433 Teams

OPHTHALMOLOGY

10

Neuro-Ophthalmology

Color index:

432 Team – Important – 433 Notes – Not important

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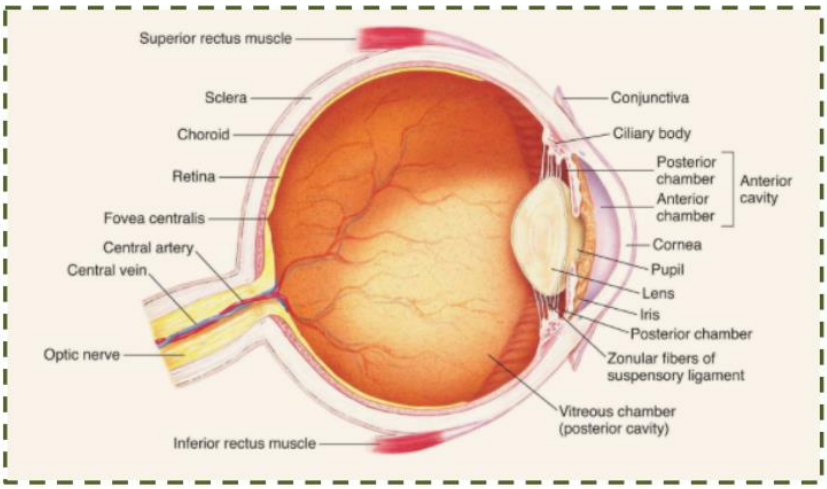
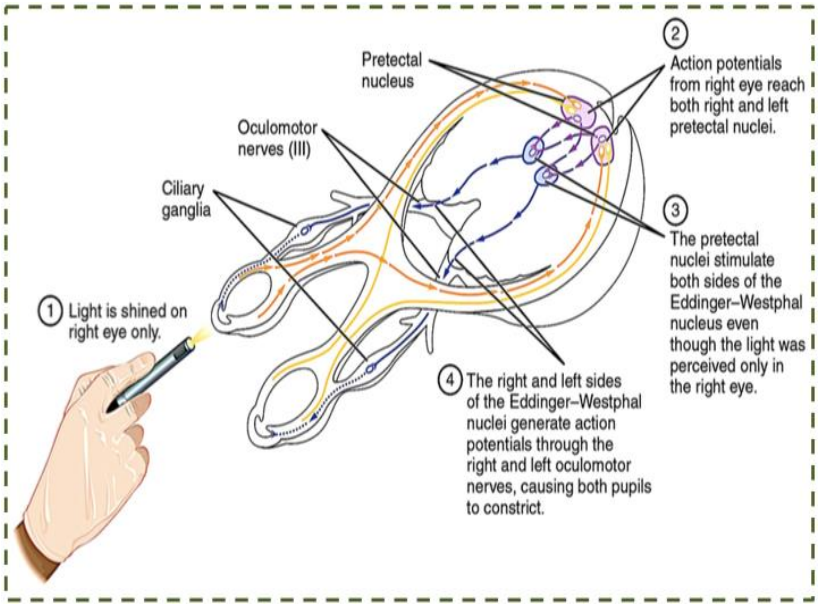
Anatomy and physiology:



Pupils:

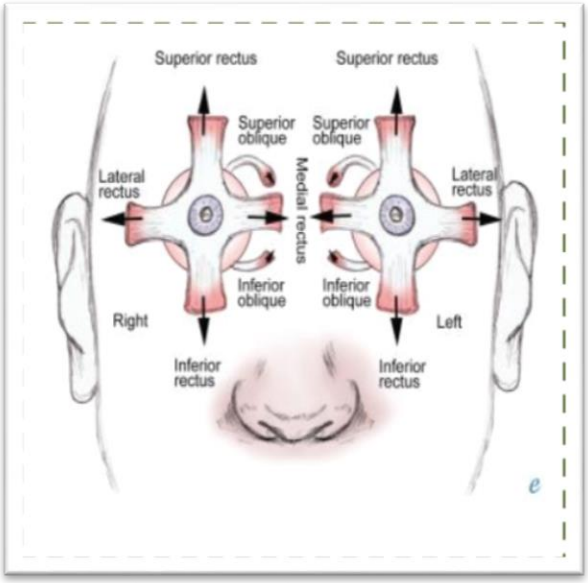
pupillary control: The physiology behind a "normal" pupillary constriction is a balance between the sympathetic and parasympathetic nervous systems.



