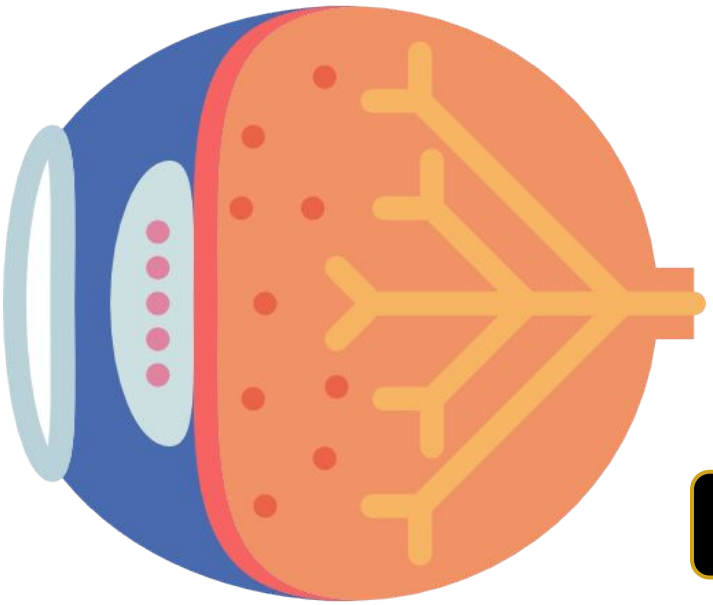
Lecture: 8



Editing file

Neuro-ophthalmology

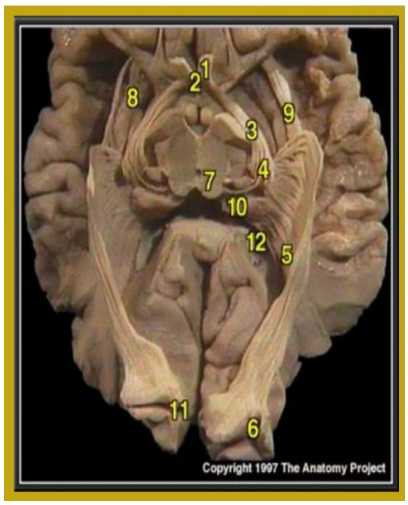
- **Presented By: Dr. Majed Alobailan**
- To outline the applied neuro anatomy including visual pathway, pupil reflex, oculomotor nerves (cranial nerves III, IV, VI, VIII).
- To identify the clinical picture of cranial nerve palsies.
- To discuss the causes of raised intracranial pressure and papilledema.
- Cerebrovascular disorders and ocular signs (e.g. carotid cavernous fistula, cavernous sinus thrombosis) .
- To know common visual field defects of neurological diseases.
- To know ocular manifestation of migraine.

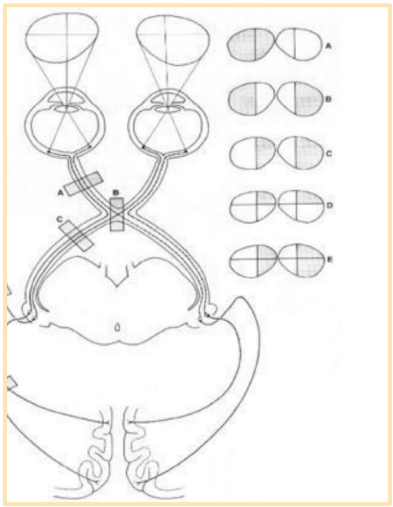
Important | Doctor's notes | Golden notes | Extra | Book

OPTHALMOLOGY TEAM

Afferent Anatomy

❖ What is neuro-ophthalmology? It is a sub-specialty that deals with the afferent & efferent visual system.





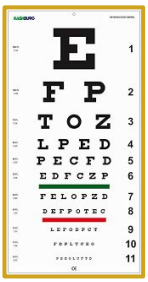
Name structure number 2 ? Optic chiasm
Name structure number 3 ? Optic tract Name structure number 5 ? Optic radiation

- What is afferent (sensory) visual system? The vision sense (optic nerve).
- What is efferent visual system? pupil, eyelid and ocular motility.
- In afferent visual system:
  - Eyeball, optic nerve going through optic canal, optic chiasm, optic tract, optic radiation and occipital cortex.
  - The optic nerve exit the eye from the posterior aspect, and runs through the orbit. It has a lazy s-shape → exit the orbit through the orbital canal and enters the optic chiasm: from the optic chiasm we have the optic tract, optic radiation and occipital cortex.
- In the visual pathway, we don't say lateral or medial, but we say nasal or temporal.
  - The temporal fibers are responsible for nasal visual field, and the nasal fibers are responsible for the temporal part of the field; There is a cross relationship.
- The afferent system starts at the retina (the nerve fiber layer of retina) forming the optic nerve then optic chiasm → optic tract (carries nerve fibers layer from both crossed & uncrossed fibers) → optic radiation → occipital cortex.
- There are some fibers that decussate (cross to the other side).
  - The nasal fibers cross to the other side, while the temporal fibers remain in the same side.
  - The amount of the crossed fibers is more than the uncrossed fibers; 53% of fibers cross & 47% of fibers remain uncrossed.

Afferent Visual System Examination

Examinations

Visual acuity

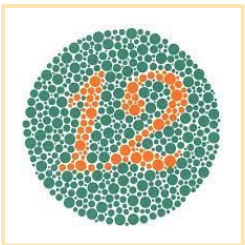


Uses : to measure visual acuity 20/20 feet (6/6 meters) mean the patient can see at 20 feet what a normal person can see at 20 feet. It is used for older children and adults (less than 6 years, we use allen chart)

In snellen chart you ask the pt to name each letter.

Color vision

Ishihara test



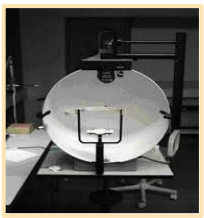
Tested Monocular, it's a screening test for color vision deficit.

Visual field

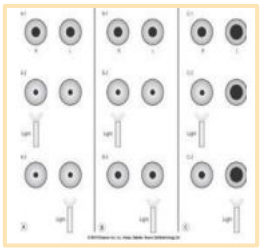
Confrontation test (by counting fingers 1,2 and 5)

Humphrey (automated visual field test) ask the pt to click the button whenever they seen any light
More commonly used then goldmann

Goldmann (kinetic visual field)



Pupil examination



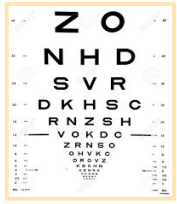
Fundoscopy

- Direct ophthalmoscopy
- Indirect ophthalmoscopy
- Slit lamp and lens
- Look at the nerve itself.

- When we examine the visual system we have to do it mono-ocularly meaning we cover one eye and test the other separately and then we switch to the other eye and do the same.
- Why do we need to do all the five examinations? because someone with optic nerve injury might have 20/20 visual acuity but they might have a major defect in the peripheral visual field.

