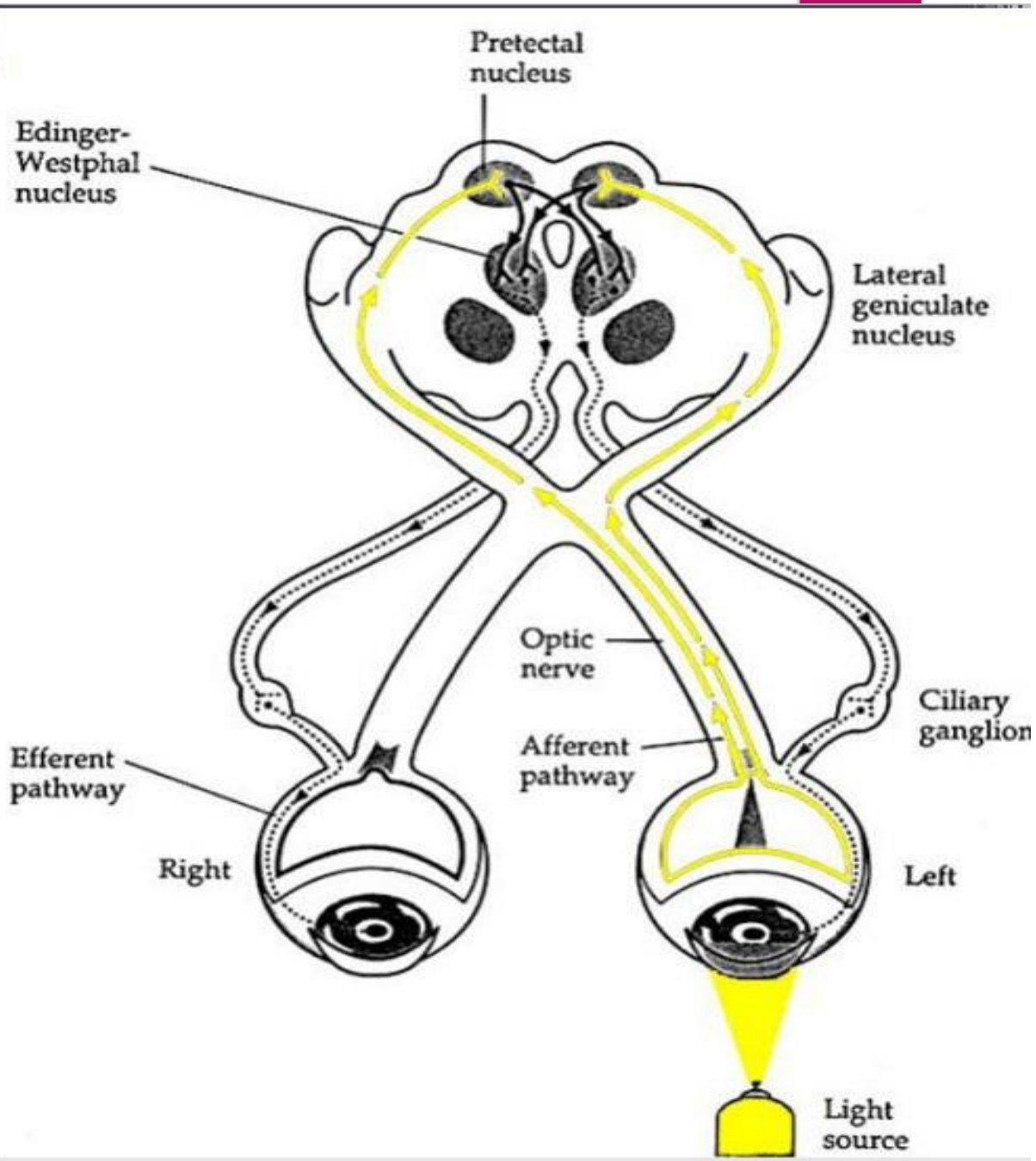
- determine which pupil is abnormal;
- search for associated signs.

Anatomy & pupillary reflexes

- Afferent and efferent arms .
- Normal pupillary reflexes .
- Afferent Pupillary Defect.

Afferent Pathway of Pupillary Light Reflex

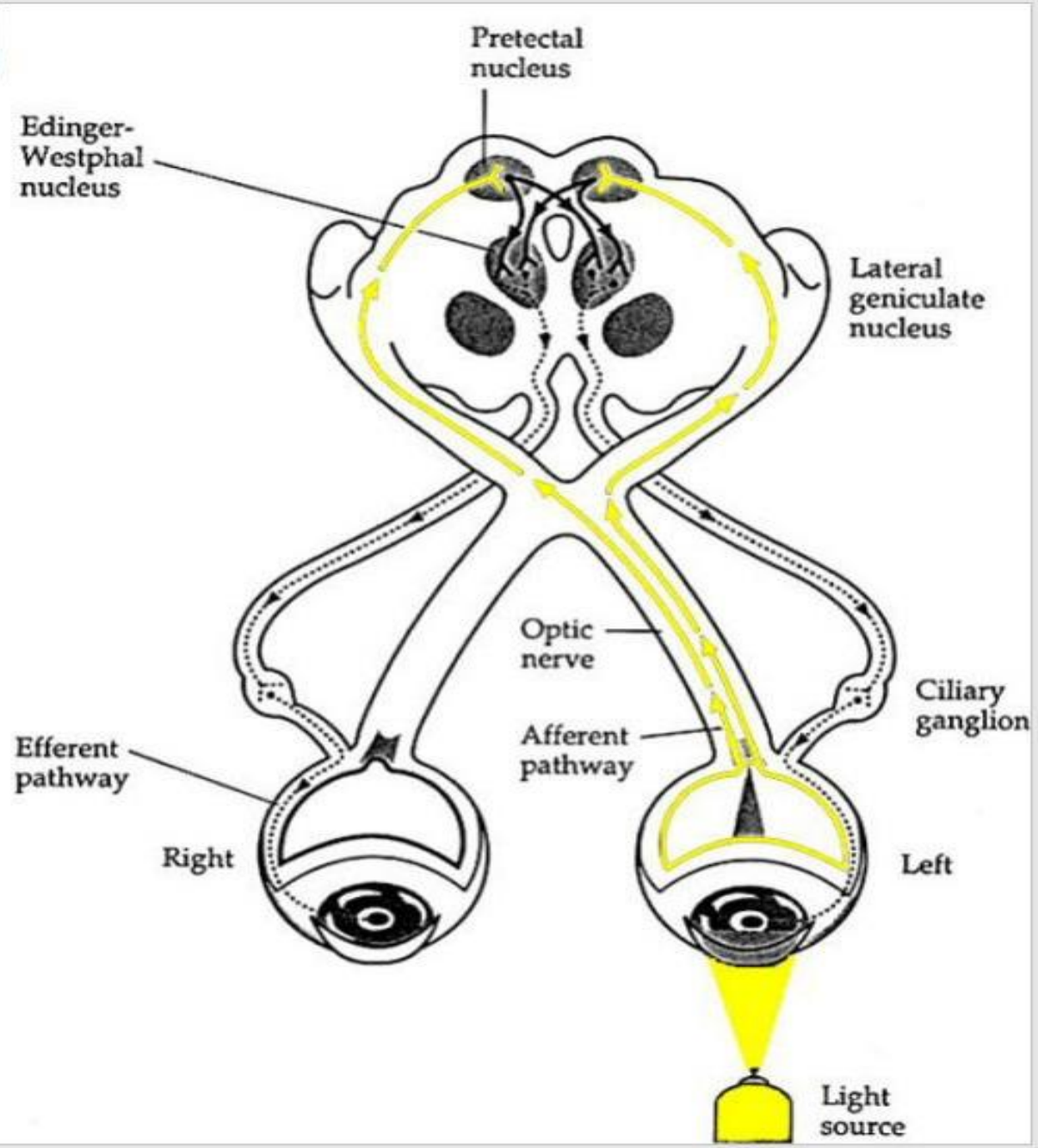
- Light enters the **pupil** and stimulates the **retina**.
- Retinal ganglion cells transmit the light signal to the **optic nerve**
- The optic nerve enters the **optic chiasm** where the nasal retinal fibers cross to contralateral **optic tract**. while the temporal retinal fibers stay in the ipsilateral optic tract
- Fibers from the optic tracts project and synapse in the **pretectal nuclei** in the dorsal midbrain in the collicular region
- The pretectal nuclei project fibers to the ipsilateral **Edinger-Westphal nuclei** and also to the contralateral Edinger-Westphal nucleus via the posterior commissure



Efferent Pathway of Pupillary Light Reflex

- The Edinger-Westphal nucleus projects pre-ganglionic parasympathetic fibers, which exit the midbrain and travel along the **oculomotor nerve** (CN III) and then synapse on post-ganglionic parasympathetic fibers in the **ciliary ganglion**
- Ciliary ganglion post-ganglionic parasympathetic fibers (short ciliary nerves) innervate the sphincter muscle of the pupils resulting in pupillary constriction.

The physiological result is that light shined in one eye will result in pupillary constriction in both the ipsilateral pupil (**direct pupillary light reflex**) and the contralateral pupil (**consensual pupillary light reflex**).

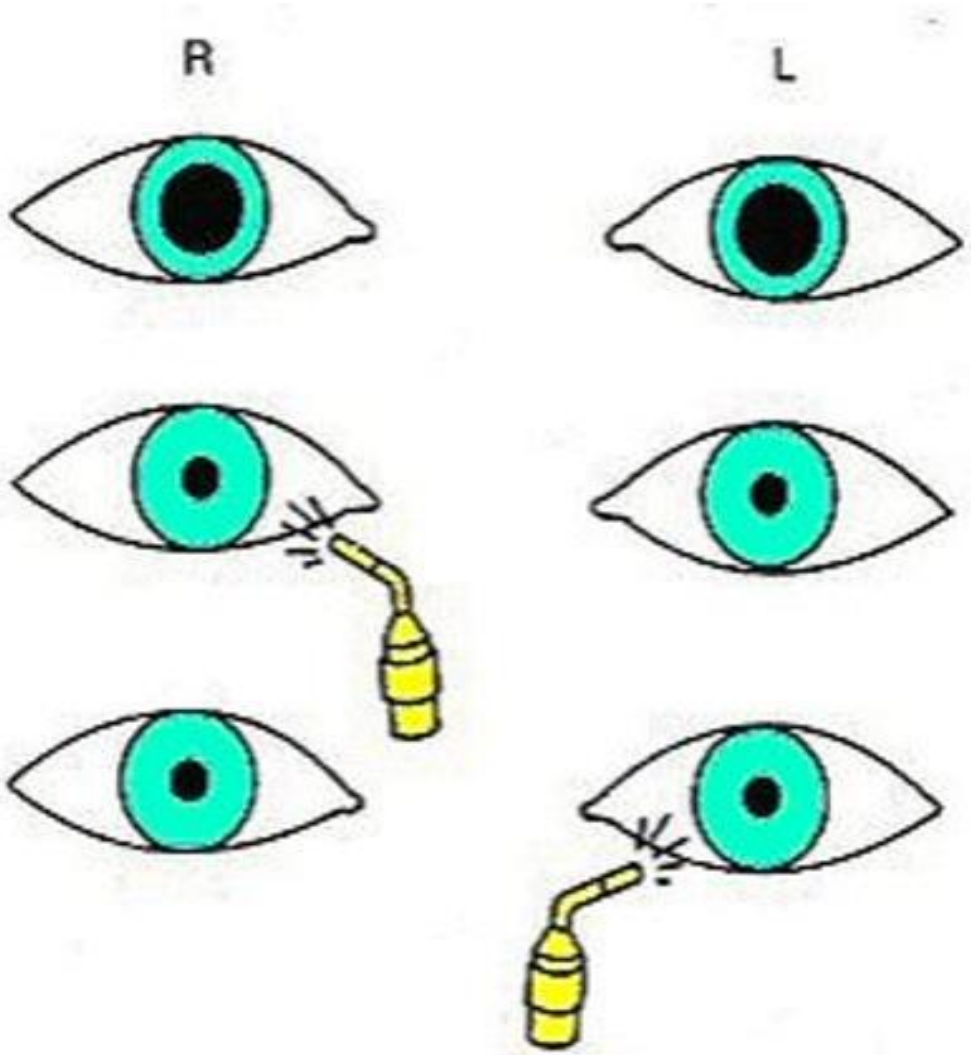


Normal Pupillary Light Response :

Row 1: pupils in a dark room without light stimulation.

Row 2: intact direct and consensual responses for right eye

Row 3: intact direct and consensual responses for left eye



Disorders of the pupil may result from: • ocular disease; • disorders of the controlling neural pathway; • pharmacological action. The parasympathetic
fibers reach the eye through the third cranial nerve.

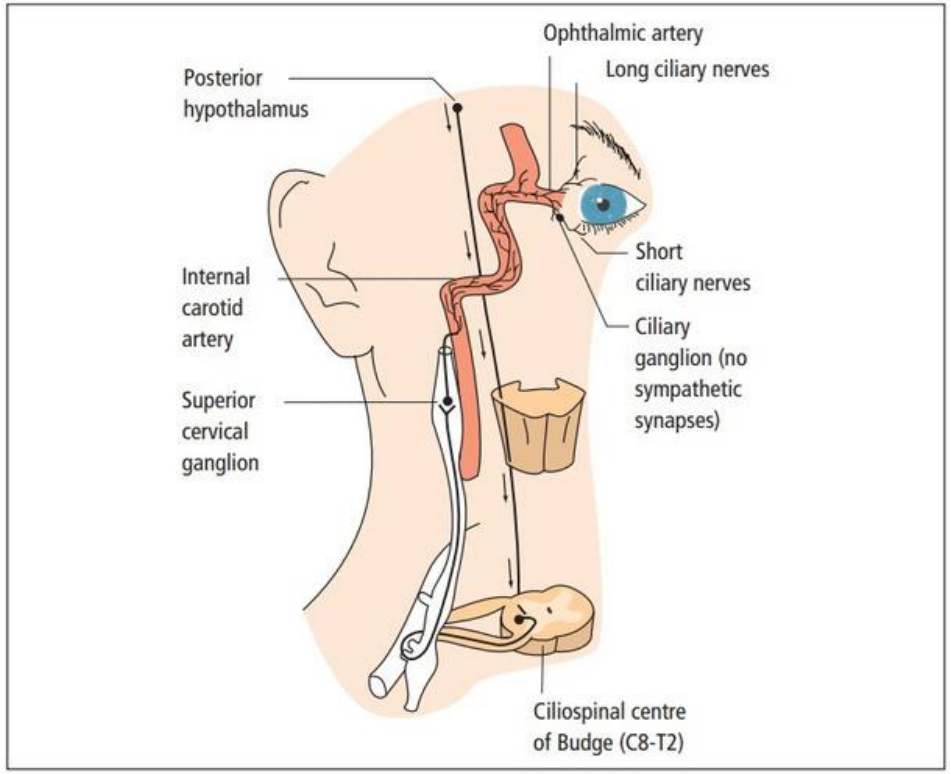
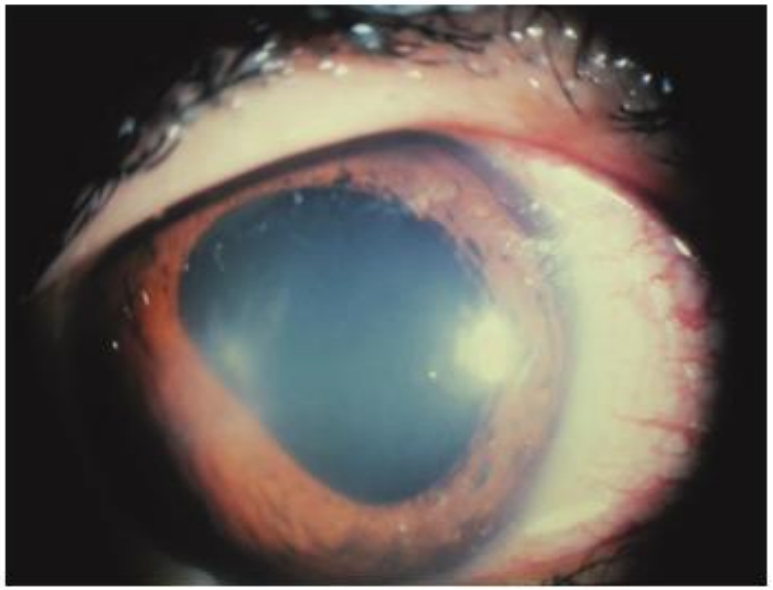


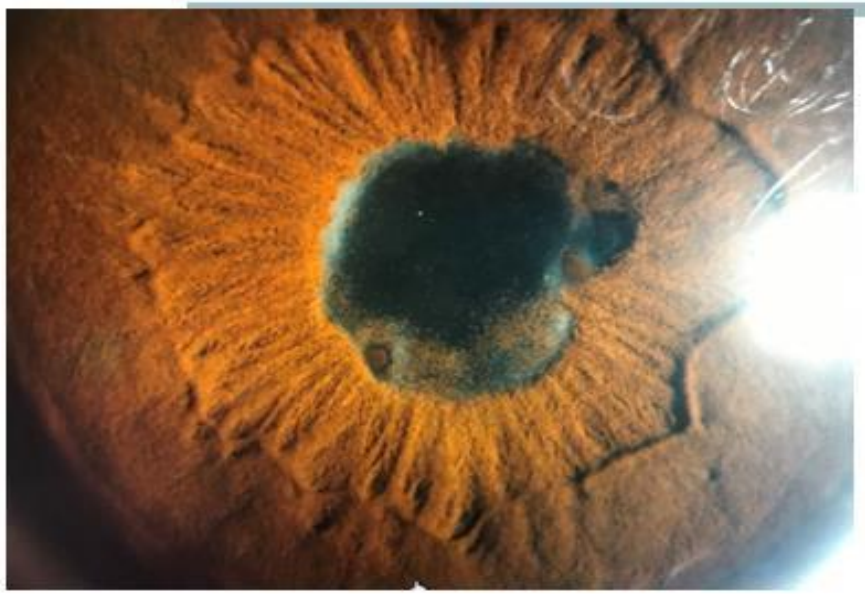
Figure 13.1 The pathway of sympathetic pupillary control. (Adapted with permission from Kanski, J. J. (1994) *Clinical Ophthalmology*. Butterworth–Heinemann, Oxford.)

