Neuro-ophthalmology review

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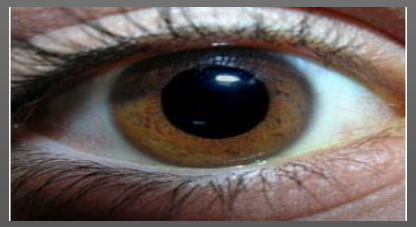
Neuro-ophthalmology deals with visual problems caused by disorders of the brain or the optic nerve connection

Part 1:

Pupillary Disorders



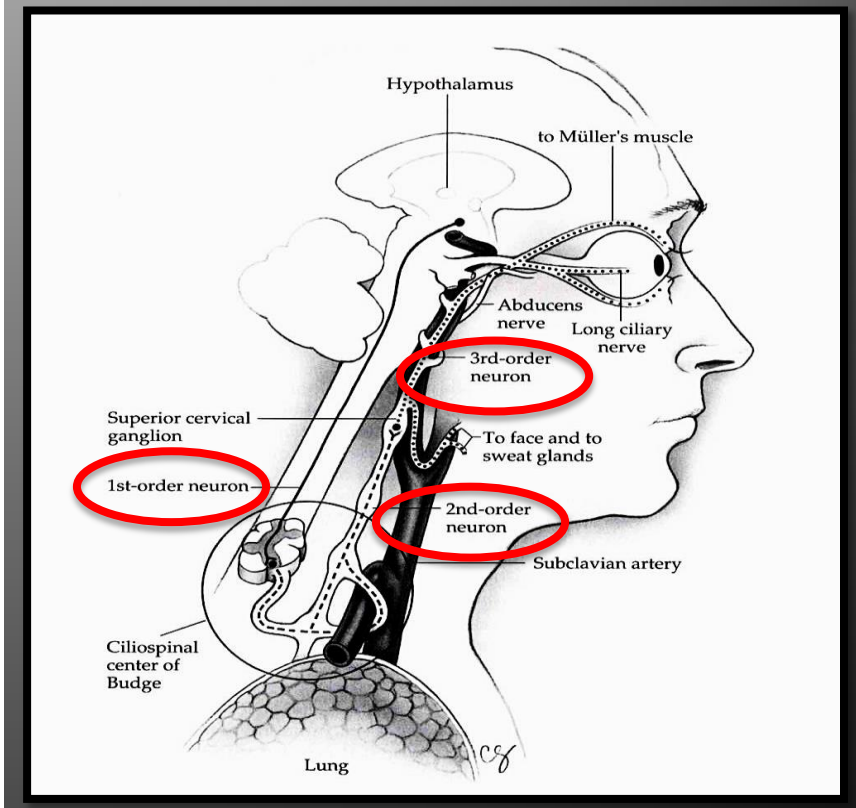
The anatomy and physiology of the pupil

- 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.

- Pupil constrict to light and near stimuli.

