Neuro-Ophthalmology

Objectives: Not Given!

[Color index : Important | Notes | Extra]

Resources: Slides+Notes+Lecture notes of ophthalmology + OphthoBook.
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❖ Introduction :

- Neuro-ophthalmology deals with ocular problem caused by disorder of brain, optic nerve, Cranial Nerves and pupil pathway.
- Our eyes simply receive visual information - we actually see with our brain. In turn, the brain controls the position and focus of the eyes, directing our visual attention.
- 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 **converge** and the pupils **constrict** (the near response).
- The pupils are **normally equal** in size but some 20% of people may have noticeably unequal pupils (**anisocoria**) with no associated disease.

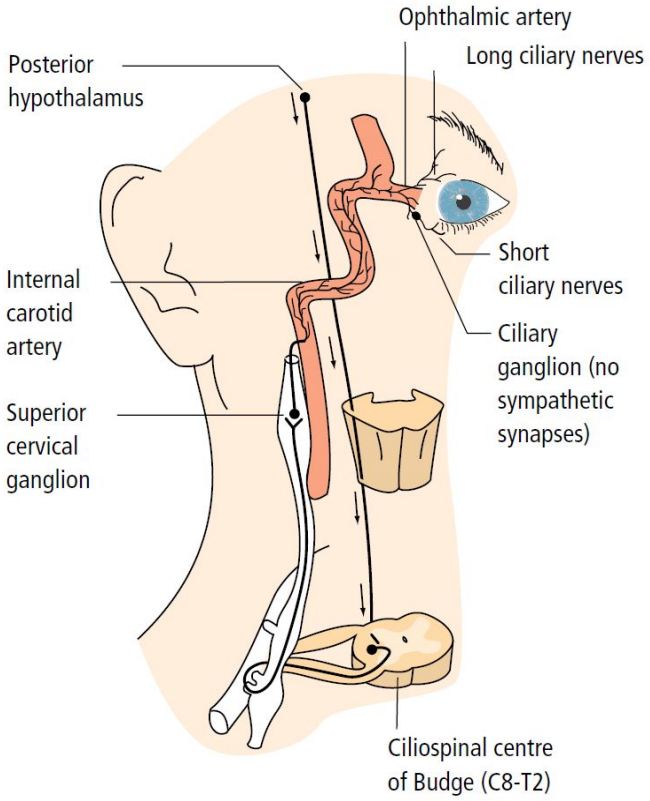
The key to diagnosis of pupillary disorders is to:

- ➢ Determine which pupil is abnormal.
- ➢ Search for associated signs.

Disorders of the pupil may result from:

- ➢ Ocular disease.
- ➢ Disorders of the controlling neural pathway.
- ➢ Pharmacological action.

The parasympathetic fibres reach the eye through the third cranial nerve. The sympathetic pathway is shown in Figure.



Part 1: Pupillary Disorders

Anatomy and physiology:

❖ Pupils:

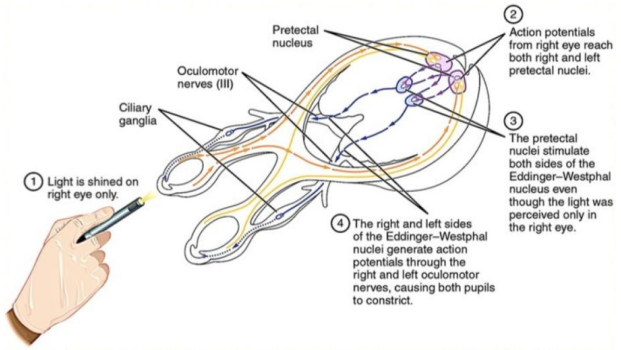
Pupillary control: The physiology behind a "normal" pupillary constriction is a balance between the sympathetic and parasympathetic nervous systems.

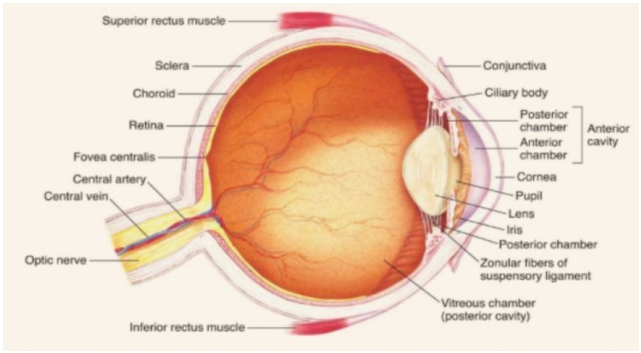
Several diseases of the eye can cause pupil irregularity and alter pupil reactions:

- ➢ Anterior uveitis.
- ➢ Intraocular surgery - Direct muscle injury.
- ➢ Blunt trauma. Rupture the sphincter muscle, causing irregularity or fixed dilation (Traumatic mydriasis).
- ➢ Acute or chronic High IOP - Acute glaucoma.

➢ **Parasympathetic "CHOLINERGIC"** innervation leads to **pupillary constriction**.

- Originate from **Pretectal nucleus** at **midbrain** and stimulate **both Edinger-westphal nucleus.**
- Divided into **superior** and **inferior** division**.**
- **Inferior division** go to **ciliary ganglia** (parasympathetic ganglia) and finally reach to muscle.
- **Sphincter pupillae muscle:** Supplied by **parasympathetic fibers** of **Oculomotor nerve** and lead to: **Constriction of pupil** (Pupil constrict to light and near stimuli).
- In accommodation reflex there is miosis, conversion and lens accommodation.





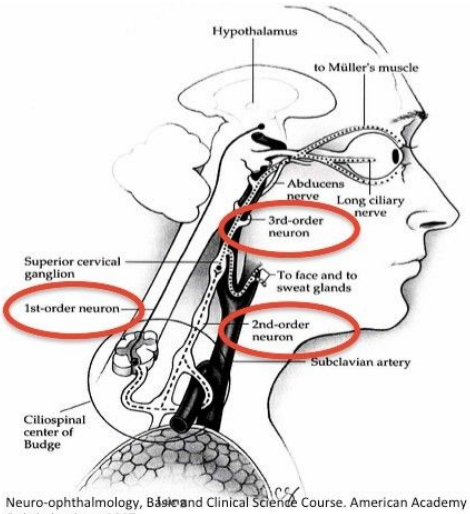
➢ **Sympathetic "Adrenergic"** innervation leads to **pupillary dilation.**

- Originate from **hypothalamus** and go through **superior cervical ganglia**.
- **Dilator pupillae muscle:** Supplied by **Sympathetic fibers** and lead to **Dilation of pupil,** a group of muscles in the **peripheral 2/3** of the **iris.**
- **If there is a cut through sympathetic pathway patient will develop signs of Horner syndrome.**
- **Sympathetic (adrenergic) pathway mediated throw three order neuron.**
- **Important to know that the second order neuron is close to the carotid artery.| Sympathetic innervate Müller muscle, pupillary dilator, lid retractors.**

*The first-order neuron descends from the hypothalamus to the first synapse, which is located in the cervical spinal cord (levels C8-T2, also called ciliospinal nucleus of Budge).