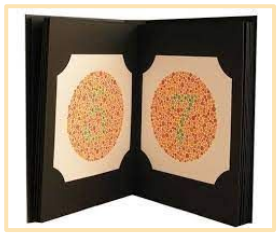
1.Visual acuity

- The visual acuity test is used to determine the smallest letters you can read on a standardized chart (Snellen chart) or a card held 20 feet (6 meters) away.
- The visual acuity can be tested by projecting letters (Snellen chart) It can also be tested by using E game chart.
- It is used to test for far and near objects
  - Distance for near object is about 30-40 cm (reading distance)
  - E game chart and Snellen chart are used for far object



Afferent Visual System Examination

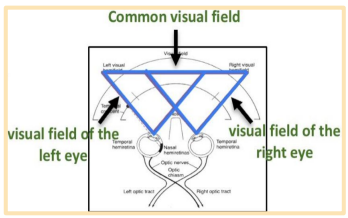
2.Color vision



- Ishihara color chart. It is a hand-held book with multiple pages and charts that has different numbers with colors.
  - Basically, you just name the number. It should be done mono-ocular
  - Count how many did the patient get correct. 10\15 – 12\15
  - Good for screening of color vision defect, but it does not tell you the type of color defect.
  - If the patient wears glasses ask them to put on their glasses before the examination

3.Visual field

- Visual field means an island of vision. So, if you close one eye, you will see an island that is a triangle in shape, expanding the further it goes.
- That is why when you examine the visual field (by confrontation test), there has to be an equal distance between you and the patient.
- The further the object go the bigger the field becomes
- By closing one eye, only 1/5 of the visual field disappears. So, if someone loses one eye, 1/5 of the visual field will be lost. Why? (in the pic)
- If you draw a triangle for each eye. There is a common triangle in the middle represented by both eyes.
- Benefits of having a common visual field:
  - 3-dimensional vision.
  - Locking mechanism: our eyes are straight because we see the image from 2 different angels, then the brain will lock them to each other.



- So. If we lose one eye, we lose only the temporal field from one direction.
- 3 types of visual field examination:

1- Confrontation test:

- Good for screening and is a good test for absolute scotoma (total blindness)
- Scotoma means blindspot, and if we wanna measure it we have to set the patient to an actual machine that can measure the visual field (perimeter) peri=scotoma + meter=measure
- It is a good screening to detect massive visual field defect e.g. A patient in ER bc of stroke or patient with cushing syndrome, or pituitary adenoma and you wanna rule out bitemporal hemianopia
- It is a qualitative test (the patient can either see or not); it doesn't calculate the density of the defect.
- **Quantitative test** will measure the density of the blind spot or the quantity of the defect.

- How to perform the examination:
  - Confrontation visual field testing involves having the patient looking directly at your eye or nose and testing each quadrant in the patient's visual field by having them count the number of fingers that you are showing.
  - You have to sit exactly in front of the patient and your eye has to be on the same level of the patient's eye.
  - You test each eye separately (mono-ocular) covering one eye and testing the other by projecting your fingers in each quadrant and ask the patient to tell you how many finger does she/he see.
  - The examiner should be one the same level of the patient, at arm's length.
  - If there is a defect, you say there is a defect in superior-temporal, inferio-temporal, superior-nasal, or inferio-nasal.
- On the other hand, you ask the patient "look at my cornea". Then ask: do you see black and white? Do you see lid margins? Do you see the eyebrow? If he\she is able to see the details, that means no central scotoma.
- In central scotoma: patient will say, I cannot see your eyes, I can see only half of your face.
- If peripheral, patient will not be able to count your fingers.

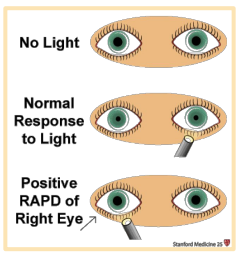
2- Goldmann test: a technician will move a target, then ask the patients if they can see it.
3- Humphrey: automated. It is a quantitative method; it measures the density of the visual field defect.

- Some patients have relative visual field defect, it is like a mesh in front of their vision; in such a case, you must do actual visual field test by using a machine (Humphrey). Most common device

4.Pupils Examination

- We examine the pupil reaction to light, near, and we also want to know the average size of the pupil and whether they're equal or not
  1. Dim light.
  2. Ask the patient to look at a specific far target, not the wall. E.g. electric plug, a clock or a letter on the E chart.
  3. Shine light from down. Look at both pupils. Are they equal in size, or not?
     Unequal pupil size: anisocoria - Unequal refractive error: anisometropia. - Unequal image size: Aniseikonia.
  4. From the side, shine light over one pupil, then observe pupil constriction if it is brisk or weak. Then do the same for the other pupil. Estimate the reaction if equal or not.

5. If you are not sure, do **swinging flash test**. Swing the light from eye to eye (you have to keep the light for 2 seconds to see the pupil unrest then you shift to the other eye).
    - Normally, if you shine a light on the pupil, it constricts then relax a little (this is called pupil unrest or hippus pupil).
    - **If both constrict** → normal. Composed of direct (the one with light) and indirect (consensual)(the other pupil).
    - **If one dilates** → this is abnormal & it means there is relative afferent pupillary defect (RAPD+).
    - We call it relative because we are comparing one pupil in relation to the other.
    - RAPD+ tells you there is an afferent visual pathway injury (optic nerve injury), regardless of the cause (e.g. optic neuritis, ischemic optic neuropathy or optic nerve tumor).
6. Finally we do near reflex (we ask the patient to look at a near target and examine the pupil reactions)

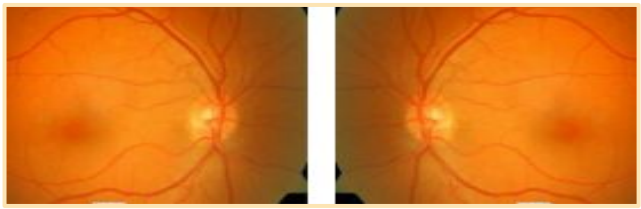


- The components of near reflex are:
    - Miosis → the pupil became smaller
    - Accommodation → a swelling the lense with change curvature to look at a near & tiny object.
    - Convergence → both eyes move toward the nose (toward each other)
    - Near reflex is a part of pupil examination. As pupils don't only constrict to light but also when looking at near objects.

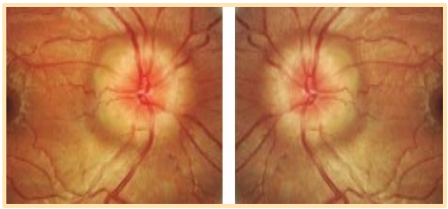
5.Fundus examination

- We look at the optic nerve, by:
    1. Slit lamp with lens.
    2. Direct ophthalmoscope (direct because we can pick it up and use it): is a handheld device with a light source & optical center used to examine the pupil. It is monocular with high magnification so larger image, smaller field & image is real (upright).
    3. Indirect ophthalmoscope: is a head-mount device that is binocular, inverted, magnification is less, you need to use a lens so larger field and smaller image (usually we use lens with 20 dioptric power).
- **The Total dioptric power of the eye is 60.**
- So, 60\20=3 so the magnification is 3 times.
- Common question in OSCE: difference between direct and indirect ophthalmoscope
    - Direct is mono-ocular (we use one eye to test the patient's eye)
    - Because it mono-ocular it doesn't give us a 3D image so we can't tell clearly if the object is elevated or not
    - The magnification in the direct is 15 times making the visual field smaller so you can only the optic nerve or fovea or only blood vessels, unlike the indirect which has a 3 times magnification thus the field is bigger so you can see optic nerve and macula together.

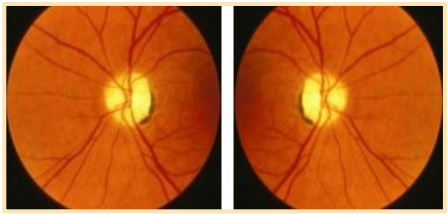
Afferent Visual System Examination



- This is what we see when we use slit lamp and hand-held lens.
- We can see:
  1. Optic nerve.
  2. Macula.
  3. Pigmented fovea
  4. Superior temporal arcade.
  5. Inferior temporal arcade.
  6. Superior nasal arcade.
  7. Inferior nasal arcade.



- Dome shaped disc
- Blood vessels are obscured bc of the fluid (disc swelling)
- young, obese female, idiopoathic increase of ICP



Very yellow pale nerve that indicates damaged atrophied nerve.