Parasympathetic innervation leads to **pupillary constriction**. A circular muscle called the *sphincter pupillae* accomplishes this task. The fibers of the *sphincter pupillae* encompass the pupil. The pathway of pupillary constriction begins at the Edinger- Westphal nucleus near the occulomotor nerve nucleus. The fibers enter the orbit with CNIII nerve fibers and ultimately synapse at the cilliary ganglion.

Sympathetic innervation leads to **pupillary dilation.** Dilation is controlled by the *dilator pupillae,* a group of muscles in the peripheral 2/3 of the iris. Sympathetic innervation begins at the cortex with the first synapse at the cilliospinal center. Post synaptic neurons travel down all the way through the brain stem and finally exit through the cervical sympathetic chain and the superior cervical ganglion. They synapse at the superior cervical ganglion where third-order neurons travel through the carotid plexus and enter into the orbit through the first division of the trigeminal nerve.

Extraocular Muscles



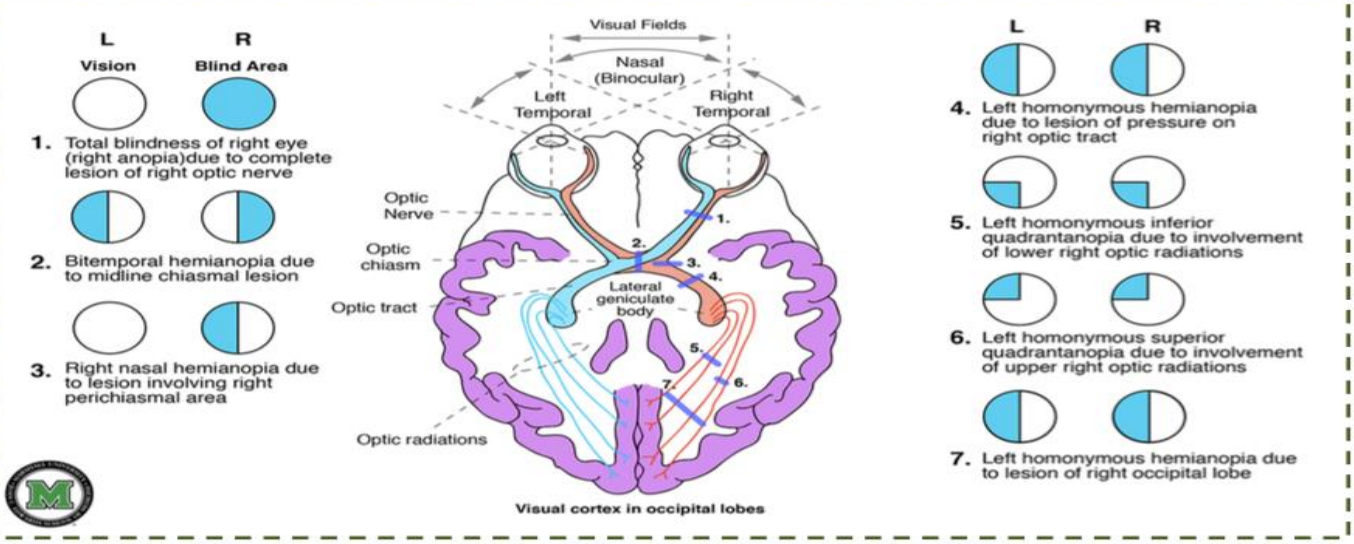
- There are six voluntary muscles that run from the posterior wall of the orbital cavity to the eyeball. These are the superior rectus, the inferior rectus, the medial rectus, the lateral rectus, and the superior and inferior oblique muscles.
- Medial and lateral rectus muscles have only horizontal actions.
- The medial rectus muscle is the primary adductor of the eye, and the lateral rectus muscle is the primary abductor of the eye.
- Superior and inferior rectus muscles are the primary vertical movers of the eye.
- The superior rectus acts as the primary elevator, and the inferior rectus acts as the primary depressor of the eye.
- This vertical action is greatest with the eye in the abducted position.
- The secondary action of vertical rectus muscles is torsion. The superior rectus is an incyclotorter (inwards rotator), and the inferior rectus is an excyclotorter (outwards rotator). The tertiary action of both muscles is adduction.

