

**MOTOR
DISTURBANCES IN
CRANIAL NERVE
SUPPLY**

PTOSIS

NEUROLOGY STATION

Degrees of Ptosis

Within the limits of human variation, usually the upper eyelid is about the level of the upper border of the cornea (see Fig 33).

- **Complete ptosis:** The upper eyelid covers the whole eye
- **Moderate:** The eyelid comes to the pupillary level
- **Severe:** The eyelid is below pupillary level
- **Mild:** A few mm of the upper cornea is covered

Congenital ptosis and Horner's syndrome produces only a mild ptosis, while severe and complete ptosis are seen with severe lesions of the III nerve.

Pathophysiology of Ptosis

Ptosis is due to weakness of elevators of the upper eyelid. The elevators of the upper eyelid are levator palpebrae superioris (LPS) and the Mullerian muscle supplied by the cervical sympathetic nerve. So, ptosis is a weakness of LPS or the Mullerian muscle.

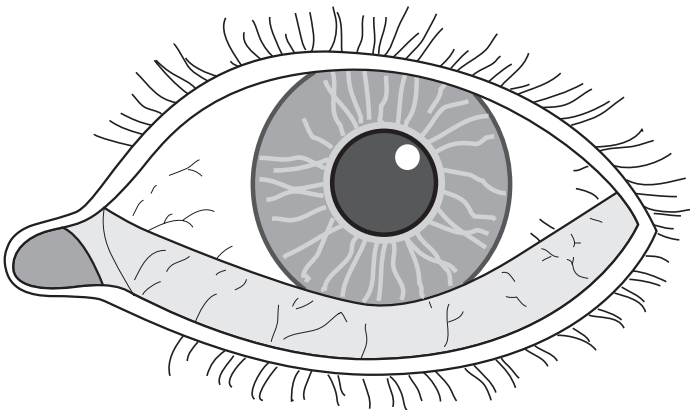


Figure 33: Eyelids in relation to cornea

Useful Differential Points for Diagnosis of Ptosis

- Duration and progress (old photographs are of immense value)
- Laterality
- Severity
- Presence of pain
- Diplopia
- Pupillary size and reaction
- Neighbouring cranial nerve symptoms and signs
- Family history
- Long tract signs

PTOSIS

Condition	Uni/bilat	Severity/ pain	Temporal profile	Family history	Diplopia	Pupils size light accom.	Other features
Horner's	unilateral	v. mild painless	variable	0	0	small Difficult to see	meiosis enophthalmos
Myopathy	bilateral symmet.	mod painless	v. slow progressive	+	0	normal	myotonia ± facioscap
Myasthenia	bilateral	variable painless	diurnal variation	0	+	normal (except cholinergic)	bulbar, limb
3rd nerve							
a) Orbital	unilateral	variable	progressive	0	+	variable	proptosis squint
b) Sup orb Fissure	unilateral	moderate painful	acute	0	+	normal	ophthalmic sensory imp.
c) Cavernous sinus	uni/bil	severe painful	acute progressive	0	+	normal	chemosis, oph. max. sens. imp.
d) Post comm. aneurysm	unilateral	severe painful	acute	0	+	dilated	0 0 hypertensive elderly
Mid brain	uni>bil	moderate painless	variable	0	+	variable	long tract, pyr, extrapyr.
Congenital	uni/bilat	mild painless	static	+	0	normal	Marcus Gunn jaw winking ±
Tabes dorsalis	bilateral	mild painless	insidious static	0	0	small irregular	0 + A incomp. AJ -
Migrainous neuralgia	unilateral	mild painful	acute	+	0	small regular	+ + lacrimation conj. Injectin
Cranial Polyneuritis	uni/bilat	moderate	acute painful	0	+	normal	other CN ^ ESR ^ CSF protein

Sites of Lesion and Causes of Ptosis

Muscle – Ocular Myopathy

Myopathy of ocular muscles occurs in ocular myopathy (Autosomal Dominant AD), and in myotonia dystrophica (AD). There is insidious, slowly progressive, bilateral and symmetrical ptosis with external ophthalmoplegia. As ophthalmoplegia is symmetrical there is usually no diplopia.

Neuromuscular Junction – Myasthenia Gravis

This is the most common presentation of myasthenia. Often the ptosis is bilateral and asymmetrical with accompanying ptosis. There is often a history of varying severity of ptosis and diplopia. Ptosis alternating from one eye to the other is almost always due to myasthenia.

Fatigability can be demonstrated by repeated blinking or a continued upward gaze. Look also for fatigability of speech by asking the patient to count from 1–110. There is a difference in the quality of the 1–10 count and the 100–110 count. There may be dysphagia, nasal regurgitation and fatigable limb weakness.