1.Ocular causes of pupillary abnormality Several diseases of the eye cause pupil irregularity and alter pupil reactions: • anterior uveitis, when posterior synechiae give the pupil an irregular appearance

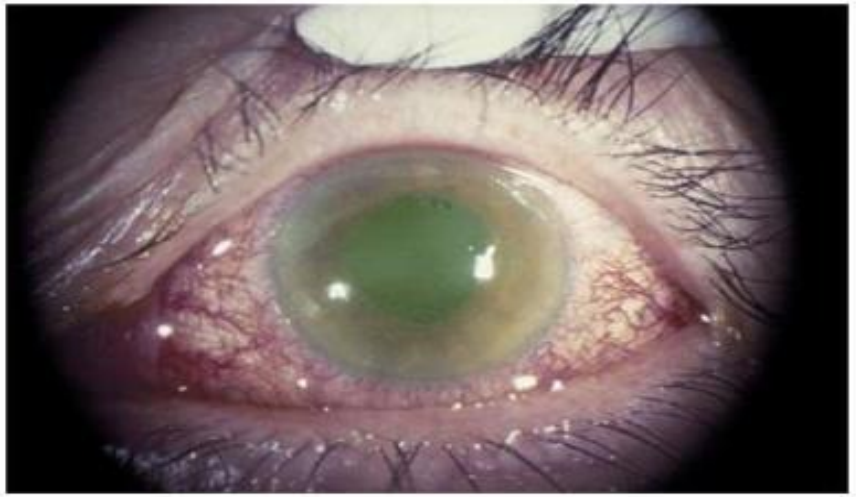
• the sequelae of intraocular surgery;
• blunt trauma to the eye, which may rupture the sphincter muscle, causing irregularity or fixed dilation (traumatic mydriasis); • an acute and severe rise in ocular pressure – as in acute glaucoma.



Traumatic mydriasis



POSTERIOR SYNECHIA SECONDARY TO HLA B27 ANTERIOR UVEITIS



Acute Angle-closure Glaucoma

2.Neurological causes of an abnormal pupil

A.Horner's syndrome Interruption of the sympathetic pathway causes: • A small pupil on the affected side due to loss of the dilator function .This is more noticeable in the dark, because the normal pupil of the fellow eye dilates more than that of the affected pupil. • A slight ptosis on the affected side. • An apparent recession of the globe into the orbit (enophthalmos). The reduced palpebral aperture size gives an impression of recession.

• Lack of sweating on the affected side, if the sympathetic pathway is affected proximal to the base of the skull. This catches fibres travelling with the branches of the external carotid, which innervate the skin of the face.

Because of its extended course, the sympathetic pathway may be affected by a multitude of pathologies. Examples include:

• Syringomyelia , an expanding cavity within the spinal cord, sometimes extending into the medulla (syringobulbia), which compresses the pathway. Typically, it also causes wasting of the hand muscles and loss of sensation. •

Small - cell carcinoma at the lung apex which catches the cervical sympathetic chain. Involvement of the brachial plexus gives rise to pain and to T1 wasting of the small muscles of the hand in Pancoast ' s syndrome. •

Neck injury, disease or surgery. • Cavernous sinus disease – catching the sympathetic carotid plexus in the sinus.

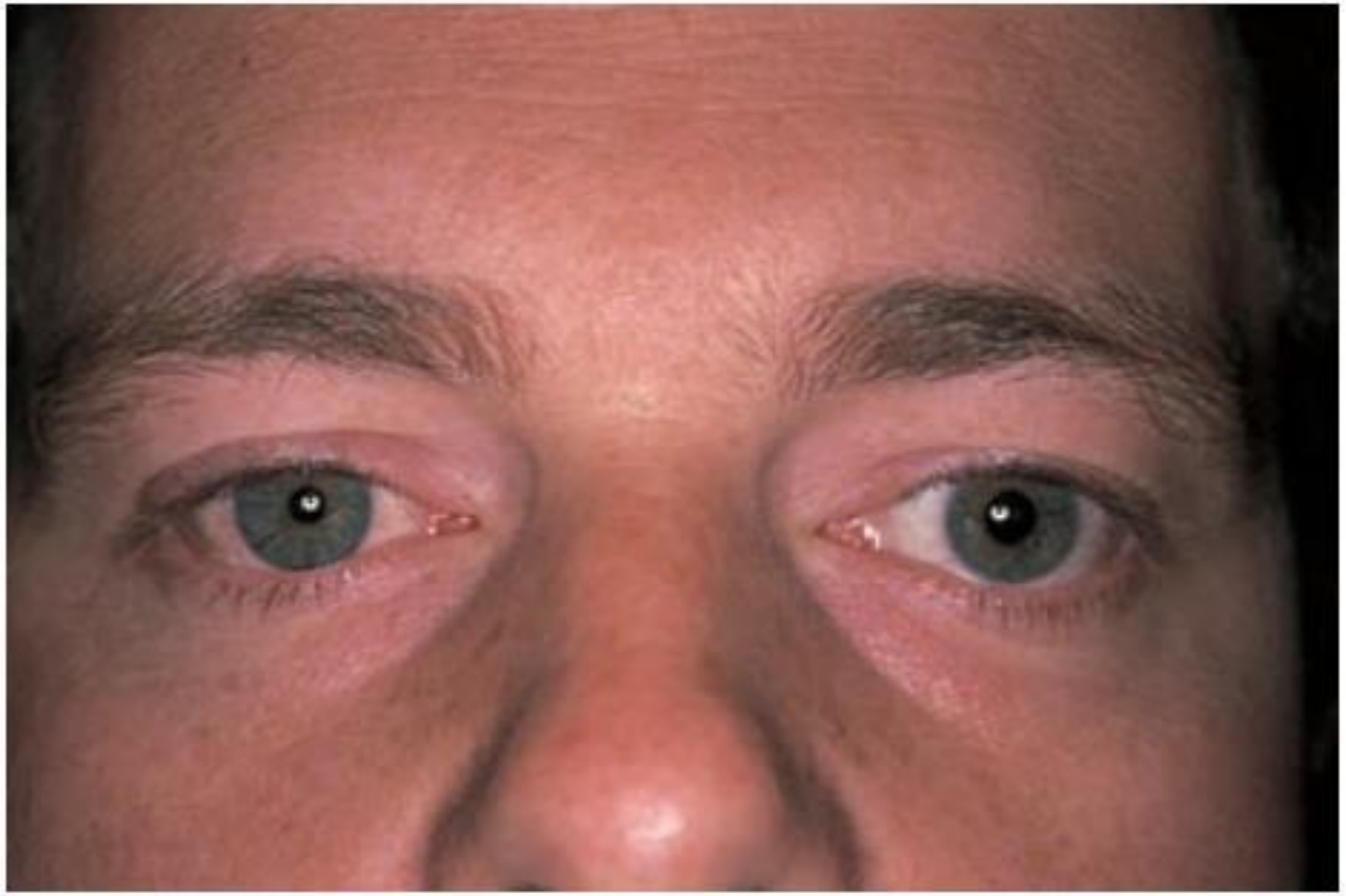


Figure 13.2 A right ptosis and miosis in Horner's syndrome.

Horner's syndrome may also be congenital, in which case the iris color may be altered when compared to the fellow eye (heterochromia).

B. Relative afferent pupillary defect (RAPD) A lesion of the optic nerve on one side blocks the afferent limb of the pupillary light reflex. The pupils are equal and of normal size, but the pupillary response to light directed to the affected side is reduced, while the near reflex is intact. Testing for an RAPD is critical in a patient suspected of having an optic nerve lesion, such as optic neuritis . RAPD may be seen in very severe disease of the retina but not with opacities of the cornea or lens.

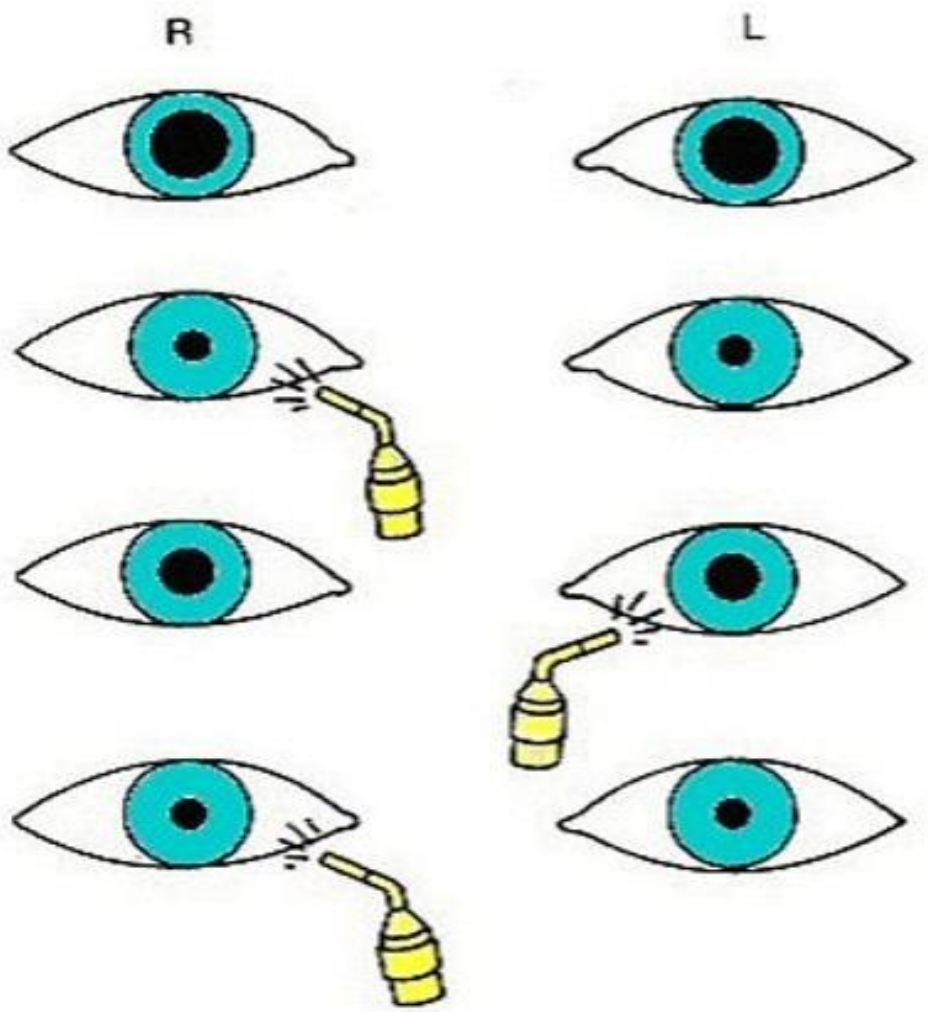
Relative afferent pupillary defect

Row 1: Unstimulated pupils in a dark room

Row 2: Stimulation of the right eye produces bilateral pupillary constriction, indicating intact afferent right limb, and intact bilateral efferent limbs.

Row 3: When moving the light source from the right to left eye, the left eye paradoxically dilates. This indicates a faulty left eye afferent limb, most likely from left optic nerve dysfunction.

Row 4: Demonstrates again that the right afferent pathway is functioning normally and that the problem is with the left eye's afferent pathway.

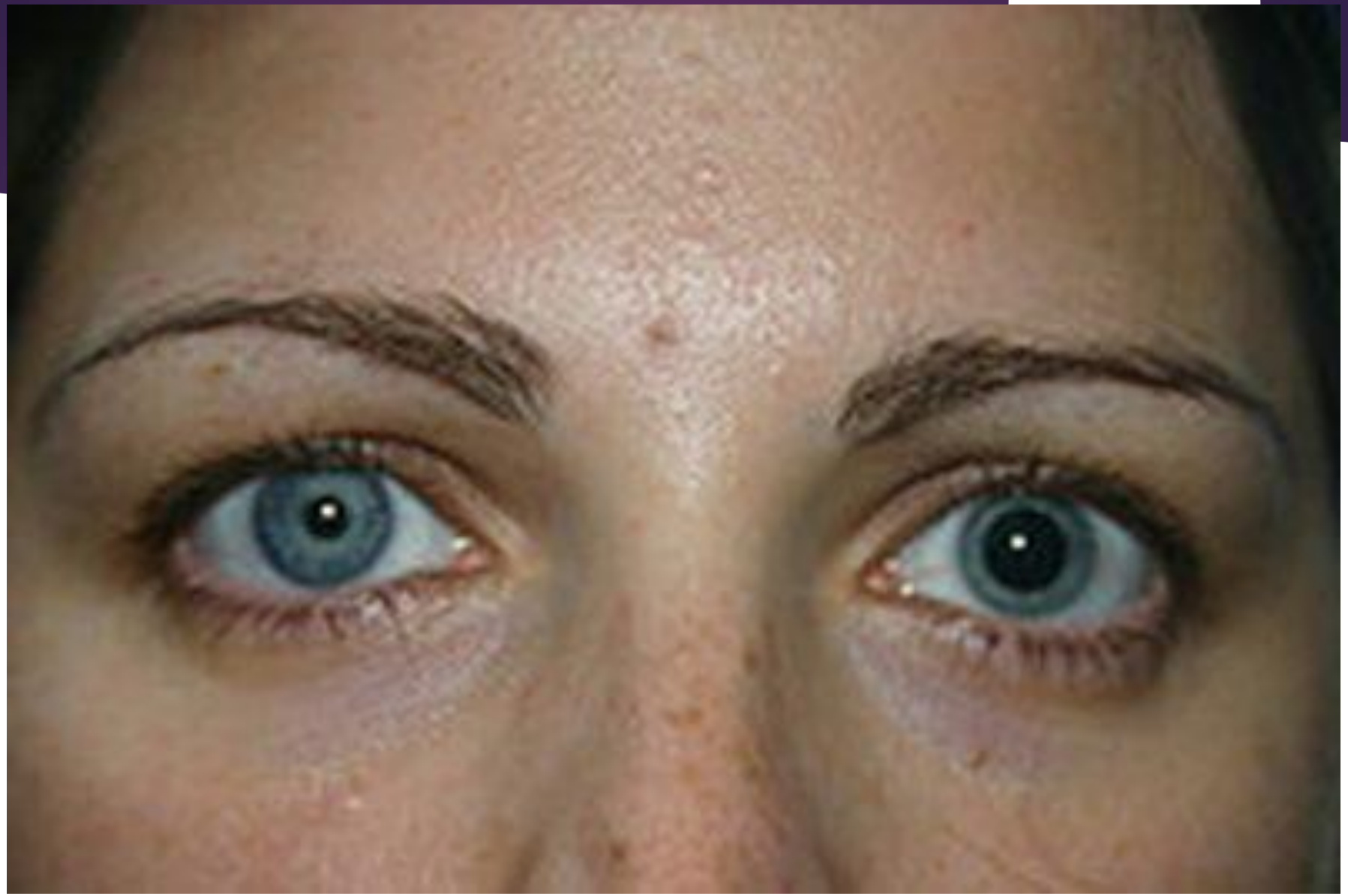


Light – near dissociation

The key feature is an impaired reaction of the pupils to light, while the near response to accommodation is retained. It is seen with Adie ' s tonic pupil, the Argyll Robertson pupil and with peri - aqueductal brainstem lesions such as Parinaud ' s syndrome. Other causes include diabetes and multiple sclerosis. There is no conditioninwhich the lightreflexisintactbut the near reflexisdefective.

