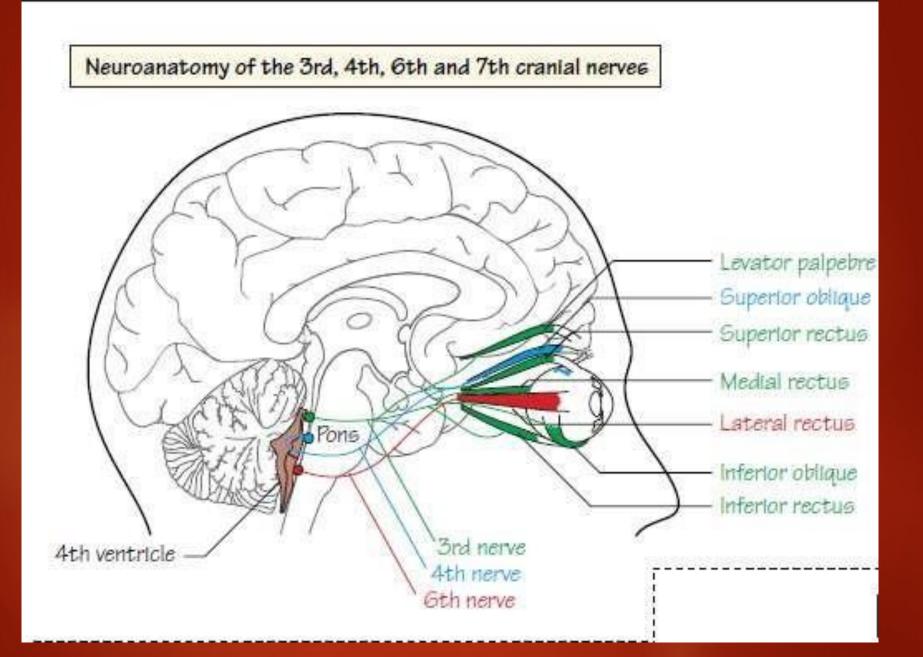


"In The Name of Allah, Most Gracious, Most Merciful"

Ocular Nerve Palsies Dr Khalid Shahab

Objectives

- I. Recall anatomy and function of cranial nerves
- II. Etiology of ocular nerve palsies
- III. Clinical feature of ocular nerve palsies



Cranial Nerve 3

- Most of the eye muscles
- Down and out
- Causes:
 - Vasculitic (DM, HTN)
 - Aneurysms





The oculomotor nerve

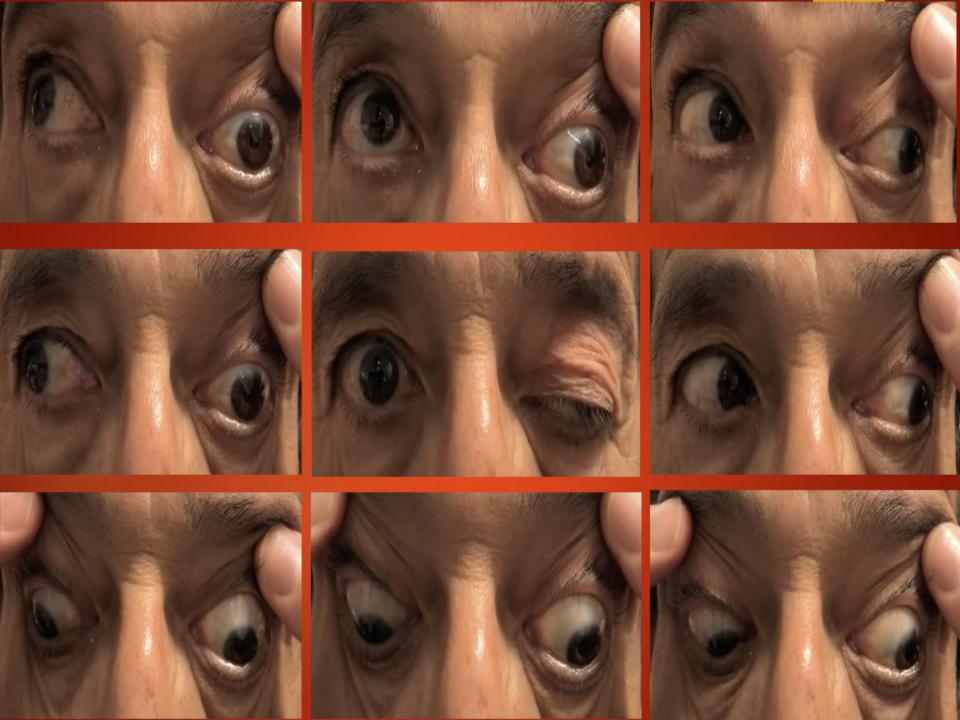
- It is the largest of the ocular motor nerves.
- It contains about 15000 axons, including motor fibers and parasympathetic motor fibers.
- The oculomotor (third cranial) nerve is entirely MOTOR in function.
- It supplies all the extraocular muscles except the LR and SO.
- It also supplies the sphincter pupillae and the ciliary muscle.