Edrophonium ('Tensilon') Test

Edrophonium ('Tensilon') is available in 1 ml vials for intravenous use only. (1 ml=10mg). A test dose (0.2 ml = 2 mg) is first given and if there are no cholinergic effects, the remainder is given.

Improvement in ptosis, diplopia, speech, swallowing or limb weakness is noted. It is essential to decide which parameter is to be observed after the intravenous injection.

The test dose of Edrophonium is also used for:

- Differentiating a cholinergic crisis from a myasthenic crisis
- Noting adequacy of the dose of the cholinergic drug – by giving it one hour after oral neostigmine or two hours after oral pyridostigmine (time of maximal effect of drug).

Cholinergic Effects

Cholinergic effects are seen with an excess of an acetyl choline effect during treatment or with edrophonium, even with a test dose. Acetyl choline at the sites of action produces the following:

- **Neuro muscular junction:** Fasciculation
- **Sweat glands – (Cholinergic):** Excessive sweating.

At the parasympathetic endings of the cranial nerves:

- **III:** Constricted pupil
- **V,VII:** Excessive salivation and lacrimation
- **IX,X:** Bronchospasm and secretions, abdominal cramps and diarrhoea, bradycardia – occasionally cardiac arrest if edrophonium is given too rapidly to a sensitive subject.

Third (III) Nerve Palsy

In Orbit

Disorder has to affect the superior division of the III nerve supplying the LPS and the superior rectus muscles. Other nerves may be affected, producing a variable external ophthalmoplegia. There will be a downward proptosis.

Superior Orbital Fissure (SOF)

It is useful to remember the structures in SOF as **LFT SONIA** and the nerve supply as LR6 SO4, ie lateral rectus supplied by the VI nerve and superior oblique by the IV nerve and all others by the III nerve.

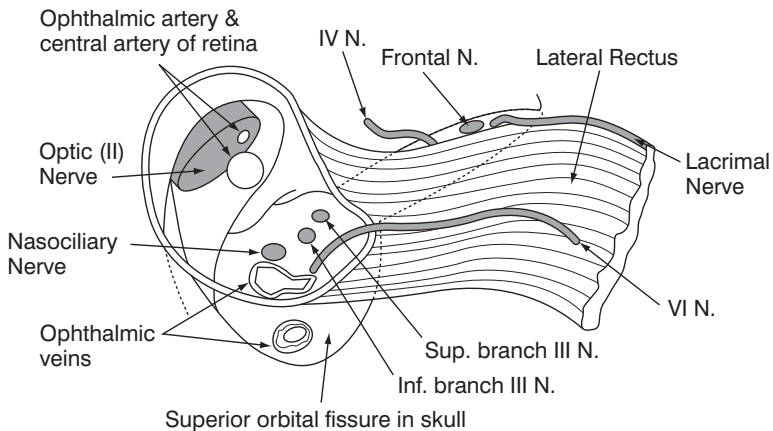


Figure 34: Third nerve in orbit and superior orbital fissure

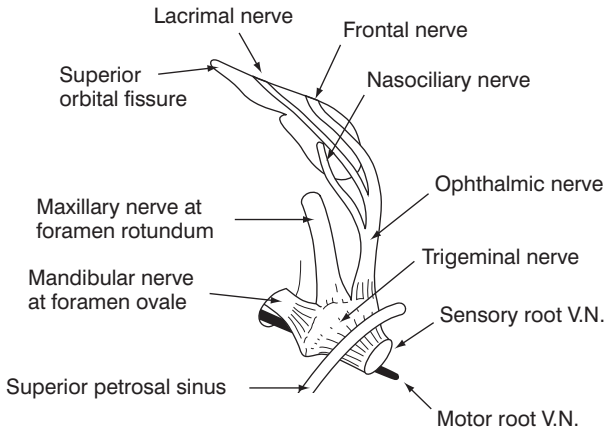


Figure 35: Superior Orbital Fissure and branches of v nerve

LFT SONIA stands for:

Lacrimal} Sensation in the ophthalmic division

Fronta} of V

Trochlear: IV nerve supplying superior oblique muscle

Superior division of the oculomotor nerve

Ophthalmic veins

Nasociliary: Branch of ophthalmic V nerve including the nose. When vesicles appear in the distribution of this nerve in the ophthalmic zoster, there is a danger of affection of the cornea.

Inferior division of the oculomotor supplying the inferior rectus and inferior oblique.