1.Adie ' s tonic pupil This is a not uncommon cause of unequal pupil size (anisocoria) in young adults, but has no serious consequences. Onset is subacute and affects females more commonly than males (2 : 1). It is due to a ciliary ganglionitis which denervates the parasympathetic supply to the iris and ciliary body. Normally, the ciliary body receives about nine times more nerve fibers than the iris sphincter. However, on recovery from the ganglionitis, while the parasympathetic innervation of the iris sphincter is greatly reduced, most of the fibers which reinnervate it are aberrant fibers which were previously directed to the ciliary body and involved in accommodation. As the sphincter is partially denervated its muscarinic receptors are supersensitive to cholinergic agonists.

The consequence is that the pupil: • is enlarged – because the sphincter is relatively denervated; • is poorly reactive to light – because few of the innervating fibers were originally destined for the sphincter. Also, because of the irregular fibre distribution, pupil movement in response to light consists of a slow, worm - like (vermiform) contraction, on biomicroscopy.



Due to muscarinic super sensitivity of the sphincter, the pupil also: • shows slow, sustained miosis on accommodation; • constricts to dilute pilocarpine (0.1%), unlike the normal pupil. This is a diagnostic test.

Because the ciliary body is also partially denervated, the ability to accommodate is impaired too and the patient may complain of blurred vision when looking from distance to near, or vice versa . Systemically the disorder is associated with loss of tendon reflexes; there are no other neurological signs.

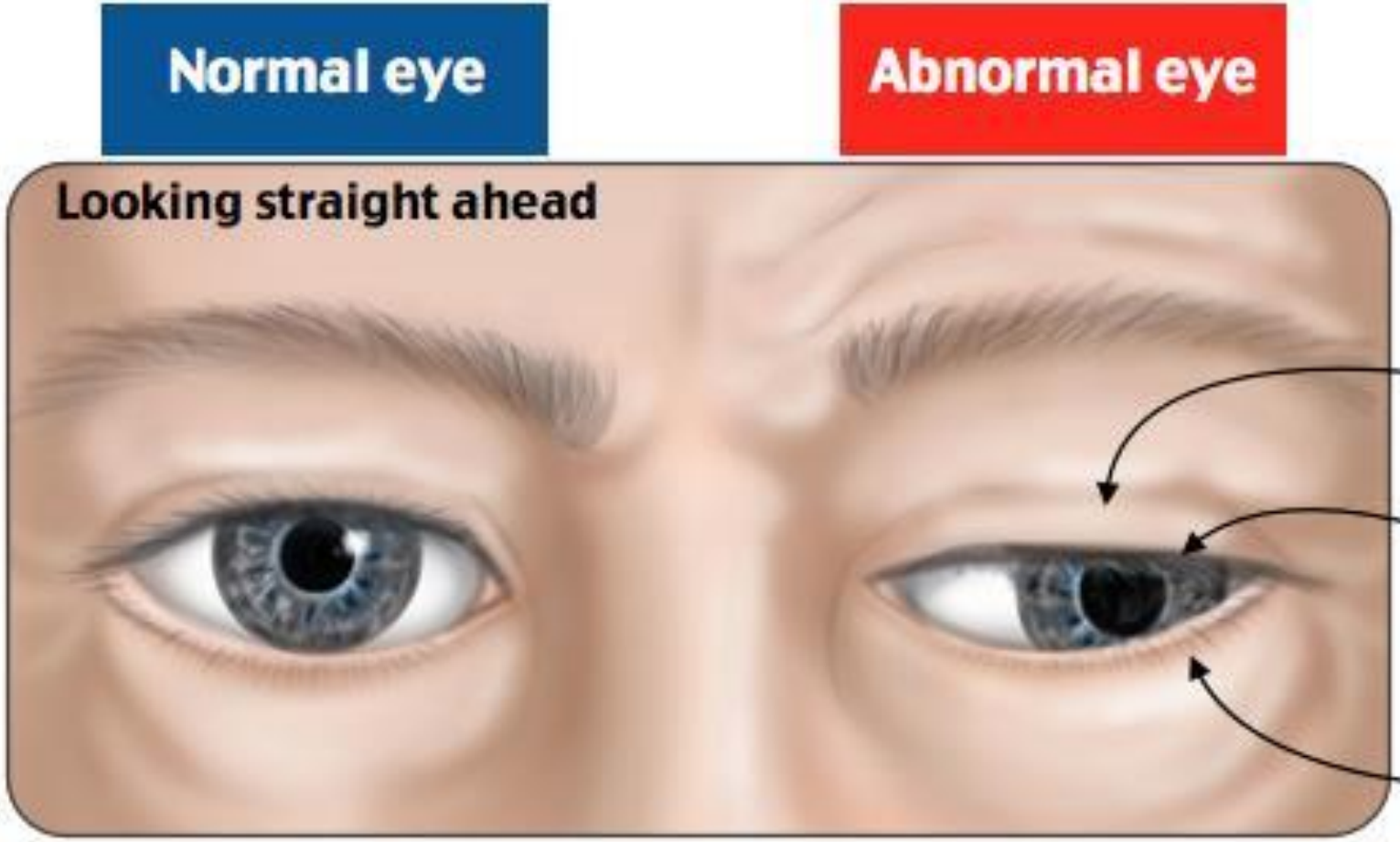
2.Argyll Robertson pupil The pupils are bilaterally small and irregular. They do not react to light but respond to accommodation. The iris stroma has a typical feathery appearance and loses its architectural detail. Classically it is seen in neurosyphilis. It is suggested that a peri - aqueductal lesion on the dorsal aspect of the Edinger Westphäl nucleus involves fibers associated with the response to light, but spares those associated with the near response.

3.Midbrain pupil Lesions affecting the pretectal nuclear complex in the dorsal region of the midbrain can disrupt retinotectal fibers while preserving the supranuclear accommodative pathway. This produces mydriasis and a light – near dissociation. Causes include demyelination, infarction, enlargement of the third ventricle and space - occupying tumours such as pinealoma, as part of a dorsal midbrain (Parinaud ' s) syndrome .

4.Other causes of pupillary abnormality

- In coma, both pupils may become miosed with preservation of the light reflex if a pontine lesion is present, but remember that patients taking pilocarpine for glaucoma or receiving morphine also show bilateral miosis. Midbrain lesions cause loss of the light reflex with mid -
- point pupils. Coma associated with a unilateral expanding supratentorial mass, e.g. a
- hematoma, results in pressure on the third nerve and dilation of the pupil.
- Intrinsic third nerve lesions also cause a dilated pupil .
- The pupil may also be affected by drugs, both topical and systemic ,and in some parts of the world bilateral mydriasis may be caused by the accidental inoculation of plant material into the eye from the Jimson weed (Datura stramonium), which contains the belladonna alkaloids atropine and scopolamine (‘ cornpickers ’ s pupil ’). Occasionally, patients are encountered who have deliberately instilled a mydriatic drop in order to simulate eye disease.

Oculomotor Nerve (CNIII) Palsy



Ptosis
Inactivation of the levator palpebrae

Mydriasis
Decreased tone of the constrictor pupillae muscle

"Down and Out"
Unopposed left superior oblique and lateral rectus muscles

Table 13.1 Drugs having a pharmacological effect on the pupil.

Agent	Action	Mechanism
Topical agents		
Dilates	Muscarinic blockade	Cyclopentolate
		Tropicamide
		Atropine (long-acting)
	Alpha-adrenergic agonist	Phenylephrine
		Adrenaline
		Dipivefrine
Constricts	Muscarinic agonist	Pilocarpine
Systemic agents		
Dilates	Muscarinic blockade	Atropine
	Alpha-adrenergic agonist	Adrenaline
Constricts	Local action and action on central nervous system	Morphine

Differential diagnosis of abnormally large pupil :

1. Adie,s pupil 3rd nerve palsy
2. Dilating drops
3. Traumatic mydriasis Iris robiosis Urrets-zavalia syndrome (iris
4. atrophy following corneal graft) Physiological anisocoria
5.
6.
7.

Differential diagnosis of abnormally small pupil :

1. horner's syndrome
2. Pilocarpine drops
3. Uveitis / posterior synechia
4. Chronic unilateral aphakia
5. Physiological anisocoria

KEY POINTS

- Take a good history to help exclude an ocular cause for the pupillary changes and to see if a medical condition exists which may contribute to the pupillary problem.
- Determine whether it is the small or the large pupil that is abnormal.
- Search for associated signs that may help make a diagnosis.

Optic nerve

The optic nerve is formed by the axons arising from the retinal ganglion cell layer , which form the nerve fiber layer of the retina.

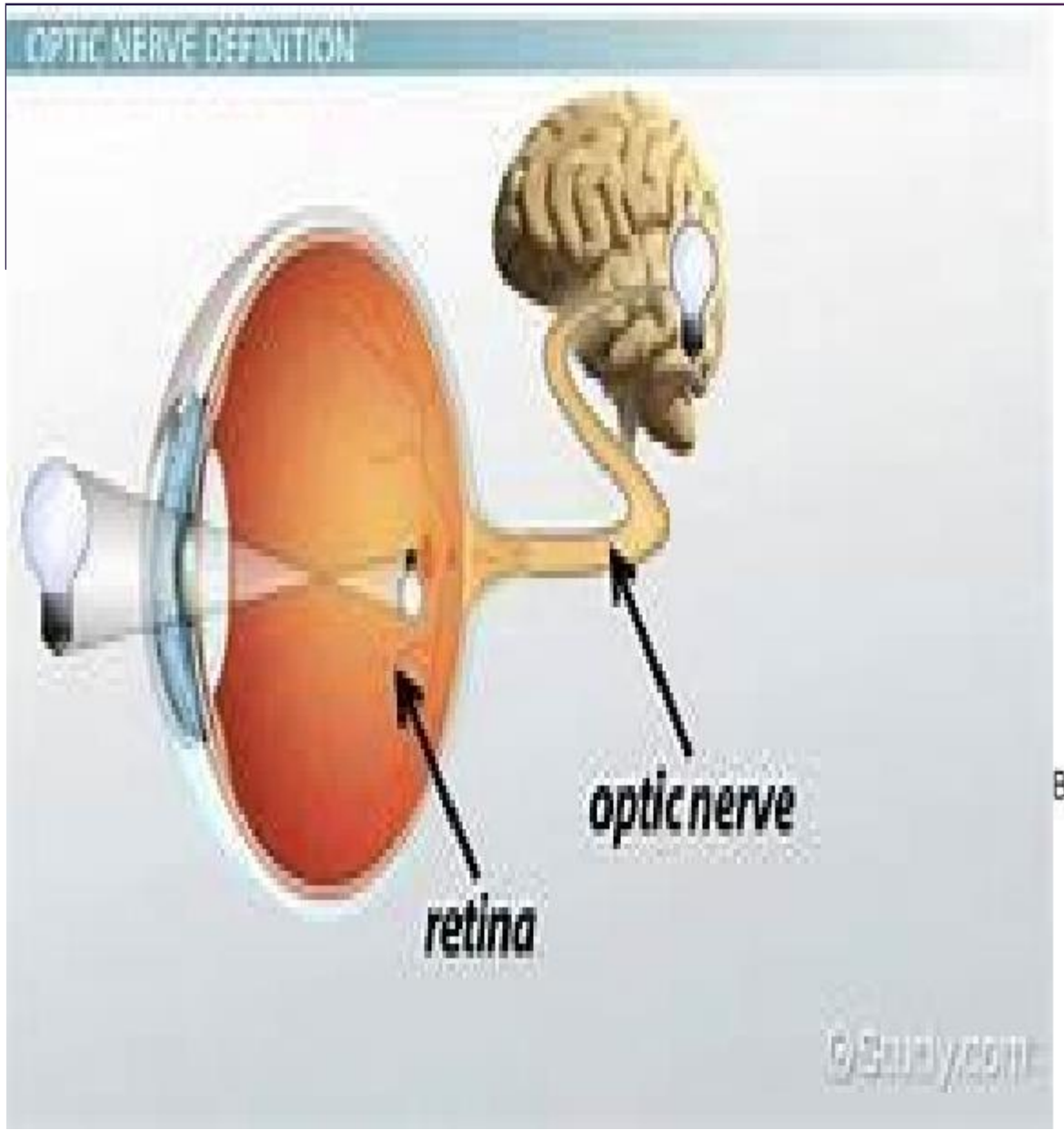
The optic disc or nerve head is the point where the axons from the retinal ganglion cells leave the eye.

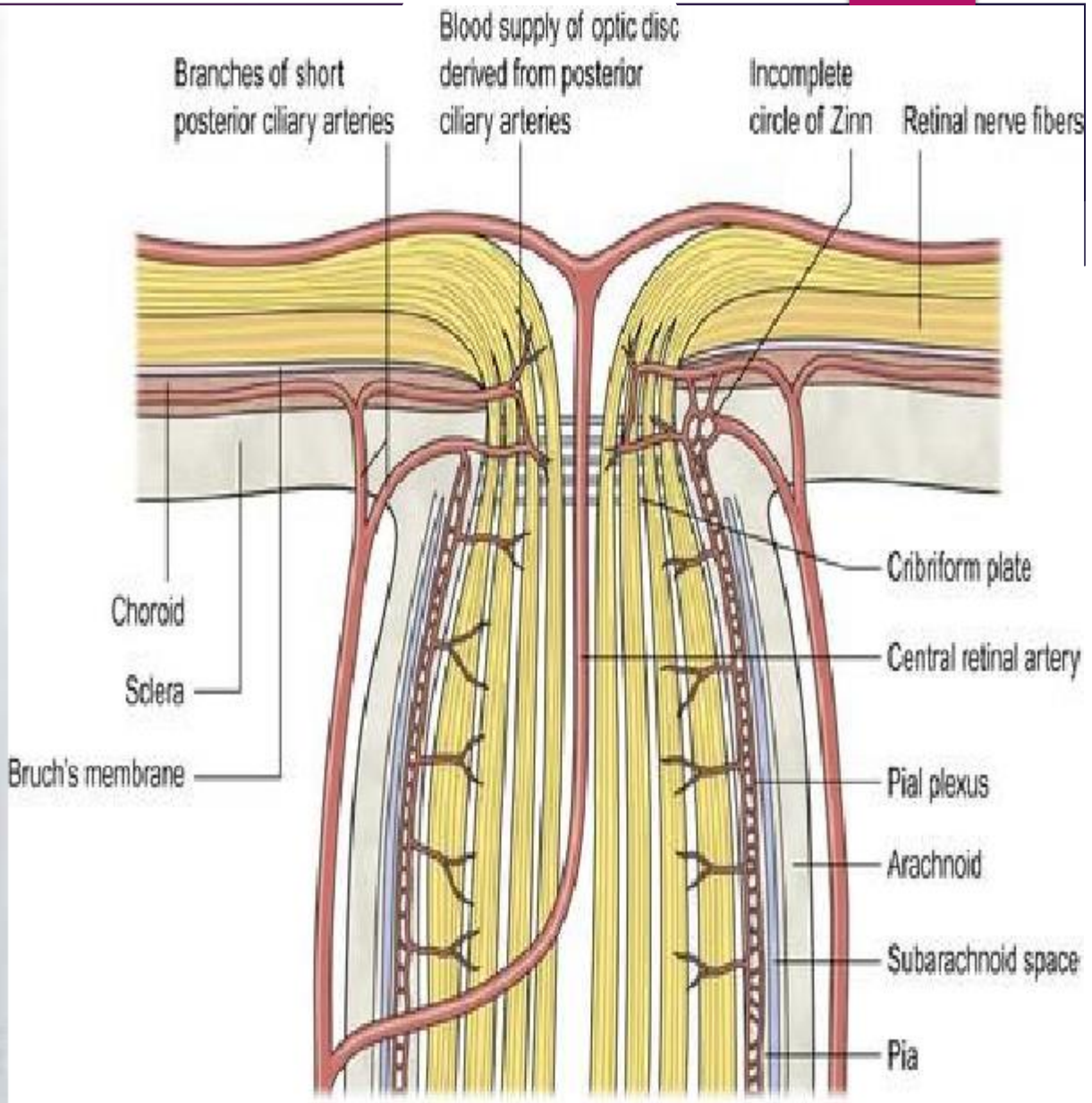
The nerve head appears as a white circular structure in the back of the eye. There are no photoreceptors on this structure. physiological blind spot.

it passes out of the eye through the cribriform plate of the sclera . That allow the nerve fibers to pass through many holes and into the extraocular (outside of the eyeball)

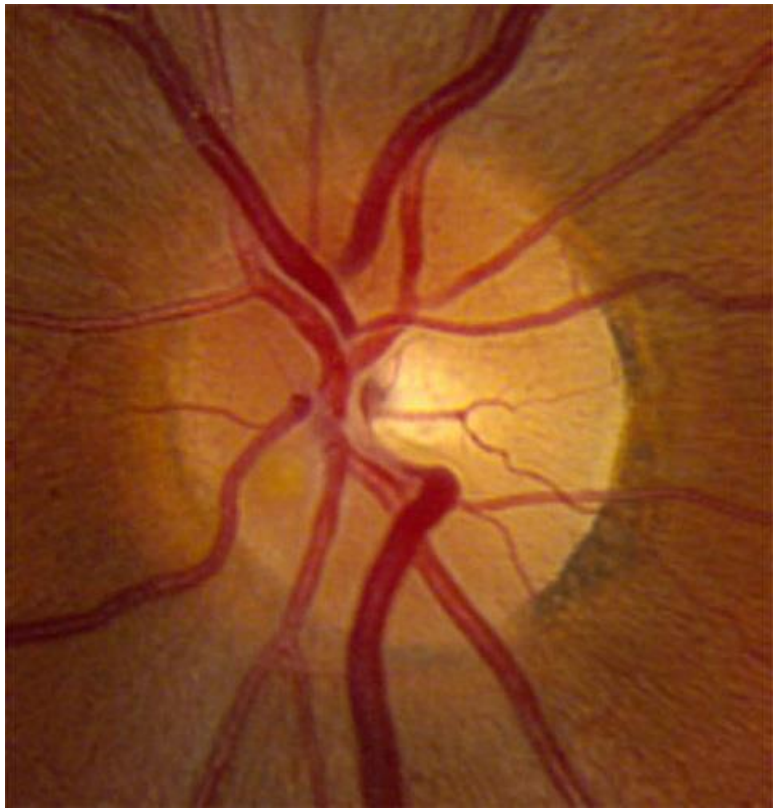
space. As the fibers pass through, they become covered by a sheath formed by dura , arachnoid and pia , continuous with that surrounding the brain . It is bathed in CSF.