- What is the normal color of the nerve? slightly orange, reddish.
- The optic nerve disc has a cup (depression) and neuro-retinal rim (remaining part).
- Can you draw the margins of the nerve with a pencil? Yes → normal. | No → abnormal (indicates swelling).
- What do we call a bilateral optic nerve swelling? bilateral disc edema (NOT always papilledema).
  - It might swell because of papillitis (optic neuritis) inflammation of optic nerve or infiltration of the optic nerve in case of some tumors such as leukemia or lymphoma.
  - Papilledema by definition is a bilateral optic nerve head (disc) swelling because of high ICP.
  - It is an emergency because high ICP might be secondary to a tumor & i**t requires an immediate imaging.**
  - If the imaging is normal, what might be the cause?
    - Fever → (infection, meningitis).
    - Headache and usually obese women on OCP → benign idiopathic intracranial HTN (pseudotumor cerebri), very common in our community.
- What are the causes of optic nerve atrophy?
  Post-optic neuritis - Post-increased ICP - Tumor - Inflammation - Compression.
- Thus, optic nerve atrophy requires imaging. (Appears pale)
- Disc pallor in eye refers to pallor of the optic disc which results from irreversible damage to the retinal ganglion cells and axons. (Atrophy)

Afferent Visual System Diagnosis

❖ Compression: From a tumor

- ◇ Intraorbital ON
- ◇ Intracranial ON
- ◇ Optic chiasm
- ◇ Optic tract
- ◇ Posterior afferent system

❖ Trauma:

- ◇ In adults the most common trauma causing optic nerve injury is head trauma due to MVA with fractured skull
- ◇ In children the most common trauma is from door handles bc their eye level is at the same level of the door handle and they can injure their eye if not careful.
- ◇ Globe by fire cracks, hand fest, tennis ball or door handle especially in children.
- ◇ Intraorbital ON.
- ◇ Optic canal.
- ◇ Optic chiasm.
- ◇ Occipital lobe.
- ◇ Example: 25 years old man had a Firecracker exploded near left eye.
  - ■ NLP OS (no light perception).

❖ Inflammation:

- ◇ Orbital pseudotumor
- ◇ Typical Optic neuritis (very common):
  - ■ Inflammatory demyelinating condition
  - ■ Linked to MS
    - ➢ 27 year old woman developed blurred vision OD and mild right periorbital pain, VA 20/50, MRI abnormal.
  - ■ Most common type in young adults
  - ■ Majority is central Visual loss /color vision loss
  - ■ Pain that worsen with eye movement. Why? Optic nerve is surrounded by EOM
  - ■ Visual field loss
- ◇ Other (SLE or sarcoidosis)

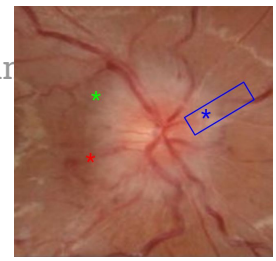
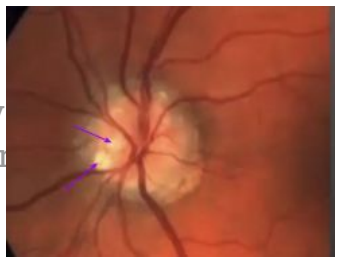
❖ Ischemic optic neuropathy

- ◇ Non-arteritic ischemic optic neuropathy.
- ◇ Giant cell
- ◇ Giant cell arteritis with ischemic optic neuropathy. (non giant cell)
- ◇ Central retinal artery occlusion. (emboli from a valvular disease, carotid plaque or post cardiac surgery)
- ◇ Other retinal emboli.

Afferent Visual System Diagnosis

❖ Congenital and genetic problems: (SKIPPED BY DOCTOR)

- ◇ Congenital retinal dystrophies.
- ◇ Optic nerve hypoplasia.
- ◇ Leber hereditary optic neuropathy (Through a mitochondrial DNA mutation). Young, monocular, +ve family hx
- ◇ Dominant and recessive optic atrophy.
- ◇ Glaucoma.
- ◇ **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. (Dr.Daniah explained this and asked us to know the difference between this condition and optic disc edema)

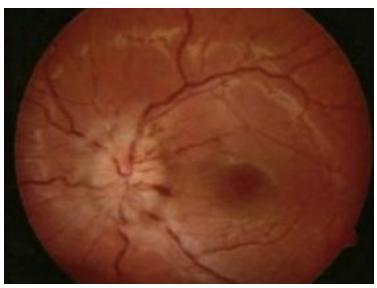
Optic disc edema	Congenital disc elevation
1- Hue of fluids (*) 2- Splinter hemorrhage indicatin ischemia (*) 3-Blood vessels can't be traced; some parts appearing and other disappearing (*) 	1- No hue of fluids; very sharp margins 2- Blood vessels are easily traced; all parts are appear 3- No hemorrhage 4- Drusens (*) 

❖ Other causes of optic neuropathy:

- ◇ Infection e.g viruses, TB, cryptococcus and syphilis
- ◇ Toxic and nutritional deficiencies

❖ Papilledema

- ◇ Bilateral swelling of the optic discs secondary to increased intracranial pressure.
- ◇ **Fundoscopy:**
  - ■ Hyperemia of the disc
  - ■ Tortuosity of the veins and capillaries
  - ■ Blurring and elevation of disc margins
  - ■ Peripapillary flame shaped haemorrhages
- ◇ **Look for spontaneous venous pulsation**. If present we can exclude increased ICP
- ◇ **Cause:**
  - ■ Intracranial mass (urgent neuroimaging including in some cases MRV for VT)
  - ■ Severe systemic hypertension
  - ■ Idiopathic intracranial hypertension (pseudotumor cerebri) (If imaging shows no mass and blood pressure is normal then do LP to measure pressure)



Bilateral optic neuritis can cause similar findings but with NO elevation of ICP!

Afferent Visual System Diagnosis

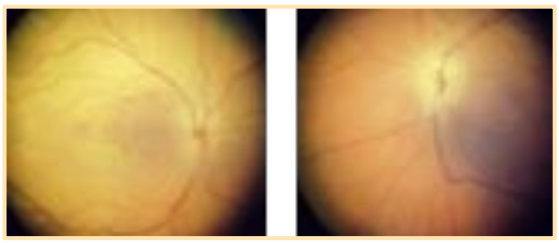
Non-arteritic ION

- Patients often have DM, HTN and other vascular risk factor.
- Most common cause in older patients (above 40 years).
- Sudden painless acute visual loss
- Altitudinal visual field loss (superior or inferior).
- **Treatment**: no treatment, ask them to control the risk factors to protect the other eye.
- **Prognosis**: Most of time is irreversible visual loss

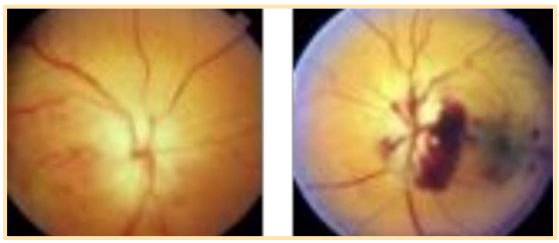
Arteritis ION

- (> 55) years old (older than non-arteritic ION).
- Associated with giant cell arteritis.
- Severe visual loss. Present with severe irreversible visual loss (counting finger) more than non-arteritic (20/200).
- Check: jaw claudication, proximal myalgia & arthralgia, scalp tenderness/pulseless, headache.
- Elevated erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP). If increased with GCA presentations then high steroids!
- Temporal artery biopsy is the gold standard for diagnosis.
- Treatment: systemic steroids, given immediately if suspected even before the biopsy, patient should not leave the ER without it to protect the other eye (within the first day they will go blind) .
- Binocular involvement occurs in third of cases, often within the first day.

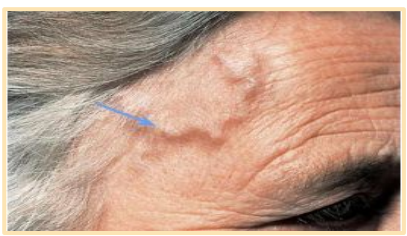
Giant Cell Arteritis, day 2



Giant Cell Arteritis, day 4



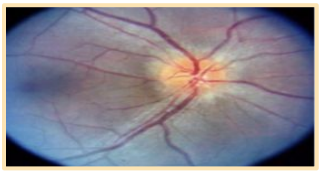
This is GCA. If you palpate the temporal artery in the affected side, it would be pulseless.