Abducens: VI nerve supplying the lateral rectus.

The superior oblique is a depressor of the adducted eye. However, if the III nerve is paralysed, adduction is not possible. An attempt to look down produces internal rotation of the eye, when the IV nerve is intact in the presence of a III nerve palsy.

Cavernous Sinus

All structures finding their exit through the SOF are also found in the cavernous sinus. The cavernous sinus has one additional structure which exits through the foramen rotundum – maxillary branch of the V nerve (see Fig 36 on page 102).

So it is important to look for impaired sensation over the maxilla to differentiate a cavernous sinus lesion from an SOF lesion. Additional features with thrombosis of the cavernous sinus include proptosis, chemosis, conjunctival injection, engorged retinal veins and sometimes papilloedema.

Infranuclear

Posterior communicating aneurysm is the common lesion at this site producing an acute, painful, unilateral ptosis with a dilated pupil in an elderly hypertensive patient.

Mid Brain

In addition to the III nerve palsy, other structures affected include:

- **Pyramidal tract:** This produces a contralateral hemiparesis (crossed hemiplegia) also known as Weber’s syndrome
- **Red nucleus:** Action tremor
- **Medial lemniscus:** Contralateral hemi-anaesthesia
- **Superior cerebellar peduncle:** Ipsilateral cerebellar signs
- **Superior colliculus:** Impaired upward gaze.

See Fig 8 on page 21.

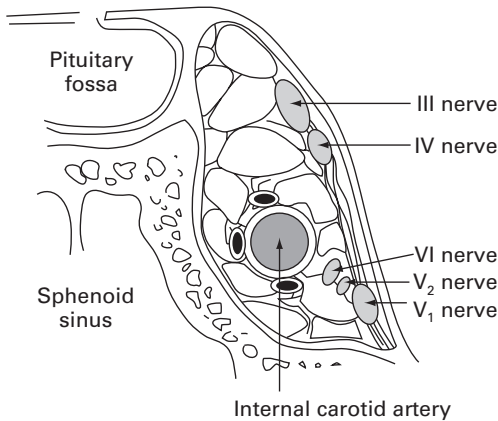


Figure 36: Cavernous sinus

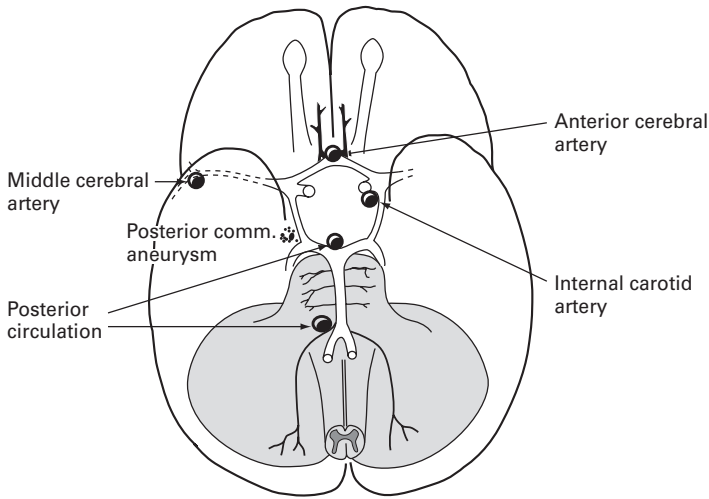


Figure 37: Posterior communicating aneurysm

Horner's Syndrome

This produces only a mild ptosis with only a partial coverage of the cornea by a few mm. Irrespective of the site and cause of the lesion the mild ptosis is accompanied by:

- **Meiosis (small pupil):** The pupil reacts to light and accommodation but the reaction may be difficult to note as the pupil is already small. The opposite reaction (dilation) is better seen when the eye is shaded or the patient looks far – this is the opposite of the usual technique for light and accommodation responses.
- **Enophthalmos:** This is the opposite of exophthalmos. The cornea is covered above by the upper eyelid and below by the lower eyelid.
- **Conjunctival injection:** This is due to a loss of vasoconstrictor activity.
- **Absence of sweating:** The area involved depends on the site of the lesion.

Sites of Lesions Producing Horner's Syndrome

Sympathetic pathway starts in hypothalamus, but clinically lesions are seen in the brain stem, cervical cord, T1 nerve root, neck and internal carotid artery – extra and intracranially (see Fig 38 on page 104).