Thecentral retinal vein and artery enter the eye in the center of the optic nerve .


OPTIC NERVE DEFINITION
optic nerve
retina


Branches of short posterior ciliary arteries
Blood supply of optic disc derived from posterior ciliary arteries
Incomplete circle of Zinn
Retinal nerve fibers
Choroid
Sclera
Bruch's membrane
Cribriform plate
Central retinal artery
Pial plexus
Arachnoid
Subarachnoid space
Pia

The normal optic nerve head has distinct margins, a pinkish rim and, usually, a central, pale, cup. The central retinal artery and vein enter the globe slightly nasally in the optic nerve head, referred to ophthalmoscopically as the optic disc. The optic disc may be involved in many disorders but has a limited repertoire of responses. Ophthalmoscopically it may become swollen, or it may become pale.



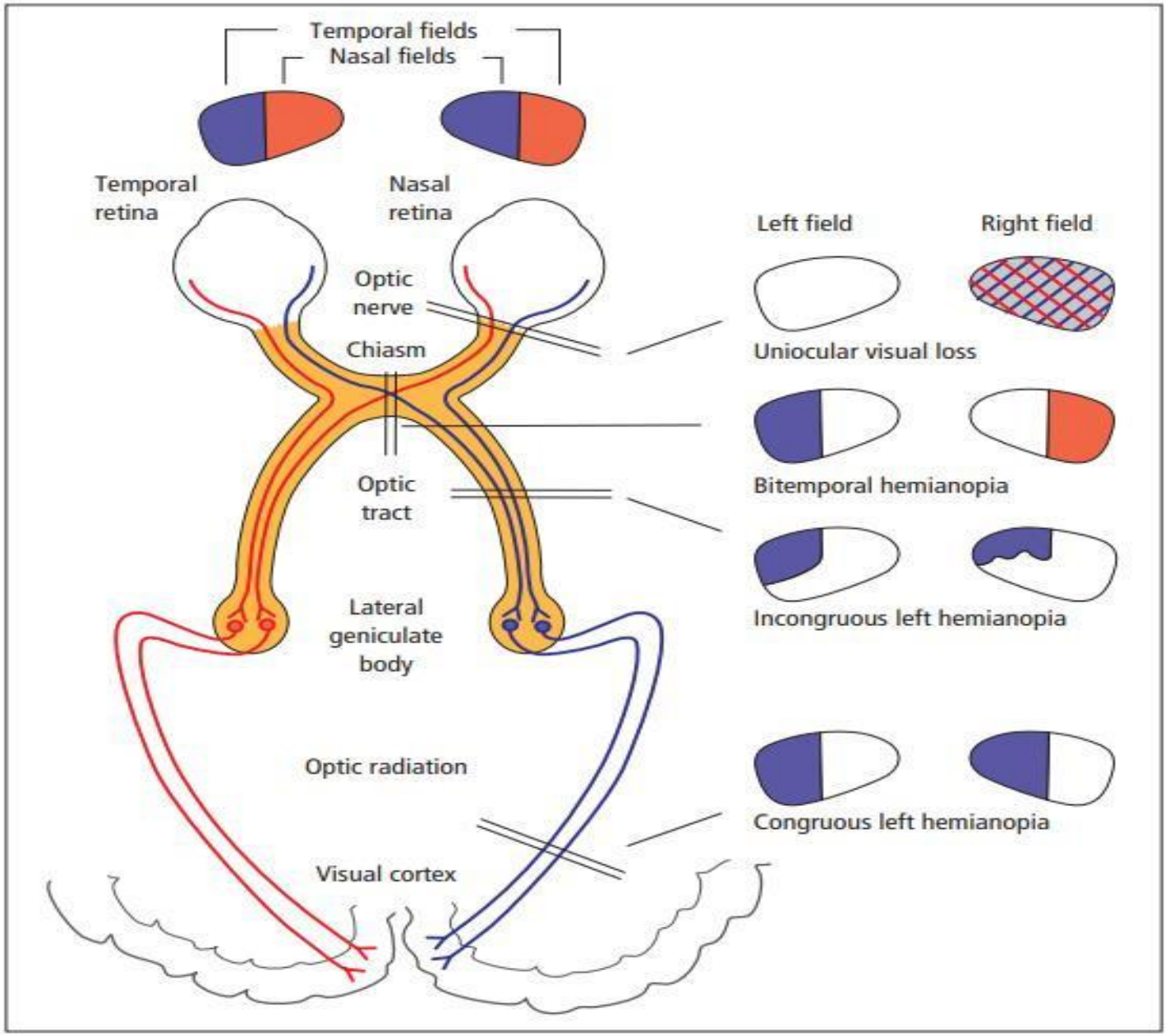


Figure 14.1 Anatomy of the optic pathway and the field defects produced by lesions at different sites.

The swollen optic disc The swollen disc is an important and often worrying sign. Papilledema is the term given to disc swelling associated with raised intracranial pressure (ICP), accelerated hypertension and optic disc ischemia. Papillitis , a condition with a similar fundoscopic appearance, is due to optic neuritis (inflammation) affecting the nerve head. Visual loss always occurs with optic neuritis but is uncommon with the papilledema of hypertension and/or raised intracranial pressure; it is, however, a feature of ischemic papilledema.

- Some normal optic nerve heads may appear to be swollen, due a crowding of nerve fibers entering the disc. This is termed pseudopapilloedema and occurs particularly in small, hypermetropic eyes where the nerve entry site is reduced in size.
- Note also that myelinated nerve fibers occurring on the nerve head may be mistaken for optic disc swelling. During development, myelination of the optic nerve begins proximally and spreads centrifugally to be completed at the lamina cribrosa at term. Occasionally, as a developmental variant, the process extends into the peripapillary retinal nerve fiber layer as a patch of light - scattering, myelinated fibers; the normally unmyelinated retinal nerve fiber layer is partly myelinated, giving it a feathery, white appearance which reflects the organization of the nerve fiber layer.
- In myopia it is common to see a pale ‘ myopic crescent ’ of peripapillary atrophy, at the temporal margin of the optic disc. In high myopia the optic disc may be surrounded by suchanatrophicarea (peripapillaryatrophy),whichmaybeconfusedwithdiscswelling.

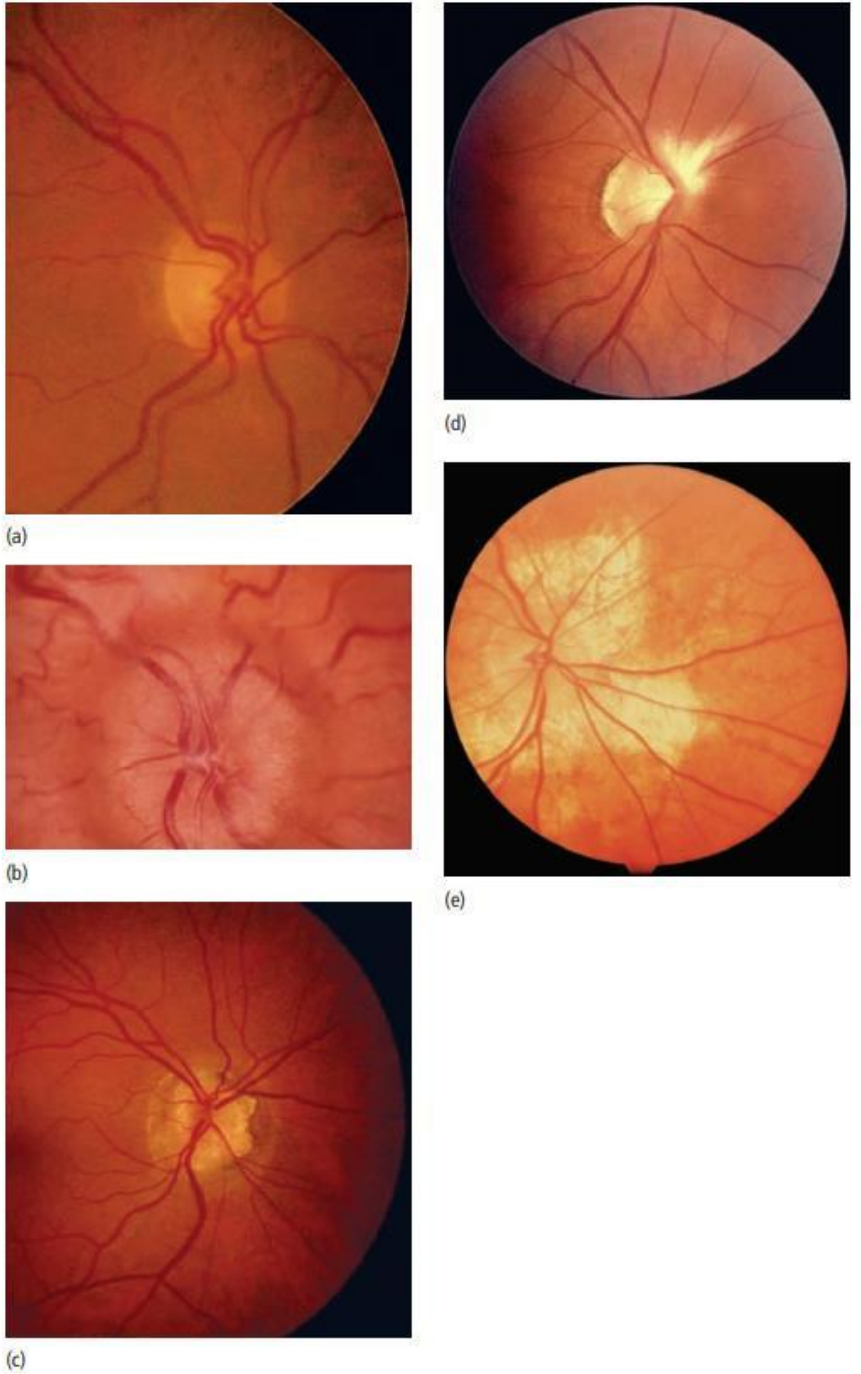
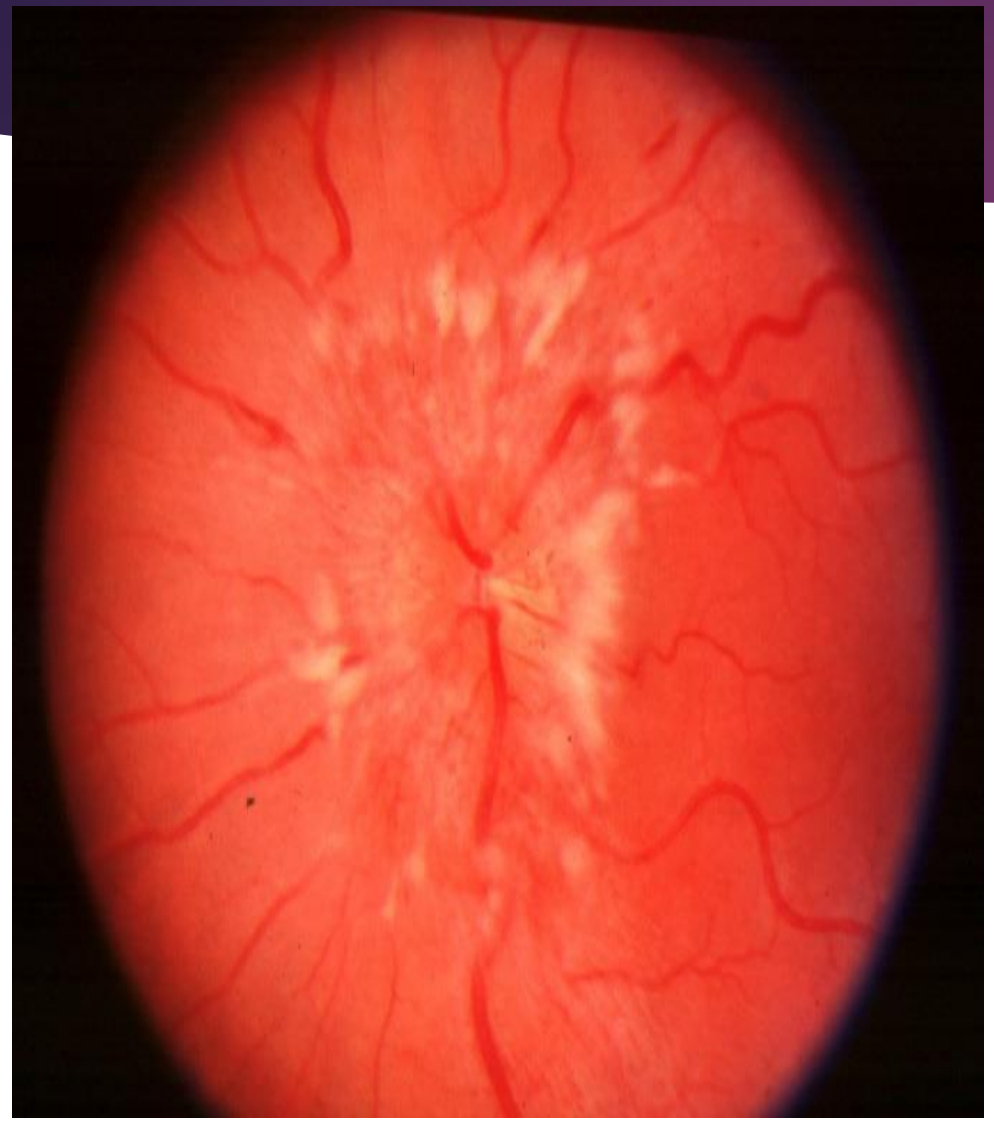


Figure 14.2 (a) A normal disc. (b) A swollen disc secondary to raised intracranial pressure. Note the lack of a sharp outline to the disc and the dilated capillaries on the disc. (c) The appearance of optic disc drüsen; note how the solid yellow lesions cause irregularity of the disc margin. (d) Myelination of the nerve fibres around the nerve head may be mistaken for a swollen optic disc. (e) A myopic optic disc. Note the extensive peripapillary atrophy.



You can notice:

- Disc hyperemia
- Blurred margins
- Nerve fiber layer (NF swelling
- Disc elevation
- Tortous veins

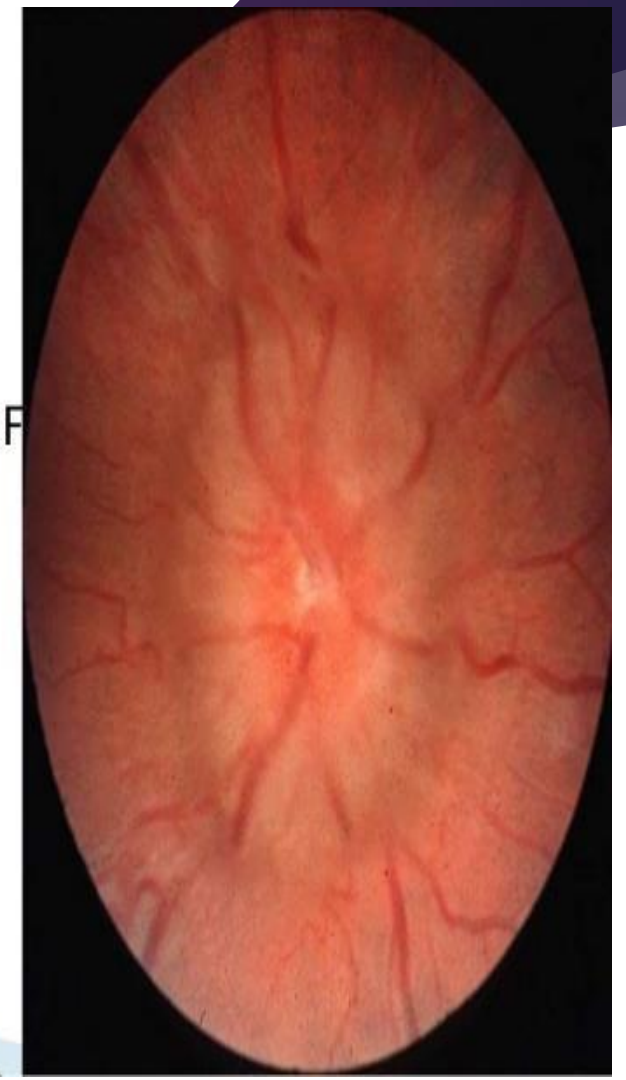


Table 14.1 Causes of a swollen optic disc.