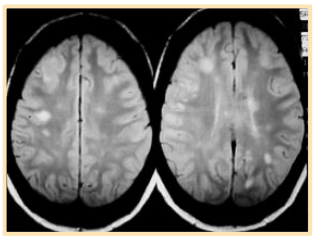


Afferent Visual System Diagnosis

Optic neuritis case

- 27 years old women developed blurred vision OD and mild right periorbital pain
  - Pain with eye movement
- VA 20/50
- Pupil examination: optic nerve is slightly hyperemic and the margins of the disc are blurred
- MRI abnormal: showing multiple white plaques (demyelinating lesions)
- Diagnosis: multiple sclerosis.
- **Treatment and prognosis:** Good recovery, IV steroids may speed up the recovery process but does not influence the final outcome.
  Oral corticosteroids are contraindicated in optic neuritis because it increases recurrency

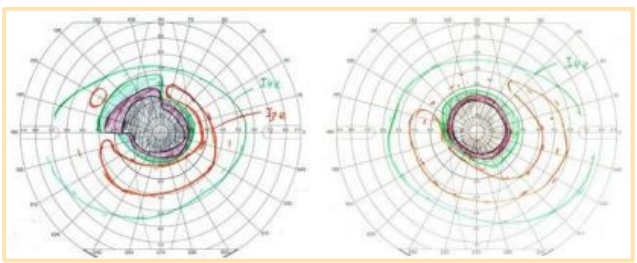




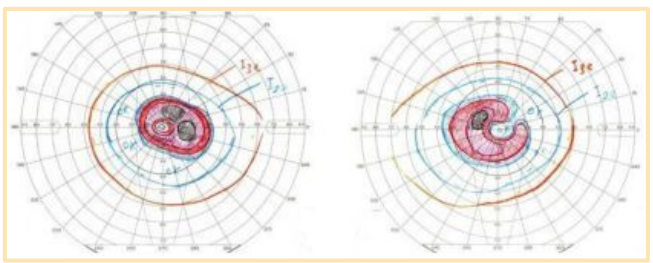
Extra case from 436

- 14 years old girl, vision OS began to decline gradually without pain.
- First visit with VA OD 20/40 and OS CF at 3'
- Pale, flat optic discs OU
- VA 1-month later CF at 3' OU
- VA 10 months later 20/20 OU
- Diagnosis LHON-like optic neuropathy.

VF 1 month later



VF 10 month later



Afferent visual system tests

❖ During exam:

- ◇ Visual field test.
- ◇ A and B scans (ultrasound of the eyeball)
- ◇ Optical Coherence Tomography (OCT) it can show the anatomy of retina in ten layers (it can be used in macular edema to show the exact location of the abnormality
- ◇ Electroretinography (ERG) like ECG it measures the function of photoreceptor
- ◇ Visual Evoked Potential (VEP) test the conductivity of the optic nerve
- ◇ *the doctor said you don't have to know these tests

❖ Neuroimaging:

- ◇ CT scan (not that useful)
- ◇ MRI scan (best test to examine the optic nerve)

❖ Blood test:

- ◇ Vasculitis (ESR, CBC, ANA, VDRL) bc the optic nerve can be affected by SLE, sarcoidosis ,TB, syphilis.
- ◇ LFT (SGOT[1], SGPT[2], Alkaline phosphatase).
- ◇ Urine analysis
- ◇ Creatinine, BUN
- ◇ Electrolytes

❖ Ultrasound:

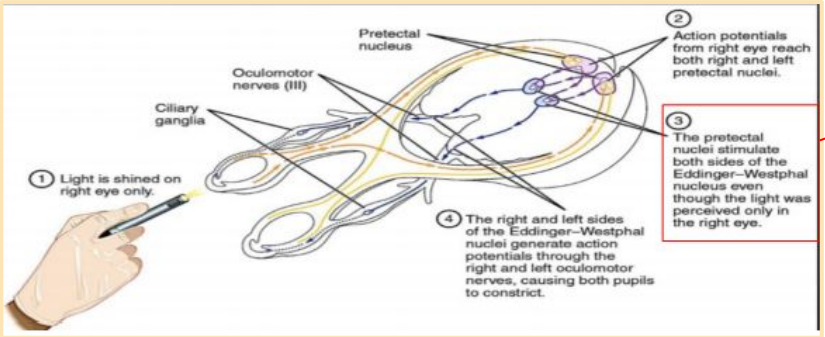
- ◇ Carotid doppler.

❖ Genetic evaluation.

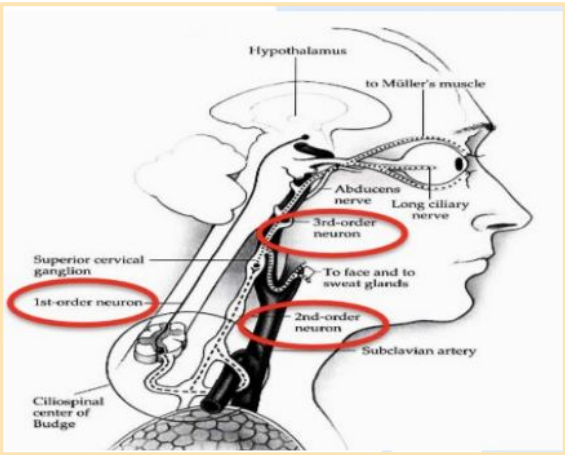
[1] Serum glutamic oxaloacetic transaminase
[2] Serum glutamic pyruvic transaminase

Pupillary Disorders

- **The anatomy and physiology of the pupil:**
  - The pupil size 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
- **Parasympathetic (cholinergic) pathway:**
  - It is the main cause of afferent pupillary defect and relative afferent pupillary defect



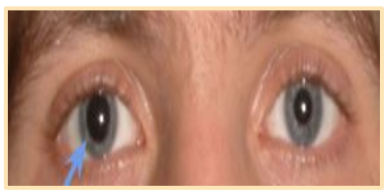
Both sides must receive equal amount of stimuli; this can be tested through the indirect or consensual reflex.



Sympathetic innervation of pupils starts from **hypothalamus** and descends down as **1st order neuron** until **Ciliospinal center** (found at level C8 to T2) then continues as **2nd order neuron** through **brachial plexus** where it crosses the apex of lungs (this is clinically important: Pancoast tumor can lead to horner syndrome due to this anatomical relation), then it ascends to **superior cervical ganglion** (ganglion location is important, it is found in the angle of mandible near to the bifurcation of common carotid artery, from there **3rd order neuron** enters the cranium with internal carotid artery; dissection of the artery). After that, it will enter the **cavernous sinus,** near **6th CN** then joins the **trigeminal nerve (V1)** from there it goes to orbit to supply the orbital structures. **What does it supply**? Pupils, muller's muscle (eyelid elevation; not major muscle), retaractors of lower eyelids.

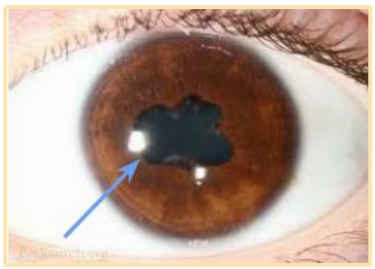
❖ Anisocoria

- ◇ 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.