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

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

- **Trochlear nerve:** the fourth cranial nerve supplies the superior oblique muscle.
- **Abducent nerve:** the sixth cranial nerve supplies the lateral rectus muscle.
- **Oculomotor nerve:** the third cranial nerve begins as a nucleus in the midbrain that consists of several subneclei that innervates the individual extraocular muscles, the eyelids, and the pupils. It supplies the superior, inferior and medial rectus muscles and the inferior oblique muscle.

The Visual Pathway



- Visual field defects: if unilateral then think about optic nerve pathology, if bilateral then the pathology is at the optic chiasm or beyond.

- Chiasmal defects are always bitemporal.

- Homonymous visual defects could be due to stroke or tumors.

Pupillary Disorders:

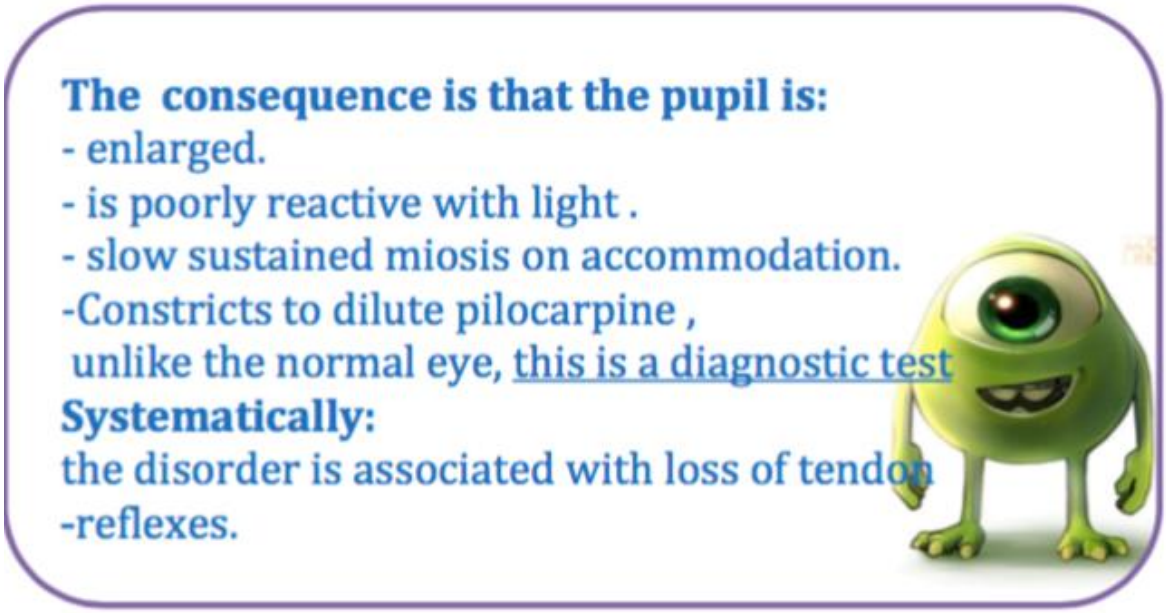
When the larger pupil does not constrict as well as the small pupil is response to light stimulus, then the large pupil is abnormal. **Possible causes:** previous ocular surgery, ocular trauma, use of medications like Cycloplegics e.g. Atropine, third nerve palsy, or **tonic pupil (Adie's pupil).**

When the small pupil does not dilate as well as the large pupil in dim light, then the small pupil is abnormal. **Possible causes:** previous ocular surgery, ocular trauma or inflammation, use of medications e.g. Pilocarpine, or **Horner syndrome.**

Tonic Pupil (Adie's Pupil)

Adie's tonic pupil is not uncommon cause of unequal pupil size (anisocornia) in young adults, but has no serious consequences. Onset is subacute and affects females more than males. It is due to *ciliary ganglionitis* which denervates the parasympathetic supply to the iris and ciliary body.

