THE ORBIT

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OUTLINE

- Defenitions
- Anatomy of the orbit.
- Proptosis.
- Thyroid eye disease.
- Diplopia.
- Ddx of orbital diseases.
- Orbital tumors.

DEFINITION;

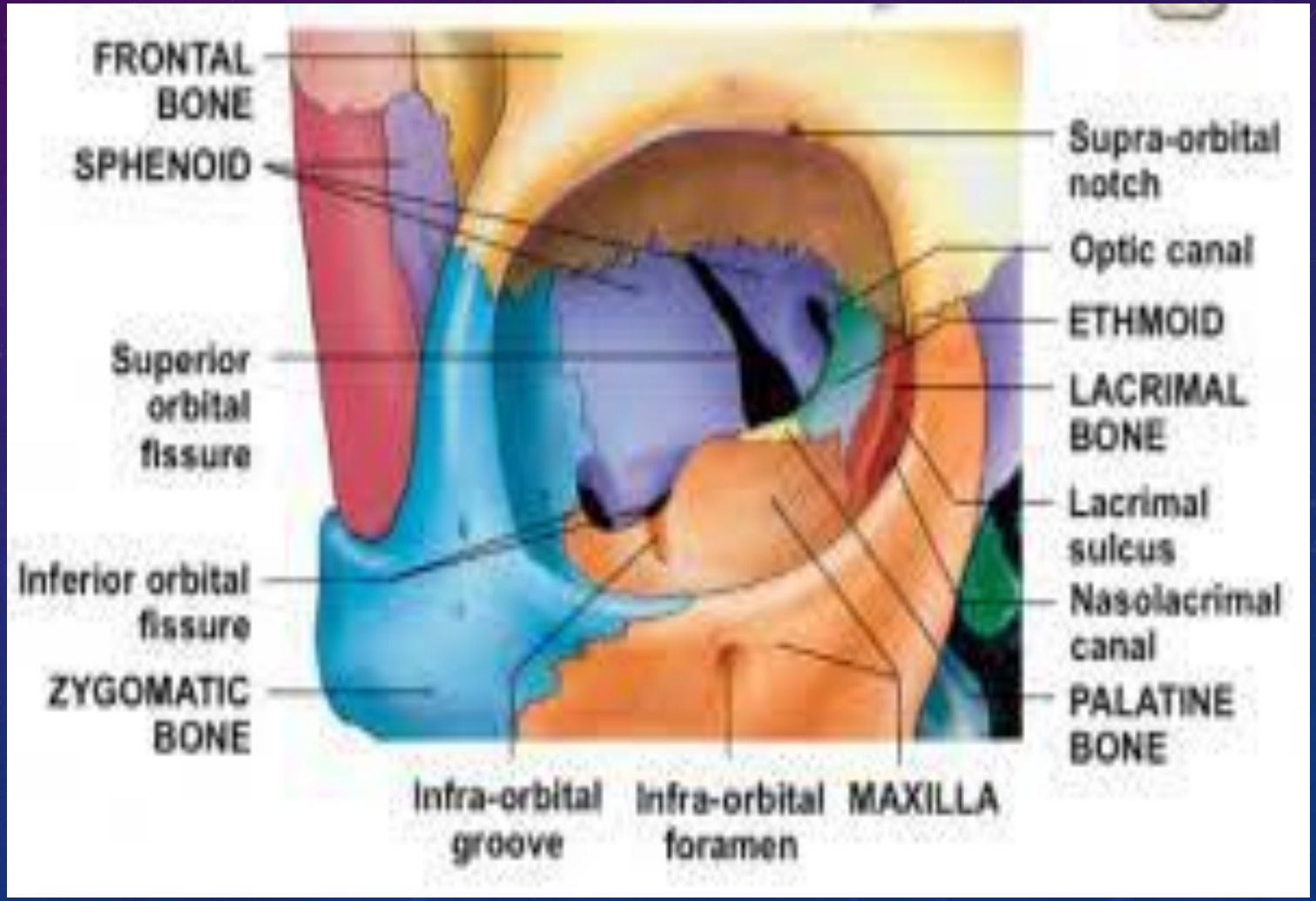
A bony, very small, crowded cavity

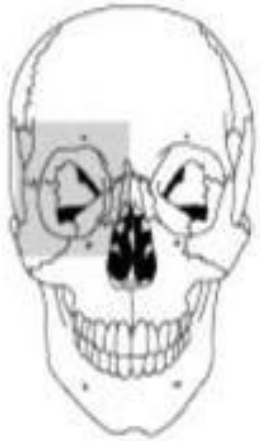
ANATOMY

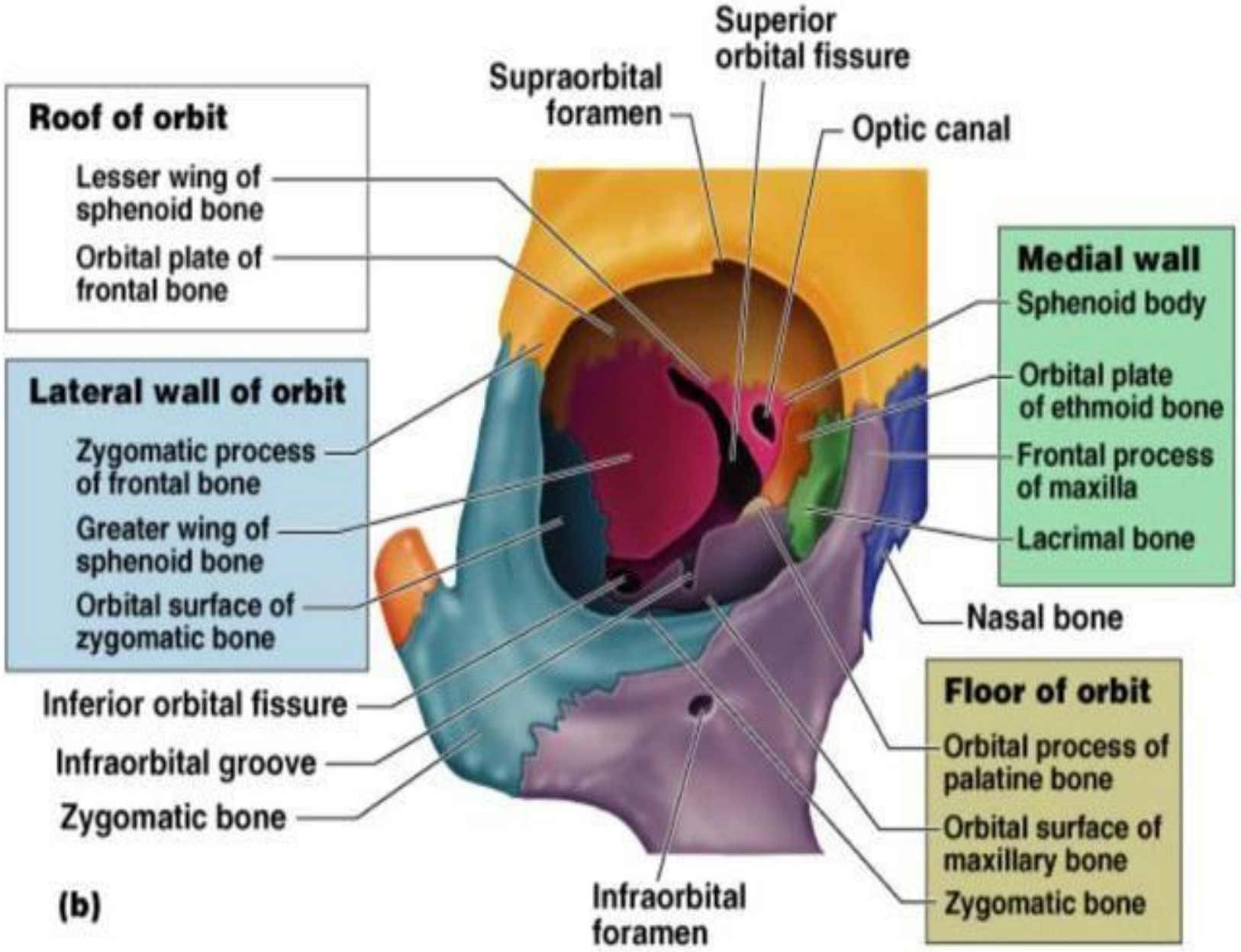
- The orbital cavity is a bony socket.
- Protects and contain the globe, optic nerve, ocular muscles, blood vessels and lacrimal gland.
- It’s shape is a four-sided pyramid.
- It’s size is 30 CC.
- Important nearby structures Paranasal sinuses / brain

- The orbit consists of **7** bones:

1. Frontal bone. Superior
2. Ethmoid bone. Medial
3. Lacrimal bone. Medial
4. Sphenoid bone. Superiolateral
5. Maxillary bone. Inferiomedial
6. Palatine bone. Inferior
7. Zygomatic bone. Superiolateral