Right relative afferent pupillary defect.
In exam you should mention which side is affected

Pupillary Disorders

The large pupil is abnormal	The small pupil is abnormal
• Previous ocular surgery injury to iris • Ocular trauma • Use of medication like cycloplegics e.g. atropine, cyclopentolate • **Third nerve palsy** you should exclude • **Tonic pupil (Adie's pupil):** ○ Sluggish, segmental pupillary responses to light better response to near followed by slow redilation. Young female. Unilateral (80%). How to diagnose? Instillation of weak cholinergic agents (0. 1% pilocarpine) will cause constriction of the tonic pupil (due to denervation hypersensitivity) but no response in the normal side. Benign condition reassurance	• Previous ocular surgery • Ocular trauma or inflammation • Use of medication e.g.pilocarpine • Horner syndrome This is posterior synechiae; iris adhesion to the lens due to chronic inflammation (uveitis is one of causes) 

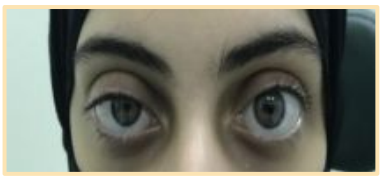
❖ Simple (physiological) anisocoria

- ◇ 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 (it is not fixed to one pupil, it alternates)
- ◇ There are no associated findings suggestive of a sympathetic or parasympathetic lesion. You must exclude any evidence of neurological lesion!

❖ Examples of pathological anisocoria

1. Holmes-Adie syndrome:

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



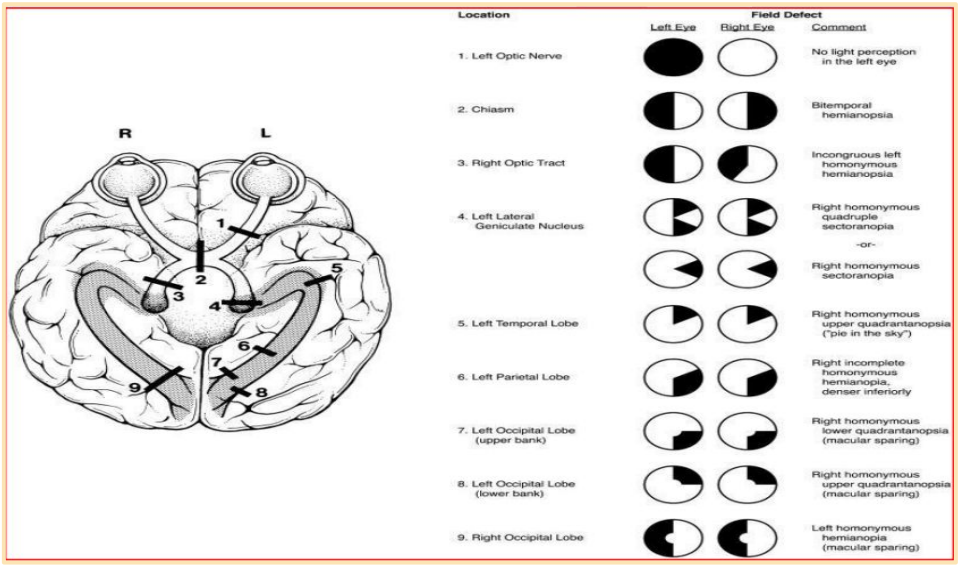
2. Horner syndrome:

- ■ Small pupil (miosis)
- ■ Ptosis
- ■ Anhydrosis
- ■ Caused by a lesion anywhere along the sympathetic pathway
- ■ Carotid dissection (Internal carotid dissection, life threatening, must be excluded, do neuroimaging: MRI & MRV), carotid aneurysm and tumor can be associated with this syndrome.

Visual Pathway Disorder

❖ Terms:

- ◇ Anopia/anopsia: Any blind area in the visual field
  - ■ If it's complete we call it complete anopsia
  - ■ Hemianopia: when ½ or ⅔ of the visual field is affected.
- ◇ Quadrantanopia/Quadranopsia: quarter of the field of vision.
  - ■ In quadranopsia, we have to specify superior or inferior.
  - ■ If it is more than quarter but less than half → we still call it quadrantanopia/quadranopsia.
  - ■ We also have to specify if there's macular sparing: mouth eaten part.



- If I ask you to name the visual field defect:
  1. Look at the printout and localize black part (right or left).
  2. Decide if both defects are homonymous (means both are on the same side) or not on each visual field: If they're not on the same side we call it heteronymous
  3. Then name the visual field defect (don't forget to specify if it's superior or inferior in case of quadranopia).
- For localization we start with the opposite word of the defect, e.g. if the lesion was on the left then the localization will be on the right
- Same goes in quadrantanopia; if the lesion was upper then localization will be lower and vise versa
  - Quadrant lesion means it's on one of the lobes either temporal or parietal

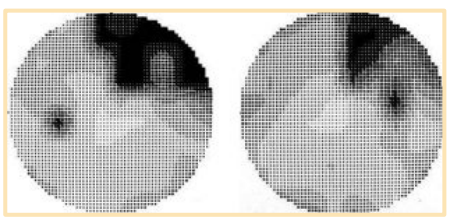
Visual Pathway Disorder

❖ Localization of visual field defect (the numbers correspond to the figure on the last page):

1. Optic nerve or eyeball → complete loss of the field (anopia)
2. Chiasm (beneath it lies the pituitary gland):
   - Any mass (e.g. parasellar tumors, pituitary macroadenoma, lactenoma, acromegaly or cushing syndrome) will press against the chiasm (chiasm compression) on the optic nerve (binasal decussating fibers) resulting in what is **called bitemporal** heteronymous hemianopia (we can't say right or left here).
3. Optic tract: Since causing hemianopia it will be in the optic tract.
   - The side of the lesion **should be opposite** to the side of the affected visual field.
   - Hemianopia visual field defect always indicates a lesion in the optic tract.
   - Therefore, in left homonymous hemianopia
   - The lesion is in the right side.
4. Lateral Geniculate Body:
   - It has a dual vascular supply and usually come in either wedge shaped defect in the middle taking the upper & lower part quadrants or wedge sparring with upper and lower defect.
   - **Sectoranopia (sector = part of)**
5. Temporal lobe:
   - Right homonymous superior quadrantanopia
   - The lesion is in the left.
   - We call it **pie in the sky.**
   - Since the defect is superior, so in the brain should be inferior (in the temporal lobe).
6. Parietal lobe:
   - Right homonymous inferior quadrantanopia
   - The lesion is in the left.
   - Some people call it **pie in the floor**
   - Since the defect is inferior, so the lesion should be superior (in the parietal lobe).
7. Occipital lobe:
   - Occipital lobe lesions give congruous visual field defect (identical or symmetrical visual field defect).
   - Why there is a macular sparing? because the occipital lobe has a dual blood supply: the middle cerebral artery (MCA) and the posterior cerebral artery. MCA accounts for Macular fibers.
   - Macular sparing means MCA is not occluded
   - If the defect is more peripheral, it indicates posterior cerebral artery involvement.

- When there is a stroke that hits the MCA and PCA we will notice NO macular sparing BUT with other symptoms too since its a stroke

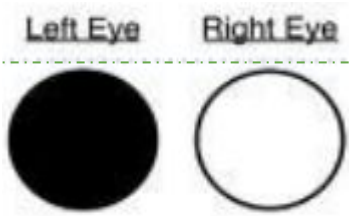
After Temporal Lobectomy



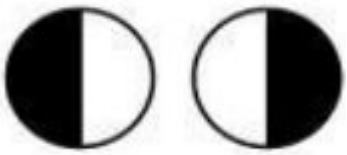
- Visual defect: right homonymous superior quadrantanopia
- Lesion: left temporal lobe
- Black dot: normal blind spot account for the optic disc

Visual Pathway Disorder

- **Very IMP! common question, name the defect & the location of the lesion**

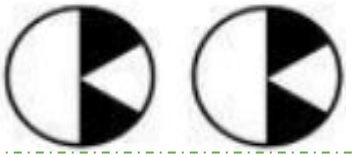


- **Visual field defect:** Complete loss of the field (complete anopia)
- **Location:** Optic nerve or one eyeball loss