The pupil will constrict with near vision, but very slowly. That's why we call it a "tonic pupil" - it's tonically slow. the parasympathetic pathway is much shorter than the convoluted sympathetic pathway, so potential causes for damage are more benign. The parasympathetic plexus sits right behind the eye and can be damaged after an otherwise benign viral infection. It is a diagnosis of exclusion



Horner Syndrome

Interruption of the sympathetic pathway causes:

- A small pupil on the affected side due to loss of the dilator function.

- A slight ptosis on the affected side from decreased Muller's muscle action.

- An apparent recession of the globe into the orbit *(Enophthalmous).*
"posterior displacement of the eyeball"

- A lack of sweating "anhydrosis" on the affected side, if the sympathetic pathway is affected proximal to the base of the skull.

This syndrome is caused by lesion anywhere along the sympathetic pathway; carotid dissection, carotid aneurysms or tumor.

Neuromotility Disorders:

1-Third Cranial Nerve "Oculomotor" Palsy

A third cranial nerve palsy is caused by a lesion of the oculomotor nucleus within the midbrain or by compression of the peripheral course of the nerve by aneurysm or tumour. It leads to **drooping of the eyelid "ptosis", dilatation of the pupil that is unresponsive to light and accommodation, and an ability to move the eyeball upwards, downwards or inwards "adduction".**

Most third nerve palsies are caused by ischemic events at the nerve secondary to **hypertension or diabetes.** The one thing you really need to worry about in these patients is a **compressive aneurysm pushing on the nerve.** These aneurysms occur at the junction of the posterior communicating artery and the internal carotid artery. Compressive lesions usually affect the parasympathetic nerve component: a blown pupil is a potential emergency. Whenever you have pupillary involvement, you need an **MRI and angiography to rule out a dangerous aneurysm or tumour .**

The patient presentation: the eye is deviated down and out, ptosis, pupillary dilatation and paralysis of accommodation.

2-Fourth Cranial Nerve "Trochlear" Palsy

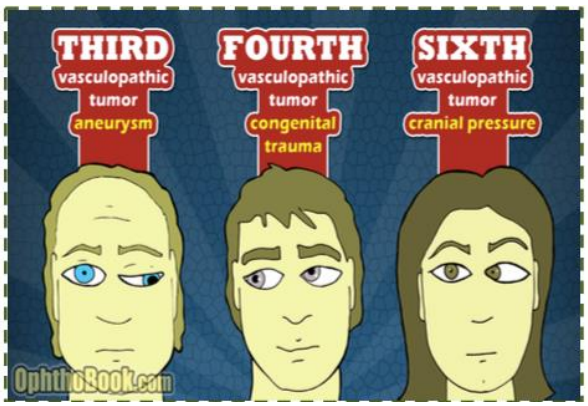
Trochlear paralysis is the hardest cranial nerve palsy to diagnose. These patients have an **upward deviation of the affected eye and a "cyclotorsion" twisting of the eye that makes them tilt their head away from the lesion.** A trochlear nerve lesion is caused by either trauma, an ischemic event, or can be congenitally present with later decompensation. The fourth cranial nerve is the skinniest nerve and runs the longest distance inside the cranial vault. This long passage makes it more susceptible to injury and neoplasm. The fourth nerve is also susceptible to being pulled from the root where it exits from the back of the brainstem. More fourth palsies occur in elderly males from trauma and more congenital palsies are found in the pediatric population. Ask about history of **closed-head injuries and check old photographs for head-tilt - this would indicate an old/congenital palsy that has recently decompensated.**

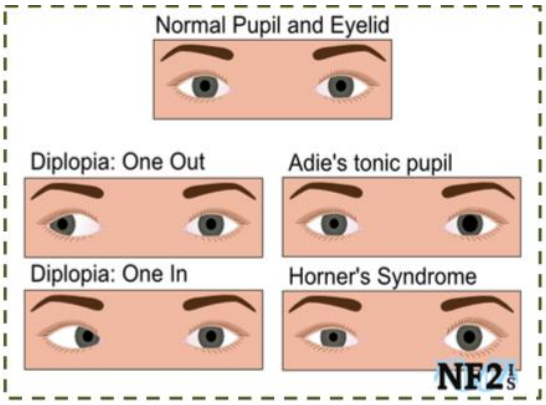
3-Sixth Cranial Nerve "Abducent" Palsy

A sixth cranial nerve palsy is caused by a lesion of the abducens nucleus in the pons or by compression of the peripheral course of the nerve by an aneurysm or tumour. It leads to **inability to move the eye outwards "abduction".**

Patients will go cross-eyed "*Esotropia = one or both eyes turn inward*", so to compensate they may turn their head to avoid double vision.