Anatomy

- The NUCLEAR COMPLEX of the 3rd nerve is situated in the midbrain at the level of the superior colliculus, ventral to the Sylvian aqueduct.
- Edinger-Westphal nuclei supply the parasympathetic preganglionic neurons that project to the ciliary ganglion.
- The FASCICULAR PORTION of the oculomotor nerve descends as its moves ventrally and laterally through the mesencephalon.
- The fascicles pass through the medial longitudinal fasciculus (MLF), the red nucleus, and the medial portion of the cerebral peduncle.

Course of CN III

- The oculomotor nerve exits the brainstem coursing downward and laterally through the intracranial, prepontine space, reaching the top of the clivus at a point just lateral to the posterior clinoid process.
- During the passage through the subarachnoid space, the nerve passes just below the posterior cerebral artery and above the superior cerebellar artery.
- After passing between these arteries, the nerve travels adjacent to the posterior communicating artery.
- Once the nerve penetrates the dura, it continues forward in the lateral wall of the cavernous sinus. It is positioned just above the fourth cranial nerve.
- In the anterior cavernous sinus, the oculomotor nerve divides anatomically into two distinct divisions: superior and inferior.



3rd Nerve Palsy in Adult

The most common causes of third nerve palsy in adults :

■ **ISCHAEMIA** D.M, HrT, ↑ Lipids, Vasculitis

Intracranial ANEURYSM at junction of ICA & PCoA

Head TRAUMA

Intracranial TUMORS direct or mass effect damage

Presentation

Symptoms

- Double vision—horizontal and/or vertical.
- Droopy lid.
- Enlarged pupil.
- Headache

Signs

- Ptosis.
- Exotropia and hypotropia.
- Fixed dilated pupil.
- Limitation of elevation, depression, and adduction. May present with any of these or combination.