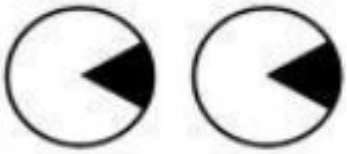


- **Visual field defect:** Bitemporal hemianopia
- **Location:** Optic chiasm

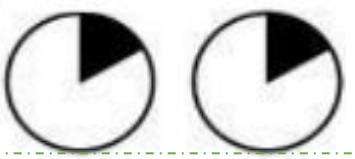
- **Visual field defect:** Left homonymous hemianopia
- **Location:** Right optic tract



- **Visual field defect:** Right homonymous quadruple sectoranopia
- **Location:** Left lateral geniculate body

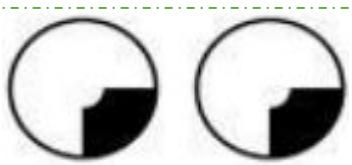


- **Visual field defect:** Right homonymous quadruple sectoranopia or Right homonymous quadruple wedge shape defect
- **Location:** Left lateral geniculate body



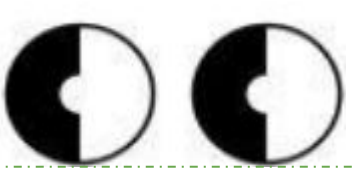
- **Visual field defect**: Right homonymous upper quadrantanopia
- **Location:** Left temporal lobe

- **Visual field defect:** Right homonymous inferior quadrantanopia
- **Location:** Left parietal lobe



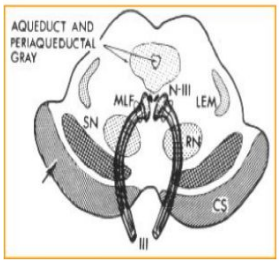
- **Visual field defect:** Right homonymous inferior quadrantanopia with macular sparing
- **Location:** Left occipital lobe

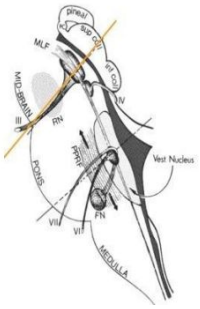
- **Visual field defect:** Right homonymous superior quadrantanopia with macular sparing
- **Location:** Left occipital lobe

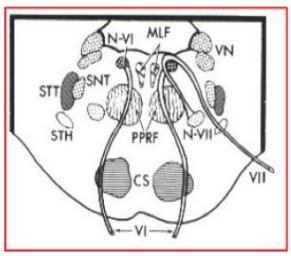


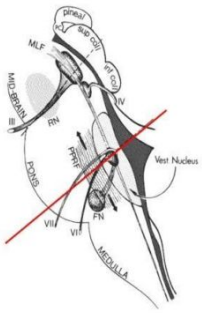
- **Visual field defect:** Left homonymous hemianopia with macular sparing
- **Location:** Right occipital lobe

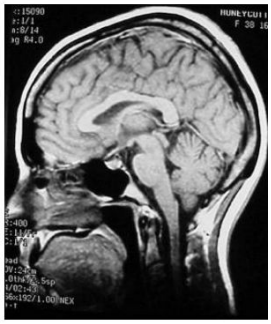
Efferent Neuro-anatomy





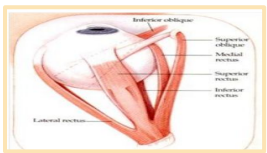


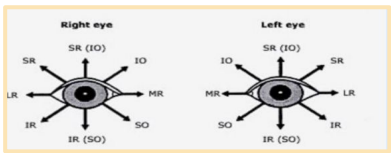




- Remember we said that the efferent visual system consists of pupil size, eyelid position and ocular motility.
- The ocular motility is controlled by cerebellum, brainstem, interneuronal nuclei as well as the peripheral cranial nerves.
- The cranial nerves controlling the eyelids and ocular motility are 3 nerves which are 3rd , 4th & 6th cranial nerves.
- All cranial nerves originate from brainstem (the midbrain and pons), then goes anteriorly to the orbit to control ocular motility.
- If i ask you to look at your left; the 6th CN in your left eye will push the left eye laterally (away from your nose) → lateral rectus muscle and the 3rd CN in your right eye will push the right eye to the same direction → medial rectus muscle
- So both eyes (6th and 3rd nerve) will move together same speed and same direction, why? because of the interneuron connection in the midbrain (like medial longitudinal fasciculus)
- If I ask you to look at something, why you look at it and not look beyond it? this is controlled by the cerebellum and the interneuron nuclei.
- There are two types of eye movement:
  1. Fast movement: shifting from one target to other.
  2. Slow movement: tracking a moving object.

The Extraocular Muscles





- Four recti & two oblique muscles.
  - Superior, inferior, medial and lateral rectus
  - Superior oblique (pass through the trochlea)
  - Inferior oblique (insert behind the macula)
- All are supplied by oculomotor nerve except, superior oblique (trochlear nerve) & lateral rectus (abducens nerve) (SO4 LR6)
- **Anatomy & physiology**: The main nerve supplying extraocular muscles is oculomotor (third nerve)

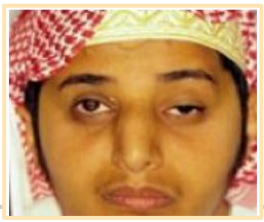
EOM	Primary action	Innervation	Nucleus
Superior rectus	Elevation (maximal on lateral gaze)	Third cranial nerve, oculomotor	Midbrain
Inferior rectus	Depression (maximal on lateral gaze)	Third cranial nerve, oculomotor	
Medial rectus	Adduction	Third cranial nerve, oculomotor	
Inferior oblique	Excyclotorsion	Third cranial nerve, oculomotor	
Superior oblique	Incyclotorsion	Fourth cranial nerve, trochlear	
Lateral rectus	Abduction	Sixth cranial nerve, abducens	Pons

Efferent Examination

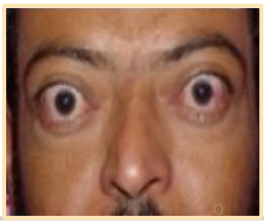
Just look at the patient? "inspection"

- ➔ Are eyes straight?
- ➔ No, the left eye is not straight (inward deviation)
- ➔ Deviated towards the nose (**esotropia**).
  - ◆ Exotropia (out), hypertropia (up), hypotropia (down)

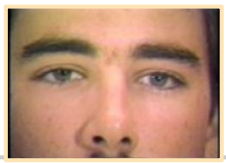
- ➔ What are the lid positions?
- ➔ Left ptosis, the patient is lifting his eyebrow to compensate.
  - ◆ Horner's syndrome (mild ptosis) sympathetic injury to Muller's muscle paralysis and in 3rd nerve palsy it's parasympathetic injury due to levator palpebrae paralysis.



- ➔ Are the eyes proptotic?
- ➔ Exophthalmos, lid retraction, lid lag.
- ➔ Injected eyes.
- ➔ Scleral show, normally it is not seen.
- ➔ It's common with thyroid diseases (thyrotoxicosis)

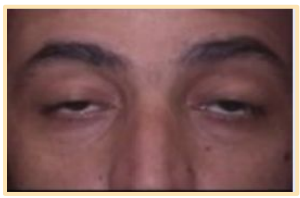


- ➔ Are there any spontaneous eye movements?
- ➔ Nystagmus (involuntary **rhythmic** spontaneous eye movement).
- ➔ You should test it in all direction



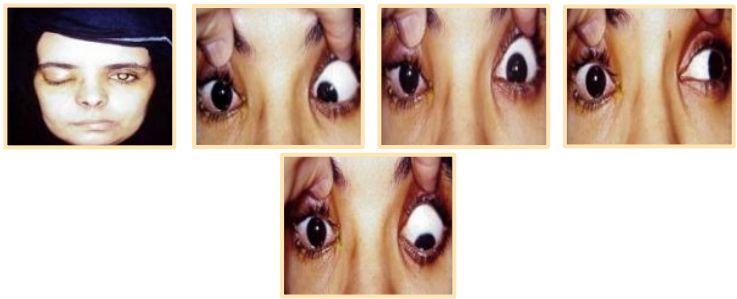
Movements of both eyes in all directions

Have the patient move eyes in all directions, not just the direction where you think there is a problem. 9 positions. Vertical, horizontal , and X



Hold lids if necessary (only after looking first without holding lids).

