Lecture 7

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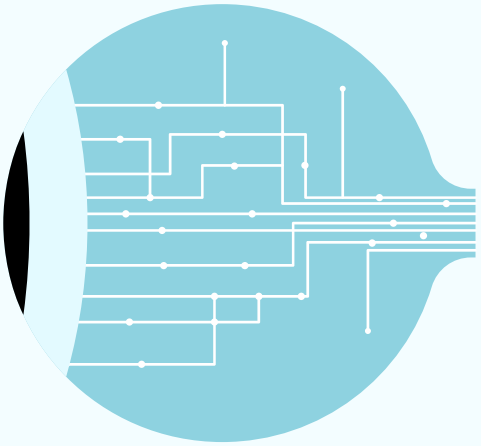


Neuro-ophthalmology

Anything not found in doctor Dania's slides will be labelled with 439 only

Presented by: Dr. Dania Alshowair

Objectives of the course:

- Review the clinical anatomy relevant to neuro-ophthalmology
- Identify and diagnose the common causes of pupillary, neuro-motility, neuromuscular and visual pathway disorders
- Understand the basis of management in neuro-ophthalmic disorders.

Color index:

Original content | Important | Golden Notes | Doctor's notes[439] | Doctor's notes[441] | Extra | Textbook

Afferent Anatomy

439 only

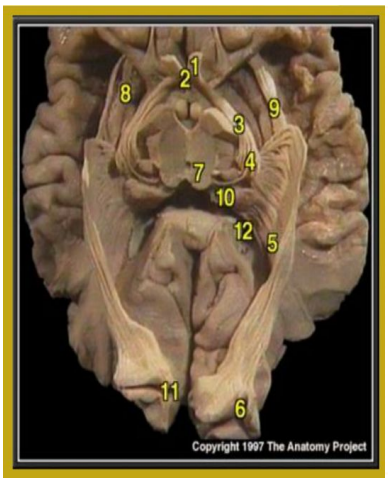
- **What is neuro-ophthalmology?**
  Subspecialty deals with visual problems that are related to the nervous system.

- **What is afferent (sensory) visual system?**
  The perception of vision (optic nerve and pupil reaction). Starts from the retina.

- **What is efferent visual system?**
  pupil size, 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.
  -

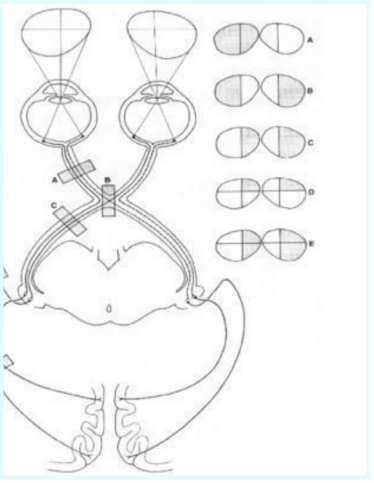
- In the visual pathway, we don't say lateral or medial, but we say nasal or temporal.
  - The temporal fibers are responsible for nasal visual field, and the nasal fibers are responsible for the temporal part of the field; There is a cross relationship.
- The afferent system starts at the retina (the 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; 53-55% of fibers cross & 47-45% of fibers remain uncrossed.



Name structure number 2 ?
Optic chiasm

Name structure number 3 ?
Optic tract

Name structure number 5 ?
Optic radiation



Afferent Visual System Examination

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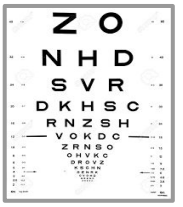
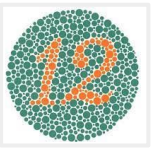
1.Visual acuity

- The visual acuity can be tested by projecting letters (Snellen chart) or by using E game chart (up, down, right, left).
  It is used for older children and adults (< 6 years, we use allen chart)



2.Color vision

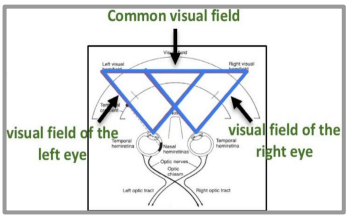
- Ishihara color chart



  - It should be done <u>mono-ocular</u>
  - 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 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.
- 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) .
- Advantages of having a common visual field between the 2 eyes :
  - 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.



- **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.
   - **Physiological (normal) blindspot:** is present in all people (both eyes) because the optic disc/nerve has <u>no</u> photoreceptors (so the optic nerve itself is a blindspot).
   - 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.

Afferent Visual System Examination

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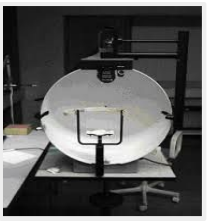
3.Visual field count…

- 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 (superior-temporal, inferior-temporal) and ask the patient to tell you how many finger does she/he see.