Pupil based 3rd CN palsy

Pupil involving 3rd N. palsy

Pupil sparing 3rd N. palsy

aneurysm

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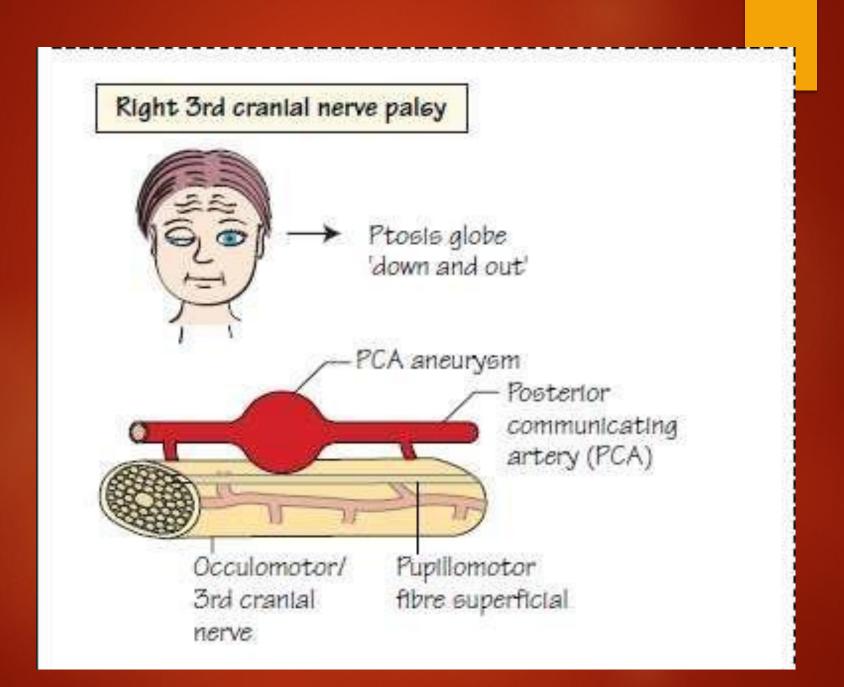
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superficial parasympathetics

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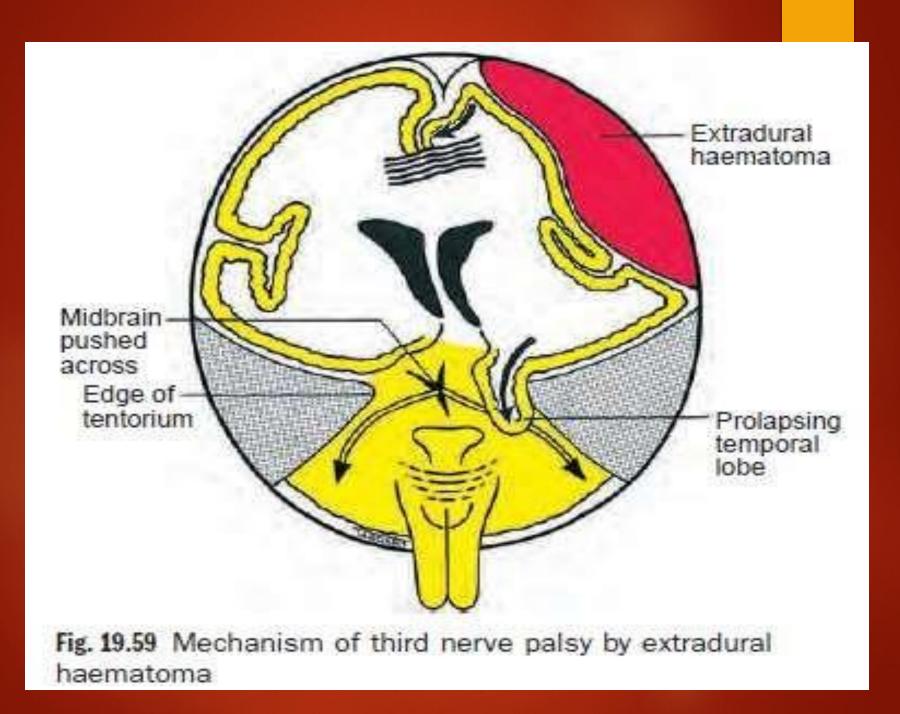
Pupil involving 3rd N. palsy Require Neuroimaging

Causes:

- 1. Aneurysm at the junction of PCoA & INC
- Most common in elderly patients rare before 20 yrs.
- Associated **headache** is important
- Emergent angiographic neuroimaging is rule (CTA or MRA).
- 2. Head trauma with extradural or intradural hematoma, due to herniation of the temporal lobe, which compress the nerve as it pass over tentorial edge.

Initially it cause irritative **Miosis** then followed by **Mydriasis** and complete third nerve palsy.