Ophthalmic nerve > supraorbital & supratrochlear nerves > forehead

Maxillary > infraorbital nerve > cheek & upper gums

- The major nerves and vessels enter through:

✓ ***Optic canal;***

- At the posterior apex of the orbit.
- Transmits optic nerve. & surrounding meninges (CSF)

✓ ***The superior and inferior orbital fissures;***

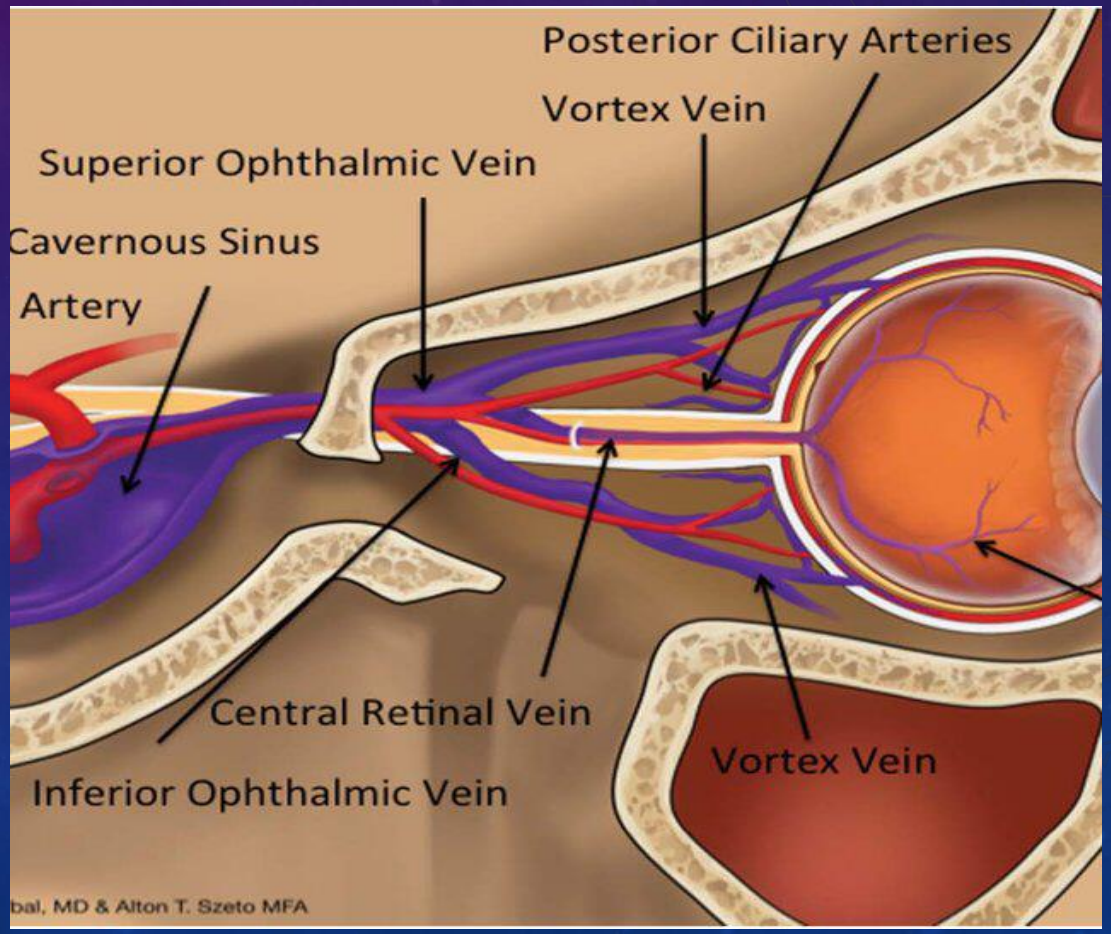
- Transmits blood vessels and cranial nerves from the brain to the orbit (and from face and venous drainage back to the brain).

- ***<u>The optic canal:</u>***

➢ At the apex of the orbit, within the sphenoid bone.

➢ The structures that enter through it:

1. Optic nerve.
2. Ophthalmic artery.
3. Central retinal vein.

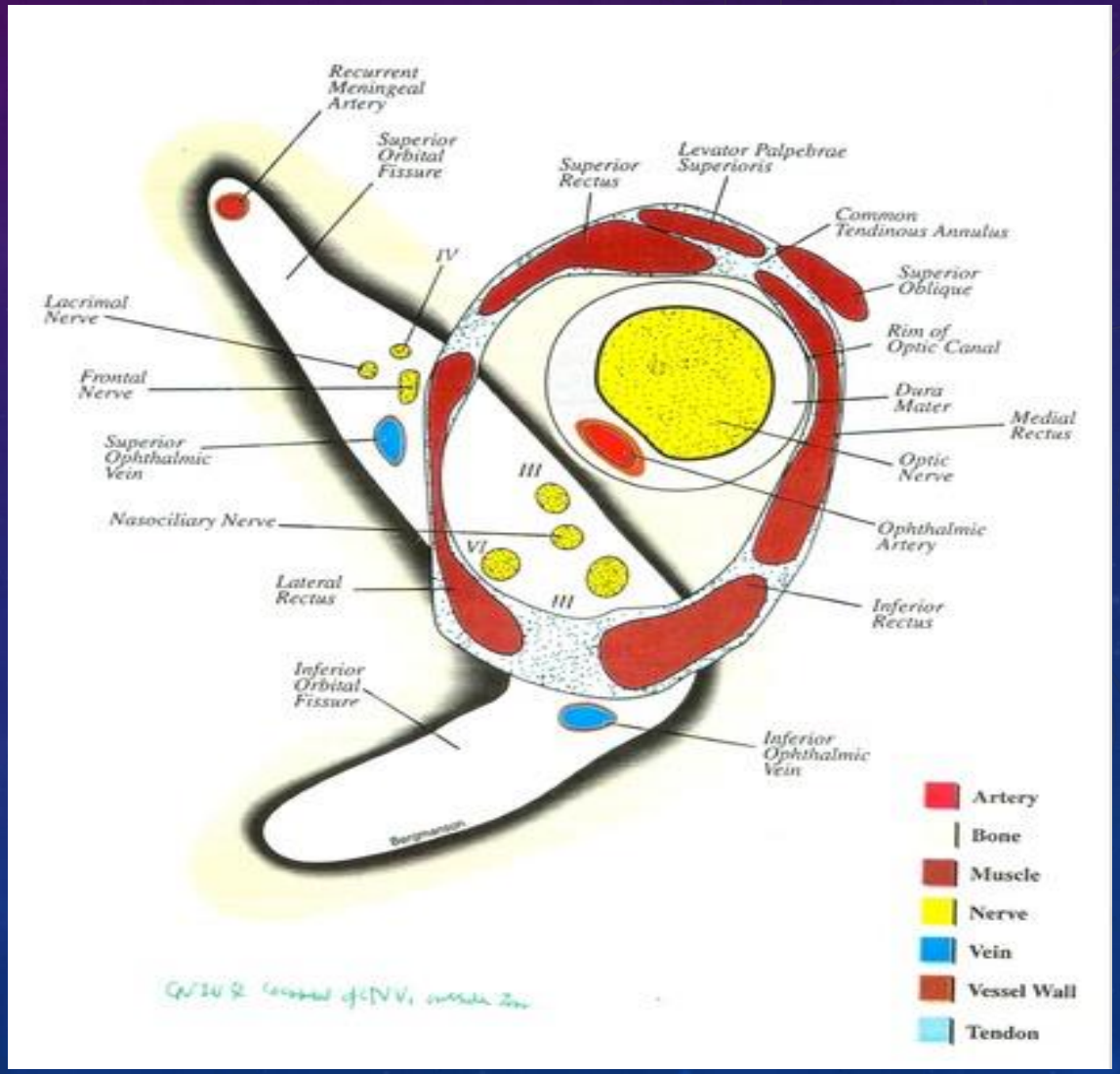


- ***<u>The superior orbital fissure:</u>***

➢ Bounded by the lesser and greater wings of the sphenoid.

➢ The structures that pass through it:

1. Cranial nerves (CN) (3, 4, 6)
2. Lacrimal nerve.
3. Frontal nerve.
4. Nasociliary nerve.
5. Superior ophthalmic vein.
6. Orbital branch of middle meningeal artery.
7. Recurrent branch of lacrimal artery.
8. Superior orbital vein.