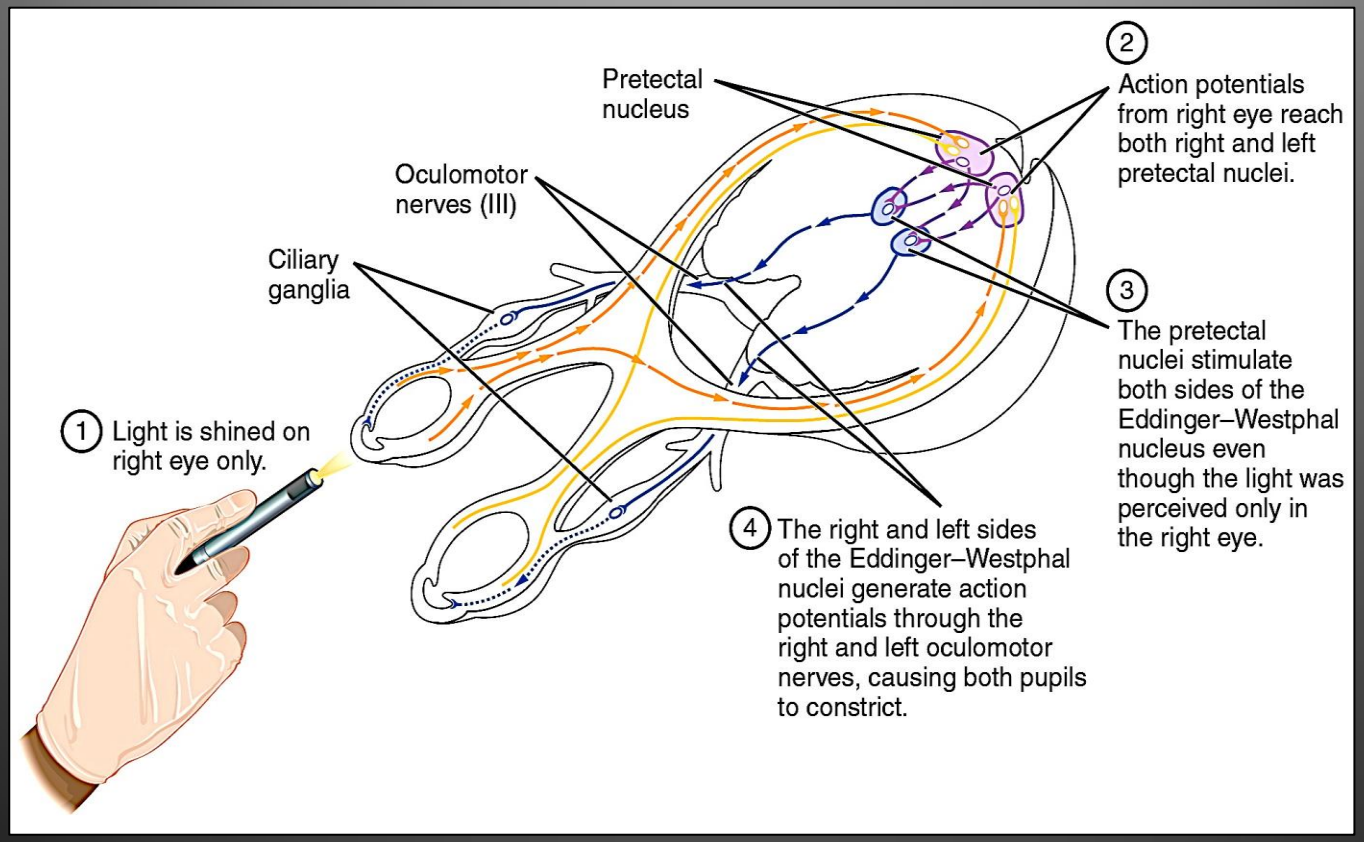
Sympathetic (adrenergic) pathway

- Pupillary dilation is mediated through sympathetic (adrenergic) pathway that originate in the hypothalamus