 20% of cases are vasculopathic but milder pupillary involvement of 1 mm anisocoria



Pupil sparing 3rd N. palsy

- Equal pupil size and reactivity
- This is the typical finding for **ischemic** cranial neuropathy
- Often associated with pain, which improves (and usually fully resolves) within 3-6 MONTHS.
- A complete & pupil-sparing third nerve palsy is almost always BENIGN.
- □ Often associated with **DM**, **HrT**, **Hyperlipidemia**.

Complete !!

a patient over 50 years of age with appropriate vascular risk factors but without history of cancer, present to the consultation with an acute, isolated, pupil-sparing, complete third nerve palsy, does neuroimaging is required or not?



- BUT general medical evaluation is required :
- Glucose level
- BP check
- Serum Lipid level
- No improvement after 3-6 m or progression to other cranial nerve palsy here NEUROIMAGING is mandatory.

Partial !!

- Pupil sparing, partial 3rd N. palsy, is it BENIGN or not?
- This distinction is crucial given that some proportion of partial third nerve palsies with normal pupillary function are related to compressive lesions and may later progress to involve the pupil.
- Neuroimaging is indicated to exclude compressive lesion.

Fascicular lesion

The causes of nuclear and fascicular lesions are similar, except that demyelination may affect the fasciculus.

•Benedikt syndrome involves the fasciculus as it passes through the red nucleus and is characterized by ipsilateral third nerve palsy and contralateral extrapyramidal signs such as hemitremor.

•Weber syndrome involves the fasciculus as it passes through the cerebral peduncle and is characterized by ipsilateral third nerve palsy and a contralateral hemiparesis.

•Nothnagel syndrome involves the fasciculus and the superior cerebellar peduncle and is characterized by ipsilateral third nerve palsy and cerebellar ataxia.

 Claude syndrome is a combination of Benedikt and Nothnagel syndromes.

Moral

Pupil involvement IS BAD

Trochlear 4th CN palsy

- The trochlear cranial nerve supplies only the superior oblique muscle.
- The nucleus Motor (entirely crossed) is located at the level of the inferior colliculi ventral to the Sylvian aqueduct. It is caudal to, and continuous with, the third nerve nuclear complex.
- The trunk leaves the brainstem on the dorsal surface, just caudal to the inferior colliculus.
- It then curves laterally around the brainstem, runs forwards beneath the free edge of the tentorium, and like the third nerve passes between the posterior cerebral artery and the superior cerebellar artery.
- □ It then pierces the dura and enters the cavernous sinus.

Key features

It is a very long and slender nerve, increasing its vulnerability.

It is the only cranial nerve to emerge from the dorsal aspect of the brain.

It is the only decussated cranial nerve besides the optic nerve, innervating the SO muscle contralateral to its nucleus.

It has the fewest axons of any of the cranial nerves.

Causes of 4th CN palsy

1. Idiopathic

- Most are congenital
- Symptoms may appear late in adult life
- Old photograph will help
- 2. Trauma
- Frequently cause bilateral 4th nerve palsy
- 3. Microvascular
- 4. Aneurysms and Tumor

Features

- Asymptomatic
- Diplopia vertical (difficulty in reading and going downstairs)
- Head tilt position opposite to the side of the lesion.
- **Hypertropia** on the affected side.
- Limitation of depression, most marked in adduction
- **Extorsion**, greatest in abduction.
- Bilateral involvement should always be excluded, particularly following head trauma.
- Parks three-step test
- I. Step one In the primary position. In a fourth nerve palsy, the involved eye is higher
- II. Step two The eyes are examined in right and left gaze to determine where the hypertropia is greater. In superior oblique weakness the deviation is worse on opposite gaze.
- III. Step three The Bielschowsky head tilt test (BHTT)