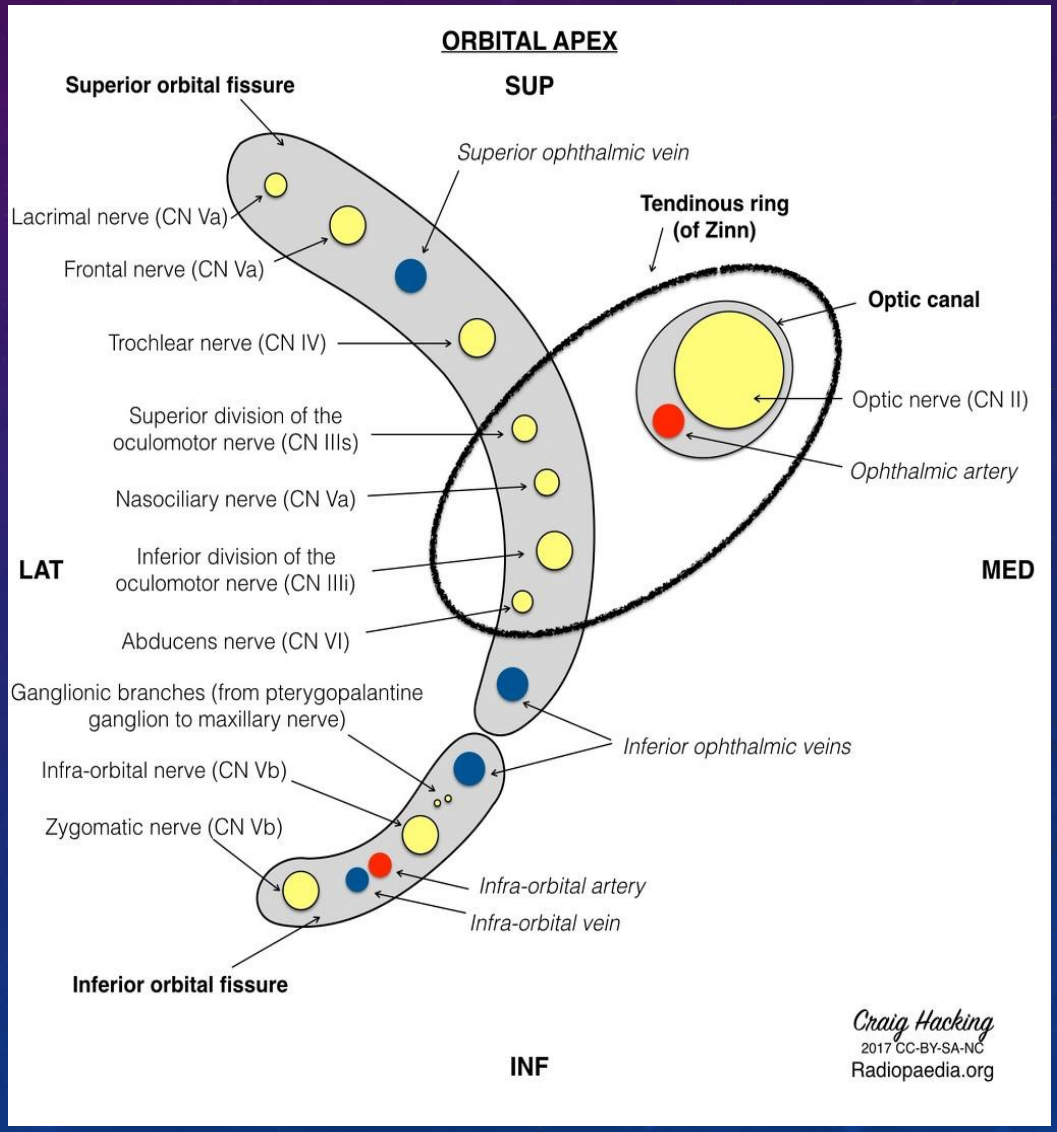
The supraorbital & infraorbital veins drain into the superior ophthalmic & inferior ophthalmic veins; respectively. The latter two drain into the cavernous sinuses which facilitates the spread of infection from the orbital cavity to the brain.

- ***The inferior orbital fissure:***

➢ Formed by the greater wing of the sphenoid, the maxilla and the palatine bone.

➢ The structures that enter though it:

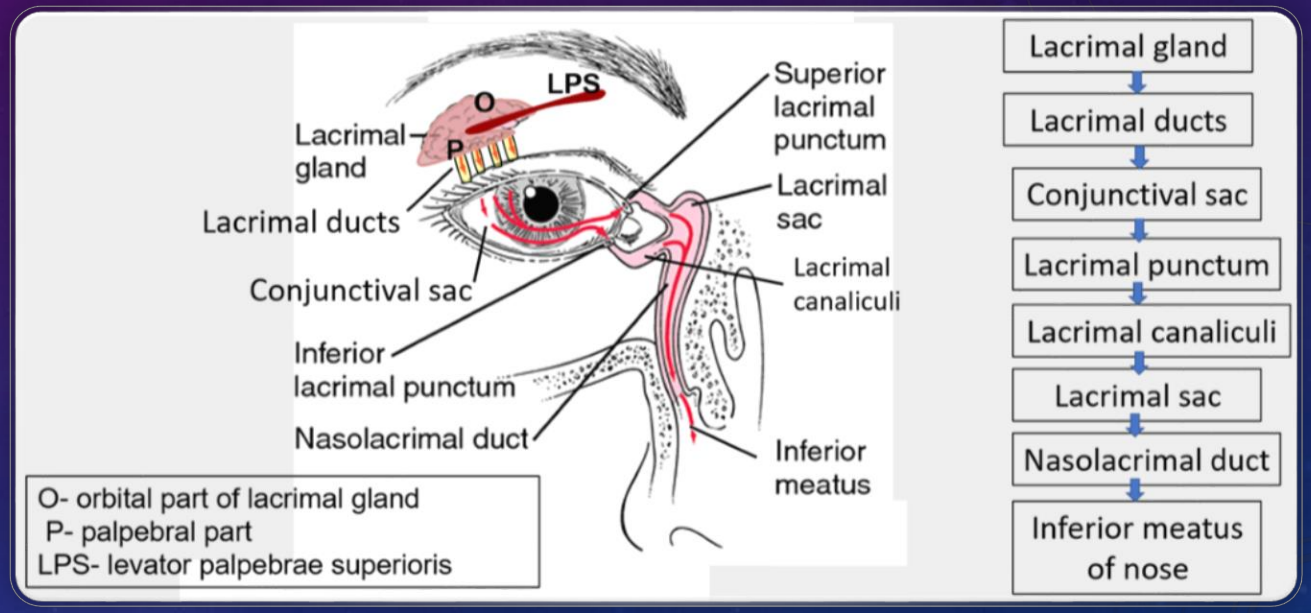
1. Infraorbital nerve.
2. Zygomatic nerve.
3. Parasympathetic fibers (to lacrimal gland).
4. Infraorbital artery (from maxillary artery).
5. Infraorbital vein.
6. Inferior ophthalmic vein.



•***<u>The lacrimal gland:</u>***

•Lies anteriorly in the superolateral aspect of the orbit.

•lacrimal sac lies On the anterior part of the medial wall.