Condition	Distinguishing features
Raised intracranial pressure	Vision and field usually normal save for large blind spot. Obscurations (short episodes of visual loss usually on changing posture). Field may be contracted in chronic disease. Colour vision normal. No RAPD. No spontaneous venous pulsation of the vein at the disc (but some people with normal intracranial pressure do not have this). Dilated capillaries and haemorrhages on disc. Other symptoms and signs of raised intracranial pressure
Space-occupying lesions of the optic nerve head	Various solid or infiltrative lesions at the nerve head, e.g. optic disc drüsen (calcified axonal material), gliomas, sarcoidosis and leukaemia, may produce disc swelling. These may be associated with reduced vision and field defects
Papillitis (optic neuritis affecting the optic nerve head)	A swollen optic disc. Exudates around the macula may occasionally be seen. Vision is profoundly reduced. Colour vision is abnormal. RAPD present. A central field defect is present
Accelerated (malignant) hypertension (see Chapter 12)	Reduced vision, haemorrhagic disc swelling. Retinal haemorrhages, exudates and cotton-wool spots away from the nerve head. Check blood pressure!
Ischaemic optic neuropathy	Sudden visual loss, field defect. Colour vision may be normal. RAPD may be present. Spontaneous venous pulsation at the optic disc may be present. May be sectorial swelling only. Haemorrhages on disc and disc margin. Cotton-wool spots may be seen around disc, particularly if caused by giant cell arteritis
Central retinal vein occlusion (see Chapter 12)	Sudden marked visual loss, tortuous veins, gross retinal haemorrhage

Disc swelling must be distinguished from pseudopapilloedema such as optic nerve head drüsen, myelinated nerve fibres and the peripapillary atrophy of high myopia.

RAPD, relative afferent pupillary defect; see Chapter 2.

1.Papilledema due to raised intracranial pressure History The crucial feature

of disc swelling due to raised intracranial pressure is that
there is rarely an associated visual loss, although some patients, with

advanced papilledema, may develop fleeting visual loss lasting seconds
when they alter posture from lying to standing (obscurations of vision).

Other important clues to the presence of raised intracranial pressure include:

- headache, worse on waking and made worse by coughing;
- nausea, retching;
- diplopia (double vision), usually due to a sixth nerve palsy;
- other neurological signs, if the raised pressure is due to a cranial space - occupying lesion – visual fi eld loss; cranial nerve palsy;
- a history of head trauma suggesting a subdural hemorrhage;
- a history of medications sometimes associated with raised intracranial pressure (e.g. oral contraceptives, tetracyclines).

Signs • The optic disc is swollen, its edges are blurred and the superficial capillaries are
dilated and thus abnormally prominent. There is no spontaneous venous pulsation of the central retinal vein. This has a physiological basis. The central retinal vein is exposed to cerebrospinal fluid (CSF) as it leaves the optic nerve to join the veins of the orbit. Normally, venous pressure in the retinal veins at the nerve head is just above ocular pressure. Venous pulsation occurs because the vein collapses briefly with each rise in ocular pressure with arterial inflow. When the CSF pressure is higher than the ocular pressure, as occurs in papilledema due to raised intracranial pressure, the pressure in the veins at the disc rises above the ocular pressure and so spontaneous venous pulsation is lost. Absence of spontaneous venous pulsation is seen in 5 – 20% of those with normal nerve heads, but in this case venous collapse at the nerve head can be induced by light pressure on the globe.

- A large blind spot will be found on visual field testing, corresponding to the swollen nerve head. In chronic papilledema the fi eld may become constricted. A field defect may, however, be caused by the space - occupying lesion causing the papilledema.

- Abnormal neurological signs may indicate the site of a space - occupying lesion.

Investigation CT and MRI scanning will identify any space - occupying lesion

or
enlargement of the ventricles. Following neurological consultation (and

normally after a scan) a lumbar puncture will enable intracranial pressure to be measured. Treatment is dependent on findings.

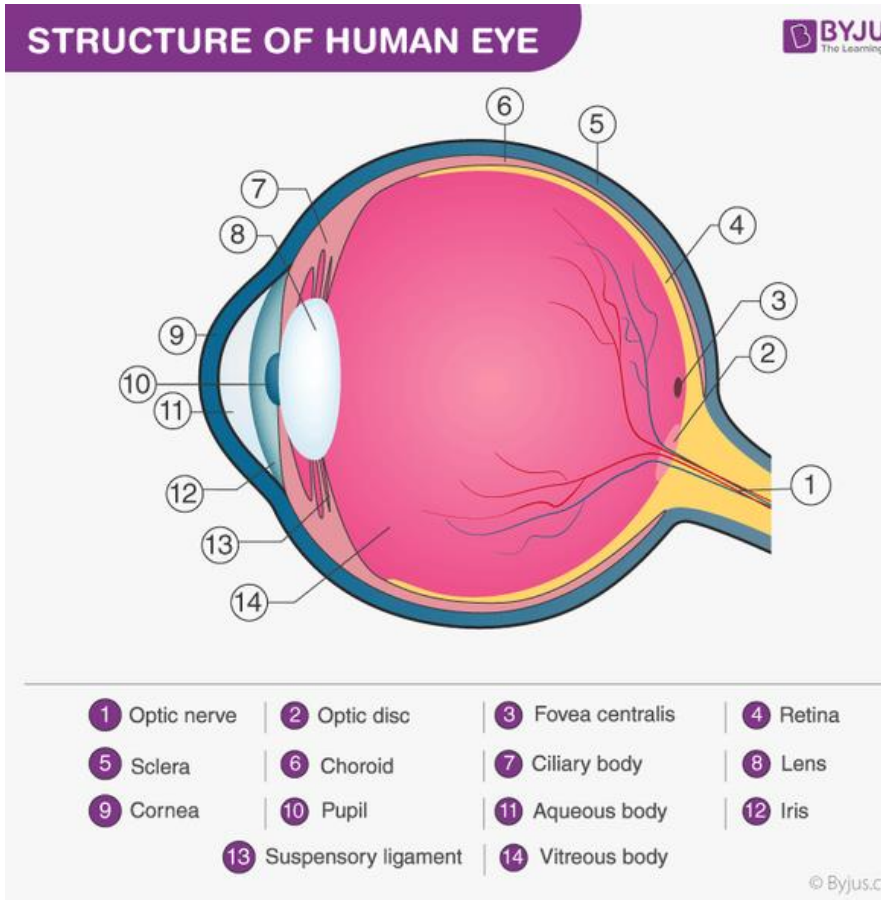
Idiopathic intracranial hypertension treatment Intracranial pressure may be elevated and disc swelling present with no evidence of intracranial abnormality and no dilation of the ventricles on the scan. This is termed idiopathic intracranial hypertension (previously, benign intracranial hypertension) and usually presents in overweight women in the second and third decades. It may also be caused by exposure to certain drugs such as the contraceptive pill and tetracyclines. Patients complain of headache and may have obscurations of vision and sixth nerve palsies. No other neurological problems are present. Although acute permanent visual loss is not a feature of papilledema, if the nerve remains swollen for several weeks there will be a progressive contraction of the visual fi eld.

It is thus important to reduce intracranial pressure. This may be achieved: • with medications such as oral acetazolamide; • through ventriculoperitoneal shunting; • through optic nerve decompression, where a small hole is made in the sheath surrounding the optic nerve to allow the drainage of CSF and reduce the pressure of CSF around the anterior optic nerve. Space - occupying lesions (i.e. tumors and hemorrhage) and hydrocephalus require neurosurgical management

2.Optic neuritis

Inflammation or demyelination of the optic nerve results in optic neuritis.

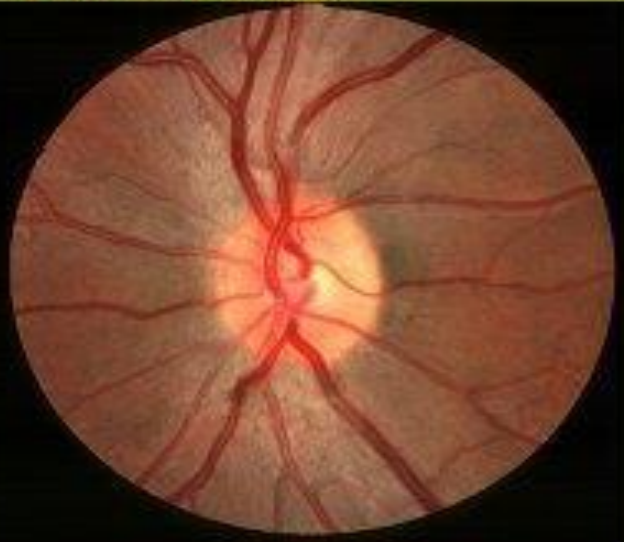
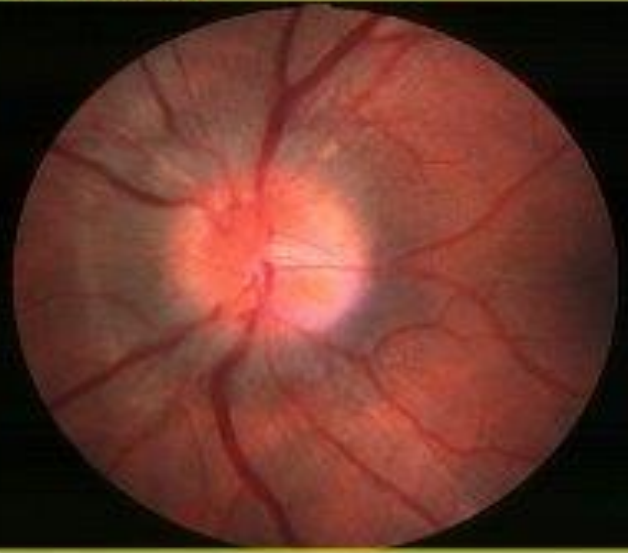
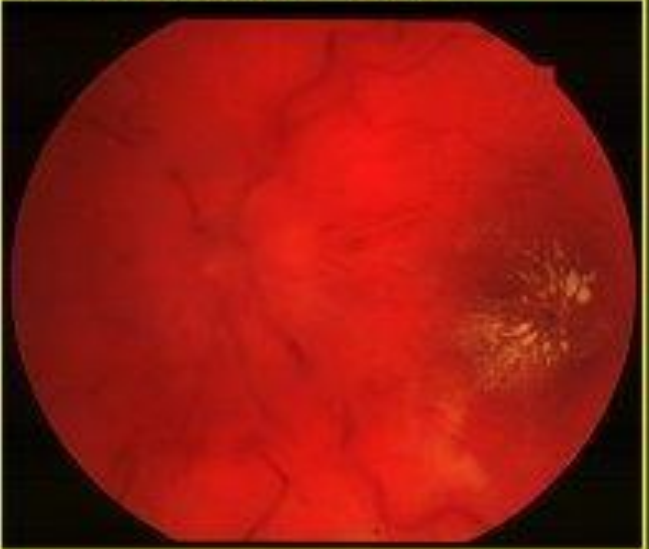
This is termed **papillitis** if the optic nerve head is affected and **retrobulbar** neuritis if the optic nerve is affected more posteriorly with no disc swelling.



Causes of optic neuritis

- Certain autoimmune diseases such as multiple sclerosis commonest cause
- 25% of MS patients come firstly with symptoms of ON
- Infections such as viruses (especially in children), measles, meningitis, syphilis, sinusitis, tuberculosis, and human immunodeficiency virus (HIV)
- Malnutrition. E.g. vit B complex deficiency.
- Intraocular inflammation (uveitis)

Classification of optic neuritis

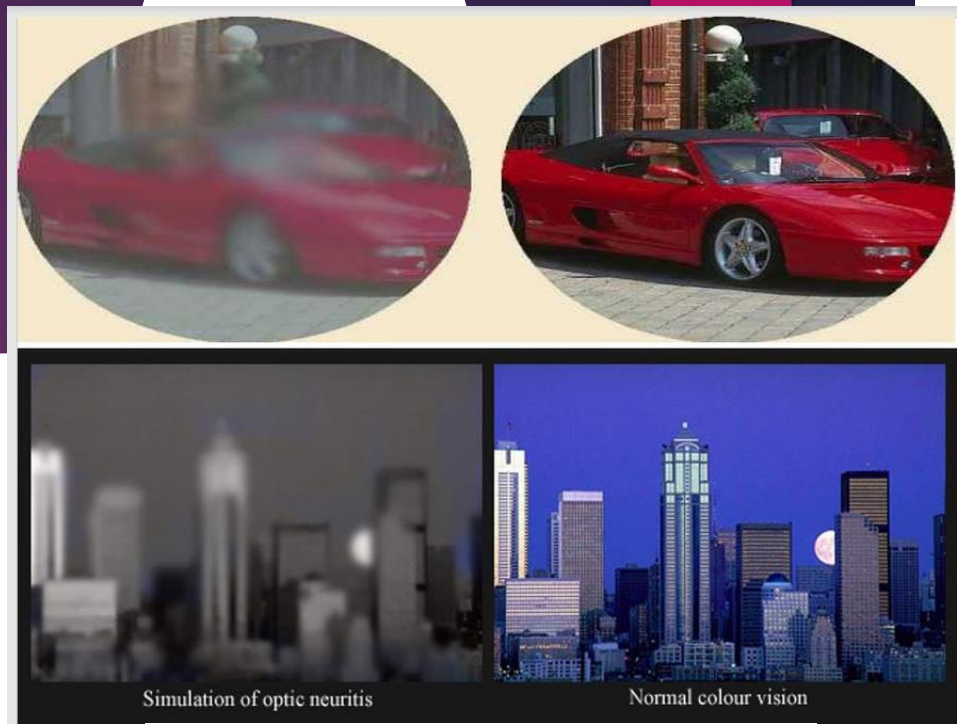
Retrobulbar neuritis (normal disc)	Papillitis (hyperaemia and oedema)	Neuroretinitis (papillitis and macular star)
		
• Demyelination - most common • Sinus-related (ethmoiditis) • Lyme disease	• Viral infections and immunization in children (bilateral) • Demyelination (uncommon) • Syphilis	• Cat-scratch fever • Lyme disease • Syphilis

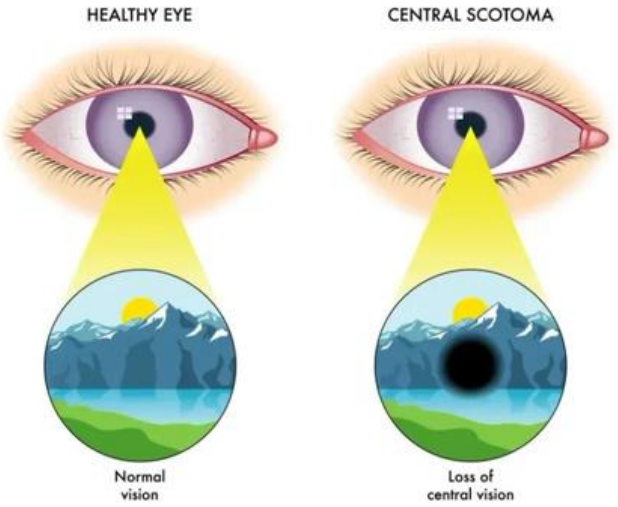
History • acute loss of vision that may progress over a few days and then slowly improve;

• pain on eye movement in retrobulbar neuritis because rectus contraction pulls on the optic nerve sheath;

• a preceding history of viral illness in some cases. Between 40% and 70% of patients with optic neuritis will have, or develop, other neurological symptoms to suggest a diagnosis of demyelination (multiple sclerosis).

Examination • reduced visual acuity; • reduced color vision; • a relative afferent pupillary defect (RAPD)
due to reduced optic nerve conduction ;
• a central scotoma on field testing;
• a normal disc in retrobulbar neuritis; a swollen disc in papillitis.