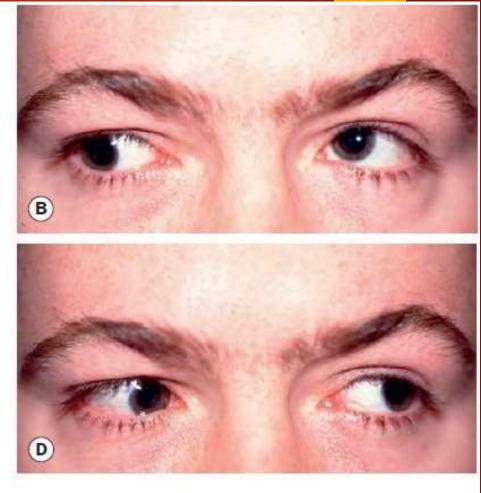
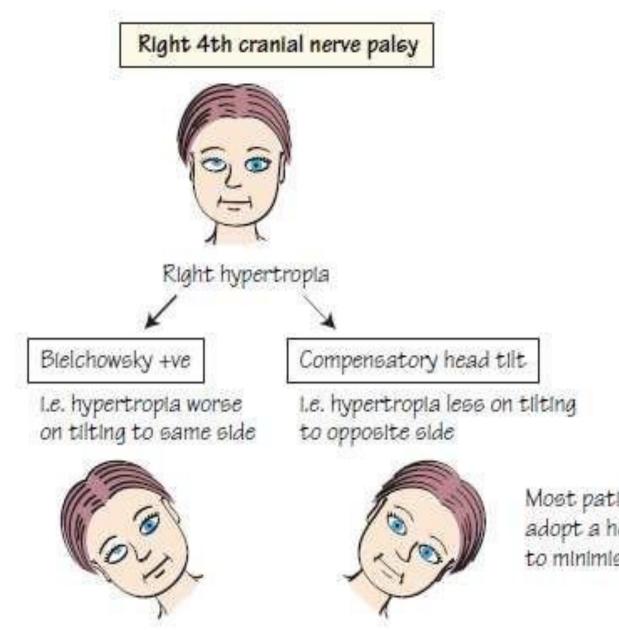


Fig. 19.64 Left fourth nerve palsy. (A) Left hypertropia (left over right) in the primary position; (B) increase in left hypertropia on right gaze due to left inferior oblique overaction; (C) limitation of left depression in adduction; (D) normal left abduction; (E) normal left elevation



Most patients will adopt a head tilt to minimise diplopia

INVESTIGATION

- Diagnostic evaluation for an isolated, non traumatic fourth nerve palsy usually yields little information because most cases have congenital, ischemic, or idiopathic causes.
- Recovery can be achieved within 3-6 months.
- Medical evaluation for those at risk of vascular accident
- Neuroimaging if no improvement noticed within 3-6 month



Figure 8-11 Congenital left fourth nerve palsy. A, Note the left hypertropia and right head tilt as a child. B, Forty years later, the right head tilt is still present, but the patient describes more difficulty maintaining single, binocular vision. C, After eye muscle surgery, the diplopia and head tilt have resolved. (*Courtesy of Lanning B. Kline, MD.*)

Abducens 6th CN Palsy

- The nucleus of the 6th nerve lies at the mid-level of the pons, ventral to the floor of the fourth ventricle.
- Motor (entirely uncrossed) fibers arise
- Important anatomical relations;
- 7th nucleus is close to 6th nucleus
- It runs close to petrous portion of temporal bone
- Then pass through cavernous sinus
- Then to the orbit through SOF

Causes

- 1. Ischaemia
- 2. Tumor (acoustic neuroma, meningioma)
- 3. Trauma
- 4. Elevated Intracranial pressure
- 5. Infection, middle ear infection, meningitis
- 6. Demyelination
- 7. Dural arteriovenous fistula
- 8. Intracranial hypotension including after lumbar puncture

- 6th CN palsy is most common single EOM palsy.
- Congenital 6th N. palsies almost never occur in isolation.
- Leukemia & brainstem glioma are important considerations in children.
- In adolescents & young adults, demyelination may be the cause.
- Gradenigo syndrome: Chronic inflammation of the petrous bone may cause an ipsilateral abducens palsy and facial pain especially in children who have experienced recurrent infections of the middle ear.