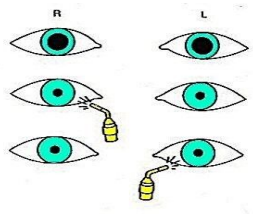
*The second-order neuron travels through the brachial plexus, over the lung apex (some tumor in the lung may damage the sympathetic pathway of the pupil) . It then ascends to the superior cervical ganglion located near the angle of the mandible and the bifurcation of the common carotid artery.

*The third-order neuron then ascends within the adventitia of the internal carotid artery, through the cavernous sinus, where it is in close relation to the sixth cranial nerve. The oculosympathetic pathway then joins the ophthalmic (V1) division of the fifth cranial nerve (trigeminal nerve). In the orbit and the eye, the oculosympathetic fibers innervate the iris dilator muscle as well as Müller's muscle, a small smooth muscle in the eyelids responsible for a minor portion of the upper lid elevation (2-3 mm) and lower lid retraction. (The main eyelid muscle elevator is Levator palpebrae, supplied by 3rd nerve), So the pupil size of is controlled by a balance between parasympathetic innervation to the sphincter muscles and sympathetic innervation of the dilator muscles of the iris.



Neuro-ophthalmology, Basic and Clinical Science Course. American Academy of Ophthalmology, 2007

Examination of the Pupils:



- Best conducted in **dim light room** using a bright light.
- The patient should be relaxed and **fixing on a distant object (to get rid of accommodation because the accommodation cause miosis).**
- The size, shape and position of each pupil should be noted in light and dark condition.
- **Check light reflex:**
    - **Direct pupil reflex:** When focus the light on one eye, **that eye** will constrict (if normal).
    - **Consensual pupil reflex:** When you focus the light in one eye, **the other eye** will constrict.

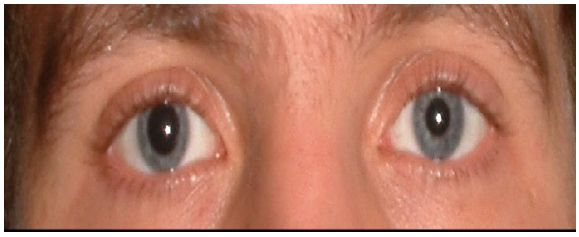
- **Looking for a relative afferent pupillary defect (RAPD):**
    - RAPD is A lesion of the optic nerve on **one side** blocks the **afferent** limb of the pupillary light reflex. The pupils are equal and of normal size, but the pupillary response to light directed to the affected side is reduced, while the **near reflex** is **intact**.
    - Do swinging light reflex **(Marcus gunn reflex)**, both eyes should **always constrict** when you focus the light, **if Dilated** when you focus the light, this is **+RAPD and means there is optic nerve damage.** Check this video

20% of the population has ***simple (Physiologic) Anisocoria***. **Criteria of Physiologic Anisocoria:**

1. Less 1 mm different sizes.
2. Same amount in the dark and light. You don't need to investigate them.

➔ In physiological anisocoria There are no associated findings suggestive of a sympathetic or parasympathetic lesion.

➔ Intermittency or variability is a hallmark.



How to know which one is abnormal? Look to the corneal light reflex (U should evaluate the patient in dim and light).

➔ When the small pupil does not dilate as well as the large pupil in DIM LIGHT, then the small pupil is abnormal.

➔ When the larger pupil does not constrict as well as the small pupil in response to a light stimulus, then the large pupil is abnormal. (This condition called **Anisocoria "unequal pupil size"**)

➔ **Causes of Dilation of pupil:**

- Previous ocular surgery.
- Ocular trauma.
- Use of medication like cycloplegics e.g. **atropine, cyclopentolate.**
- **Third** nerve palsy **(mid dilated fixed pupil, not respond to light).**
- **Tonic pupil (Adie's pup).**

Tonic pupil (Adie's pupil):

- Benign condition.
- Young female / F:M = 2:1, subacute onset.
- 80% **unilateral** dilation of pupil.
- It is due to *ciliary ganglionitis*[1] which denervates the parasympathetic supply to the iris and ciliary body.
- **Systemically the disorder is associated with loss of tendon reflexes. There are no other neurological signs.**

➔ **Physical Examination:** **The consequence is that the pupil:**

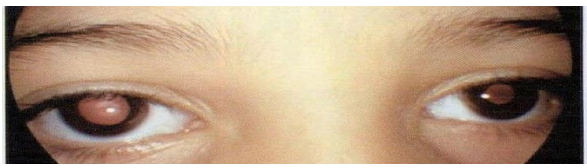
- **Enlarged pupil** – because the sphincter is relatively denervated.
- **Sluggish**, **segmental** pupillary responses to **light**[2]**.**
- Better response to near followed by slow redilation. **Called light near dissociation.**

Due to muscarinic supersensitivity of the sphincter, the pupil also:

- Shows slow, sustained miosis on accommodation.
- Instillation of weak cholinergic agents **(0. 1% pilocarpine)** will cause **constriction of the tonic pupil** (denervation hypersensitivity**) the normal eye won't change.** This is a diagnostic test.

Holmes-Adie syndrome:

- Includes tonic pupil, diminished deep tendon reflexes and orthostatic hypotension.



- The pupil will constrict with near vision, but very slowly. That's why we call it a "tonic pupil" - it's tonically slow.
- The parasympathetic pathway is much shorter than the convoluted sympathetic pathway, so potential causes for damage are more benign. The parasympathetic plexus sits right behind the eye and can be damaged after an otherwise benign viral infection.
- It is a diagnosis of exclusion. Patient most of the time will be asymptomatic. However they might complain of photophobia because of dilated pupil. Sometimes they will have abnormality of accommodation in near vision. We can provide them pilocarpine will release the photophobia and help them with the accommodation It takes few months and the pupil will go back and constrict.

[1] On recovery from the ganglionitis, reinnervation is incomplete and the partially denervated receptors of the iris and ciliary body become super sensitive to muscarinic stimulation.
[2] Poorly reactive to light – because few of the innervating fibres 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.

➔ Causes of Constriction of pupil:

- Previous ocular surgery
- Ocular trauma or inflammation
- ☆ IN the picture: The margin oh the pupil (iris) is attached to the lens (posterior synechiae) or to the cornea (anterior synechiae) (adhesion). This prevent pupil dilation.
- Use of medication e.g. **pilocarpine**
- **Horner syndrome.**

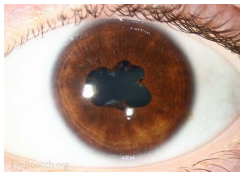


Table 13.1 Drugs having a pharmacological effect on the pupil.