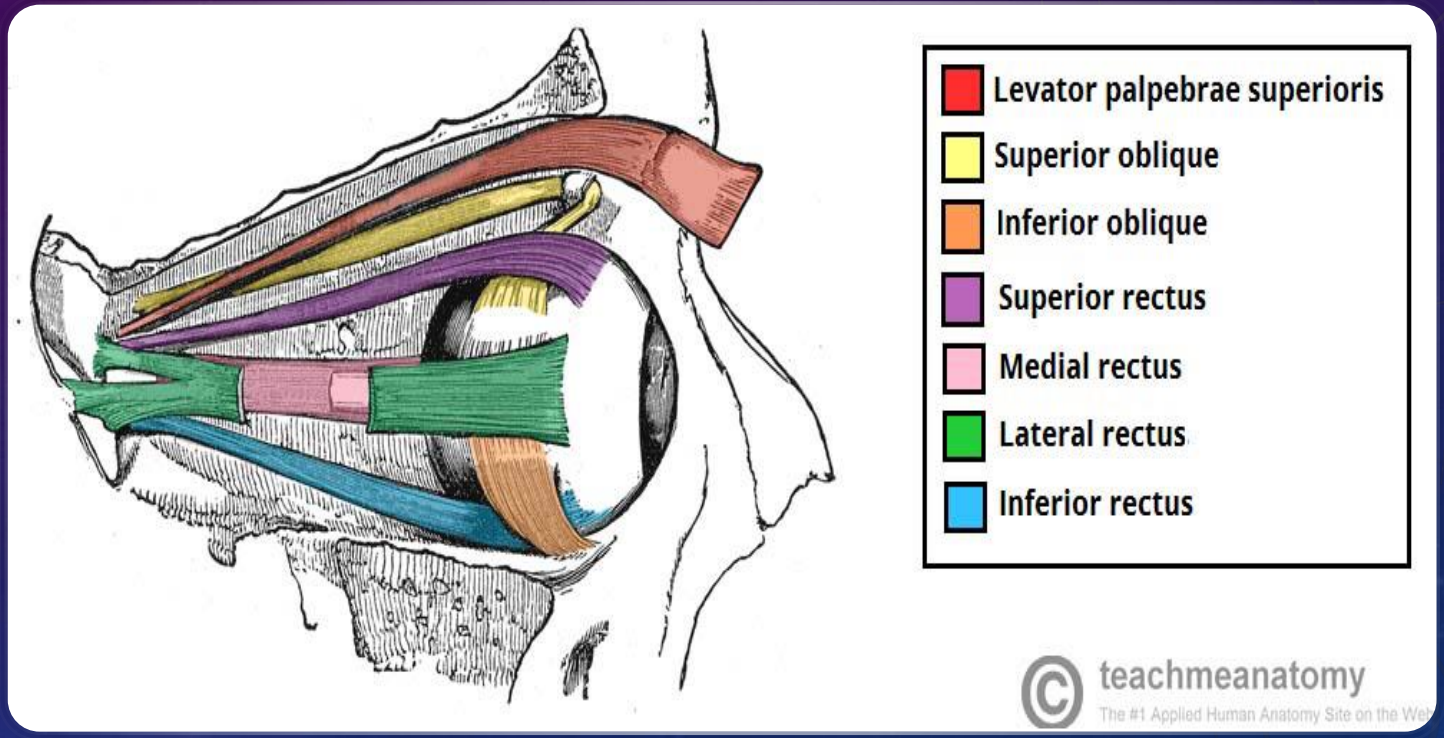
- ***Function of the orbit?***

➢ Protection of the globe.

➢ Provides attachment of the muscles which stabilize ocular movement.

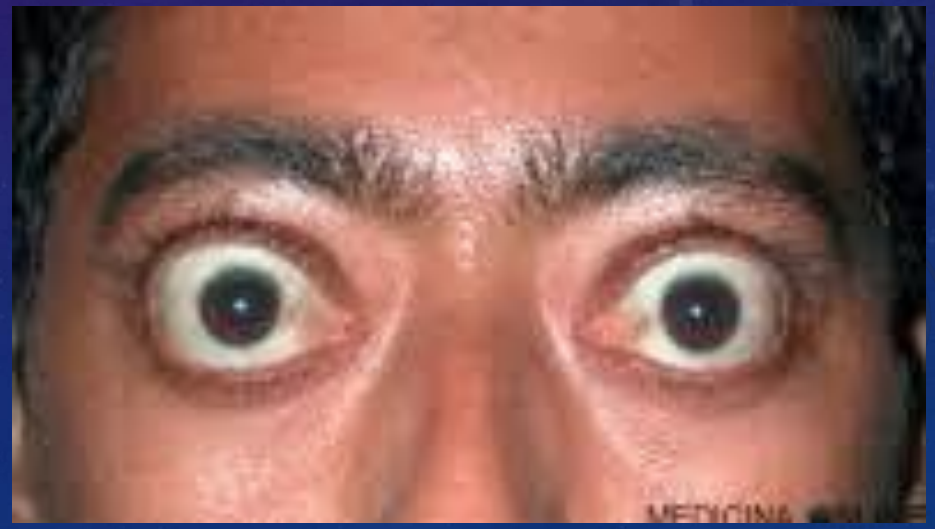
(The 6 ocular muscles originate at the apex around the optic nerve and insert into the globe.)

➢ A conduit for the transmission of nerves and blood vessels.



PROPTOSIS

- Proptosis (exophthalmos) is bulging of the eye anteriorly by a space-occupying lesion.
- It could be bilateral (Graves' disease) or unilateral (orbital tumors).



- ***Causes of Proptosis?***

1. Infectious.
2. Inflammatory.
3. Neoplastic.
4. Vasculitis.
5. Orbital vascular disease.
6. Trauma.
7. Pseudoprosptosis (pseudoexophthalmos).

- 1. ***Infectious;***

1- painful
2- develops rapidly over 1-2 days

- Orbital cellulitis (*M.C.C for unilateral Proptosis in children).*
- *Mucormycosis.*
- *Sinus disease (Mucocele)*
- *Rapid onset & painful.*

1- Very severe / rapidly progressive
2- Mostly seen in immunocompromised patients (usually in patients with uncontrolled diabetes & patients with kidney transplant on immunosuppressants)

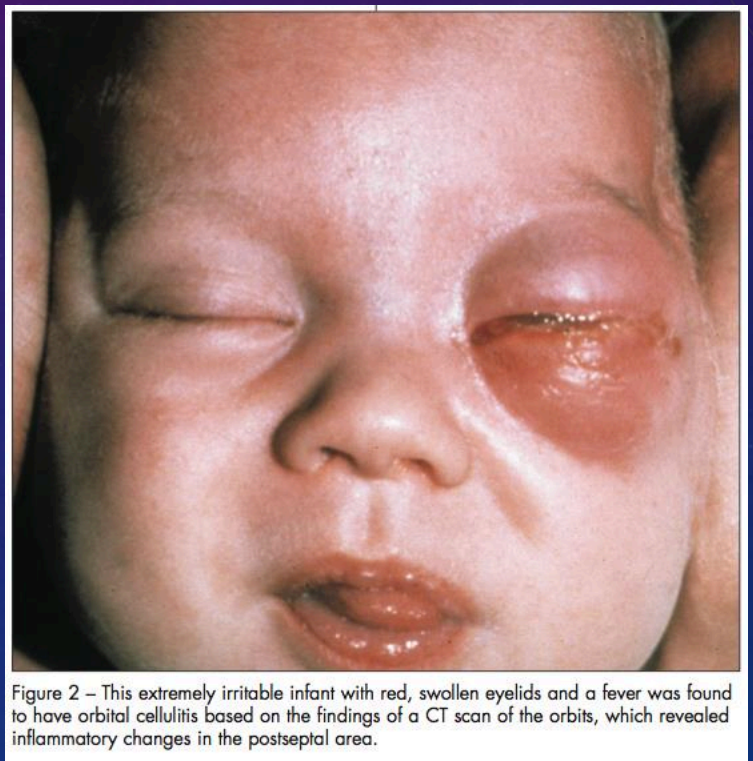


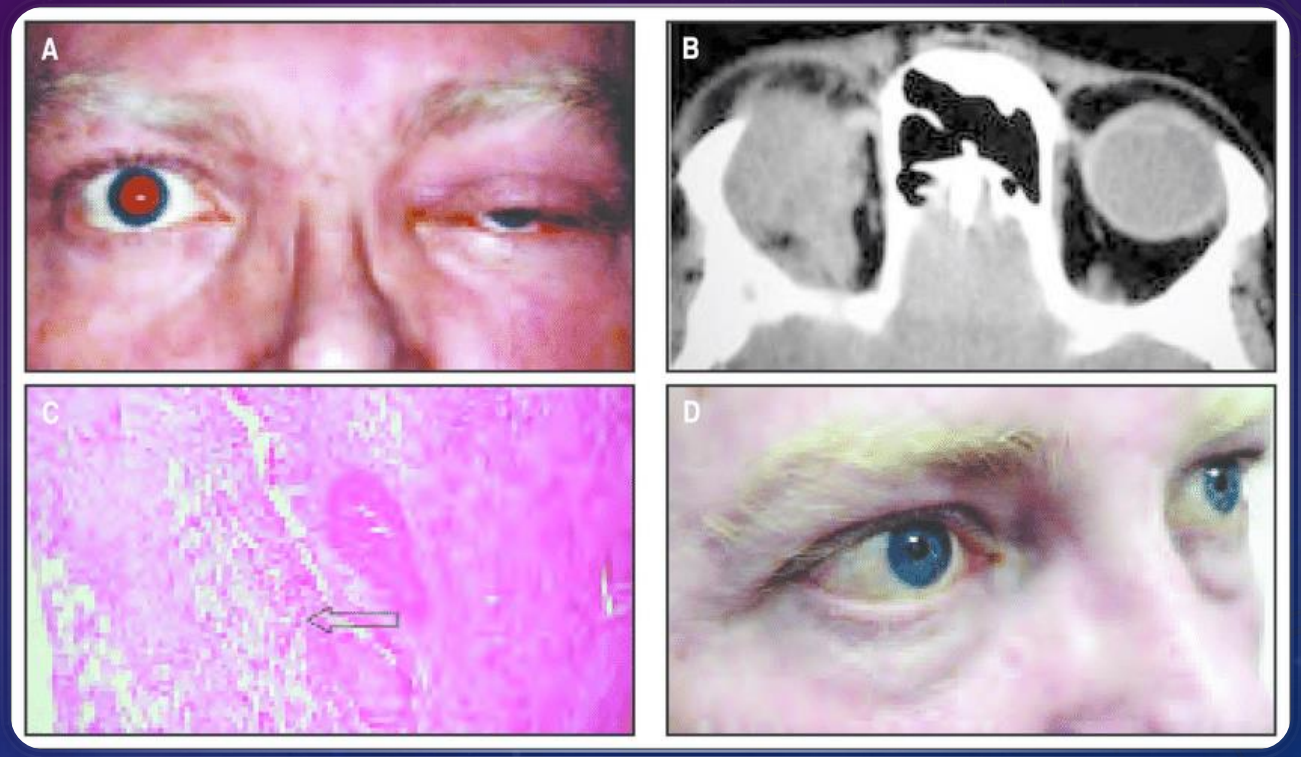
Figure 2 – This extremely irritable infant with red, swollen eyelids and a fever was found to have orbital cellulitis based on the findings of a CT scan of the orbits, which revealed inflammatory changes in the postseptal area.