- The appearance of a 6th N. palsy after seemingly minor head trauma should RAISE concern for preexisting pathology such as tumor compression making the nerve more susceptible to injury.
- Isolated 6th nerve palsies in adults older than 50 years are usually ischemic.
- Impaired abduction in patients younger than 50 years requires careful scrutiny, because few such cases are caused by ischemic cranial neuropathy.
- Younger patients should undergo appropriate neuroimaging.
- A posteriorly draining carotid-cavernous fistula may have a 6th nerve palsy as its only presenting ocular sign.

Presentation

- Double vision
- Esotropia in the primary position
- Limitation of abduction on the side of the lesion
- Normal adduction
- A compensatory face turn is towards the side of the paralysed muscle in unilateral palsy.

Evaluation of 6th N. palsy

Complete neurological examination and exclusion of other cranial N. palsies.

Hearing and corneal sensation should be checked in all patients with sixth nerve palsy.

In a child or any patient with relevant symptoms, an otorhinolaryngological consultation.

Medical evaluation is required for patients above 50 years.

Investigations

- 1. Lumber puncture
- 2. Chest x-ray
- Hematological to exclude collagen vascular disease, sarcoidosis and syphilis.
- 4. Neuroimaging is required for
- No improvement of palsy after 3-6 months in 50 years older patients.
- In patient younger than 50 years age

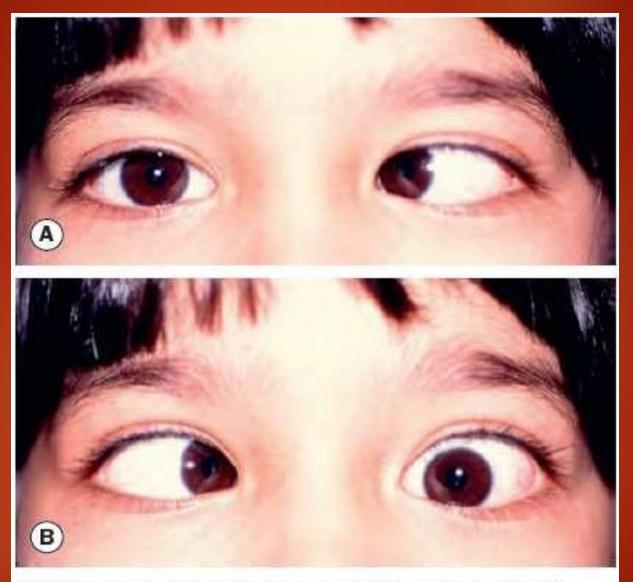


Fig. 19.71 Acute left sixth nerve palsy in a child. (A) Left esotropia in the primary position; (B) marked limitation of left abduction

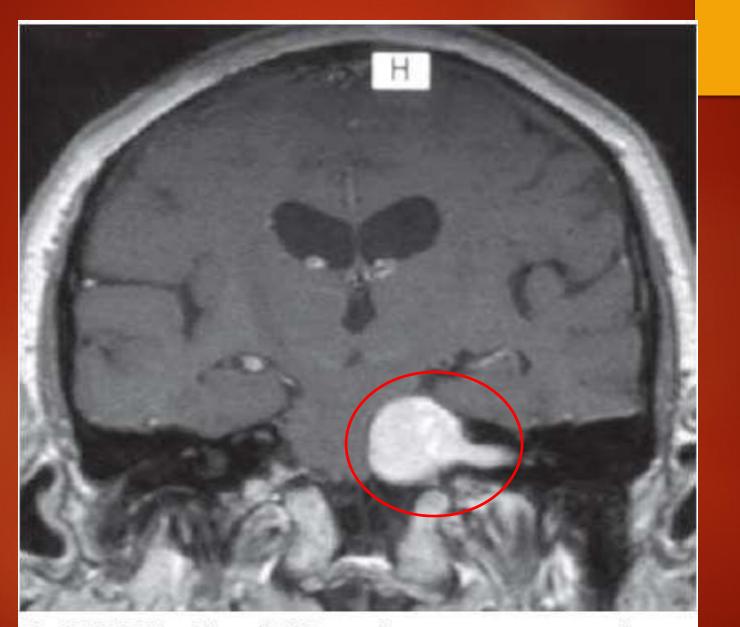


Fig. 19.70 MRI with gadolinium enhancement – coronal image of an acoustic neuroma