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

Horner syndrome:

- **Cause:** interruption of sympathetic pathway **(it can be Carotid dissection, carotid aneurysm and tumor)!!**
- **Horner's syndrome may also be congenital, in which case the iris colour may be altered when compared to the fellow eye (heterochromia).**

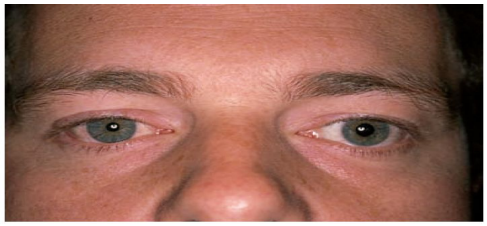
- **Signs:** at the side that affected you will see **miosis – anhydrosis – ptosis – enophthalmos**. In general no other symptoms.
- **Miosis** : due to loss of dilator function[3].
- **Anhidrosis**: lack of sweating[4].
- **Enophthalmos** (posterior displacement of the eyeball): due to paralysis of levator palpebrae muscle.
- **Ptosis:** due to paralysis of muller's muscle.

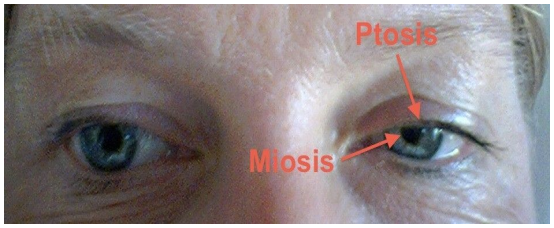
The sympathetic pathway may be affected by multitude of pathologies like:

- Syringomyelia, expanding cavity within the spinal cord.
- Small cell carcinoma at the lung apex. Involvement of the brachial plexus cause shoulder and arm pain and to T1 wasting of the small muscles of the hand (Pancoast's syndrome).
- Neck injury, disease or surgery.
- Cavernous sinus disease - catching the sympathetic carotid plexus in the sinus.

Do we need to image the patient urgently or give him the next available appointment?

1. **Acute or chronic:**
   - **Acute within 2 weeks: immediate neuroimaging.**
   - **Chronic within several months or he has a surgery: follow up.**
2. **Painful or painless:**
   - **Painful: immediate neuroimaging. (Sometime carotid dissection presents with painful Horner syndrome)**





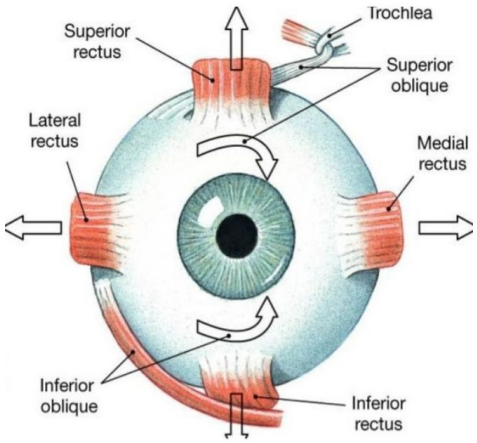
[3] This is more noticeable in the dark, because the normal pupil of the fellow eye then dilates more than that of the affected pupil.

[4] If the sympathetic pathway is affected proximal to the base of the skull. This catches fibers travelling with the branches of the external carotid, which innervate the skin of the face.

Part 2: Neuro Motility Disorders:

❖ **Extraocular Muscles:** There are **six voluntary muscles** that run from the posterior wall of the orbital cavity to the eyeball. These are:

★ **4 recti muscles:**



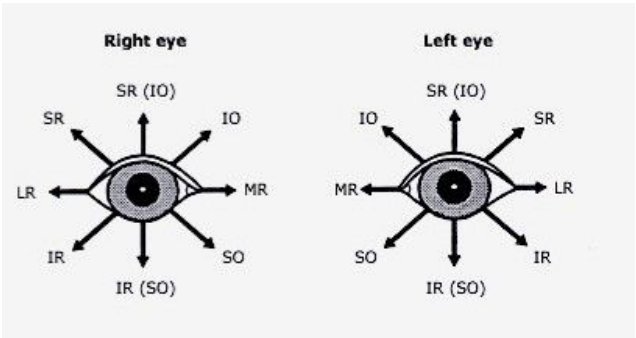
- **Superior rectus** acts as the primary **elevator**
- **Inferior rectus** acts as the primary **depressor** of the eye.
- **Medial rectus** muscle is the primary **adductor** of the eye
- **Lateral rectus** muscle is the primary **abductor** of the eye.
  - ➢ **Medial and lateral rectus** muscles have only **horizontal actions.**
  - ➢ Superior and inferior rectus muscles are the primary **vertical** movers of the eye.

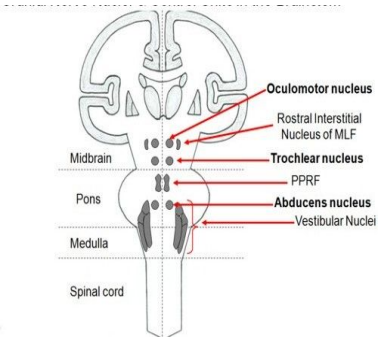
★ **2 Oblique muscles: Superior and Inferior oblique muscles.**

- ❑ This vertical action is greatest with the eye in the abducted position.
- ❑ The **secondary action** of vertical rectus muscles is **torsion**. The **superior rectus** is an **incyclotorter** (inwards rotator), and the **inferior rectus** is an **excyclotorter** (outwards rotator). The tertiary action of both muscles is adduction.
- ❑ **4th Trochlear nerve:** supplies **the superior oblique muscle**.
- ❑ **6th Abducent nerve:** supplies **the lateral rectus muscle**.
- ❑ **3rd Oculomotor nerve:** begins as a nucleus in the midbrain that consists of several subnuclei that innervates the individual **extraocular muscles, the eyelids, and the pupils.** It supplies the superior, **inferior and medial rectus muscles and the inferior oblique muscle.**

Cranial Nerve III			
Superior Rectus	**Inferior Rectus**	**Medial Rectus**	**Inferior Oblique**
Elevation (maximal on lateral gaze)	**Depression (maximal on lateral gaze)**	**Adduction**	**Excyclotorsion (Away from the nose)**
Cranial Nerve IV		**Cranial Nerve VI**	
Superior Oblique		**Lateral Rectus**	
Incyclotorsion (Toward the nose)		**Abduction**	

Oculomotor and trochlear nerves exit at the midbrain, while Abducent from pons.





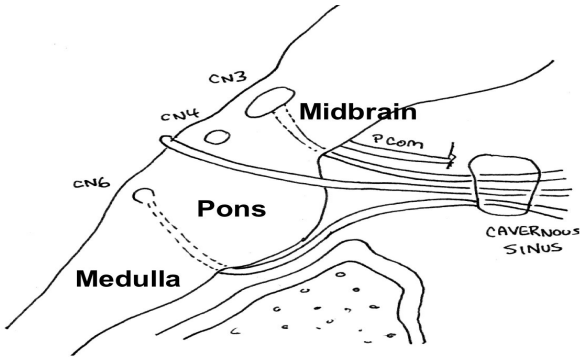
Yoke muscles are the primary muscles in each eye that accomplish a given version (eg, for right gaze, the right lateral rectus and left medial rectus muscles). Each extraocular muscle has a yoke muscle in the opposite eye to accomplish versions into each gaze position.