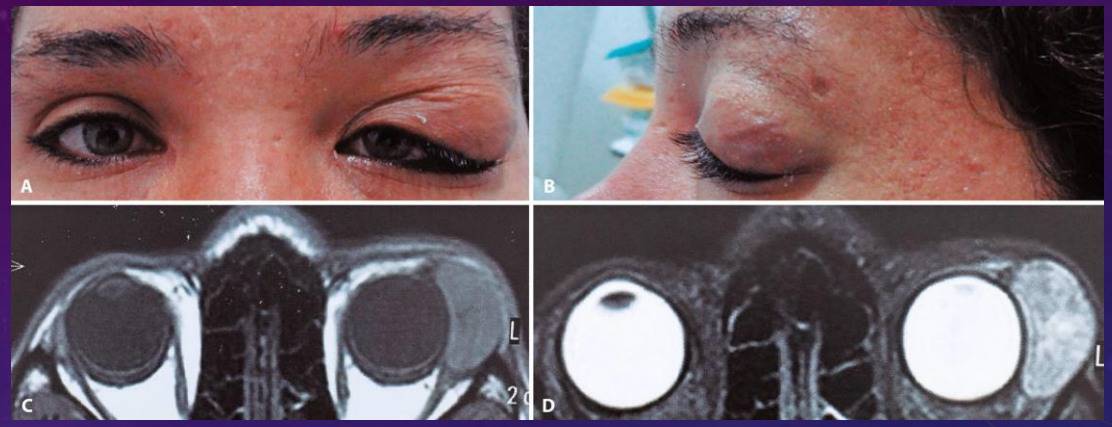
- 2. ***Inflammatory;***

➢ Thyroidopathy (*M.C.C for proptosis in adults is Graves' disease).* 90% > caused by hyperthyroidism

➢ Orbital inflammatory syndrome (orbital pseudomotor, benign orbital inflammation).

** Sarcoidosis





- 3. ***Neoplastic:***

➢ Lacrimal gland tumors. (*Eye displaced to one side; lesion outside muscle cone)*

Contralateral side

➢ Lymphoma.

➢ Leukemia.

+ optic nerve glioma

➢ Meningioma. (*forward in direction; intra-conal lesion)*

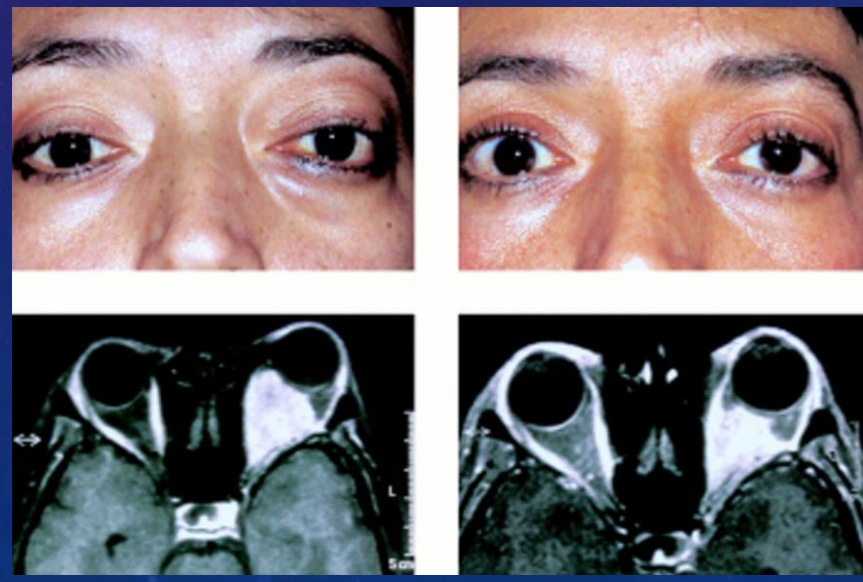
➢ Glioma.

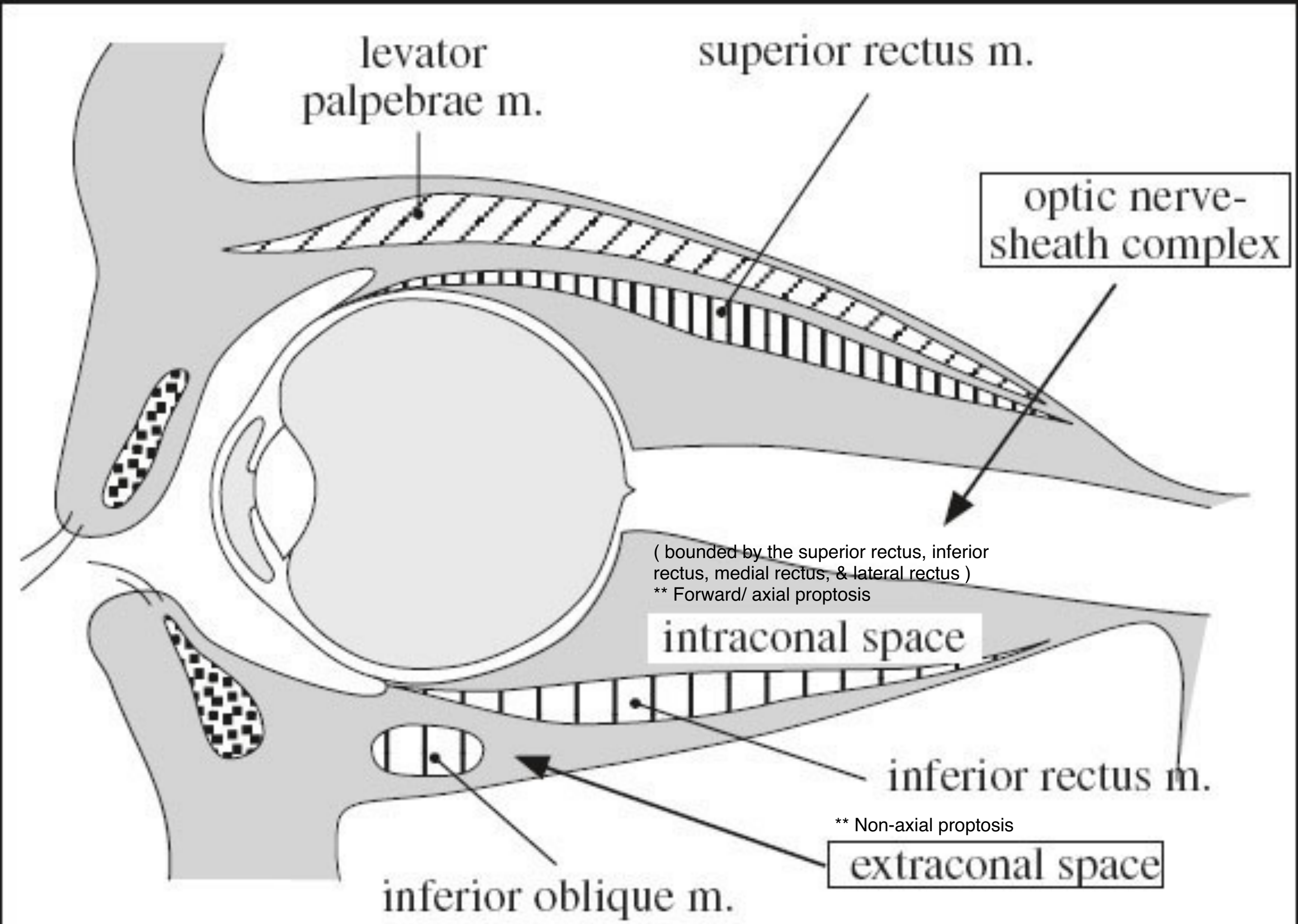
➢ Ossifying fibroma.

➢ Metastatic. (*breast in women, lung and prostate in men, GI, kidney)*

6-9 y.o

➢ **in children, rapidly developing Proptosis; rhabdomyosarcoma.