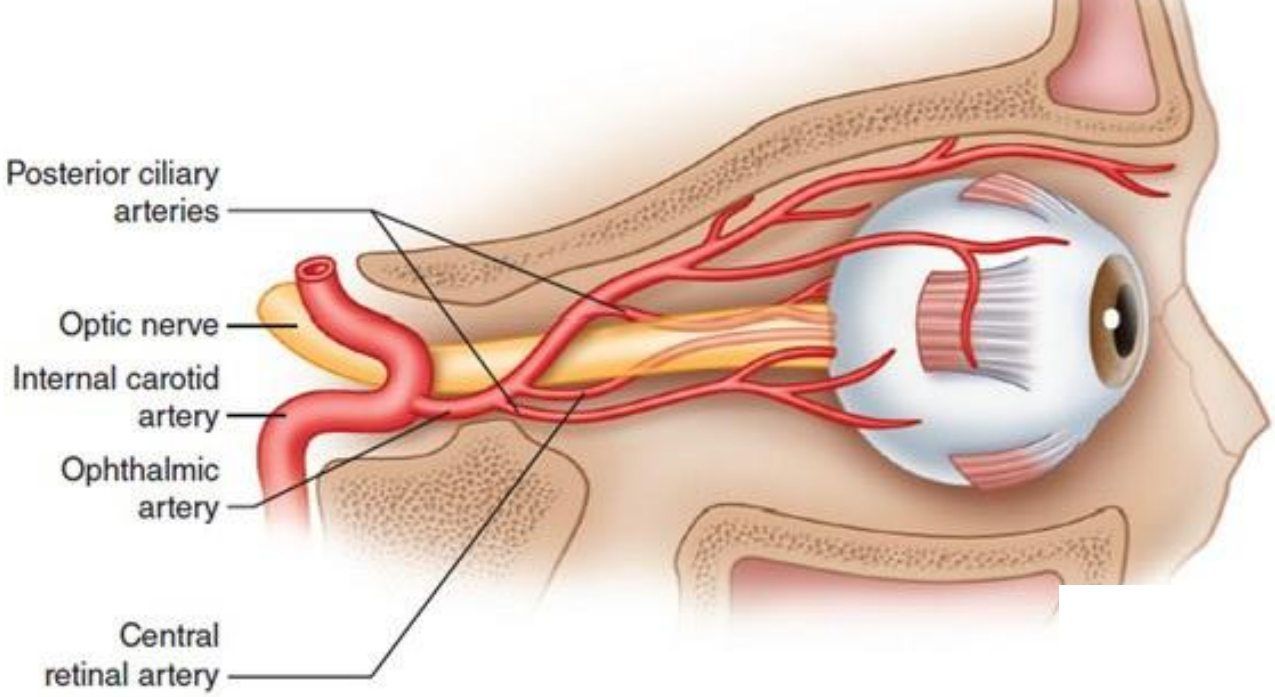
Investigation &Treatment:

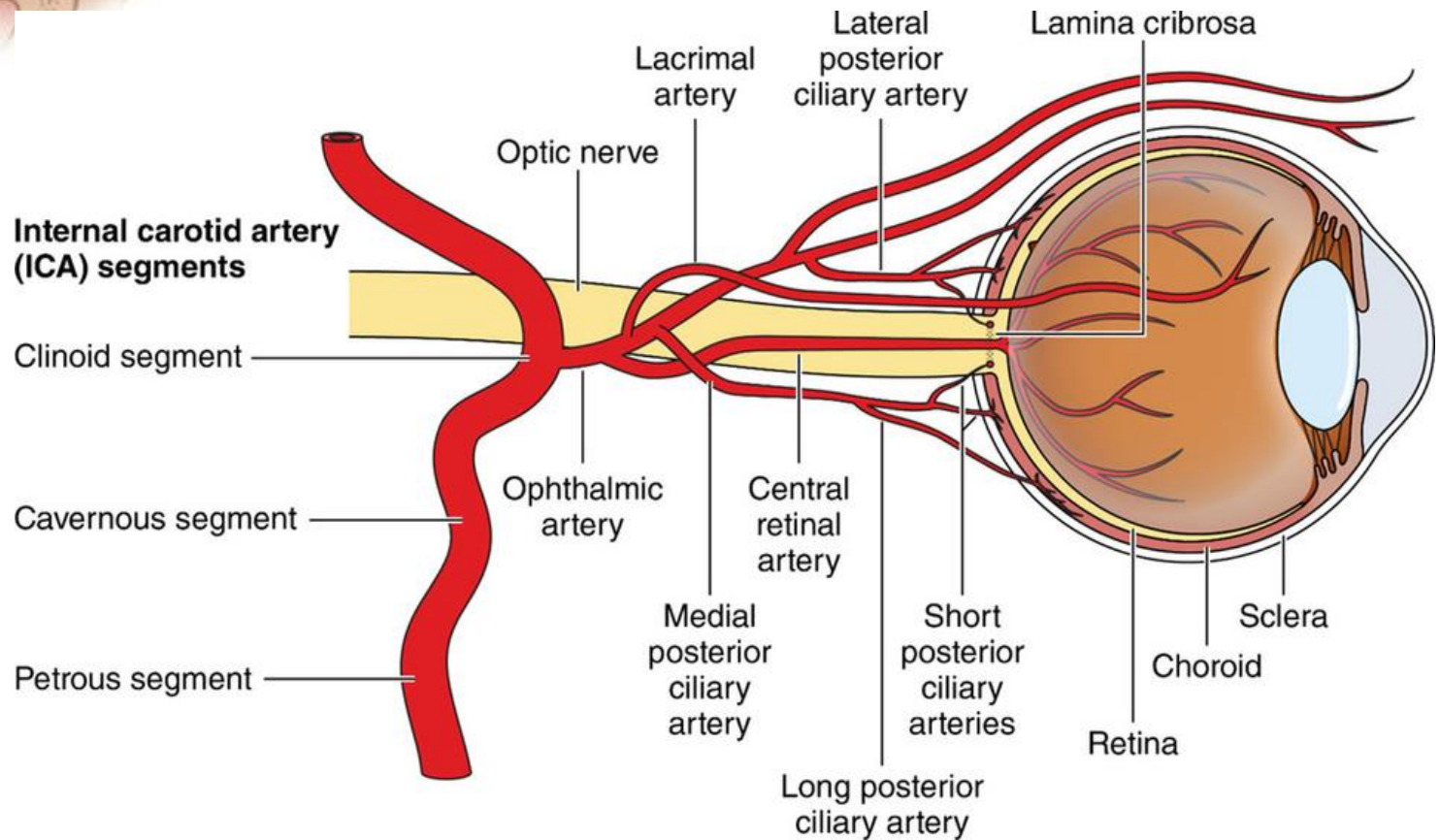
MRI scan will help to identify additional ‘ silent ’ plaques of demyelination, but the patient must be suitably counselled before a scan is performed. The diagnosis of multiple sclerosis is essentially a clinical one, and the patient may not wish to have the presence of other possible plaques confirmed with a scan. There may be a role for steroid treatment to speed up visual recovery.
You have to treat hte cause.

Prognosis:

Vision slowly recovers over several weeks, although often it is not quite as good as before the attack. Repeated episodes may lead to optic atrophy and a decline in vision. Very occasionally, in atypical cases, vision may not recover.

3.Ischemic optic neuropathy Pathogenesis The anterior optic nerve may become ischemic if the posterior ciliary vessels are compromised as a result of degenerative vaso - occlusive or vasculitic disease of the arterioles This results in an anterior ischemic optic neuropathy .

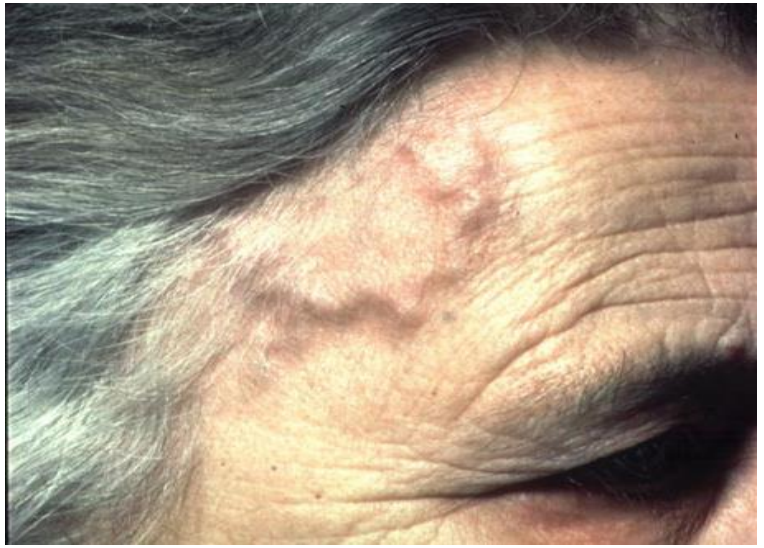

Posterior ciliary arteries
Optic nerve
Internal carotid artery
Ophthalmic artery
Central retinal artery


Internal carotid artery (ICA) segments
Clinoid segment
Cavernous segment
Petrous segment
Optic nerve
Lacrimal artery
Lateral posterior ciliary artery
Lamina cribrosa
Ophthalmic artery
Central retinal artery
Medial posterior ciliary artery
Short posterior ciliary arteries
Long posterior ciliary artery
Retina
Choroid
Sclera

Symptoms The patient complains of a sudden loss of vision or visual field, often on
waking, since vascular perfusion of the eye is decreased during sleep. If accompanied by pain or scalp tenderness, the diagnosis of giant cell arteritis must never be forgotten. Ischemic optic neuropathy is the usual cause of blindness in the disease.

Giant cell arteritis : This is an autoimmune vasculitis occurring in patients generally over the age of 60. It affects arteries with an internal elastic lamina, which therefore includes the ophthalmic artery. It may present with any combination of:

- sudden loss of vision;
- scalp tenderness (e.g. on combing);
- pain on chewing (jaw claudication);
- shoulder pain;
- malaise.

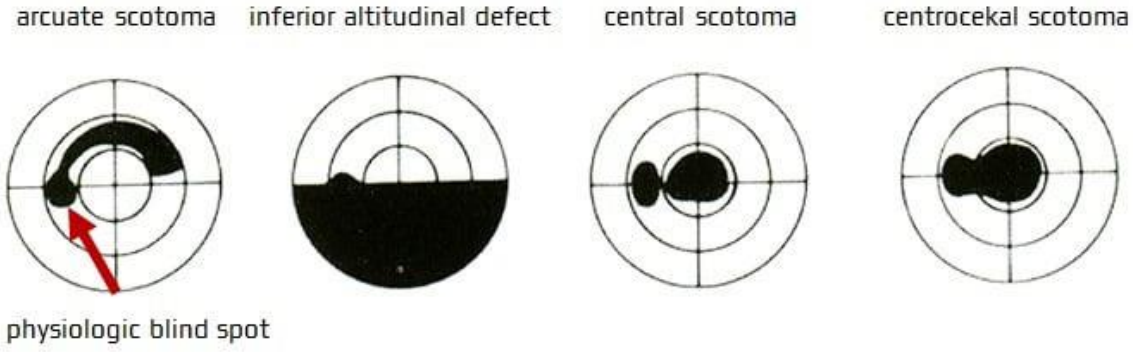


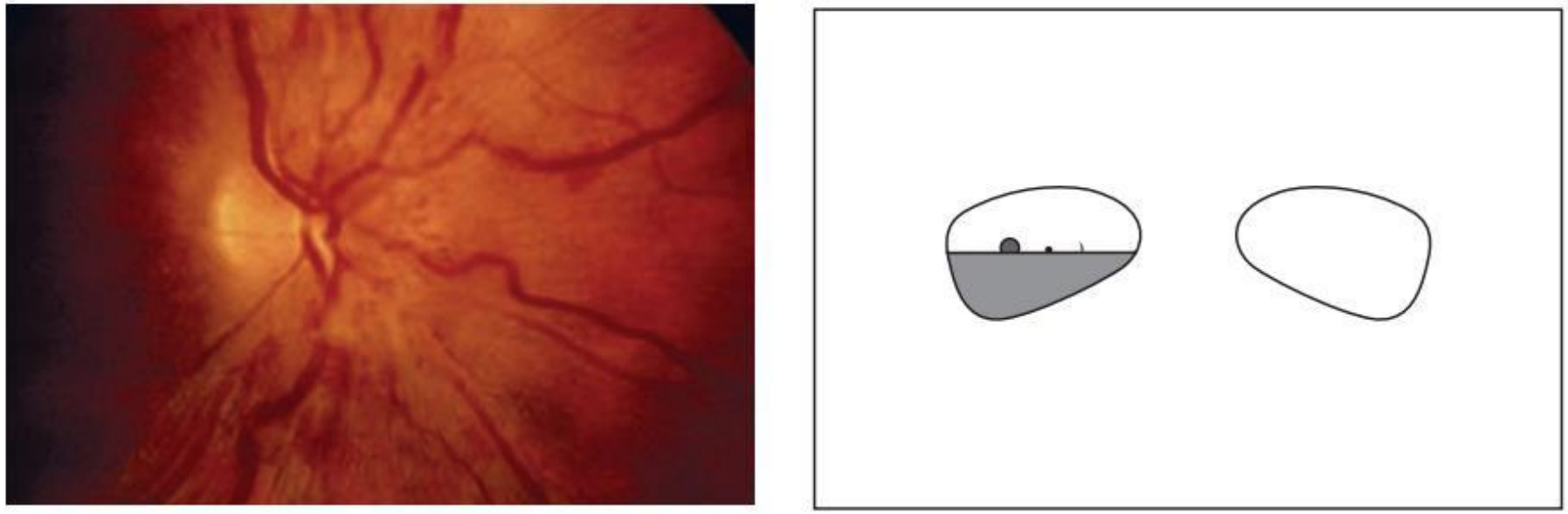
Signs • a reduction in visual acuity; • a field defect, typically an absence of the lower or upper half of the visual field (altitudinal scotoma);

• a swollen and hemorrhagic disc with normal retina and retinal vessels (remember the blood supply to the anterior optic nerve and retina are different) – in arteritic ischemic optic neuropathy the disc may be pale;

• a small normal fellow disc with a small cup in non - arteritic disease;

• a tender temporal artery, a sign suggestive of giant cell arteritis.

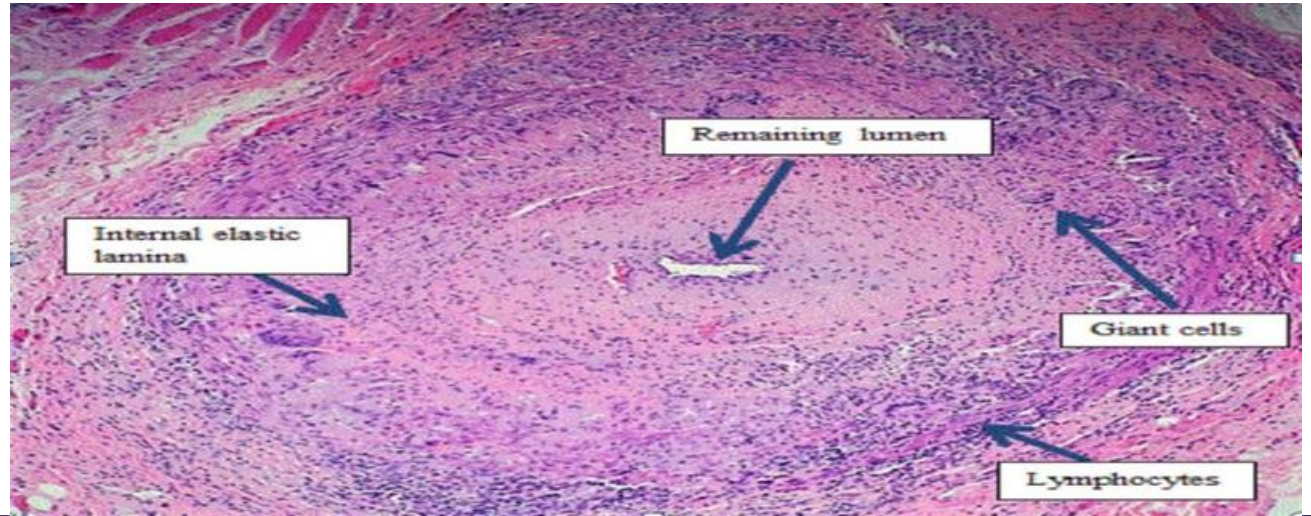




(a) (b)

Figure 14.3 (a) The clinical appearance of the optic disc and (b) one form of field defect (altitudinal) seen in ischaemic optic neuropathy.

A. Investigations If giant cell arteritis is present the erythrocyte sedimentation rate (ESR) and C - reactive protein are usually grossly elevated (e.g. ESR = 100 mm/h) (although 1 in 10 patients with giant cell arteritis have a normal ESR). Temporal artery biopsy is often helpful, but again may not lead to a diagnosis, particularly if only a small specimen is examined, because the disease may skip a length of the artery. Giant cell arteritis can also present as a central retinal artery occlusion when the vessel is affected secondarily to arteritis of the ophthalmic artery.