The patients might present with **horizontal diplopia "worse at distance".** This condition could be caused by intracranial tumors, trauma, microvascular diseases, or raised intracranial pressure.





Neuromuscular Disorders:

Ocular Myasthenia Gravis

Myasthenia gravis is a rare autoimmune disease in which the body develops autoimmune antibodies to the nicotinic acetylcholine receptors located at the neuromuscular junction of striated muscle. This leads to fatigable muscles and often involves the eye, causing diplopia and ptosis. Once the number of functional Ach receptors drops below 30% normal, then the patient becomes symptomatic and easily fatigued.

The diplopia and ptosis is usually worse on prolonged upgaze: you can test this by having your patient look at your raised finger to see who tires out first. More definitive diagnosis can be made via the Tensilon test where you give edrophonium chloride (an anticholineresterase) and look for an improvement in symptoms as their Ach levels build up.

Systemically, these patients can have problems with mastication, talking, drinking, and swallowing. Aspiration pneumonia and respiratory failure from inability to clear secretions is the big killer with this disease.

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

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

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

- Tensilon test is diagnostic.

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

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

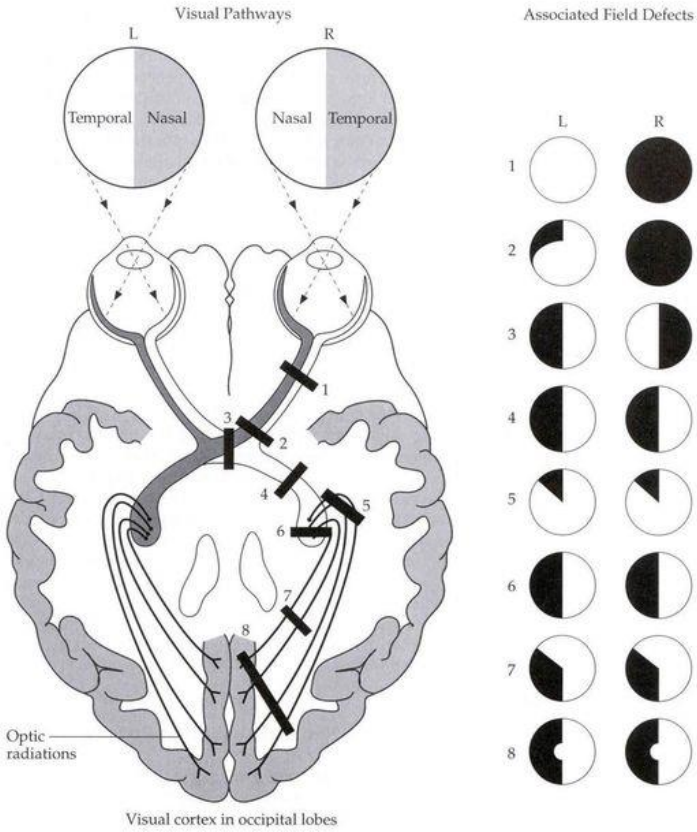
- The pupils are not affected.

Visual Pathway Disorders:

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

Optic Nerve Disease

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



1. Optic Neuritis

Inflammation or demyelination of the optic nerve results in optic neuritis. This is termed *papillitis* if the optic nerve head is affected and *retrobulbar neuritis* if the optic nerve head is affected more posteriorly with no disc swelling.

The cardinal signs in these patients are: Reduced visual acuity, reduced color vision, RAPD*, Central scotoma on field testing, normal disc in retrobulbar neuritis, swollen disc in papillitis.

Pain with eye-movement, enhancement of the optic nerve on MRI, and potential association with multiple sclerosis. This occurs in younger patients.

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

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

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

IV steroids may speed up the recovery process but does not influence the final outcome.

- Optic neuritis is more is females.

- Oral steroids if given alone might increase the risk of recurrence.