- 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 test (perimetry)** will measure the density of the blind spot or the quantity of the defect.(automated visual field test) ask the pt to click the button whenever they seen any light More commonly used then goldmann
   - 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

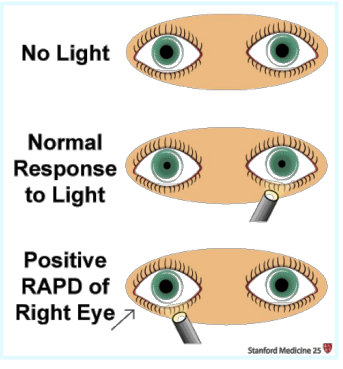
Afferent Visual System Examination

4.Pupils Examination

1. The size, shape and position of each pupil should be noted in light and dark conditions
2. Pupillary light reflex is best conducted in dim light room using a bright light
3. The patient should be relaxed and fixing on a distant object
4. Check light reflex looking for a relative afferent pupillary defect (RAPD)

- Shine light from down. Look at both pupils.Unequal pupil size: anisocoria,
- From the side of the patient, 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.
- 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).
- **In RAPD+ , when we shine light to one pupil, why does the other pupil dilate ?**
  If you shine the light over the affected pupil it will only take 50% of light and constrict accordingly (direct reflex), and the other pupil will also constricts by 50% (consensual reflex). If we move the light to the normal pupil : this optic nerve will take 100% of the stimulus > so both eyes will further constrict (direct & indirect reflex). When you move the light again to the affected pupil : it will again only take 50% of the light > so it dilates (because the stimulus reduced from 100% to 50%)

- **The components of near reflex are:**
    - Miosis → the pupil became smaller
    - Accommodation → The natural crystalline lens will increase its curvature. Occurs when looking to a near object.
    - Convergence → both eyes move towards the target (In opposite direction, 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.



Afferent Visual System Examination

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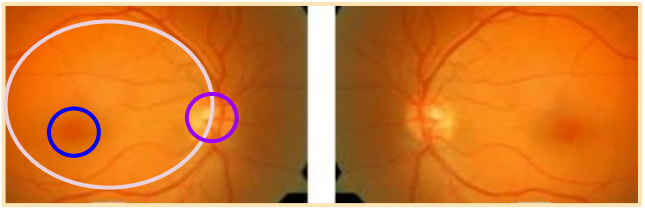
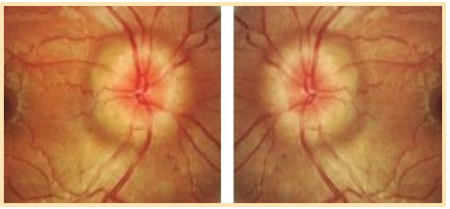
5.Fundus examination

- 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 (15 times) , smaller field & image is virtual (upright).
  3. **Indirect ophthalmoscope:** is a head-mount device that is binocular, magnification is less, you need to use a lens, so larger field and smaller image (usually we use lens with 20 dioptric power),real and inverted.
- **The Total dioptric power of the eye is 60** (lens = 20 , cornea = 40)
- The standard lens we are using in indirect ophthalmoscope is +20D. So the magnification is calculated 60\20=3. Which means the indirect offers 3 times magnification.

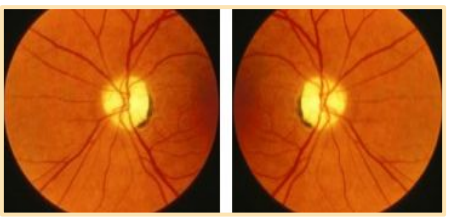
- Common question in exam: difference between direct and indirect ophthalmoscope (details in 1st lecture)



- This is what we see when we use slit lamp and hand-held lens.
  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
- Bilateral swelling
- Blood vessels are obscured bc of the fluid (disc swelling)



Very yellow pale nerve that indicates damaged atrophied nerve (optic atrophy)

Afferent Visual System Examination

439 only

5.Fundus examination count...

- **What is the normal appearance of the nerve on ophthalmoscope ?** slightly orange, reddish in color, has clear margins, a cup (depression) and neuro-retinal rim (remaining part).

- **Abnormalities of the optic disc :** disc elevation or disc atrophy

- **Fovea :**
  - Is the maximum point of fixation (gives us the color vision and the acuity).
  - If you injure the fovea, you will have a dramatic drop in vision.
  - It could be burned by laser used for retina, blue light laser, or the sun (causing **solar retinopathy**)

- **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.
  - **Causes of increased ICP :** tumors, infections, or benign idiopathic intracranial HTN
  - 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).
    - Benign idiopathic intracranial HTN (old name is pseudotumor cerebri) → very common in our community. It is the most common cause of disc elevation . Usually obese women and they present with nausea, vomiting, pulsatile tinnitus and headache. Can be caused by many medications : antibiotics (tetracyclines), vitamin A and vitamin E and OCP (they all increase ICP)

- **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 Examination

- **Optic nerve diseases:**
  - Usually unilateral
  - Afferent pupillary defect
  - Central visual loss
  - Loss of Color vision
  - Optic disc edema

- **Compression:** **not mentioned by the doctor**
  - Intraorbital ON
  - Intracranial ON
  - Optic chiasm
  - Optic tract
  - Posterior afferent system