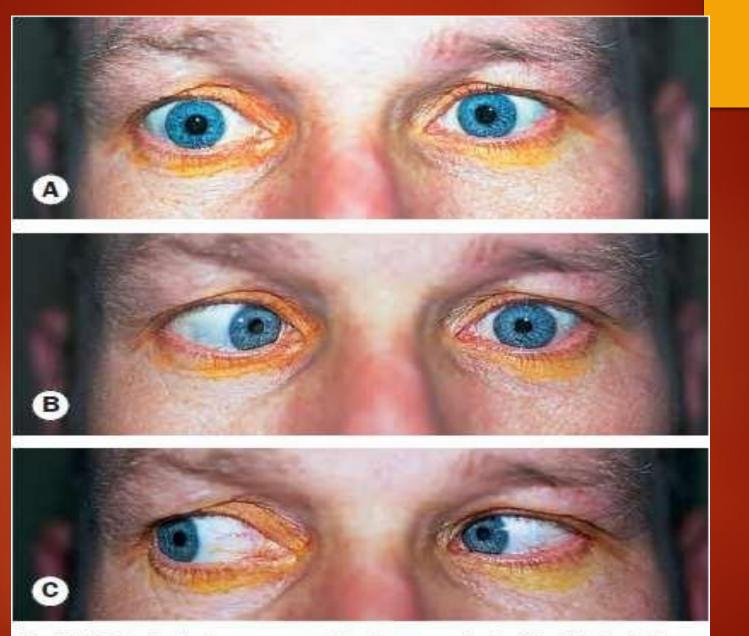
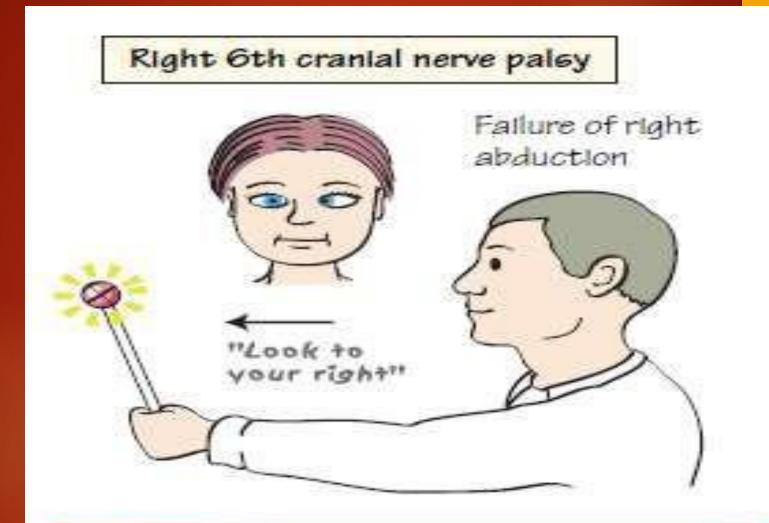


Fig. 19.72 Left sixth nerve palsy in an adult. (A) Slight left esotropia in the primary position; (B) limitation of left abduction; (C) normal left adduction



Fig. 19.73 Acute bilateral sixth nerve palsy (Courtesy of C Barry)



WARNING

Acquired 6th nerve palsy in patient with shunted hydrocephalus = blocked shunt = emergency

Paresis of More Than One Cranial Nerve

- Benign microvascular disease rarely causes simultaneous involvement of more than 1 ocular motor cranial nerve.
- Simultaneous involvement of contiguous nerves (CNs III, IV, V, VI, and sympathetic nerves) strongly suggests a lesion of the cavernous sinus.
- Bilateral involvement of the cranial nerves suggests a diffuse process such as
- 1. Infiltrative disease (eg, carcinoma, leukemia, or lymphoma),
- 2. A midline mass lesion that extends bilaterally (eg, chordoma, chondrosarcoma, or nasopharyngeal carcinoma),
- 3. A meningeal-based process, an inflammatory polyneuropathy (eg, Guillain-Barre syndrome or its variant, the Miller Fisher syndrome, or sarcoidosis), or myasthenia gravis.