Muscle*	Primary	Secondary	Tertiary
Medial rectus	Adduction	—	—
Lateral rectus	Abduction	—	—
Inferior rectus	Depression	Excycloduction	Adduction
Superior rectus	Elevation	Incycloduction	Adduction
Inferior oblique	Excycloduction	Elevation	Abduction
Superior oblique	Incycloduction	Depression	Abduction

*The superior muscles are incycloductors; the inferior muscles, excycloductors. The vertical rectus muscles are adductors; the oblique muscles, abductors.

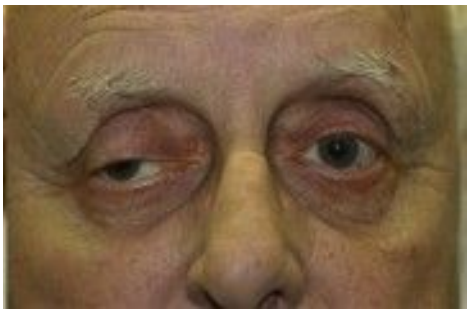
❖ Quick introduction to Isolated nerve palsy: "u can skip it if u want"

- **Pathogenesis**:
  - Disease of the third, fourth and sixth nerves and their central connections gives rise to a paralytic strabismus.
  - Each nerve may be affected at any point along its course from brainstem nucleus to orbit.
- **History and examination:**
  - The patient complains of diplopia. There may be an abnormal head posture to compensate for the inability of the eye to move in a particular direction.
  - *A sixth nerve palsy results in failure of abduction of the eye.*
  - *A fourth nerve palsy results in:*
    - Defective depression of the eye when attempted in adduction.
    - It produces the least noticeable eye - movement abnormality.
    - Patients may notice vertical double vision with some torsion of the image, particularly when going downstairs or reading.
  - *A third nerve palsy results in:*
    - Failure of adduction, elevation and depression of the eye.
    - Ptosis.
    - In some cases, a dilated pupil due to involvement of the autonomic fibres.



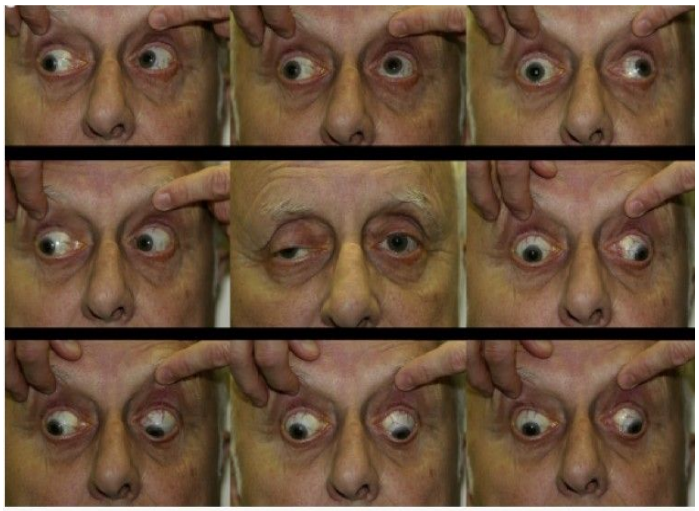
3rd CN (oculomotor) palsy:

- **65 yrs** old presented to **ER** complaining **of double vision.**
  - **The patient presentation: the eye is deviated down and out, ptosis, pupillary dilatation and paralysis of accommodation.**



- Begins as a nucleus in the midbrain that consists of several subnuclei that innervate the individual extraocular muscles, the eyelids, and the pupils.
- Third cranial nerve palsy is caused by a lesion of the oculomotor **nucleus** within the midbrain or by compression of the **peripheral** course of the nerve by aneurysm or tumour.
- It leads to **drooping of the eyelid (ptosis), dilatation of the pupil that is unresponsive to light and accommodation, and Inability to move the eyeball upwards, downwards or inwards "adduction". Patient will come with horizontal diplopia.**

➔ Physical Examination:



Neurosurg Clin N Am 23 (2012) 607–619

This patient have right 3rd nerve palsy. How did we know?
He can **abduct his right eye** only, which is **lateral rectus muscle function**

➔ **If you want to rule out 4th cranial nerve palsy along 3rd nerve palsy what will you do?**

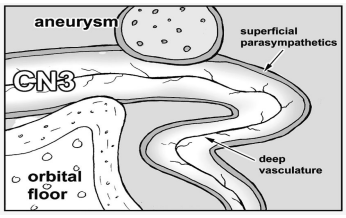
Ask the patient to look **down**, if the eye **intorted** the **4th cranial nerve is intact**

Check for pupil involvement?

Absence of pupillary involvement suggests a benign process that can be observed over a couple of weeks. **A fixed, dilated pupil** requires extensive neurologic evaluation.

➔ **What is the best investigation for PCA aneurysm? Magnetic resonance angiography.**

Etiology of Third cranial nerve (oculomotor)palsy:



- Microvascular ischemia (DM and HTN).
- Intracranial aneurysm (posterior communicating artery).
- Trauma.
- Brain tumor.

➔ Medical 3rd nerve palsy:

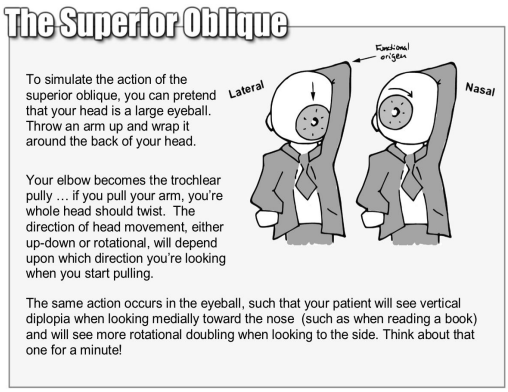
- **Isolate motor** part damaged due to Vascular diseases such as **diabetic and hypertension.**

➔ Surgical 3rd nerve palsy:

- Pressure on pupil constrictor fibers of CN III due to tumor or **Posterior communicating artery aneurysm (most common cause)** and Internal carotid artery **lead to Unilateral dilated pupil.** Compressive lesions usually affect the parasympathetic nerve component: a blown pupil is a potential emergency. Whenever you have pupillary involvement "is a crucial diagnostic sign", you need an **MRI and angiography to rule out a dangerous aneurysm or tumour. 'dangerous' (PCA located in circle of Willis).**

4th CN (Trochlear) palsy:

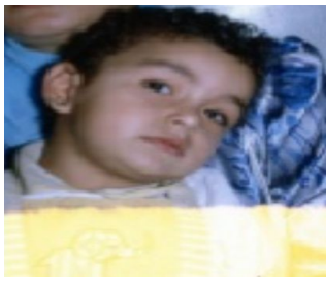
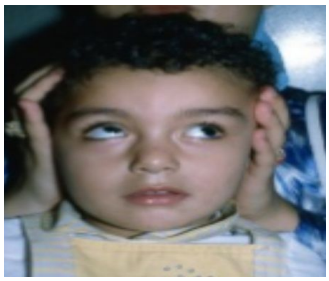
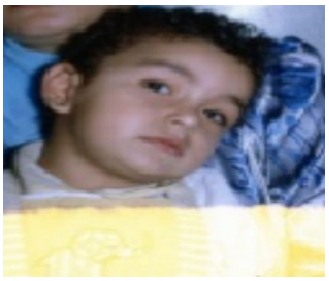
- These patients have an **upward deviation of the affected eye and a "cyclotorsion" twisting of the eye that makes them tilt their head away to the opposite shoulder and vertical diplopia. Also the Patients have difficulty in down gaze.**



- The fourth cranial nerve is the skinniest nerve and runs the longest distance inside the cranial vault. This long passage makes it more susceptible to injury and neoplasm. The fourth nerve is also susceptible to being pulled from the root where it exits from the back of the brainstem. Trochlear paralysis is the hardest cranial nerve palsy to diagnose.

➔ Etiology:

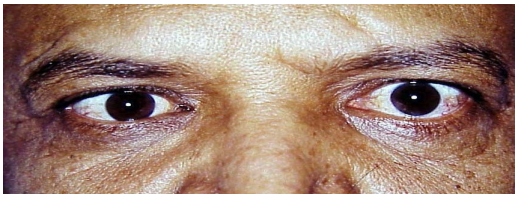
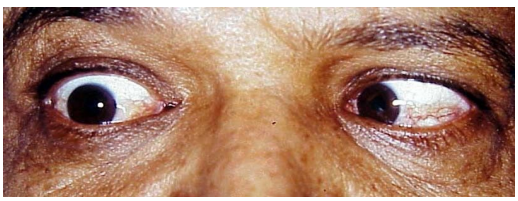
- Trauma.
- Idiopathic.
- Congenital. **More fourth palsies occur in elderly males from trauma and more congenital palsies are found in the pediatric population.**
- 1/3rd Trauma | 1/3rd Congenital | 1/3rd Ischemic (diabetic) | 1/3rd Tumor.

★ This kid is smart, he is trying to prevent double vision by tilting his head.

6th CN "Abducens" palsy:

- A sixth cranial nerve palsy is caused by a lesion of the abducens **nucleus** in the pons or by compression of the **peripheral course** of the nerve by an aneurysm or tumour.
- It leads to **inability to move the eye outwards "abduction". False -ve sign ? 6th due to ^ IOP you won't know exactly where is the lesion so false localizing sign.**

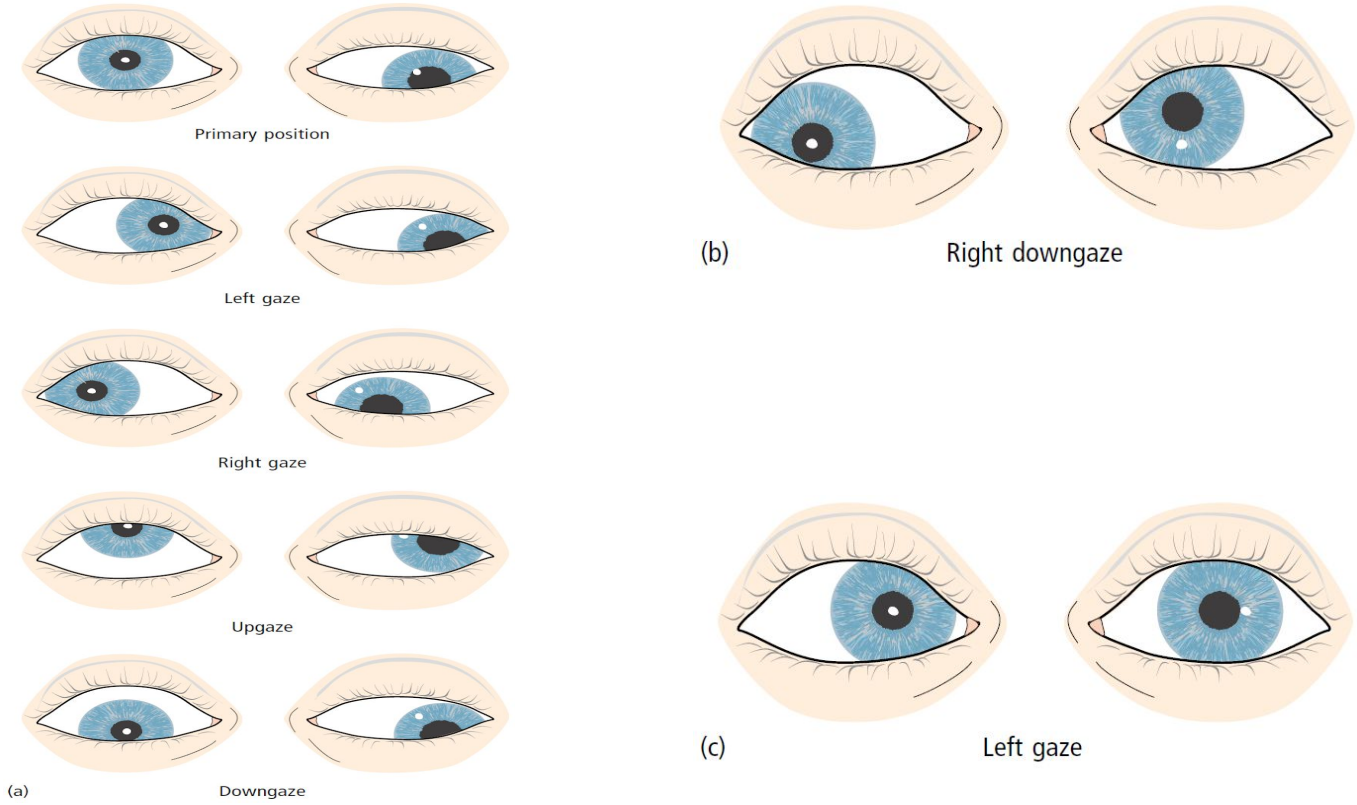
❖ **Horizontal diplopia (worse at distance)** Why? Conversion in near you don't need lateral rectus meanwhile in distance it will get worse.

- Esotropia (crossed eye).
- Face turn in the direction of the paralyzed muscle.
- Limited Abduction on the side of the lesion.

➔ Causes:

- Intracranial tumors
- Trauma **(most common cause because it's long nerve)**
- Microvascular diseases **(mostly DM)**
- Increased intracranial pressure[5] **(we call it false localizing lesion, because high pressure press on Dorello canal "a canal surrounding sixth cranial nerve")**

[5] Where the nerve enters the cavernous sinus, it makes an abrupt 90-degree bend. Something about this abrupt turn makes the 6th nerve especially susceptible to high intracranial pressure.