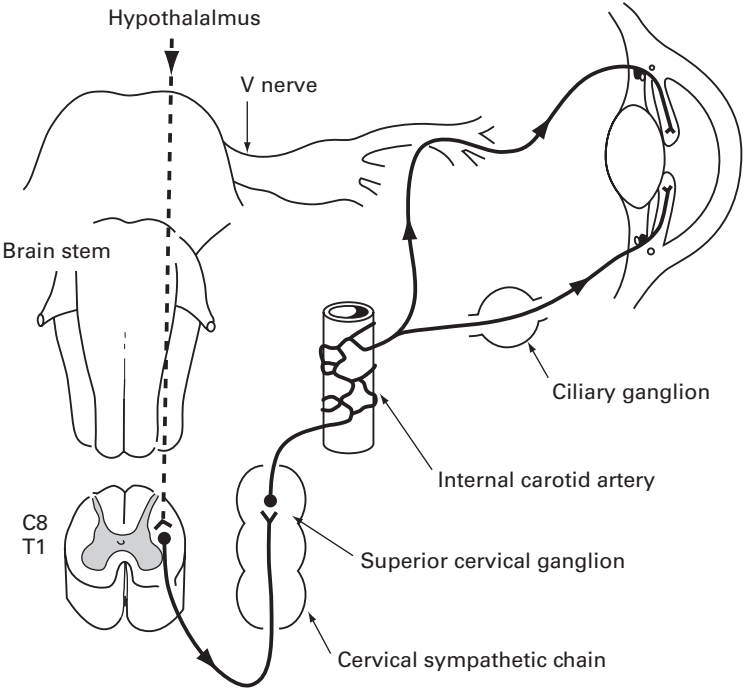


Figure 38: Sympathetic pathway

Brain Stem

Posterolateral lesions of the brain stem as in lateral medullary syndrome of Wallenberg, multiple sclerosis, brain stem encephalitis and pontine glioma, may produce a Horner’s. It is often associated with contralateral hemianaesthesia due to proximity to the spinothalamic tract (see Fig 39 on page 105).

Cervical Cord

This is due to affection of the lateral column at T1 level. Pain and temperature fibres around the central canal are also affected, producing dissociate sensory loss in the arms with hyporeflexia. Ptosis is mild and bilateral with symmetrical small reacting pupils.

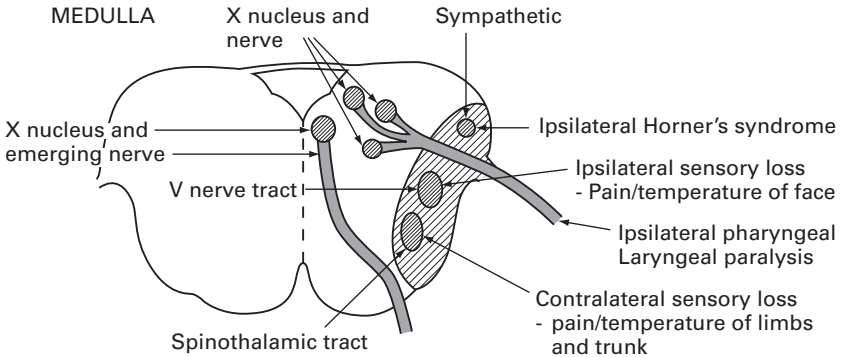


Figure 39: Lateral medullary syndrome

T1 Nerve Root

This is usually due to Pancoast's syndrome produced by an apical malignancy or thoracic outlet syndrome. C7/T1 disc prolapse is very rare.

Neck

Trauma, thyroid carcinoma and thyroid surgery, and malignant cervical lymphadenopathy are important common causes. Nasopharyngeal malignancy at the base of the skull may involve structures in the jugular foramen including cranial nerves IX–XI.

Carotid Artery

Lesions at this site may be seen in a migraine, internal carotid artery thrombosis, dissecting aneurysm of carotid and direct puncture of carotid.

The site of lesion is often apparent from the associated features. The area of impaired sweating could also localise the site of lesion. Lesions of the brain stem may impair sweating in the head, neck, arm and upper trunk of the same side.

The lower cervical region lesions impair sweating only in the face while lesions beyond the superior cervical ganglion do not impair sweating at all.

Others

See the Ptosis table on page 98. This includes congenital ptosis, tabes dorsalis and migrainous neuralgia.

HISTORY TAKING STATION

The history will, in most instances, be able to give an indication of the site, cause and nature of the lesion (see page 99).

Useful differential points are given on page 98.

The history will also give an indication of the investigations carried out, namely:

- Edrophonium test
- Electrophysiological studies
- Scanning – CT head, Ct thorax, MRI scan of head and chest
- Angiography if an aneurysm was suspected.

The advice given and treatment will indicate the prognosis and line of management contemplated.