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

Parasympathetic (cholinergic) pathway

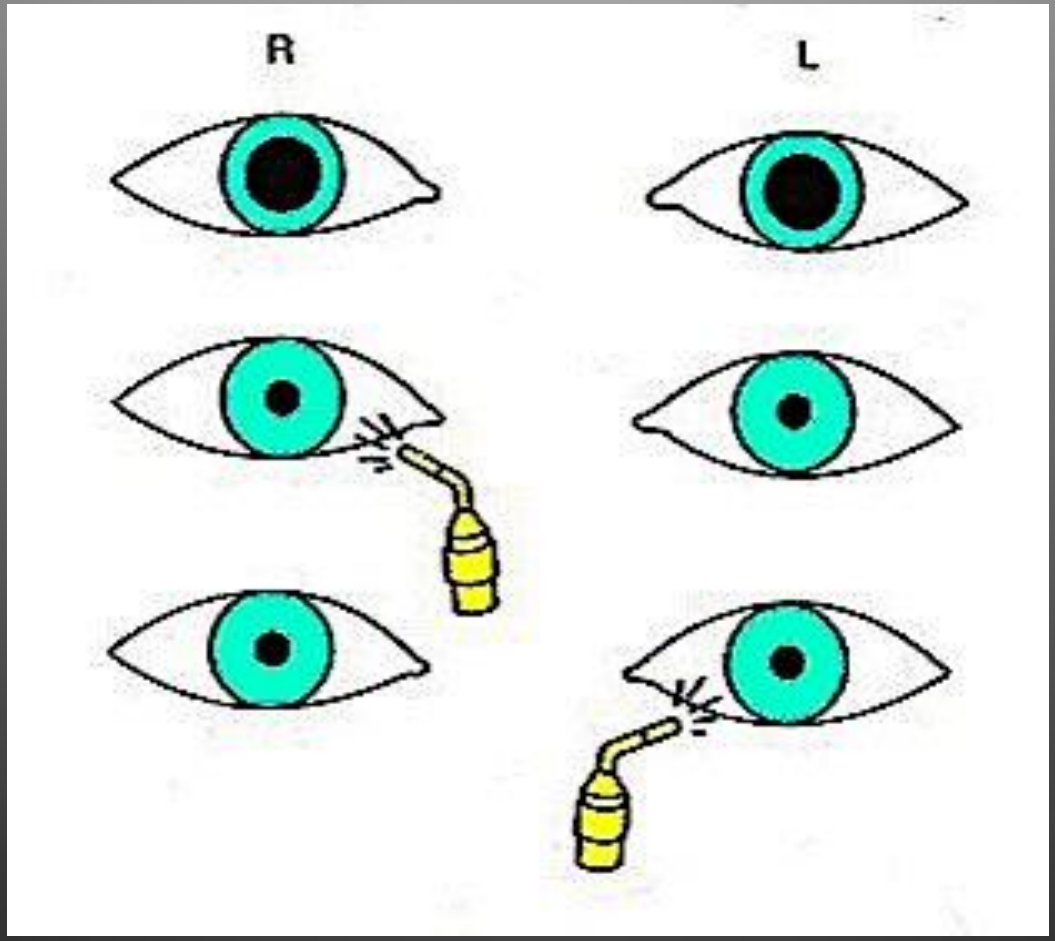


http://oerpub.github.io/epubjs-demo-book/resources/1509_Pupillary_Reflex_Pathways.jpg

Examination of the pupil

- Best conducted in dim light room using a bright light
- The patient should be relaxed and fixing on a distant object
- The size, shape, and position of each pupil should be noted in light and dark conditions
- Check light reflex looking for a relative afferent pupillary defect (RAPD)

Pupillary light reflex



http://casemed.case.edu/clerkships/neurology/NeurLrngObjectives/light%20reflex.jpg

Anisocoria

Which pupil is abnormal ?

Anisocoria

- 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.

Simple (physiological) anisocoria

- 20 % of normal people
- Difference in pupil size of less than or equal to 1 mm
- The degree of anisocoria is equal in dim and bright conditions
- Intermittency or variability is a hallmark
- There are no associated findings suggestive of a sympathetic or parasympathetic lesion.

The large pupil is abnormal

- Previous ocular surgery
- Ocular trauma
- Use of medication like cycloplegics e.g. atropine, cyclopentolate
- Third nerve palsy
- Tonic pupil (Adie's pupil)

Tonic pupil (Adie's pupil)

- Sluggish, segmental pupillary responses to light
- better response to near followed by slow redilation.
- Young female
- Unilateral (80%)
- Instillation of weak cholinergic agents (0. 1% pilocarpine) will cause constriction of the tonic pupil (denervation hypersensitivity)
- Benign condition

Holmes-Adie syndrome

- Includes tonic pupil
- Diminished deep tendon reflexes
- Orthostatic hypotension.

The small pupil is abnormal

- Previous ocular surgery
- Ocular trauma or inflammation
- Use of medication e.g. pilocarpine
- Horner syndrome

Horner syndrome

- Small pupil (miosis)
- ptosis
- anhydrosis

- Caused by a lesion anywhere along the sympathetic pathway
- Carotid dissection, carotid aneurysm and tumor can be associated with this syndrome.