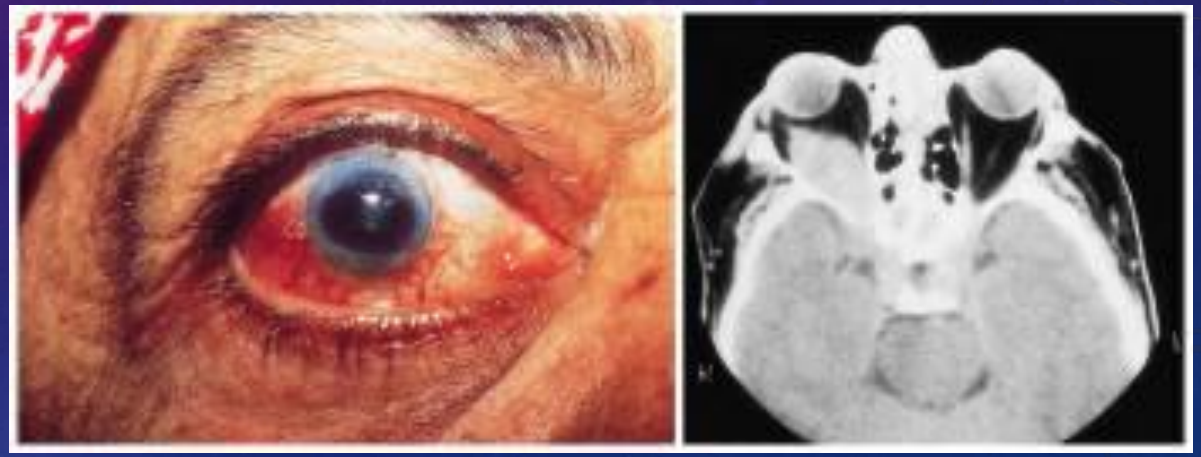


Sagittal Orbital Tomogram through Midorbit

- ***4. Vasculitis;***

➢ Wegner granulomatosis.

➢ Churg-Strauss syndrome.



Because

- 5. ***Orbital vascular disease;***

➢ Orbital varix (*venous malformation)*

(Transient proptosis; induced when increasing cephalic venous pressure, i.e: valsalva maneuver)

➢ Orbital arteriovenous malformation Arterial flow into the draining venous channels (pulsating proptosis)

(Carotid-cavernous sinus fistula, arterio venous malformation)

- 6. ***Trauma;***

➢ Traumatic/ iatrogenic orbital hemorrhage.

➢ Orbital fractures (*acute phase*).

➢ Facial fractures.

Later: enophthalmos

Edema due to trauma

- 7. ***Pseudoproptosis;***

➢ Buphthalmos. Large eye globe secondary to congenital glaucoma

➢ Contralateral enophthalmos.

➢ Ipsilateral lid retraction.

➢ Axial myopia. Large anterior-posterior diameter

➢ Contralateral blepharoptosis.

The orbit

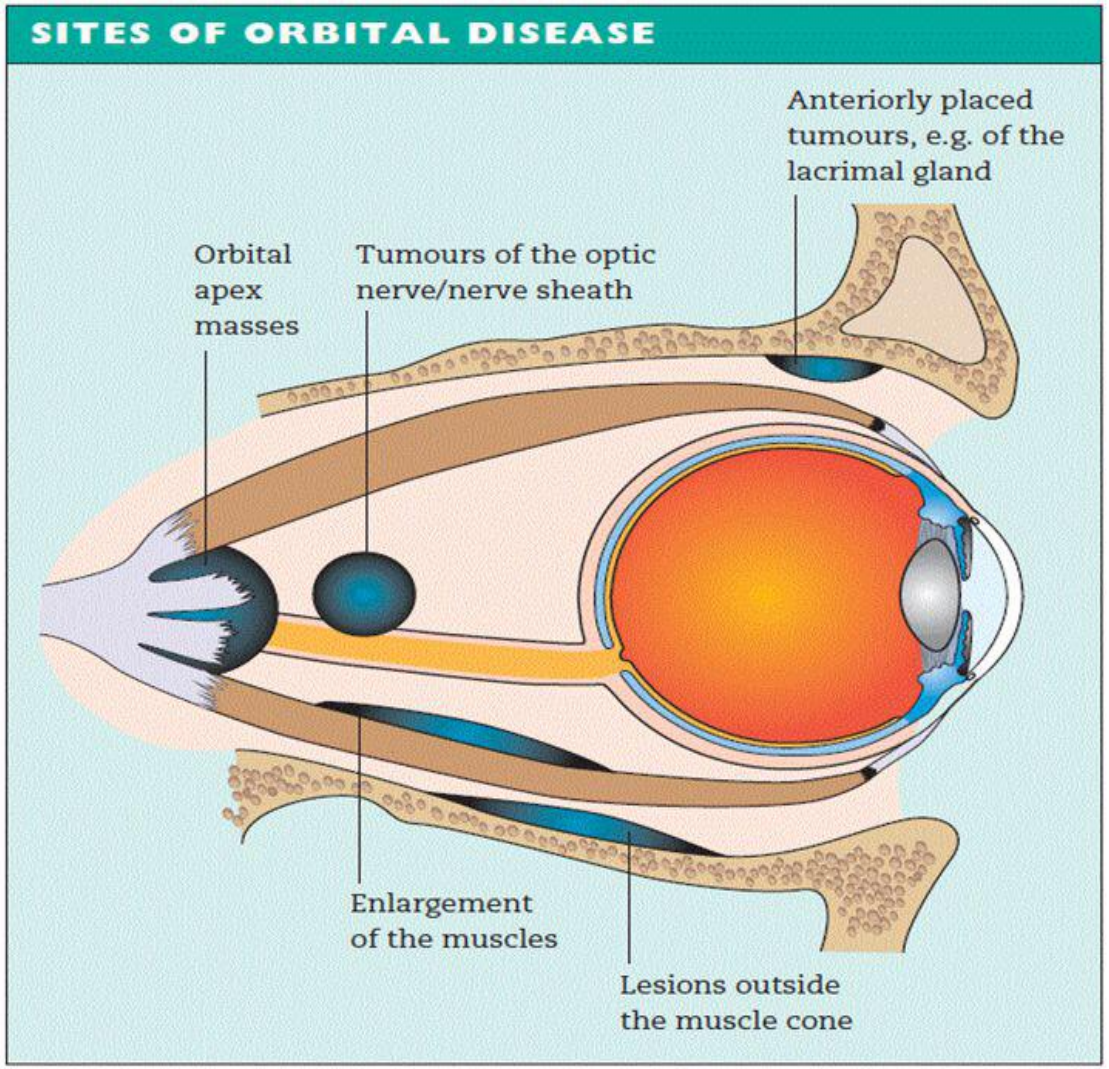
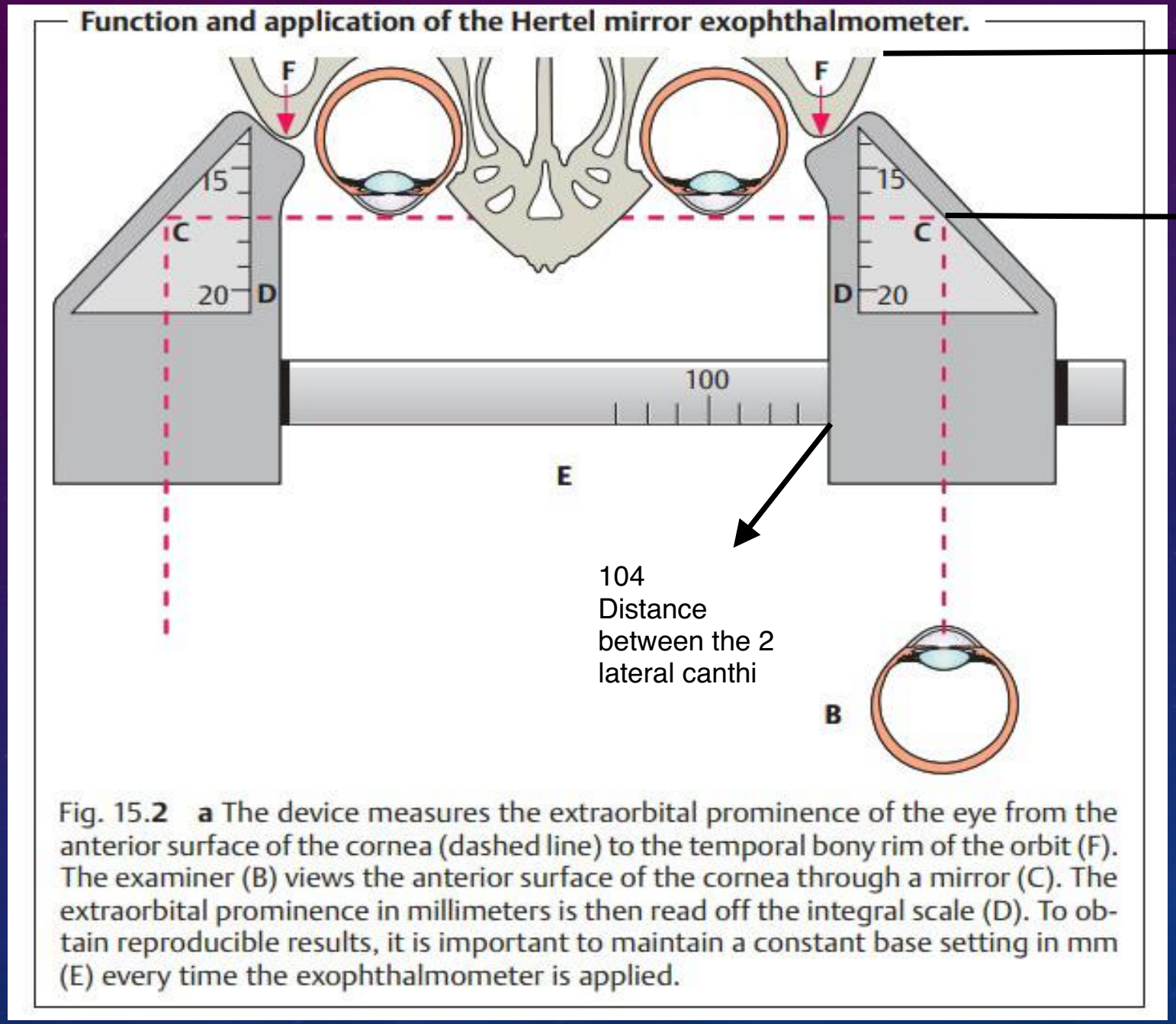