- **Trauma:** **not mentioned by the doctor**
  - 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:**
  - Typical Optic neuritis (Most common type in young adults)
    - Inflammatory demyelinating condition
    - Linked to MS
      - 27 year old woman developed blurred vision OD and mild right periorbital pain, VA 20/50, MRI abnormal.
    - Majority is central Visual loss /color vision loss
    - Pain that worsen with eye movement. Why? Optic nerve is surrounded by EOM (mainly medial rectus)
    - Fundoscopic changes : hyperemic disc and the margins are blurred.
    - **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 (could be given after 3-5 days after IV treatment)

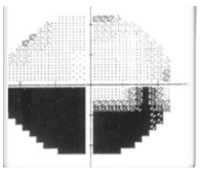
- **Ischemic optic neuropathy**
  - Non-arteritic ischemic optic neuropathy.
  - Arteritic ischemic optic neuropathy.
  - Central retinal artery occlusion. (emboli from a valvular disease, carotid plaque or post cardiac surgery)

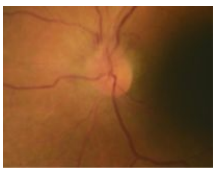
Afferent Visual System Diagnosis

Afferent Visual System Diagnosis:

Non-arteritic ION

- Patients often have DM, HTN, OSA and other vascular risk factor.
- Most common cause in older patients (above 40 years).
- Sudden painless acute visual loss
- Altitudinal visual field loss





- **Treatment**: no treatment, ask them to control the risk factors to protect the other eye.

Arteritis ION

- > 55 years old (older than non-arteritic ION).
- Associated with giant cell arteritis.

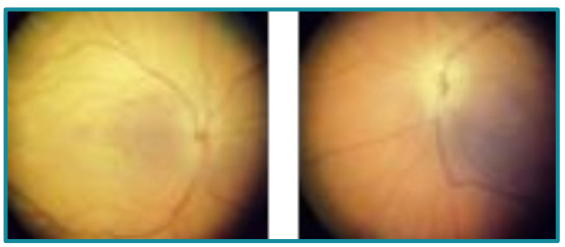
- **Symptoms :**
  - Severe visual loss. Present with severe irreversible visual loss (counting finger) more than non-arteritic (20/200).
  - jaw claudication
  - Proximal myalgia & arthralgia
  - Scalp tenderness/pulseless
  - Headache.

- **Diagnosis :**
  - 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. (take long segment 2.5-3 cm because the disease is involving segments of the arteries)

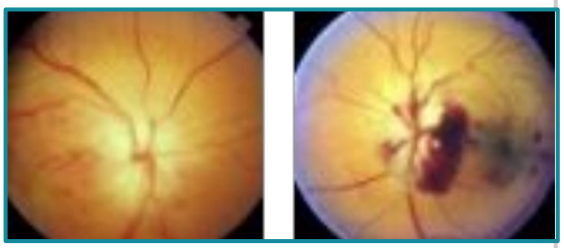
- **Treatment:** systemic steroids, given immediately if suspected even before the biopsy, patient should not leave the ER without it to protect the other eye (within the first day they will go blind) .

- Binocular involvement occurs in third of cases, often within the first day.

Giant Cell Arteritis, day 2



Giant Cell Arteritis, day 4



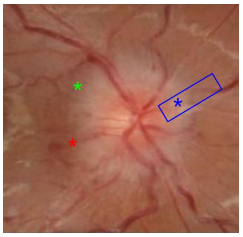
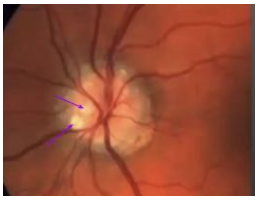
This is GCA. If you palpate the temporal artery in the affected side, it would be pulseless.

Afferent Visual System Diagnosis

Afferent Visual System Diagnosis:

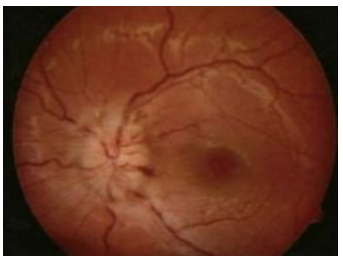
- **Congenital and genetic problems:**
  - Congenital retinal dystrophies.
  - Optic nerve hypoplasia.
  - Dominant and recessive optic atrophy.
  - Glaucoma.

  - (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 indicating ischemia (*) 3-Blood vessels can't be traced; some parts appearing and other disappearing (*) 	1- Optic disc margins blurred and the cup is absent 2- No edema, hemorrhage 3- May be associated with hyperopia 4- Drusens (*) 

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

- **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 = pseudopapilledema
  - **Cause:**
    - Intracranial mass
    - Severe systemic hypertension
    - Idiopathic intracranial hypertension (pseudotumor cerebri)



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

Afferent Visual System Tests

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Afferent Visual System Tests

- **During exam:**
  - Visual field test.
  - A and B scans (ultrasound of the eyeball) : when you have a patient with cataract and you can't see the fundus (optic nerve)
  - 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
  - MRI scan