Central	Preganglionic	Postganglionic
Hypothalamus/thalamus/brainstem	Cervicothoracic spinal cord	Superior cervical ganglion
Ischemia (infarction)	Trauma	Forceps trauma
Wallenberg syndrome	Tumor	Ganglionectomy
Tumor	Syrinx	Cervical adenopathy
Hemorrhage	AVM	Jugular venous ectasia
Demyelination	Cervical disc herniation	Internal carotid artery
Cervical spinal cord	Epidural spinal anesthesia	Dissection
Tumor	Brachial plexus	Direct trauma
Trauma	Forceps trauma	Surgical trauma (carotid endartectomy)
Syrinx	Pleural apex	Thrombosis
AVM	Apical lung tumor (Pancoast)	Tumor (nasopharyngeal carcinoma at skull base)
	Surgical trauma (chest tube placement, cardiothoracic surgery)	Cavernous sinus
	Anterior neck	Tumor (pituitary lesion, meningioma)
	Trauma	Carotid cavernous fistula
	Surgical trauma (radical neck dissection, internal jugular catheterization)	Carotid aneurysm
	Tumor (thyroid)	Inflammation
		Infection
		Thrombosis
		Vascular headaches
		Migraine headache
		Cluster headache

AVM, arterovenous malformation.

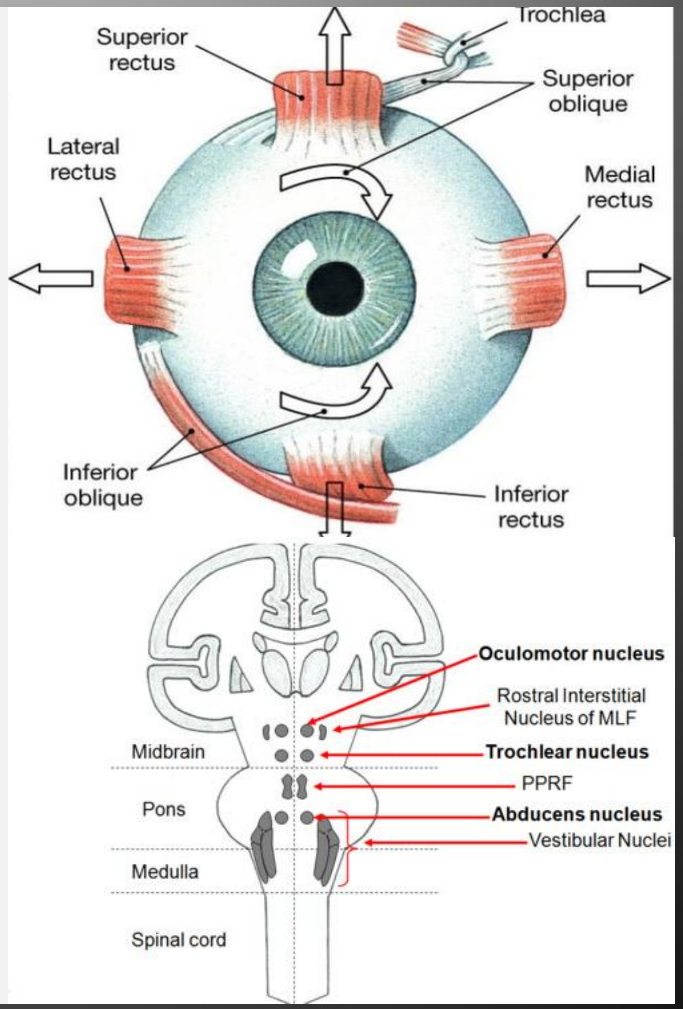
Walton, Kelly A., and Lawrence M. Buono. "Horner syndrome." *Current opinion in ophthalmology* 14.6 (2003): 357-363.