B. Investigation of the patient with non - arteritic ischaemic optic neuropathy includes:

- a full blood count to exclude anemia;
- blood pressure check;
- blood sugar check;
- ESR and C - reactive protein to check for giant cell arteritis. Both hypertension and diabetes may be associated with the condition. It may also be seen in patients suffering acute blood loss, e.g. hematemesis, where it may occur some days after the acute bleed. Hypotensive episodes may also give rise to ischemic optic neuropathy. Occasionally, clotting disorders or autoimmune disease may cause the condition.

Treatment If giant cell arteritis is suspected, treatment must not be delayed while the diagnosis is confirmed. High - dose steroids must be given, intravenously and orally, and the dose tapered over the ensuing weeks according to both symptoms and the response of the ESR or C - reactive protein. The usual precautions must be taken, as with any patient on steroids, to exclude other medical conditions that might be unmasked or made worse by the steroids (e.g. tuberculosis, diabetes, hypertension and an increased susceptibility to infection). Steroids will not reverse the visual loss but can prevent the fellow eye being affected. There is unfortunately no treatment for non - arteritic ischaemic optic neuropathy other than by the management of underlying conditions.

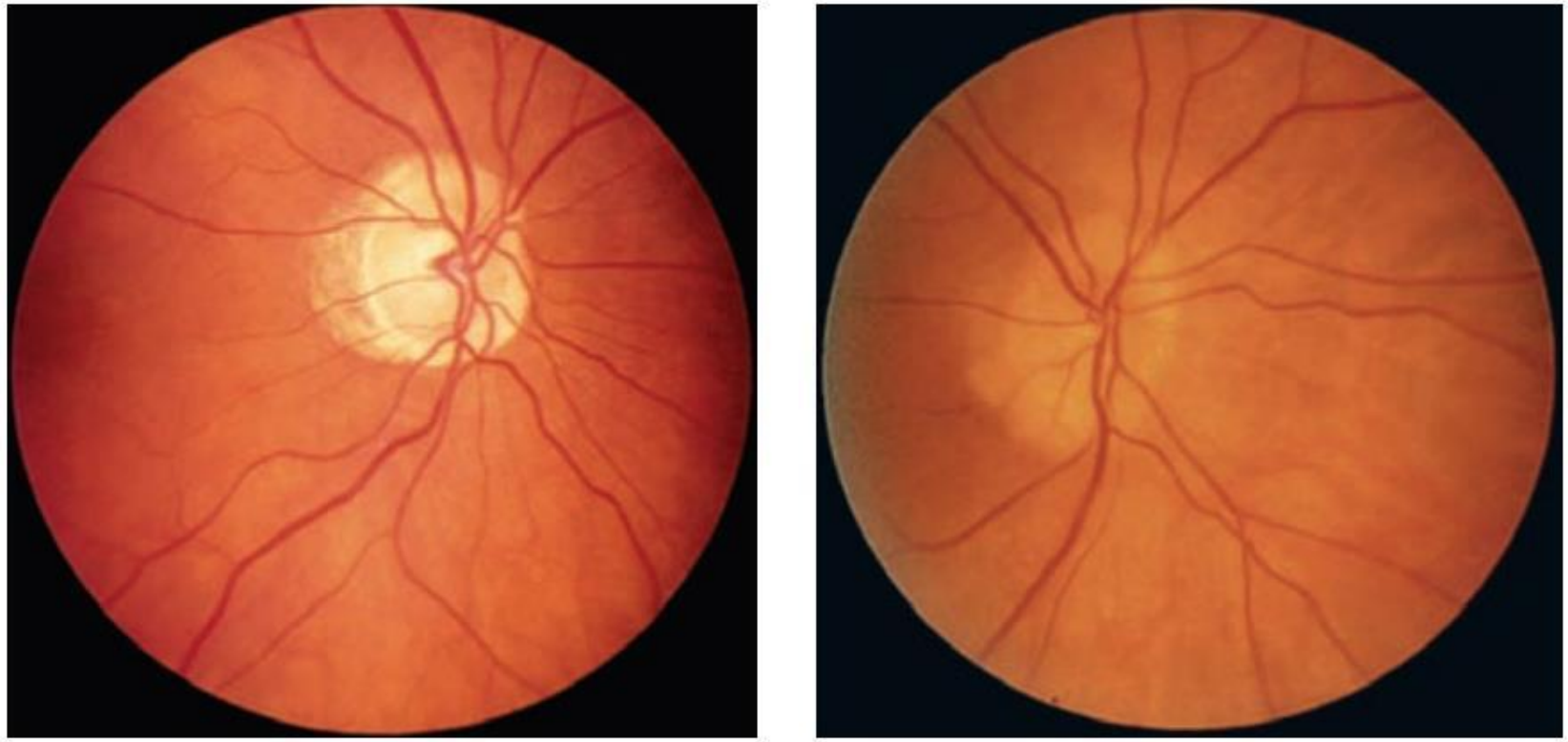
Prognosis The second eye may rapidly become involved in patients with untreated giant cell
arteritis but this can be prevented by prompt initiation of systemic steroid therapy. Steroid therapy may have to be continued on a prolonged basis and monitored on the ESR or C - reactive protein result. There is also a significant rate of involvement of the second eye in the non - arteritic form (40 – 50%). It is unusual for the vision to get progressively worse in non - arteritic ischaemic optic neuropathy, and the visual outcome in terms of both visual field and acuity is very variable. In both conditions, vision does not recover once it has been lost.

Optic atrophy

A **pale optic disc** represents a **loss of nerve fibres at the optic nerve head.** The **vision** is usually reduced and **color vision affected.**

On examination, the usual **vascularity** **of the disc is lost.**
Comparison of the two eyes is of great help in **unilateral cases,** as the contrast makes pallor much easier to see. A **relative afferent pupillary defect** will usually be present.

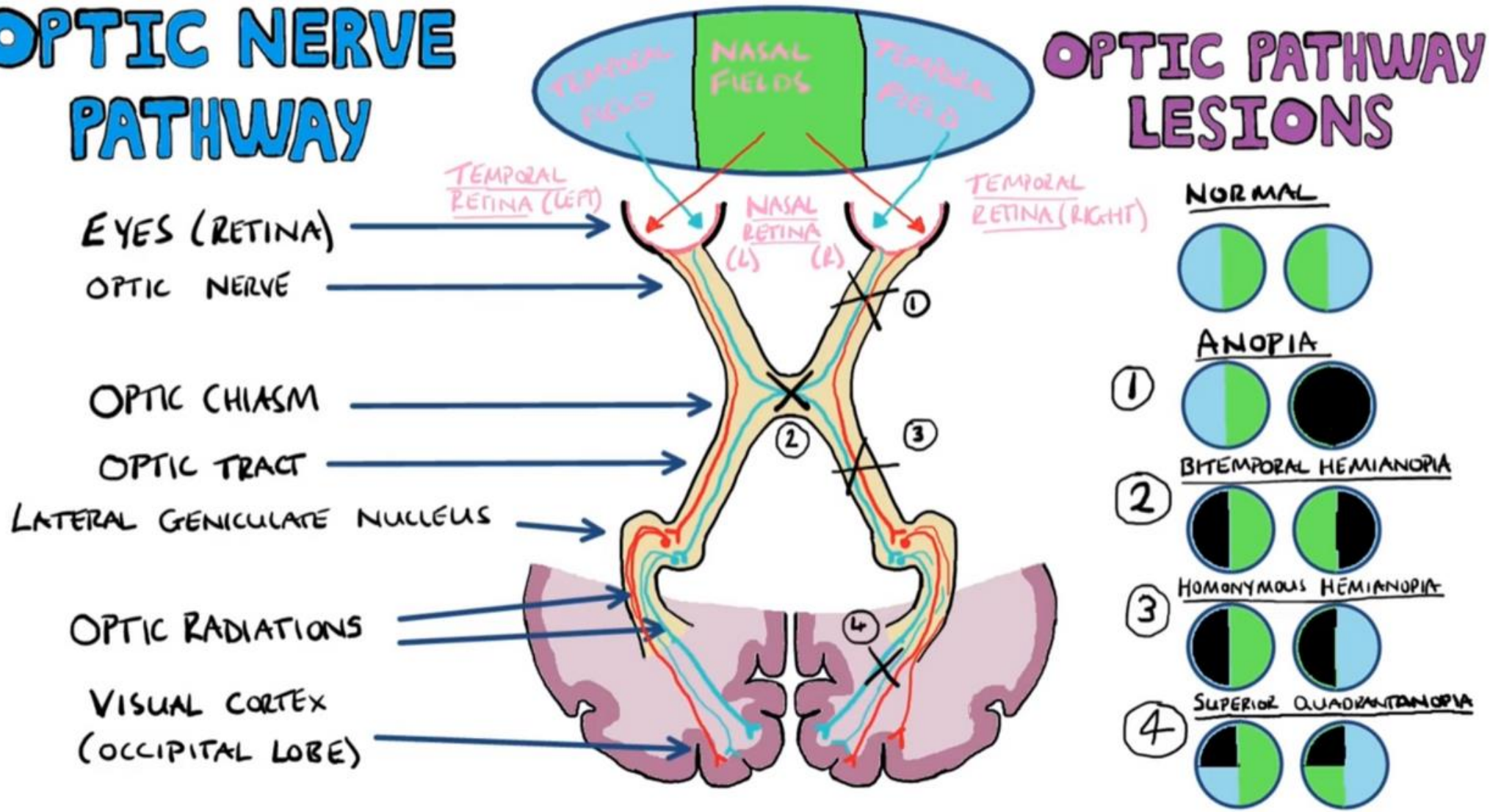
Treatment:
Optic nerve **can't regenerate**, so visual loss is irreversible



(a) (b)

Figure 14.4 (a) A pale optic disc compared to (b) a normal optic disc.

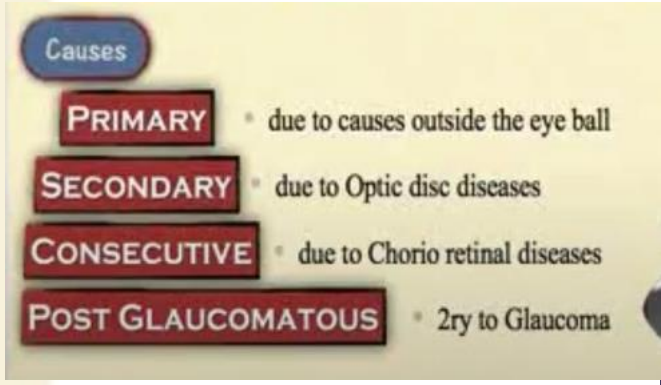
OPTIC NERVE PATHWAY



OPTIC PATHWAY LESIONS

Table 14.2 Causes of a pale optic disc.

Cause	Distinguishing features
Compression of the optic nerve	History of orbital or chiasmal disease. If sectorial, field loss may give a clue to the location of a compressive lesion
Ischaemic optic neuropathy	A history of sudden (unilateral) visual loss in the past
Retinal artery occlusion	The retinal vessels are attenuated
Retinal vein occlusion	Engorged retinal veins; retinal haemorrhage
Glaucoma (see Chapter 10)	The optic disc is pathologically cupped
Papilloedema and chronic elevation of ICP	The visual fields are contracted
Optic neuritis	There may be a history of previous loss of vision. Symptoms and signs compatible with multiple sclerosis may be present
Inherited optic nerve disease	Dominant and recessive optic neuropathy are associated with onset of blindness in the first few years of life. Leber's hereditary optic neuropathy results from an inheritable mutation of mitochondrial DNA. It typically affects males in early adulthood. It is bilateral. There is a bilateral central scotoma.
Inherited retinal disease	Retinal disease may result in optic disc pallor. It is, for example, a feature of retinitis pigmentosa and rod–cone dystrophies
Toxic optic neuropathy	Optic neuropathy may follow chemical toxicity, for example heavy metals, toluene from glue-sniffing and some drugs (e.g. isoniazid used in the treatment of tuberculosis). Again, information should be sought in the history
Tobacco/alcohol/ nutritional vitamin amblyopia	Optic neuropathy here (where all three factors are often involved together) is due to a combination of vitamin deficiency (B_{12}) and cyanide toxicity



The chiasm

The chiasm Compressive lesions at the chiasm **produce** a **bitemporal hemianopia** as the fibers representing the **nasal retina (temporal field) are compressed** as they **cross in the center of the chiasm**. Patients may present with rather vague visual symptoms, for example:

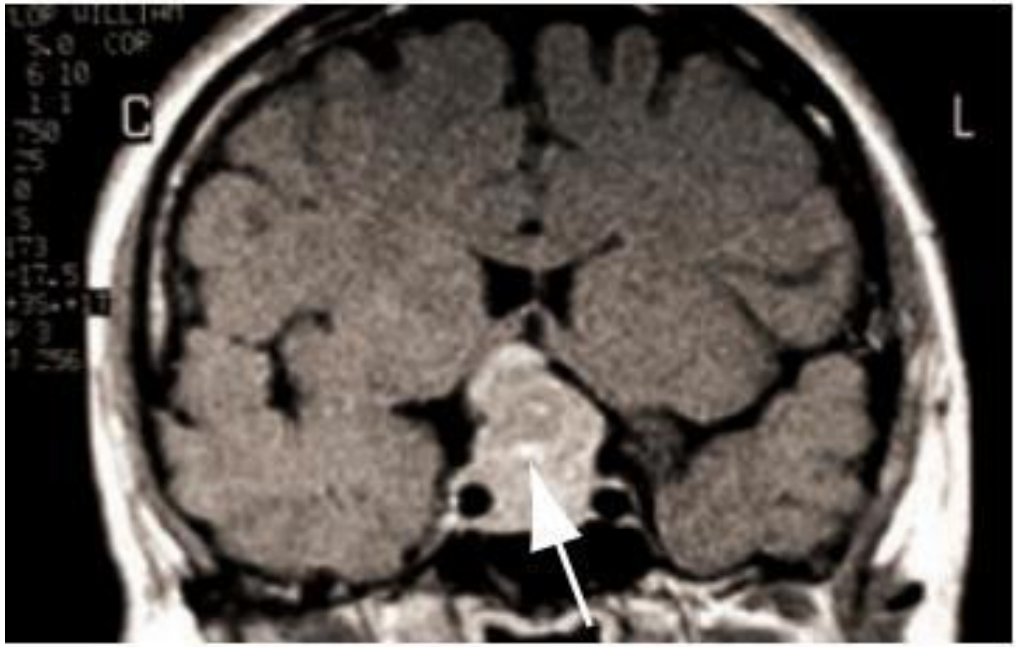
- Missing objects in the **periphery** of the visual fi eld.
- When testing vision with a Snellen chart patients may **miss the temporal letters with each eye.**
- The bitemporal field loss may cause difficulty in **fusing images,** causing the patient to **complain of diplopia** although eye position and movements are normal. • There may be difficulty with tasks requiring stereopsis such as pouring water into a cup or threading a needle.

The **most common lesion** is **a pituitary tumo**r , and the patient should be **asked whether they have symptoms relating to hormonal disturbance.**

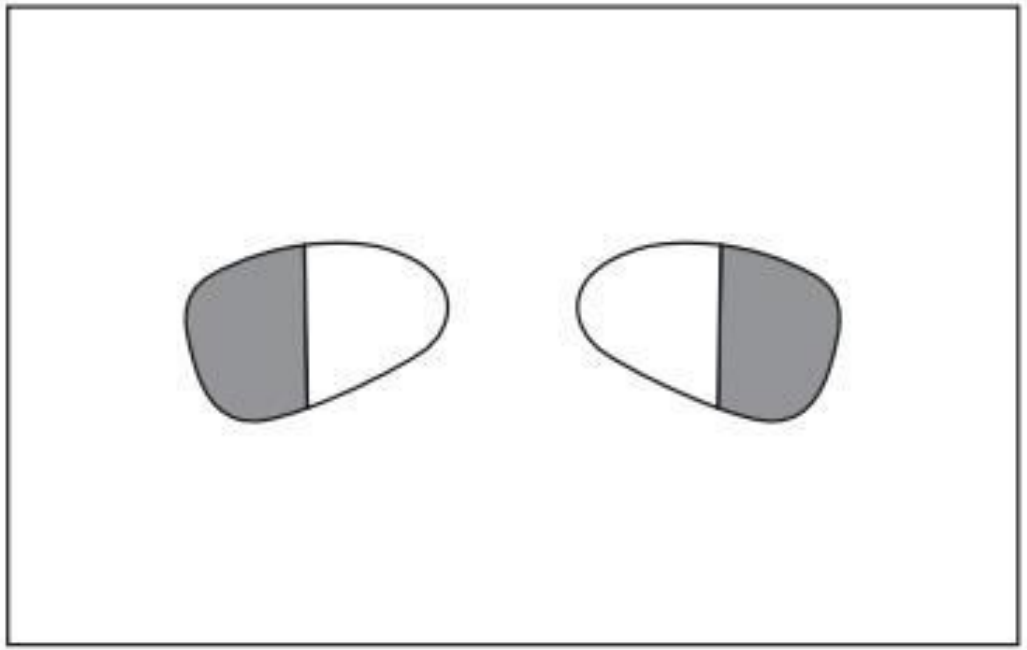
Treatment depends on the type of tumor found; some are amenable to medical therapy but many require surgical excision. A **meningioma and craniopharyngioma** may also **cause chiasmal compression.**

Optic tract, radiation and visual cortex

Lesions of the optic tract and radiation **(usually vascular or neoplastic)**, produce a **homonymous hemianopia field defect** , that is, loss confined to the right - or left - hand side of the field in of both eyes .This pattern of field loss results from the crossing of the fibers representing the nasal retina in the chiasm .Information from these fibers is projected to the primary visual cortex **from** the lateral geniculate body (LGB) **via the optic tract and radiation.** If the extent of field loss is similar in both eyes a congruous defect is said to be present. This usually means that the defect has affected the optic radiation or cerebral cortex. **Neoplasia** more commonly affects the radiation in the anterior temporal lobe. The commonest cause of disease in the occipital cortex is a cerebrovascular accident. The visual loss is of rapid onset; a slower onset is suggestive of a space - occupying lesion.