- **Blood test:**
  - Vasculitis (ESR, CBC, ANA, VDRL) because the optic nerve can be affected by SLE, sarcoidosis ,TB, syphilis, behçet disease.
  - LFT (SGOT[1], SGPT[2], Alkaline phosphatase).
  - Urine analysis
  - Creatinine, BUN
  - Electrolytes

- **Ultrasound:**
  - Carotid doppler , orbital color doppler

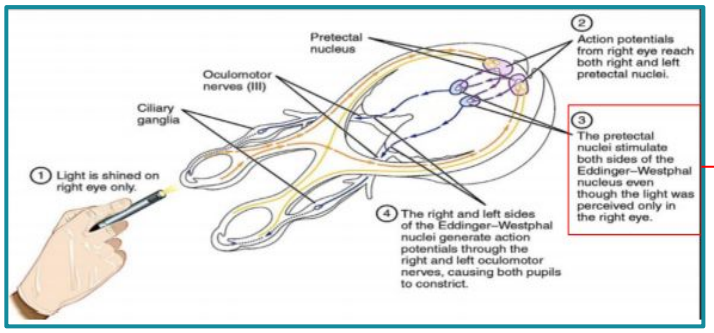
- **Genetic evaluation.**
  -

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

Pupillary Disorders

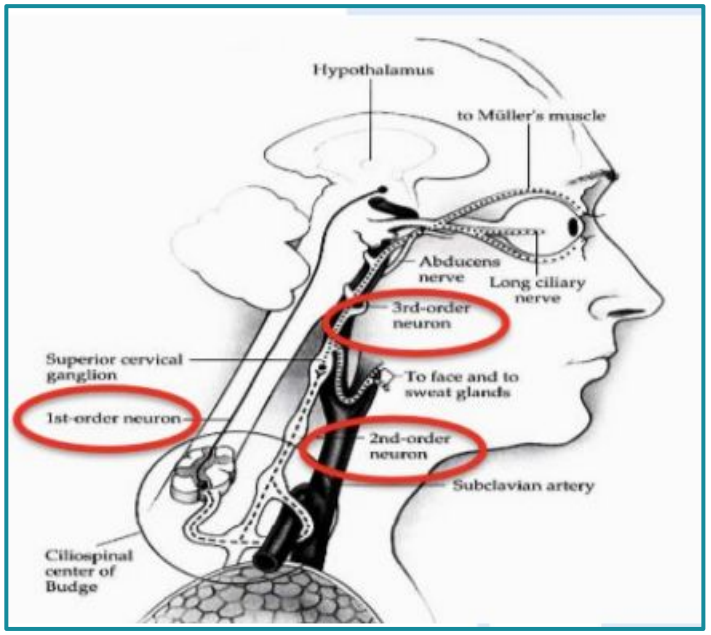
- **The anatomy and physiology of the pupil:**
  - The pupil size is controlled by a balance between parasympathetic innervation to the sphincter muscles and sympathetic innervation of the dilator muscles of the iris.
  - Pupil constrict to light and near stimuli.

- **Parasympathetic (cholinergic) pathway:**



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

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

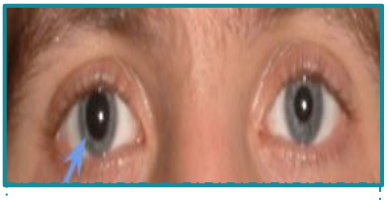


1-Sympathetic innervation of pupils starts from **Hypothalamus**
2-descends down as **1st order neuron** until **Ciliospinal center** (found at level C8 to T2)
3-then continues as **2nd order neuron** through **brachial plexus** where it crosses the apex of lungs (this is clinically important: Pancoast tumor can lead to horner syndrome due to this anatomical relation).
4-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.
5- from there **3rd order neuron** enters the cranium with internal carotid artery; dissection of the artery).
6-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**

Anisocoria is different pupillary size

  - 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.

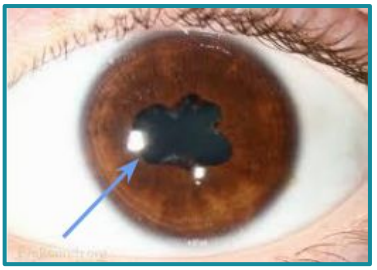


Right relative afferent pupillary defect.
In exam you should mention which side is affected

Pupillary Disorders

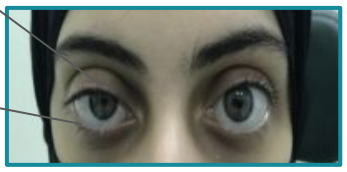
- **Simple (physiological) anisocoria**
  - 20 % of normal people
  - Difference in pupil size of less than or equal to 1 mm.
  - The degree of anisocoria is equal in dim and bright conditions
  - Intermittency or variability is a hallmark (it is not fixed to one pupil, it alternates)
  - There are no associated findings suggestive of a sympathetic or parasympathetic lesion. You must exclude any evidence of neurological lesion!

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. chief complaint
  ○ 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
	• 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)

- **Examples of pathological anisocoria**
  - **Holmes-Adie syndrome:**
    - Includes tonic pupil
    - Diminished deep tendon reflexes
    - Orthostatic hypotension.