Part 2:

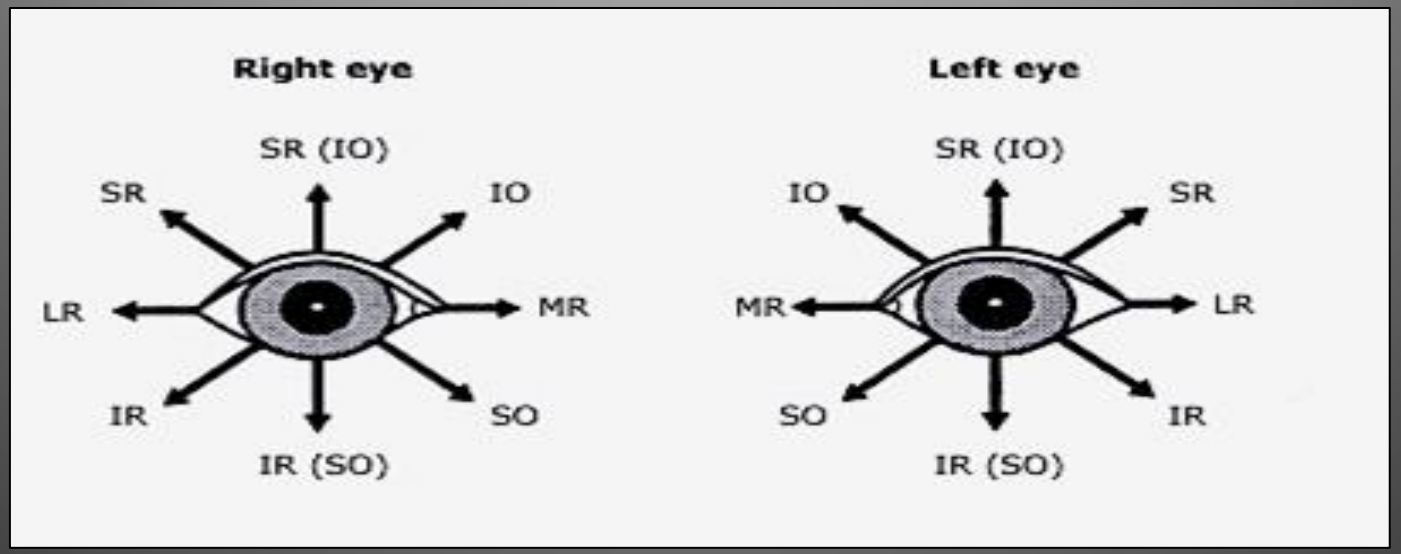
Neuromotility disorders

Anatomy and physiology

Innervation of extraocular muscles	Primary action
Cranial nerve III	
Superior rectus	Elevation (maximal on lateral gaze)
Inferior rectus	Depression (maximal on lateral gaze)
Medial rectus	Adduction
Inferior oblique	Excyclotorsion
Cranial nerve IV	
Superior oblique	Incyclotorsion
Cranial nerve VI	
Lateral rectus	Abduction



Anatomy and physiology



http://www.uptodate.com/contents/images/NEURO/58849/title=Positions+of+gaze

65 yrs old presented to ER complaining of double vision

Neurosurg Clin N Am 23 (2012) 607–619

Third cranial nerve (oculomotor)

- Begins as a nucleus in the midbrain that consists of several subnuclei.
- Innervate the individual extraocular muscles, the eyelids, and the pupils

Third cranial nerve (oculomotor)palsy

Check for pupil involvement

Third cranial nerve (oculomotor)palsy

Etiology:

- Intracranial aneurysm (posterior communicating artery)
- Micro-vascular ischemia (DM and HTN)
- Trauma
- Brain tumor

Fourth cranial nerve (trochlear) palsy

- Vertical diplopia
  - Head tilt to the opposite shoulder

Fourth cranial nerve (trochlear) palsy

- Etiology:

  - trauma

  - idiopathic

  -congenital

Neuromotility disorders

- Which muscle is affected?