Figures description:

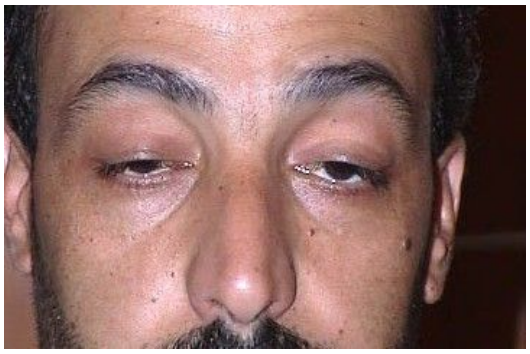
A. Left third nerve palsy: note the dilated pupil and ptosis as well as the limitation of eye movement.
B. Left fourth nerve palsy: the defect is maximal when the patient tries to look down when the left eye is adducted.
C. Sixth nerve palsy: the left eye is unable to abduct.

Table 15.1 The causes of isolated nerve palsies.

Orbital disease	e.g. neoplasia
Vascular disease	Diabetes (a 'pupil sparing' third nerve palsy, i.e. there is ptosis and extraocular muscle palsy but no mydriasis)
	Hypertension
	Aneurysm (most commonly a painful third nerve palsy from an aneurysm of the posterior communicating artery. Mydriasis is usually present)
	Caroticocavernous sinus fistula (also causes myogenic palsy)
	Cavernous sinus thrombosis
Trauma	Most common cause of fourth and sixth nerve palsy
Neoplasia	Meningioma
	Acoustic neuroma
	Glioma
Raised intracranial pressure	May cause a third or sixth nerve palsy (a false localizing sign)
Inflammation	Sarcoidosis
	Vasculitic (i.e. giant cell arteritis)
	Infection (particularly herpes zoster)
	Guillain–Barré syndrome

Part 3: Neuromuscular Disorders:

Ocular Myasthenia Gravis (OMG):



- Chronic autoimmune disease affecting the **neuromuscular junction** in skeletal muscles (nicotinic acetylcholine receptors).
- **History:** Patient is not able to stand from his bed at morning after sleeping due to muscle weakness Or he feel fatigue at the end of the day.
- Ask the patient is your double vision or ptosis worse early morning or at the end of the day? Have you notice that the double vision worse at evening?
- **Signs: Ptosis** (due exhaustion of muscle NOT due to paralysis) – **Diplopia** – **pupil is normal –** painless condition - Fatigability and variability of clinical findings are characteristic. The diplopia and ptosis is usually worse on prolonged upgaze: you can test this by having your patient look at your raised finger to see who tires out first.

- **Investigations:**

1. **Tensilon test:** inhibits acetylcholinesterase and can transiently reverse signs of weakness due to OMG, such as ptosis and extraocular muscle paresis. **(Where you give edrophonium chloride (an anticholinesterase) and look for an improvement in symptoms as their Ach levels build up).**
2. **Check for systemic weakness, difficulty in swallowing or breathing**. **you have to ask about generalized symptoms because pt can convert to systemic**
3. **Assess orbicularis strength: Ask the patient to close his eye strongly and open them**
4. **Blood test for: acetylcholine receptor antibodies (50% present in OMG)**

- Ocular myasthenia gravis patients might present with ptosis or diplopia or both. We have to ask if the symptoms change during the day (better in the morning or at night) and whether the diplopia is stable or not (horizontal or vertical).

- Assessing the orbicularis strength: by asking the patient to close both eyes strongly then we try to open them.

- Ach receptor antibodies in general myasthenia gravis = 60-80%, it is less in ocular MG.

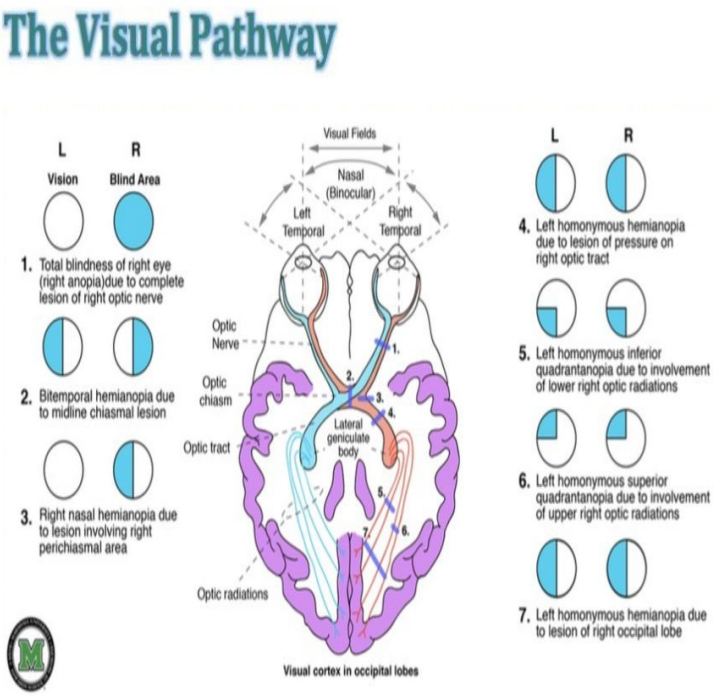
- Tensilon test is diagnostic.

- Other tests for ocular MG like ice test: ask the patient to put an ice pad over the ptosis for two minutes and then check for any improvement (measure the degree of ptosis before and after).

- Sleep test: measure the degree of ptosis then ask the patient to sleep and re-measure after the patient awakes. (improvement = positive test)

- The pupils are not affected.

Part 4: Visual Pathway Disorders:



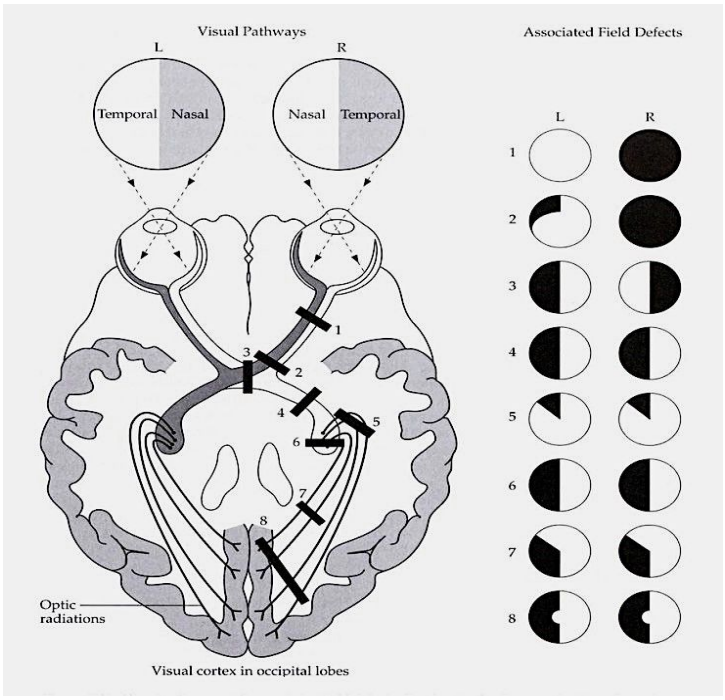
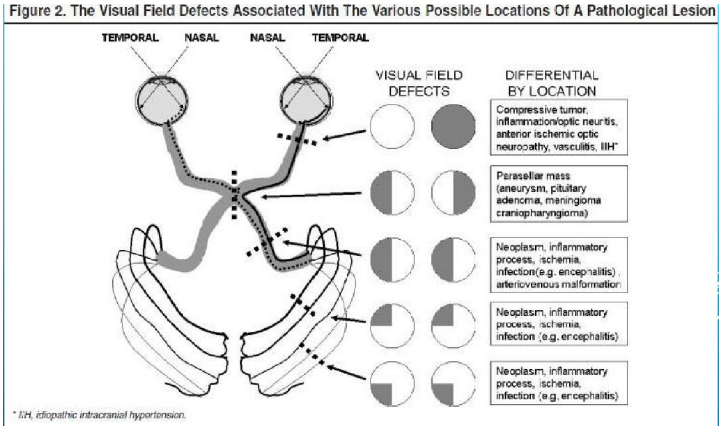