  - **Horner syndrome:**
    - Small pupil (miosis)
    - Ptosis
    - Anhydrosis
    - Caused by a lesion anywhere along the sympathetic pathway



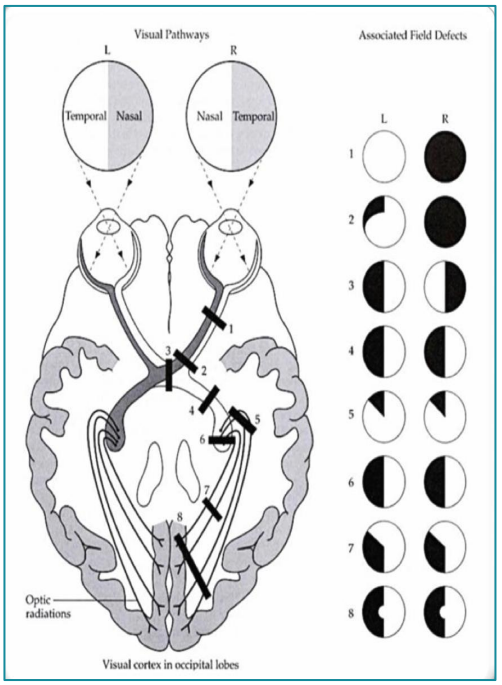
Any patient present acutely with horner syndrome you must exclude internal carotid dissection because the fibers run near the adventitia of the internal carotid artery.
Etiology post trauma.
Diagnosis by CT-Angio.

Visual pathway disorders

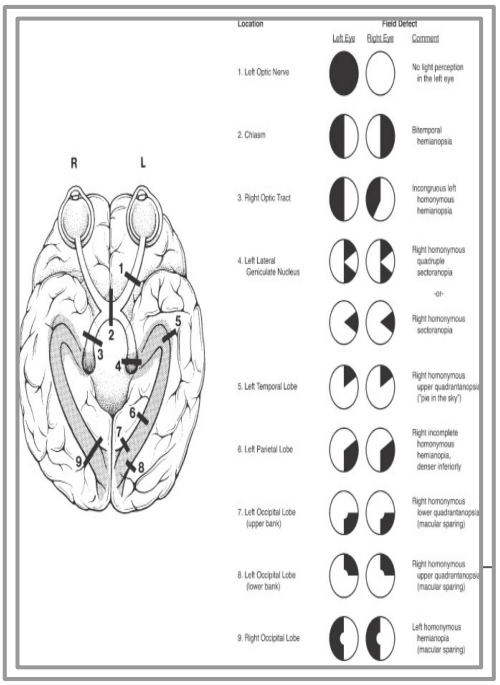
- **Terms:**
- **Anopia/anopsia:** Any blind area in the visual field
  1. If it's complete we call it complete anopsia
  2. Hemianopia: when ½ or ⅔ of the visual field is affected.
- **Quadrantanopia/Quadranopsia:** quarter of the field of vision.
  1. In quadranopsia, we have to specify superior or inferior.
  2. If it is more than quarter but less than half → we still call it quadrantanopia/quadranopsia.
  3. We also have to specify if there's macular sparing: mouth eaten part.
- **Homonymous :** same affected side in both eyes. **Heteronymous :** different affected sides
- **Incongruous :** not identical visual field defect

- **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 or 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 vice versa
    - Quadrant lesion means it's on one of the lobes either temporal or parietal
  - Macular sparing indicates occipital lesion
  - Wedge shaped space or wedge sparing indicates lateral geniculate nucleus lesion



441 slides



439 slides

Visual pathway disorders

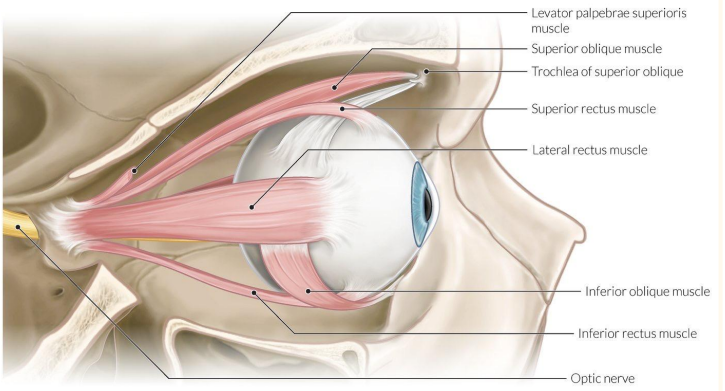
- **Localization of visual field defect** (the numbers correspond to 441 figure on the last page)