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2. Ischemic Optic Neuropathy

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

Could be **Non-arteritic IOP or Arteritic IOP.**

Non-arteritic IOP: patients often have diabetes mellitus, hypertension, and other vascular risk factors. Common in old patients and presents with altitudinal visual field loss.

Arteritic IOP: seen in patient **older than 55 years** and mostly associated with **giant cell arteritis (GCA).** They present with sudden loss of vision, scalp tenderness, pain on chewing "jaw claudication", shoulder pain and malaise.

Signs:

- Reduction in visual acuity
- Field defect
- Swollen and hemorrhagic disc with normal retina and retinal vessels – in AIOP the disc may be pale.
- Small normal fellow disc with a small cup in NAIOP.
- Tender temporal artery, suggestive of giant cell arteritis.

Investigation:

If giant cell arteritis is present, **the erythrocyte sedimentation rate (ESR) and C- reactive protein (CRP) are usually grossly elevated. Temporal artery biopsy is the gold standard for diagnosis.**

Systemic steroids should be given immediately if GCA is suspected.

3. Congenital Disc Elevation

it is a rare disease. Optic disc margins are blurred and the cup is absent but no edema or haemorrhage can be observed. May be associated with hyperopia "farsightedness" or drusen "yellow deposits under the retina made up of fatty proteins".

Other causes of optic neuropathy:

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

Papilledema

It is a bilateral swelling of the optic disc secondary to raised intracranial pressure. Could be caused by intracranial mass, severe systemic hypertension, or idiopathic intracranial hypertension (pseudotumor cerebri).

Symptoms :

- Headache, worse on awakening
- Nausea and vomiting if the raise in ICP is severe, may be followed by loss of consciousness, pupillary dilatation and death.
- Pulsatile tinnitus
- Visual symptoms often are absent
- **Signs:**
  - Disc hyperemia
  - There is no spontaneous venous pulsation of the central retinal vein.
  - Blurring and elevation of the disc margins
  - Peripapillary flame shaped haemorrhage

DDx:

- Adult optic neuritis
- Hypertension

- Papillitis: edematous or inflamed optic disc.

- Important signs in optic nerve disease: blurring of the margins, splinter hemorrhage in the peripapillary area, edema and elevation of the disc.

- The presence of hemorrhage = acute raise in the pressure.

- Papilledema is a diagnosis of exclusion should be confirmed by lumbar puncture.

- Idiopathic intracranial hypertension
- Pseudopapilledema

investigations:

- CT or MRI followed by lumbar puncture.
- B-scan ultrasonography to rule out buried disc drusen.
- Fluorescein angiography

If Negative think about idiopathic intracranial hypertension (pseudotumor cerebri) and look for risk factors: Obesity, OCP, PCOS, high vit.A. and the treatment; acetazolamide or shunt.

Pupillary examination:

It is very useful check it out:

http://www.neuroexam.com/neuroexam/content.php?p=19

Summary

Pupillary disorders:

Tonic pupil (Adie's pupil): - enlarged, is poorly reactive with light, slow sustained miosis on accommodation, Constricts to dilute Pilocarpine , unlike the normal eye, this is a diagnostic test.
Systematically: the disorder is associated with loss of tendon reflexes.

Horner's syndrome: Small pupil, enophthalmos, anhydrosis, ptosis.

Neuromotility disorders:

3rd: drooping of the eyelids "ptosis", dilatation of the pupil that is unresponsive to light and accommodation, and an inability to move the eyeball upwards, downwards, or inwards "adduction".

4th: an upward deviation of the affected eye and a "cyclotorsion" twisting of the eye that makes them tilt their heads away from the lesion.

6th: inability to move the eye outwards "abduction". Patients will go crossed-eyed "Esotropia = one or both eyes turn inward".

Neuromuscular disorders:

MG: ptosis with or without diplopia, the pupil is intact.

Visual pathway disorders:

Optic neuritis: disc edema except if retrobulbar neuritis, reduced visual acuity and color vision, central scotoma, RAPD.

Papilledema: swollen disc, blurred margins, capillary dilated, no venous pulsation.

Ischemic optic disease: field defect, swollen disc with normal margins, tender temporal artery if (Giant cell arteritis).

Congenital disc elevation: optic disc margins are blurred and the cup is absent but no edema or hemorrhage can be observed. May be associated with hyperopia.

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