3rd and 6th nerve.



Examine each eye separately if **necessary**.

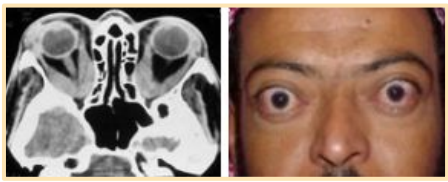
Smooth pursuit	Saccades
The reflex that helps to maintain fixation on an object in motion in the visual world while the head is stable. Also, the reflex that inhibits the vestibluo-ocular reflex.	The reflex that permits a rapid refixation from one point in the visual field to another. Fast eye movements in equal speed. For both eyes Cerebellum diseases; Dysmetria

Efferent diagnosis

❖ Orbit:

1. Extraocular muscles:

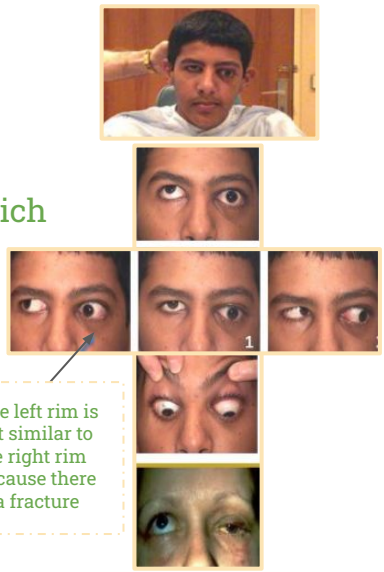
- In the CT, the muscles are enlarged, hypertrophied.
- This is thyroid orbitopathy; The muscles enlarge and can compress on the nerve and cause loss of vision



Bilateral proptosis

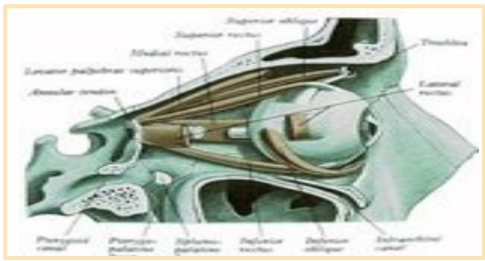
2. Trauma:

- 14-years old boy
- In picture 1:
  - There is malalignment.
  - Dropping of left eye (abnormal eyeball position which is called ocular dystopia).
  - If we draw a line from the lateral canthus to the lateral canthus, they are not in the same position; There is sagging. When ask the patient to:
    - Look left, he will do it.
    - Look right, will do with some limitation.
    - Look down, he will do it.
    - Look up, he can not do it.
- Very common with **orbital floor fracture** because of tennis ball trauma.
- This patient had a trauma by fist, so the orbital floor got fractured. Inferior rectus is entrapped in the bone; When the patient looks up, the muscle cannot relax, but looking down, it can contract.
- Needs emergency correction. .



The left rim is not similar to the right rim because there is a fracture

Diagnosis: blunt fracture of the left eye.
Modality for investigation: CT scan



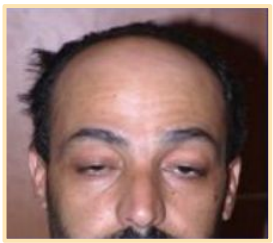
3. Mass

❖ Neuromuscular junction:

- **Ocular myasthenia gravis**: Myasthenia gravis could be an ocular condition affecting the ocular motility of the eye only without systemic manifestations.
- **Definition:** chronic autoimmune disease affecting the neuromuscular junction In skeletal muscles.
- **Clinical presentation:**
  - **Symptoms**:
    - PAINLESS ptosis (bilateral).
    - Diplopia (can present alone or together).
    - **Fatigability and variability** are characteristic (usually worse at the end of the day) unlike other palsies which are constant.
    - Ask the patient if symptoms are worse early in the morning or at the end of the day?).
    - Ask about systemic weakness, difficulty in swallowing or breathing. Majority present with ocular MG then eventually convert to general MG and may develop respiratory crisis!

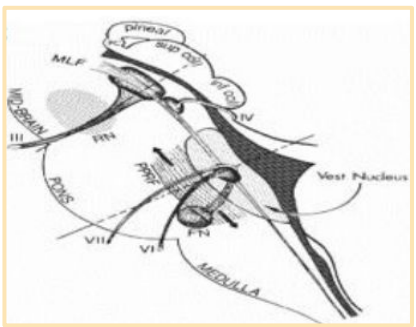
Efferent diagnosis

- **Examination:**
  - **Pupil is not affected**. This is how we can differentiate between MG and CN III palsy in which pupil is affected
  - Assess orbicularis strength (Ask patient to close eyes strongly & open them) If painful ptosis or there is pupil involvement don't say myasthenia gravis with your differential diagnosis.
- **Investigations:**
  - Blood test for acetylcholine receptor antibodies. 50% present in OMG.
  - Tensilon test: inhibits acetylcholinesterase and can transiently reverse signs of weakness due to OMG, such as ptosis and extraocular muscle paresis. Look for improvement in symptoms. You must be prepared for complications during this test including bradycardia and respiratory failure. You should have antidote (atropine) and resus kit. Diagnostic but we don't have it here.
- **Treatment:**
  - You don't need to know the details, but we can give steroids or Acetylcholinesterase inhibitors. And immunotherapy



Bilateral ptosis compensated by lifting the eyebrows (absence of forehead wrinkles indicates loss of frontalis muscle tone that could be secondary to myasthenia gravis). Eyelid did not go up.

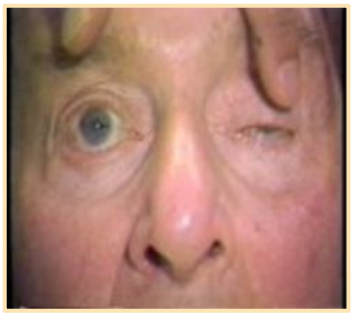
❖ Multiple cranial nerves



❖ Single cranial nerves:

1. Third (oculomotor) nerve palsy

Case

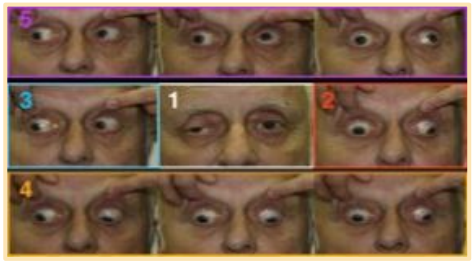


- Left eye:
  - Ptosis. (Levator Palpebrae muscle innervation)
  - Loss of adduction, infraduction & supraduction (3rd nerve affected).
  - The abduction is intact.
- 65 years old presented to ER complaining of double vision (typical presentation of CN III palsy always keeps it in your head!).
  - Ptosis and Eyes are (down & out).
  - Pupillary dilatation & no accommodation.
  - The eye rests in a position of abduction, slight depression, and intorsion.

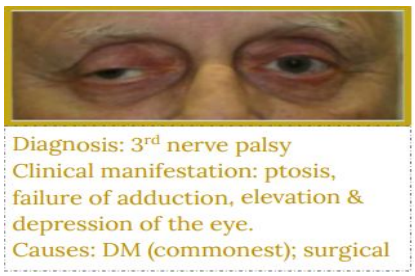
Efferent diagnosis

Case

- Examination of extraocular movement:
  1. Primary position: looking straight ahead.
  2. Looking to his left: abnormal.
  3. Looking to his right: normal.
  4. Mild infraduction limitation.
  5. Mild spraduction limitation.