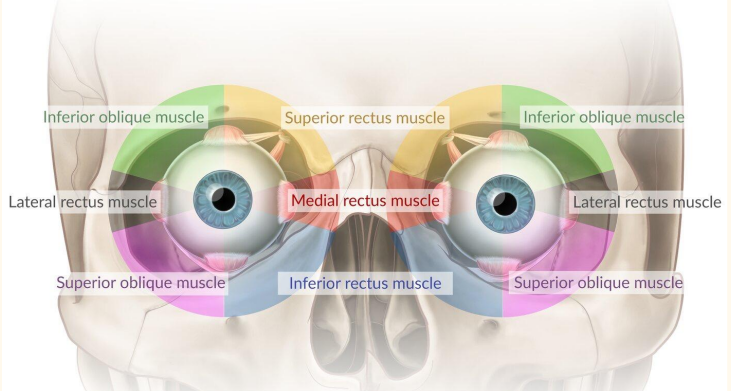
1. Optic nerve or eyeball → complete loss of the field (anopia)
2. Junction of the optic nerve and the optic chiasm→Junctional scotoma
3. Chiasm:
    - Any mass (e.g. meningioma, pituitary adenoma, craniopharyngioma, parasellar tumors, lactenoma, acromegaly or cushing syndrome) will press against the chiasm (chiasm compression) on the optic nerve (binasal decussating fibers) resulting in what is **called bitemporal** hemianopia (we can't say right or left here).
4. 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.
5. Temporal lobe:
    - Left homonymous superior quadrantanopia
    - The lesion is in the right .
    - We call it **pie in the sky.**
    - Since the defect is superior, so in the brain should be inferior (in the temporal lobe).
6. All optic radiation is involved.
7. Parietal lobe:
    - Left homonymous inferior quadrantanopia
    - The lesion is in the right.
    - Some people call it **pie in the floor**
    - Since the defect is inferior, so the lesion should be superior (in the parietal lobe).
8. Occipital lobe:
    - Occipital lobe lesions give identical visual field defect (symmetrical homonymous hemianopia)
    - 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 so the lesion in PCA
    - The defect in the left and the lesion in the right.

The Extraocular Muscles

- **Extraocular muscles:**
- **Motility is tested binocularly.**
- **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)

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





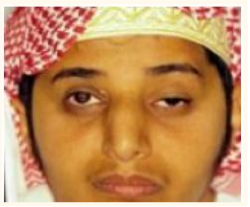
Effrent examination

439 only

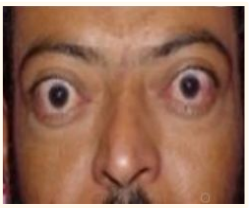
Just look at the patient? "inspection"

- ➔ Are eyes straight?
- ➔ No, the left eye is not straight (inward deviation) (**esotropia**).

- ➔ What are the lid positions?
- ➔ Left ptosis, the patient is lifting his eyebrow to compensate.
- ➔ Dilated pupils
  - ◆ 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.



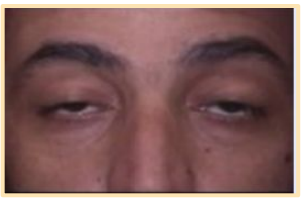
- ➔ Are the eyes proptotic?
- ➔ Exophthalmos, lid lag.
- ➔ Injected eyes.
- ➔ Scleral show, normally it is not seen **(Lid retraction)**
- ➔ It's common with thyroid diseases (thyrotoxicosis)



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

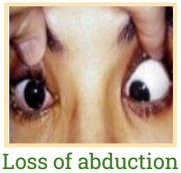
Movements of both eyes in all directions

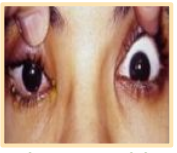
Have the patient move eyes in all directions, not just the direction where you think there is a problem. 9 positions of gazes (including central) . Vertical, horizontal , and X



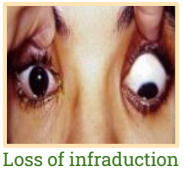
Hold lids if necessary (only after looking first without holding lids).

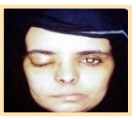
The pictures show : 3rd and 6th nerve palsy


Loss of abduction


Primary position


Loss of adduction


Loss of infraduction


Ptosis

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

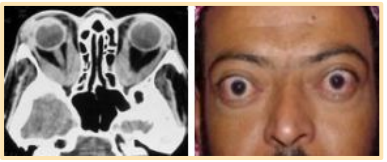
Effrent diagnosis

439 only

Orbit

1- Extraocular muscles:

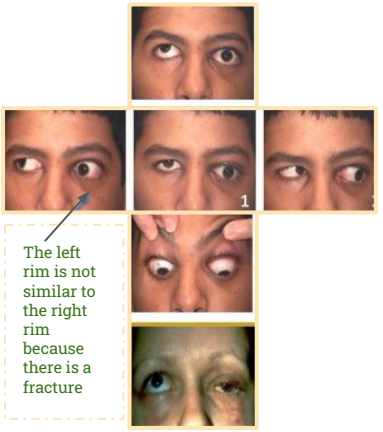
- In the CT, the muscles are enlarged, hypertrophied.
- This is thyroid orbitopathy; The muscles enlarge & can compress on the nerve and cause loss of vision if left untreated