Figure 2. The Visual Field Defects Associated With The Various Possible Locations Of A Pathological Lesion



Lesions anywhere in the visual pathway will produce visual field defect:

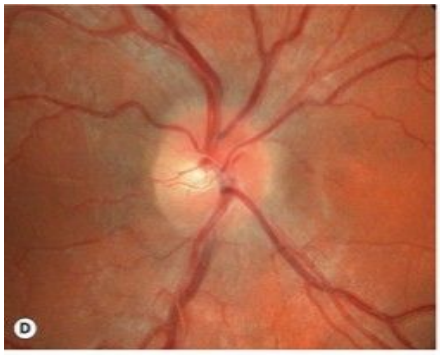
- Number 5: pie in the sky (temporal lobe is affected)
- Number 7: pie in the floor (parietal lobe is affected)
- Number 8: there is macular sparing (occipital lobe is affected) (Macular sparing because it has two blood supply).

- Visual field defects: if unilateral then think about optic nerve pathology, if bilateral then the pathology is at the optic chiasm or beyond.
- Chiasmal defects are always bitemporal.
- Homonymous visual defects could be due to stroke or tumors.
- **Left eye blindness** due to Left optic nerve damage
- **Binasal hemianopia** due to bilateral carotid artery aneurysm compressed optic chiasm
- **Bitemporal hemianopia** due to pituitary tumor compressing optic chiasm
- **Right Homonymous hemianopia** due to Left optic tract damage
- **Right superior quadrantic hemianopia** due to Left optic radiation at temporal lobe lesion (pie in the sky)
- **Right inferior quadrantic hemianopia** due to Left optic radiation at parietal lesion (pie in the floor)

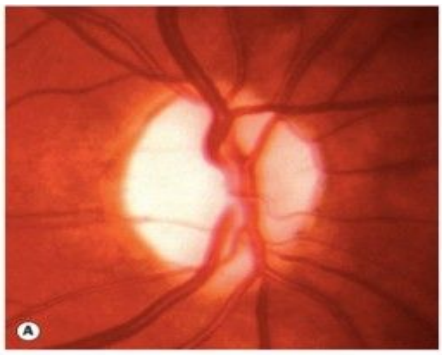
Optic Nerve Disease:

- ➢ Usually unilateral.
- ➢ Afferent pupillary defect.
- ➢ Central visual loss.
- ➢ Loss of color vision.
- ➢ Optic disc edema.
- ➢ Optic atrophy.

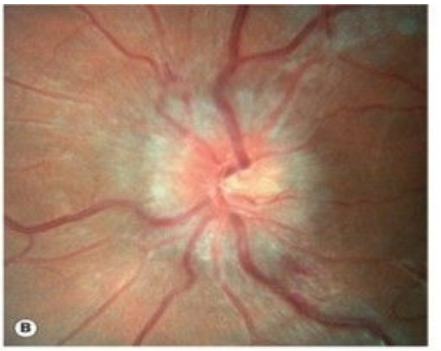
- The normal optic nerve head has distinct margins, a pinkish rim and, usually, a central, pale, cup.
- The central retinal artery and vein enter the globe slightly nasally in the optic nerve head, referred ophthalmoscopically as the **optic disc.**
- **Optic neuropathy is usually unilateral (if bilateral this is papilledema), afferent pupillary defect, central visual loss, loss of colour vision, optic disc edema or atrophy.**



Normal Optic disc



Pale disc (optic atrophy)



Disc edema

How to assess the optic nerve in the clinic: visual acuity, visual field, color test and afferent pupillary defect

1. Optic Neuritis:

- Inflammatory demyelinating condition associated with Multiple sclerosis.
- Most common type in female young adults.
- 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.

- **History:**
  - Patient will come with sudden visual loss/ visual field loss/ color vision loss[6].
  - Ocular pain while moving the eye[7]. **Why ocular pain happened?** Because **optic nerve sheath is attached to medial rectus muscle sheath.**
  - Recent history of viral illness in some cases. Present In 40-70% of MS patients.

Relative Afferent Pupillary Defect (RAPD, Marcus Gunn Pupil) [1]:

An RAPD is a defect in the direct response. It is due to damage in optic nerve or severe retinal disease.

It is important to be able to differentiate whether a patient is complaining of decreased vision from an ocular problem such as cataract or from a defect of the optic nerve. If an optic nerve lesion is present, the affected pupil will not constrict to light when light is shone in the that pupil during the swinging flashlight test. However, it will constrict if light is shone in the other eye (consensual response). The swinging flashlight test is helpful in separating these two etiologies as only patients with optic nerve damage will have a positive RAPD.

[6] May progress over a few days and then slowly improve.
[7] In retrobulbar neuritis because rectus muscle contraction pulls on the optic nerve sheath.

- **Signs:**
  - Reduce visual acuity and color vision.
  - Positive afferent pupillary defect.
  - Optic disc edema.
  - Pain with eye movement **(optic nerve sheath is in close association with ocular muscle and because it's inflamed any movement will cause pain).**
  - **Scotoma visual field defect.**
  - **Normal disc in retrobulbar neuritis; a swollen disc in papillitis.**
- **Treatment:** IV steroids my speed up the recovery process but does not influence the final outcome.
  - **(You can't give only oral, because there is high chance of recurrence.)**
  - **A patient with optic neuritis needs an MRI of the brain and orbits to look for enhancing lesions. -Oral steroids if given alone might increase the risk of recurrence.**
- **Good recovery and good prognosis[8].**

2. Ischemic Optic Neuropathy:

➔ 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, which result in an ***anterior ischemic optic neuropathy.***

➔ Ischaemic optic neuropathy is the usual cause of blindness in the disease.