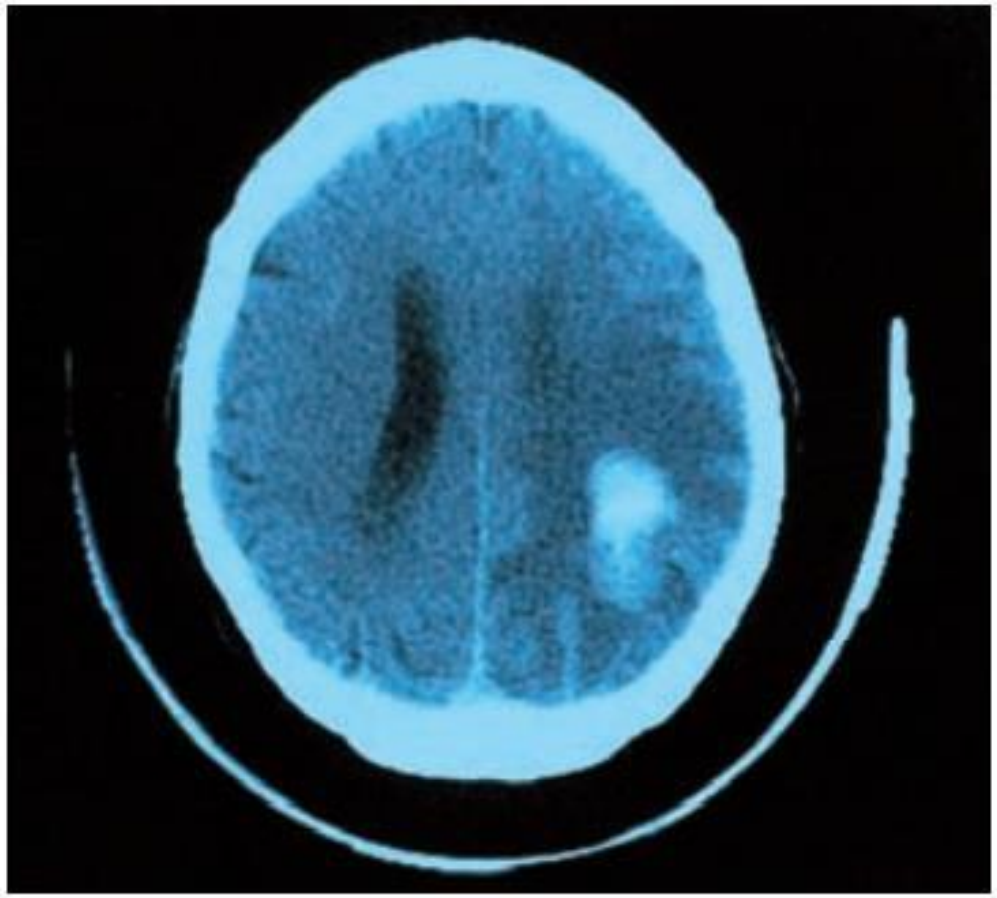


(a)

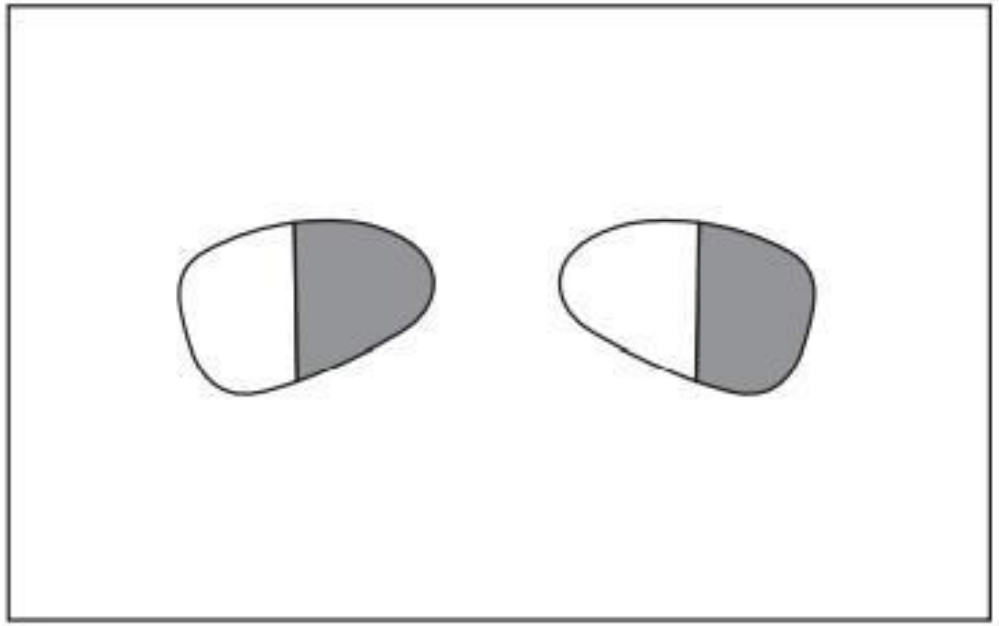


(b)

Figure 14.5 (a) The CT appearance of a pituitary tumour (arrow). (b) The bitemporal visual field loss produced.



(a)



(b)

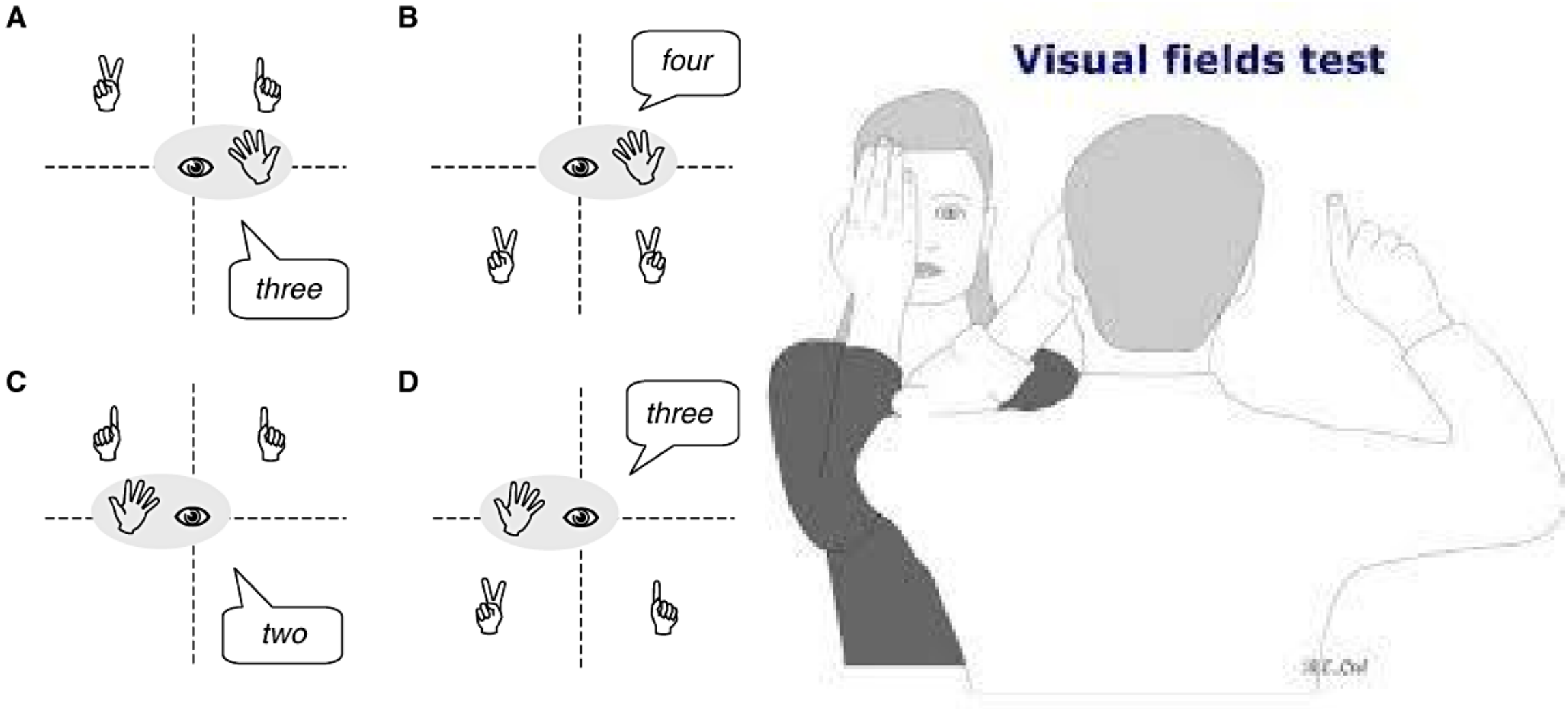
Figure 14.6 (a) A CT scan showing a left cortical infarct. (b) The complete congruous right homonymous hemianopia produced by the infarct.

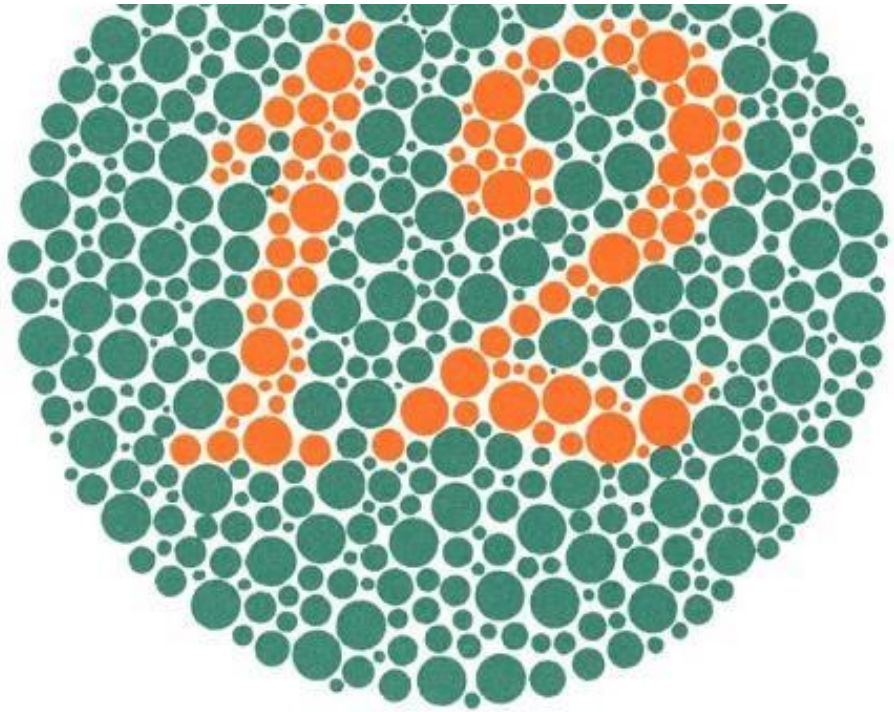
KEY POINTS

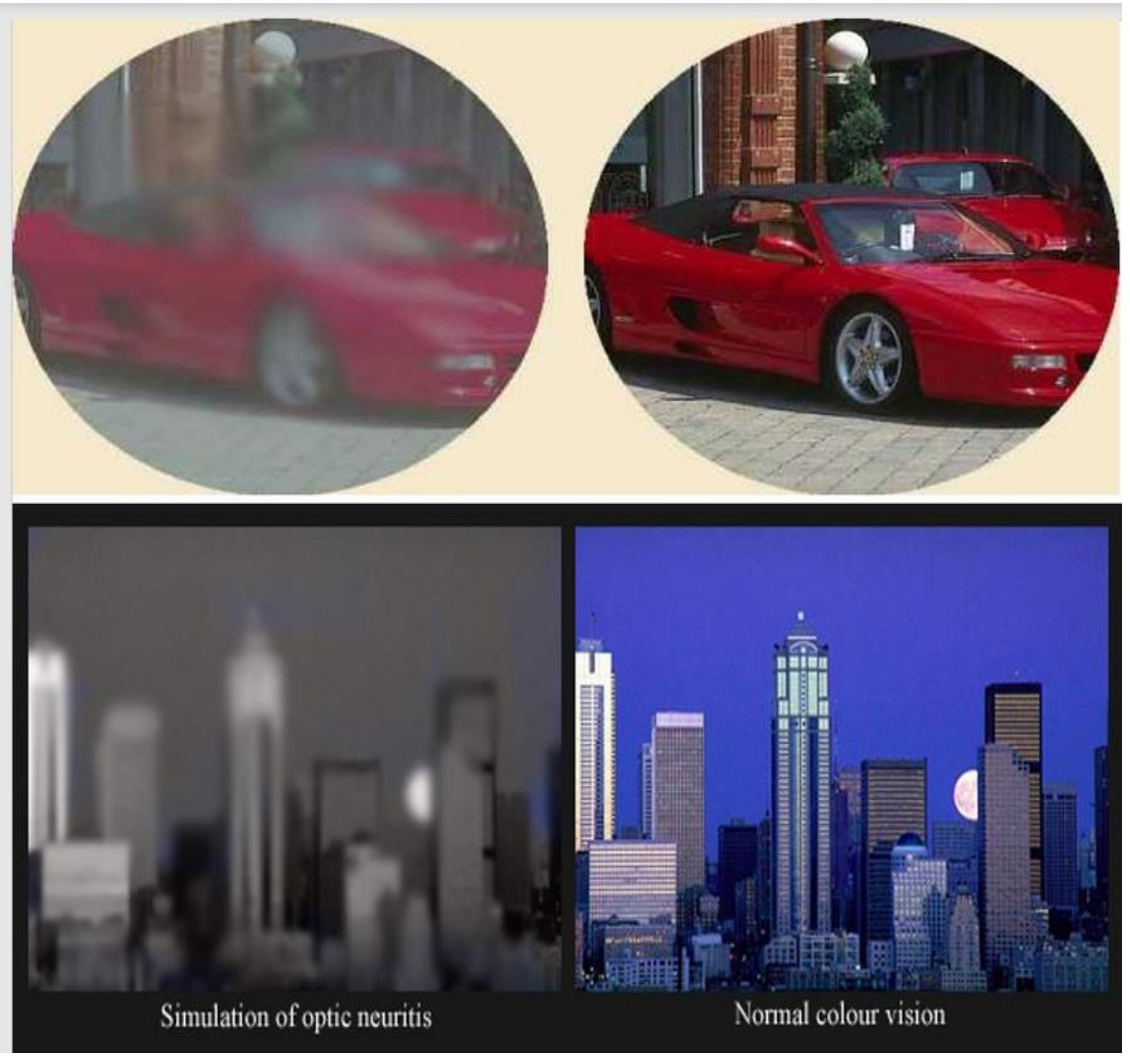
- A bitemporal visual field defect suggests a pituitary lesion.
- There are several causes of a swollen optic disc; it is not just a sign of raised intracranial pressure.
- A pale optic disc may result from retinal disease.

Neurophthalmology examination :

1. Visual acuity (near and distance)
2. Color vision and red desaturation
3. Confrontation field test
4. Pupil test (size , shape , RAPD)
5. Cover test
6. Eye movement
7. Cranial nerves
8. Fundoscopy (optic disc swelling , pale optic nerve , retinal disease)


A
three
B
four
C
two
D
three
Visual fields test




Simulation of optic neuritis
Normal colour vision

Transient visual loss differential diagnosis :

1. Carotid or cardiac emboli
2. Carotid dissection
3. Migraine (usually hemianopia , with zigzag feature)
4. Giant cell arteritis Vertebrobasilar ischemia
5. Intermittent angle-closure glaucoma
6.