Bilateral proptosis

2- Trauma

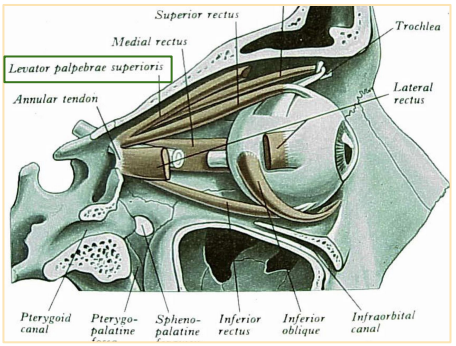
- 14-years old boy
- **In picture 1:**
- There is malalignment.
- Dropping of left eye (abnormal eyeball position = ocular dystopia).
- If we draw a line from the lateral canthus to the lateral canthus, they are not in the same position; There is sagging.
  When ask the patient to:
  - Look left, he 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



Effrent diagnosis

Neuromuscular junction:

Ocular myasthenia gravis

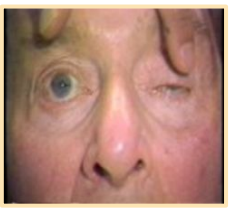
- **Ocular myasthenia gravis**: Myasthenia gravis could be an ocular condition affecting the ocular motility of the eye only without systemic manifestations.
- **Definition:** chronic autoimmune disease affecting the neuromuscular junction (Ach receptors) In skeletal muscles leading to weakness of the muscles.
- **Symptoms**:
  - PAINLESS ptosis (bilateral).
  - Diplopia
  - **Fatigability and variability** are characteristic (usually worse 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!
- **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.
  - CT chest to investigate thymoma
- **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.
Bell's phenomena (ask the patient to resist me opening his eye against forced closure) : if it's bilateral it may indicate MG or bilateral facial nerves palsies . However, unilateral indicates facial nerve palsy.

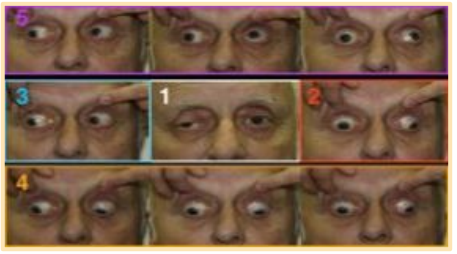
Effrent diagnosis

Single cranial nerves Cases :

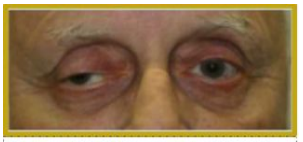
1-Third oculomotor nerve palsy



- Sign: "Left eye:"
  - Ptosis. (Levator Palpebrae muscle innervation)
  - Loss of adduction, infraduction & supraduction (3rd nerve).
  - 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.
- **Examination of extraocular movement:**
  - Primary position: looking straight ahead.
  - Looking to his left: abnormal.
  - Looking to his right: normal.
  - Mild infraduction limitation.
  - 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 very imp for ophthalmologist)
  - Micro-vascular ischemia (DM and HTN, DLP) (most common).
  - Trauma
  - Brain tumor



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

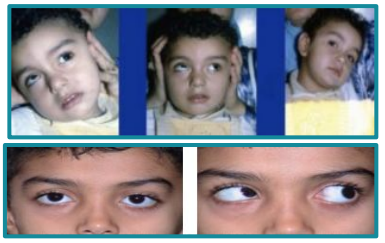
- Diagnosis:
  - Posterior communicating artery aneurysm (life threatening).
  - Do brain CT and CT angiography it's better than MRA in detecting small size aneurysms
  - **You always have to rule out aneurysm, why?** Usually parasympathetic fibers from Edinger-Westphal nucleus go the outside (superficial) with CN III pathway, so any compression will lead to CN III palsy with pupil involvement.

Effrent diagnosis

Single cranial nerves Case :

2-Fourth cranial nerve (trochlear) palsy

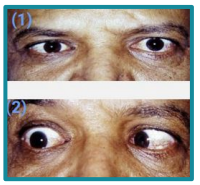
- **Clinical presentation**
  - **Vertical diplopia** (characteristic unlike CN III palsy which can have different types.)
  - Head tilt to the opposite shoulder
  - They mostly complain about actions that require downgaze vision; like: going down the stairs,eating, reading and writing.
  - You might 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
  - Medial rectus 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.
- **Etiology**
- You have to rule out:
  - Intracranial tumors.(most common)
  - Microvascular diseases
  - 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.

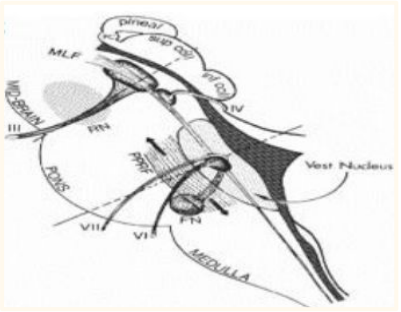


(1) Primary position
(2) Looking to his right, right lateral rectus is affected.
Diagnosis: right sixth nerve palsy

Effrent diagnosis

439 only

Multiple cranial nerves



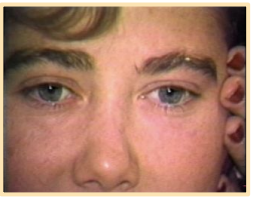
Intraparenchymal problem

1-Internuclear ophthalmoplegia:

- If you ask the patient to look right or left, the lateral rectus should abduct, and medial rectus should adduct; **What connects this movement together?**
  MLF (medial longitudinal fasciculus).