Sixth cranial nerve (abducens)palsy

- Horizontal diplopia (worse at distance)
- Esotropia
- Face turn in the direction of the paralyzed muscle
- Limited Abduction on the side of the lesion

Sixth cranial nerve (abducens)palsy

Causes

- Intracranial tumors
- Trauma
- Microvascular diseases
- Increased intracranial pressure

Part 3:

Neuromuscular disorder

Ocular myasthenia gravis

- Chronic autoimmune disease affecting the neuromuscular junction in skeletal muscles.
- Ptosis
- Diplopia
- Fatigability and variability of clinical findings are characteristic
- The pupil is not affected

Ocular myasthenia gravis

- Check for systemic weakness, difficulty in swallowing or breathing.
- Assess orbicularis strength
- Blood test for acetylcholine receptor antibodies

Ocular myasthenia gravis

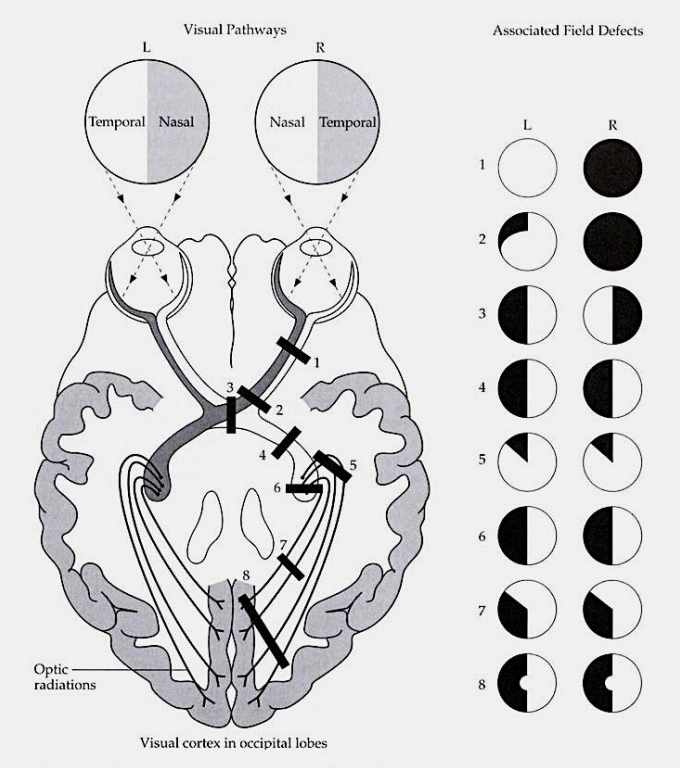
- Tensilon test: inhibits acetylcholinesterase and can transiently reverse signs of weakness due to OMG, such as ptosis and extraocular muscle paresis.

Part 4:

Visual pathway disorders

Visual pathway disorders

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



Optic nerve diseases

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

Optic nerve diseases

Typical optic neuritis

- Inflammatory demyelinating condition linked to MS
- Most common type in young adults
- Visual loss /color vision loss
- Pain that worsen with eye movement
- Visual field loss

Treatment and prognosis

- Good recovery
- IV steroids my speed up the recovery process but does not influence the final outcome.

Optic nerve diseases:

- **Ischemic optic neuropathy (ION):**
  - *Non-arteritic ION:*
  - Patients often have DM,HTN and other vascular risk factor.
  - Most common cause in older patients
  - Altitudinal visual field loss

Arteritic ION

- >55yrs old
- sever visual loss
- Associated with giant cell arteritis (GCA)
- Check for jaw claudication, proximal myalgia and arthralgia, scalp tenderness, headache
- Elevated erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP)

Arteritic ION

- Temporal artery biopsy is the gold standard for diagnosis
- Treatment: Systemic steroids is given immediately if GCA is suspected
- Binocular involvement occurs in third of cases, often within the first 2 days.