Syndromes Affecting Cran<mark>ial</mark> Nerves III, IV, & VI

A. Superior Orbital Fissure Syndrome

All the ocular motor nerves pass through the superior orbital fissure and can be affected by tumor, inflammation, or trauma involving the fissure.

B. Orbital Apex Syndrome

This syndrome is similar to the superior orbital fissure syndrome with the addition of optic nerve signs and usually greater proptosis. It is also caused by tumor, inflammation, or trauma. **C. Sudden Complete Ophthalmoplegia** Complete ophthalmoplegia of sudden onset can be due to extensive brainstem vascular disease, Wernicke's encephalopathy, Fisher's syndrome, bulbar poliomyelitis, pituitary apoplexy, basilar aneurysm, meningitis, diphtheria, botulism, or myasthenic crisis.

Cavernous sinus involvement

3rd + 4th + 5th +6th
nerves involved in
varying severity

Cerebellopontine angle involvement

 6th + 7th + 8th nerves involved in varying severity

DIFFERENTIAL DIAGNOSIS

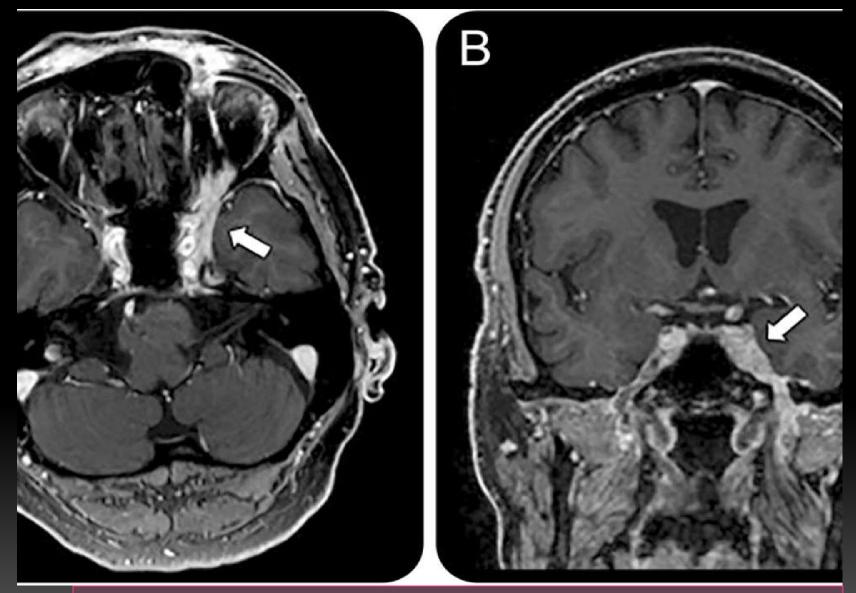
- Orbital apex syndrome
- Cavernous sinus thrombosis
- Superior orbital fissure syndrome
- Fungal infection in uncontrolled diabetes-Mucormycosis, aspergillosis
- Tolasa hunt syndrome
- Orbital tumours
- Isoloated sphenoid sinus tumours

Evaluation

- A neurologic evaluation should be undertaken if symptoms or signs indicate that more than 1 cranial nerve is involved.
- In this case, if neuroimaging results are normal, a lumbar puncture with cytopathologic examination should be considered.
- Special testing for cancer-associated protein markers may be helpful in uncovering an elusive diagnosis.



A Case of posterior cavernous sinus tumour with multiple cranial nerve palsy. Computed tomography scan showing enhancing lesion in right parasellar region extending into posterior cavernous sinus



Cavernous sinus invasion from skin tumor



PropoRordo