Fig. 4.1 Sites of orbital disease.

- ***<u>How do we measure Proptosis?</u>***

✓ it can be measured using Hertel exophthalmometer.

We have to use the same exophthalmometer as the readings differ between different exophthalmometers



- A normal range: 10-21 mm.
- Proptosis is graded as:

1. Mild: (21-23 mm)
2. Moderate: (24-27 mm)
3. Severe: (28 mm or more)

- A difference of more than 2mm between the two eyes is significant.

- ***Enophthalmos:***

A backward (retraction) displacement of the globe.

➢ Seen in:

1. Orbital fractures.
2. Horner's syndrome (pseudoenophthalmos).
3. Orbital fat atrophy.
4. Congenital abnormality.
5. Metastatic disease.

Malignant fibrous tumors of the breast

THYROID EYE DISEASE (TED)

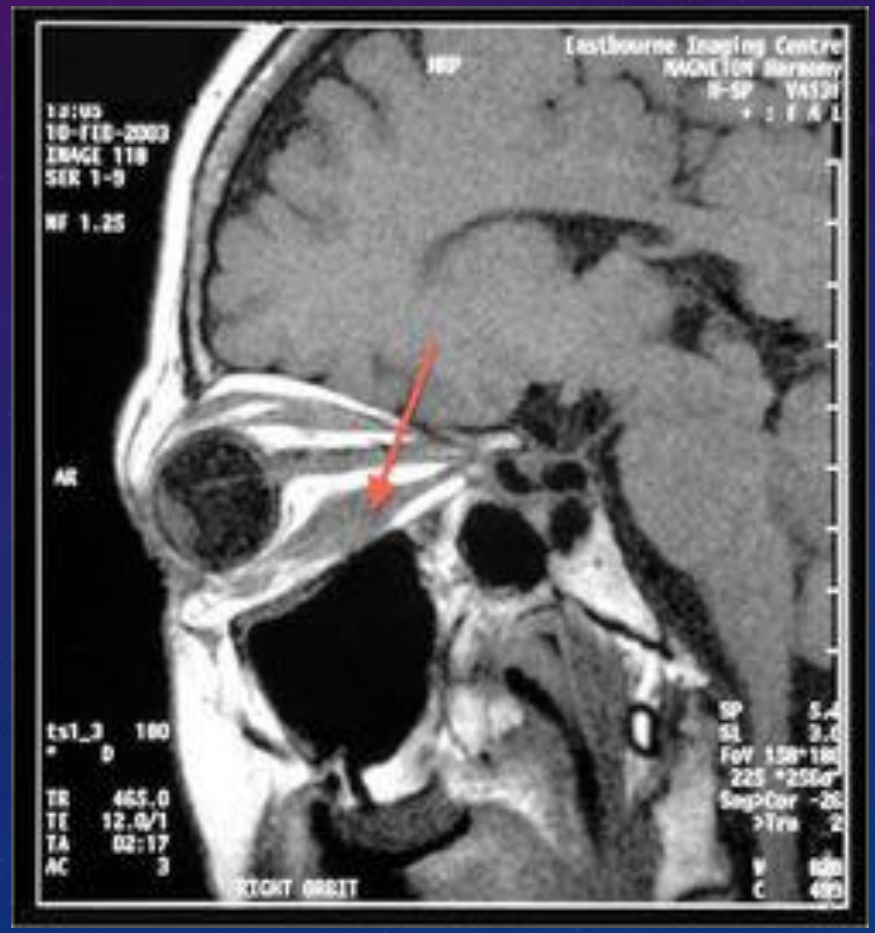
Mostly affects elderly

- The commonest cause of unilateral and bilateral Proptosis in adults.
- Marked by swelling of the extraocular muscles and fatty tissue around the eye.
- Swelling is caused by inflammation, and because the space is limited by the orbit, as swelling continues the eyeball will be pushed forward.
- Particularly in hyperthyroidism, but can happen in hypothyroidism.

Mostly affects young ages

- ✓ What's the most commonly affected muscle?

The inferior rectus muscle. then medial rectus then superior rectus then lateral rectus (Counterclockwise)



- ***<u>Signs & symptoms of Graves' opthalmopathy</u>***:

✓ Red, painful eye.

✓ Periorbital edema.

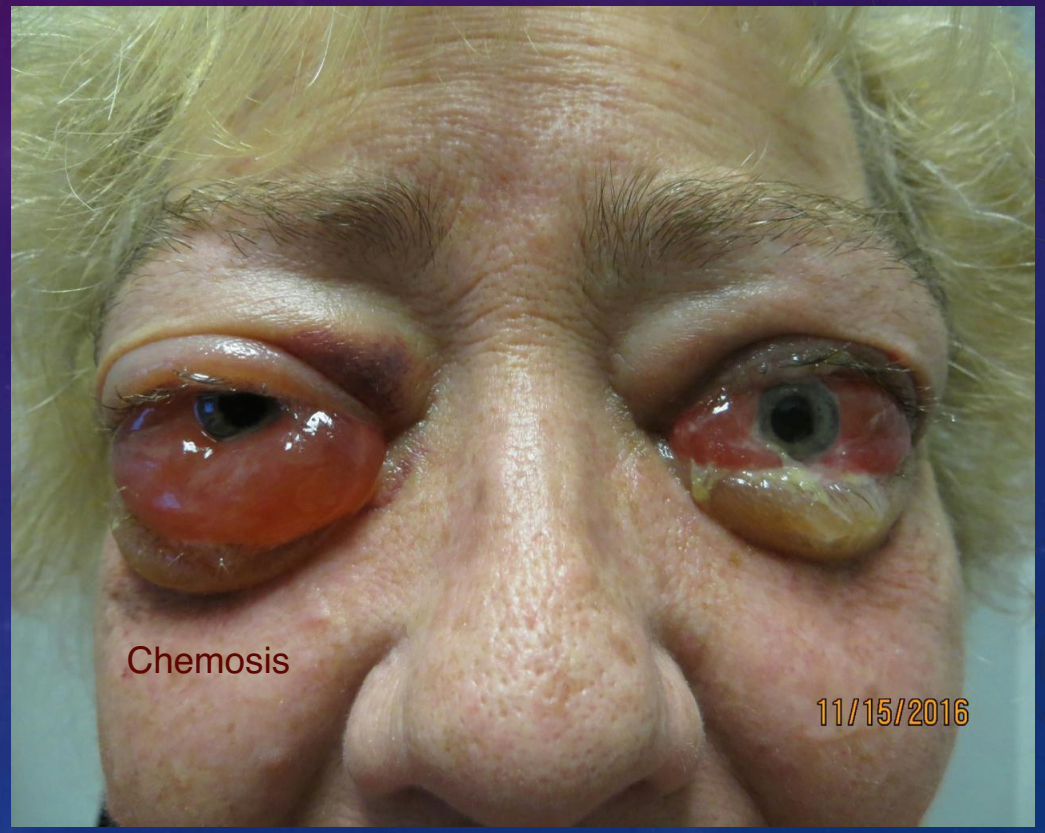
✓ Lid retraction (*sclera visible above or below cornea)*

✓ Conjunctivitis. (inflammatory not infectious)

✓ Diplopia.

✓ Proptosis.

✓ Reduced visual acuity.



- ***<u>On physical exam;</u>***

✓ Proptosis (asymmetrical).