Congenital disc elevation

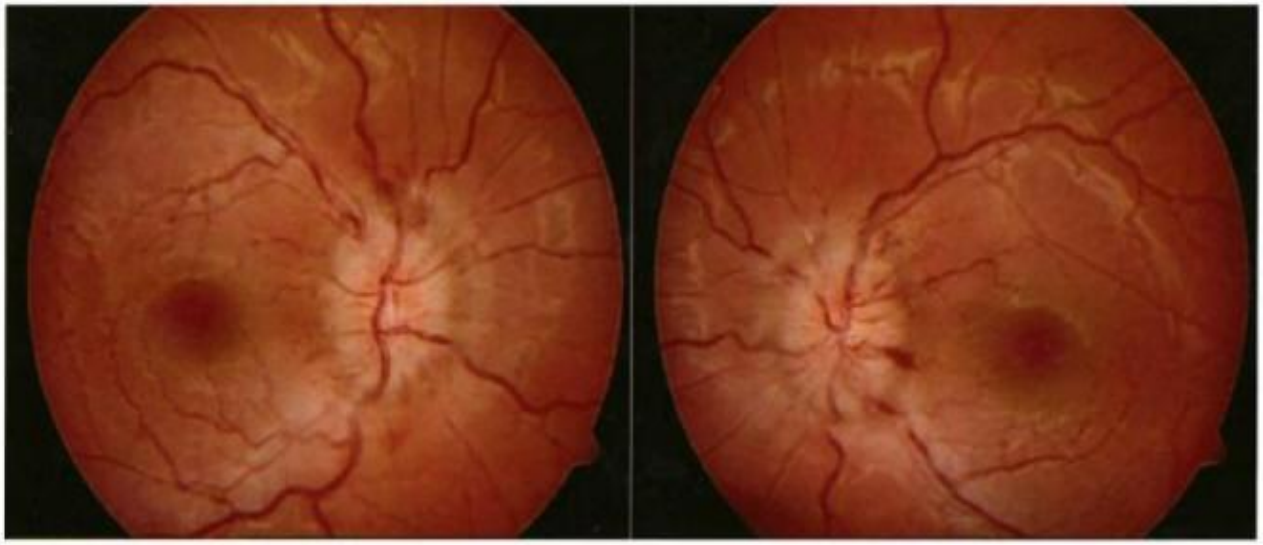
- Optic disc margins blurred and the cup is absent
- No edema or hrg can be observed.
- May be associated with hyperopia or drusen

Other causes of optic neuropathy

- Infection e.g viruses, TB, cryptococcus and syphilis
- Systemic connective tissue disease e.g SLE
- genetics : Leber's optic neuropathy (through a mitochondrial DNA mutation)
- Toxic and nutritional deficiencies
- Trauma

Papilledema

- Bilateral swelling of the optic discs secondary to increased intracranial pressure



Papilledema

- Hyperemia of the disc
-Tortuosity of the veins and capillaries
- Blurring and elevation of disc margins
- Peripapillary flame shaped haemorrhages

Papilledema

Look for spontaneous venous pulsations

Papilledema

Causes:

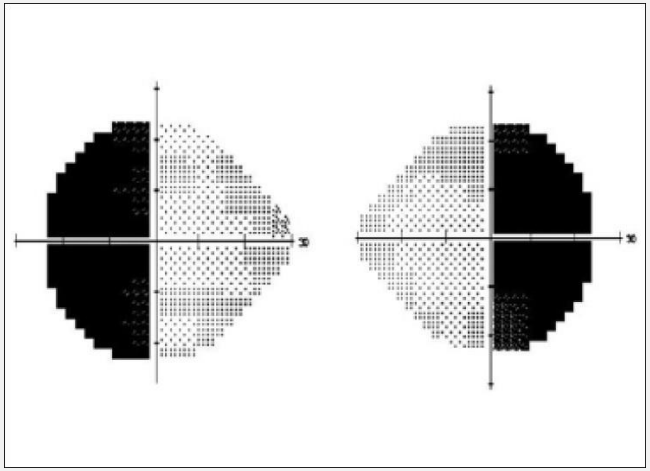
- Intracranial mass
- Sever systemic hypertension
- Idiopathic intracranial hypertension (pseudotumor cerebri)

MCQ

- a patient presented with this visual field defect.

  Which one of the following diagnosis is the most Likely?

  a. Optic neuritis
  b. tilted discs
  c. pituitary tumor
  d. 6th nerve palsy



Thank you