A. Non-arteritic IOP (more common):

- **History**: Old patient known to have **DM and HTN and other vascular disorders** come with sudden visual loss.
- **Signs**: Optic disc edema and **Altitudinal (either upper or lower field) visual field loss.** Typically an absence of the lower or upper half of the visual field (**altitudinal scotoma**).

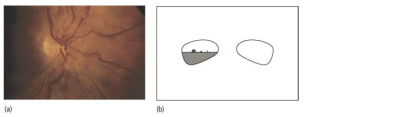


Figure 14.3 (a) The clinical appearance of the optic disc and (b) one form of field defect (altitudinal) seen in ischaemic optic neuropathy.

B. Arteritic IOP (less common):

- Seen in patient older than 55 years and mostly associated with **giant cell arteritis (GCA)** "An autoimmune vasculitis"[9][10]
- **They present with sudden severe loss of vision often on waking, since vascular perfusion of the eye is decreased during sleep, scalp tenderness** "e.g. on combing" **, headache, pain on chewing "jaw claudication", proximal myalgia and arthralgia.**

➔ **Signs:**

- Reduction in visual acuity.
- Field defect.
- Swollen and hemorrhagic disc with normal retina and retinal vessels[11] – **in AIOP the disc is swollen and very pale. (unlike NAIOP where the disk is swollen but it is not pale)**
- Tender temporal artery, suggestive of giant cell arteritis.
- Binocular involvement occurs in third of cases, often within the first 2 days.
- Small normal fellow disc with a small cup in non arteritic disease.

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[8] 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, a decline in vision and a persistent scotoma.

[9] It affects arteries with an internal elastic lamina, which therefore includes the ophthalmic artery, but NOT the retinal artery.

[10] GCA can also present as a central retinal artery occlusion when the vessel is affected secondarily to arteritis of the ophthalmic artery.

[11] (Remember the blood supply to the anterior optic nerve and retina are different)

➔ **Investigation:**

1. If giant cell arteritis is present, **the erythrocyte sedimentation rate (ESR) and C- reactive protein (CRP) are usually grossly elevated. Platelets may also be raised.**
2. If NAIOP; do CBC to exclude anemia, check BP and blood sugar. "Both hypertension and diabetes may be associated with the condition".
3. **Temporal artery biopsy is the gold standard for diagnosis.** But again mat not lead to a diagnosis. **(it should be long enough because it has skipping lesion criteria (2.5 length)**

➔ **Treatment:**

- **Systemic steroids should be given immediately if GCA is suspected.** Steroids will not reverse the visual loss but can prevent the fellow eye being affected.
- There is no treatment for non-arteritic ischaemic optic neuropathy.

3. Congenital Disc Elevation: <1%

➢ It is a rare disease. Optic disc margins are blurred and the cup is absent but no edema or haemorrhage can be observed. May be associated with hyperopia "farsightedness" or drusen "yellow deposits under the retina made up of fatty proteins". B-scan ultrasound can discover drusen (lipid collections)

★ **Other causes of optic neuropathy:**

- Infection e.g. viruses, TB, Cryptococcus and syphilis.
- Systemic connective tissue diseases e.g. SLE.
- Genetics: Leber's optic neuropathy[12].
- Toxic and nutritional deficiencies.
- Trauma.

Optic Nerve Disease:

➢ It is a bilateral swelling of the optic disc secondary to **raised intracranial pressure.**

➢ Could be caused by intracranial mass, severe systemic hypertension, or idiopathic intracranial hypertension (pseudotumor cerebri): **Female – Obesity – Tetracycline - OCPs.**

➢ **It is an Emergency, they require imaging to rule out space equipping lesion.**

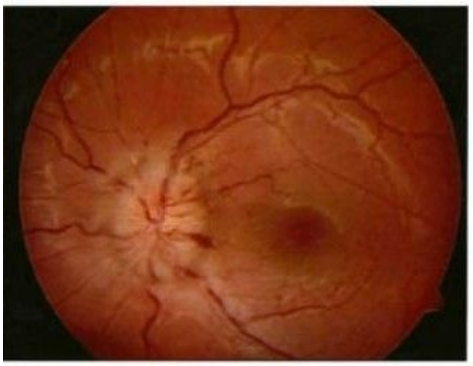
➔ **Symptoms:**

- Headache, worse on awakening and made worse by coughing.
- Nausea and vomiting if the raise in ICP is severe, may be followed by loss of consciousness, pupillary dilatation and death.
- Pulsatile tinnitus.
- Visual symptoms often are absent[13].
- Diplopia (Double vision), due to 6th nerve palsy.

[12] It is mitochondrial DNA mutation. Usually they present in young males with sudden drop of vision starts with one eye then goes to the other.

[13] In the short term there is no visual loss. However, in some patients with advanced papilloedema, a fleeting visual loss may occur, lasting seconds, when posture is altered from lying to standing (*obscurations of vision*).

➔ **Signs:**



- Disc hyperemia.
- Tortuosity of the veins and capillaries.
- Blurring and elevation of the disc margins.
- Peripapillary flame shaped haemorrhage.
- 6th nerve palsy.
- A large blind spots, due to the swelling nerve head.
- There is no spontaneous venous pulsation of the central retinal vein[14].If venous pulsations can be visualized, the cerebrospinal fluid pressure is typically less than 200 mm of water. If there is spontaneous venous pulsation: no increase in IOB, If it is absent: could be increased or normal Found in 70-80% of normal individuals.

➔ **DDx:**

- Adult optic neuritis.
- Hypertension.
- Idiopathic intracranial hypertension.
- Pseudopapilledema. Some normal optic nerve heads appear to be swollen, due a crowding of nerve fibres entering the disc. This is termed pseudopapilledema and occurs particularly in small, hypermetropic eyes where the nerve entry site is reduced in size.

➔ **Causes:**

- Intracranial mass.
- Severe systemic Hypertension.
- Idiopathic intracranial hypertension (pseudotumor cerebri).

- Papillitis: edematous or inflamed optic dist.
- Important signs in optic nerve disease: blurring of the margins, splinter hemorrhage in the peripapillary area, edema and elevation of the disc.
- The presence of hemorrhage = acute raise in the pressure.
- Papilledema is a diagnosis of exclusion should be confirmed by lumbar puncture.

Investigations	Treatments	
	Medical	**Surgical**
• CT or MRI followed by lumbar puncture **(to measure the ICP and rule out meningitis.)** • B-scan ultrasonography to rule out buried disc drusen. • Fluorescein angiography.	Diamox, diuretics.	• Optic nerve fenestration: slit cut of optic nerve sheath > fluid will come out and release the compression • Shunt: for patient who has severe headache and blurred vision

[14] This has a physiological basis. The central retinal vein is exposed to CSF in the subarachnoid space of the optic nerve, as it leaves 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 caused by arterial inflow during systole. When the CSF pressure is higher than the ocular pressure, as in papilloedema, the pressure in the veins at the disc rises above the ocular pressure and spontaneous venous pulsation is lost.