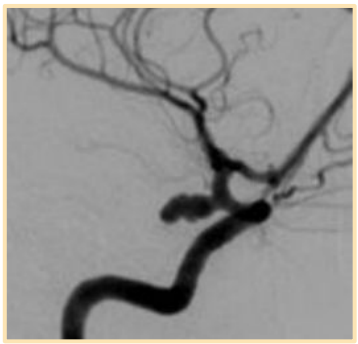


- Check for pupillary involvement:
  - To differentiate between surgical (urgent, compression, pupil involved, needs neuroimaging) and medical (pupil sparing) third nerve palsy.
  - Absence of pupillary involvement suggests a benign process that can be observed over a couple of weeks. A fixed, dilated pupil requires extensive neurologic evaluation.
- Etiology:
  - Intracranial aneurysm (posterior communicating artery)(not the most common but to ophthalmologists).
  - Micro-vascular ischemia (DM and HTN, DLP) If risk factors are controlled, the palsy will resolve by itself.
  - Trauma (Neuroimaging is indicated)
  - Brain tumor (Neuroimaging is indicated)



Diagnosis: 3rd nerve palsy
Clinical manifestation: ptosis, failure of adduction, elevation & depression of the eye.
Causes: DM (commonest); surgical

- Diagnosis: posterior communicating artery aneurysm (life threatening).
  - Right internal carotid artery injection reveals a right posterior communicating artery aneurysm with a tubular configuration. A very small anterior communicating artery aneurysm is also identified.
  - Magnetic resonance angiography (MRA) is the best investigation for PCA aneurysm. **You always have to rule out aneurysm, why?** Usually parasympathetic fibers go the outside (superficial) with CN III pathway, so any compression will lead to CN III palsy with pupil involvement.



- Pupils: innervated by parasympathetic fibers which is not part of oculomotor nerve, they run together and have the same pathway. The parasympathetic pupil-constrictor fibers from Edinger-Westphal nucleus travel within CNIII, and their loss gives you a “blown pupil”.

Efferent diagnosis

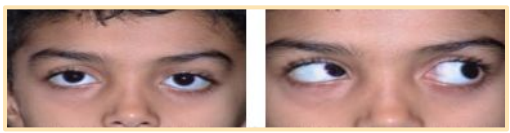
2. Fourth (trochlear) nerve palsy

◇ Clinical presentation

- It is usually difficult to be diagnosed by non-ophthalmologist.
- Patient will complain of vertical double vision only.
- **Vertical diplopia** (characteristic unlike CN III palsy which can have different types.)
  Head tilt to the opposite shoulder
- They mostly complain about actions that require downgaze vision; like: going down the stairs,eating, reading and writing.
- Head tilt to the opposite shoulder.
  - You might think the kid is shy and he doesn't want to interact, but actually he is just trying to avoid his double vision.
  - If you try to correct his head, you'll notice some hypertropia (a condition of misalignment of the eyes (strabismus).
  - If you move his head to the same side of the affected nerve it will be worse.

◇ Etiology:

- Congenital (commonest).
- Trauma even minor ones not only severe.
- Idiopathic.

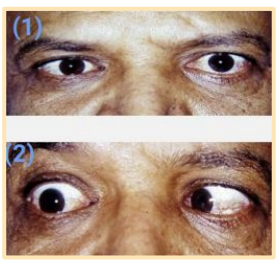


The light reflex is the left eye is lower than the right eye which means the right eye is shifted up

3. Sixth (abducens) nerve palsy

◇ Clinical presentation

- If someone has 6th nerve palsy, the lost movement is abduction (lateral rectus muscle movement). Unopposed MR.
- Mostly Diabetic Patients
- (medical recuts pulls over). If we cover the left eye, the patient will focus on the If a patient has loss of lateral rectus in the right eye, the patient will have esotropia right eye (deviated eye), then it will go back straight.
- Horizontal diplopia → two images beside each other (worse at distance).
- Because with near vision you need convergence you don't need CN VI, but "when looking far you need it for divergence (taking the eye out).
- Esotropia
  - When you do cover-uncover test, esotropia is more at distance than near.
  - Face turns in the direction of the paralyzed muscle to avoid its action.
  - Limited Abduction on the side of the lesion.



(1) Primary position
(2) Looking to his right, right lateral rectus is affected.
Diagnosis: right sixth nerve palsy.

Efferent diagnosis

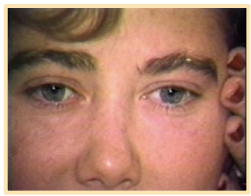
◇ **Etiology**

- You have to rule out:
  - Microvascular diseases (most commonly)
  - Intracranial tumors.
  - Trauma.
  - Increased intracranial pressure (we call it **false localizing sign** because you don't know exactly where the lesion is.)
    - The nerve passes through the Dorello canal at 90-degree angulation, this makes it susceptible to pressure due to any lesion in the brain. ex. a frontal tumor will cause pressure on this canal leading to 6th nerve palsy.

❖ Intraparenchymal problem

1. Internuclear ophthalmoplegia:

- If you ask the patient to look right or left, the lateral rectus should abduct, and medial rectus should adduct; What connect this movement together? MLF (medial longitudinal fasciculus), it is an interneuron between 3rd and 6th



- This interneuron connection can be injured by MS (most common cause in young patients), stroke (most common cause in old patients) or brainstem mass.
- What will happen if you have injured interneuron connection? The adducting eye cannot adduct, and the abducting eye will have an abducting nystagmus.

2. Gaze palsy

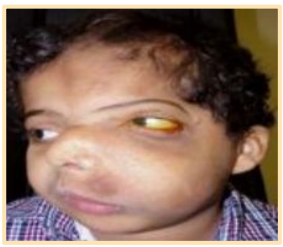
- The ocular complication of facial palsy is paralysis of orbicularis muscle → patient can't close the eye resulting in dryness.
- If you leave the dryness → abrasion → melting → perforation and eye fluid will come out.
- So, the cornea will perforate if the patient can't blink → iris will come out.
- Ointment is important, and during sleep, patient must tape the eye.

Diagnosis: right facial nerve paralysis
Ocular complications: keratoconjunctivitis, exposure keratitis

Efferent diagnosis

❖ Other

Unusual faces



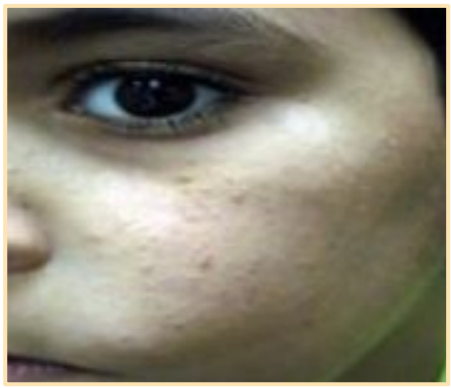
Shallow Orbit
Crouzon syndrome
Craniosynostosis

Optic Glioma
(neurofibromatosis)

Unusual postures

- The patient is trying to compensate for ptosis by lifting his chin.
- He could develop vertebral degeneration & spinal cord compression.

Unusual skin lesions



- This patient has a benign tumor of the skin. It is called adenoma sebaceum, indicating a disease called tubular sclerosis.(Tumor of sebaceous gland)
- If we see it, we have to scan with MRI for brain tumor. Those patients are prone to develop gliomas.

Doctor summary

Neuro-ophthalmology have

1. Afferent system (the optic nerve) how to examine the optic nerve?
   - Visual acuity
   - Color vision
   - Pupil exam
   - Visual field
   - Fundoscopy

➔ Any problems with these 4 indicate optic nerve pathology. how do you confirm?
   - Afferent pupillary reflex

2. Efferent system:
   - Inspection:is there lid retraction ptosis, deviation, are the pupils equal?
   - Examine:
     - Motility (range of movement, pursuit, saccadic)
     - Look at the eyelid is there ptosis proptosis

OPTHALMOLOGY TEAM

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