- 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 it will lead to abrasion → melting → perforation and eye fluid will come out.
- So, the cornea will perforate if the patient can't blink → iris will come out **(Exposure keratopathy)**
- Ointment is important, and during sleep, patient must tape the eye.

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

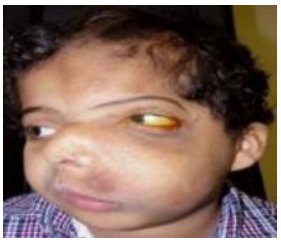
Effrent diagnosis

439 only

Other

1-Unusual faces:

- Shallow Orbit (prone to fracture)
- Crouzon syndrome
- Craniosynostosis



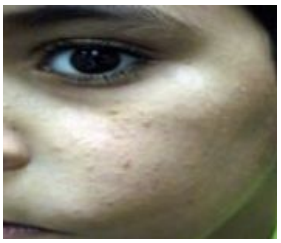
Neurofibromatosis :

- Hamartoma disease that affects the nerves and soft tissues causing tumors
- Features :
  - Pigmented lesions (cafe au lait spots)
  - Soft, pea-sized bumps/tumors on or under the skin
- Common presentation: involvement of orbit - **optic glioma** (shown in pic)

2-Unusual postures:

- The patient is trying to compensate for ptosis by lifting his chin.
- He could develop vertebral degeneration & spinal cord compression.
- Treatment: operate these patients quickly because of the risk of spinal cord injury

3-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

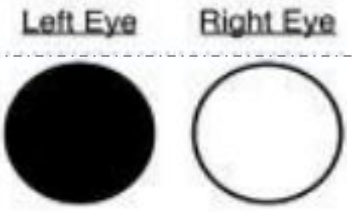
➔ Any problems with these 4 indicate optic nerve pathology. how do you confirm?

- 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

Visual pathway disorders

- **Very IMP! common question, name the defect & the location of the lesion** **Extra questions for practice, it's from 39 slides but you have to know it very well, either right or left, anything could come in the exam**



- **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

Lecture Quiz

Q1-Patient had surgery on the left temporal lobe, What type of visual field defect may he have as a complication?

A. Bitemporal hemianopia
B. Left homonymous superior quadrantanopia
C. Right homonymous superior quadrantanopia
D. Left homonymous hemianopia

Q2-80 year old hypertensive patient, presented to the ER with sudden visual loss, with history of jaw claudication, high CRP, what is the diagnosis ?

A. Optic neuritis
B. Retinal detachment
C. Giant cell arteritis
D. Central retinal artery occlusion

Q3-37 year old female with recurrent attacks of diplopia, bilateral ptosis, and fatigue that become worse at the end of the day, what is the most likely diagnosis?

A. Graves' disease
B. Third nerve palsy
C. Myasthenia gravis
D. Fourth nerve palsy

Q4-A 54 year old diabetic female presented with esotropia and binocular horizontal diplopia that worse at distance, what is the diagnosis?

A. Optic nerve disease
B. 3rd cranial nerve palsy
C. 4th cranial nerve palsy
D. 6th cranial nerve palsy

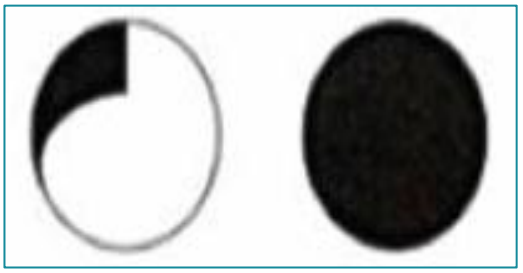
Q5-44 year old female presented with ptosis,miosis and anhidrosis, what is possible site of lesion that lead to patient condition?

A. Motor cortex
B. Lung apex
C. Occipital lobe
D. Heart valves

Answers: Q1: C | Q2: C | Q3: C | Q4: D | Q5: B

Short Answer Questions

Case 1



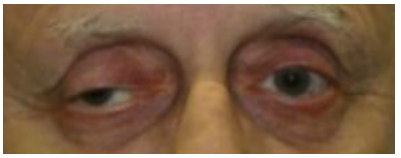
A: Name the visual defect?

B: What is the location of the lesion?

C: If the lesion was in occipital lobe, what will be the reason of macular sparing?

Case 2

A 66 year old diabetic and hypertensive patient complaining of ptosis, diplopia, failure of adduction, elevation and depression of the eye



A: What is the diagnosis?

B: Mention 2 common causes?

Answers:
Case 1
A: Junctional scotoma
B: Junction of the optic nerve and the optic chiasm
C: occipital lobe has a dual blood supply: the middle cerebral artery (MCA) and the posterior cerebral artery

Case 2
A: 3rd nerve palsy
B: 1. Microvascular ischemia (most common) 2. Posterior communicating artery aneurysm (important to rule out by CTA)

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