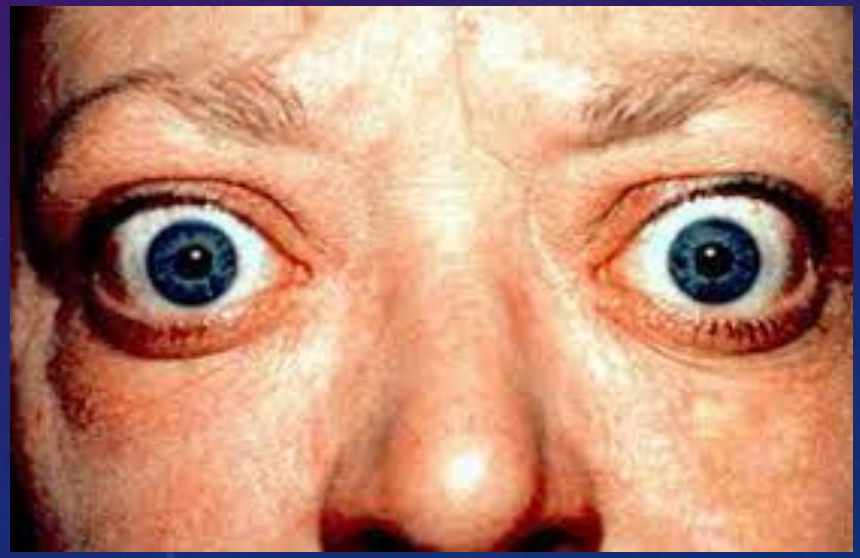
✓ Chemosis in conjunctiva.

✓ Lid retraction (staring appearance).

Why? Increased sympathetic activity.

✓ Lid lag (*behind movement of the globe on downgaze).*

✓ *+/-* restricted eye movement.



- ***Pathogenesis; (autoimmune)***

✓ TSH receptor antigen on orbital fibroblasts and adipocytes

✓ T-cell activation and cytokine release (TNF).

✓ Increased production of GAGs by fibroblasts (hyaluronic acid).

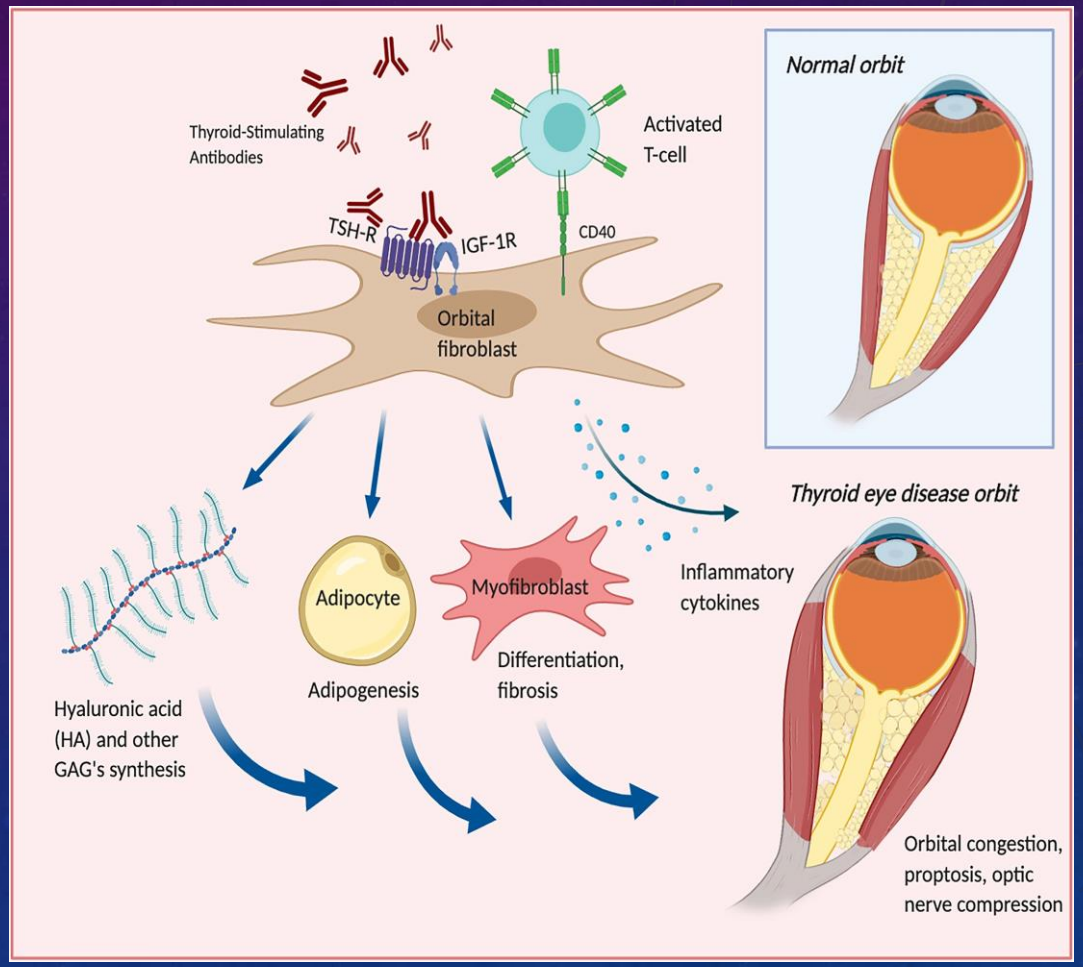
✓ Accumulate increasing osmotic pressure.

✓ Fluid accumulation and muscle swelling.

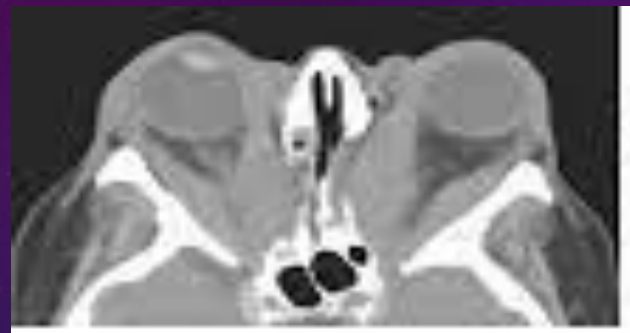
✓ Increases pressure within the orbit.

✓ Proptosis.

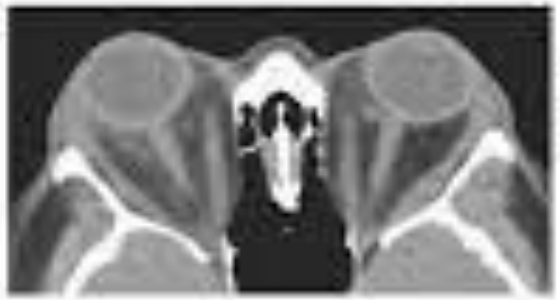
✓ Exposure of globes and limitation of eye movement.



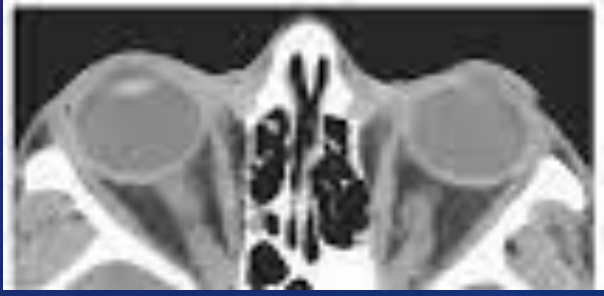
- ✓ TSH receptor mRNA and protein can be detected in orbital fibroblasts, adipocytes and pre-adipocytes in those patients.
- ✓ These cells express more TSH receptor mRNA and produce more cAMP in response to TSH than similar ones in normal people.



generalized enlargement of the extraocular muscles with marked bilateral proptosis



marked bilateral proptosis and asymmetric involvement of the extraocular muscles with expansion of the orbital fat bilaterally



Normal

- ***Complications?***

1. Excessive exposure of the conjunctiva and cornea with chemosis and corneal ulcer; *corneal perforation.*
2. Compression and ischemia of the optic nerve by the thickened muscle leading to *compressive optic neuropathy (visual field loss, blindness).*

- ***Trx of urgent complications?***

1. Systemic steroids.
2. Radiotherapy.
3. Surgical orbital decompression.

** We prefer to give it intravenously.
** If we didn't notice improvement after administering steroids for the 1st time we give them for another time and if the patient doesn't improve even after that we go for surgery.

- ***Assessment of severity;*** *ranges from 0-6/ pneumonic: NO SPECS*

Table 1 NO SPECS classification of eye changes in Graves' ophthalmopathy (Werner's classification[11])

Class	*Description*
0	**N**o physical signs or symptoms
1	**O**nly signs, no symptoms (eg, upper lid retraction, stare, and eyelid lag)
2	**S**oft tissue involvement (symptoms and signs)
3	**P**roptosis
4	**E**xtraocular muscle involvement
5	**C**orneal involvement
6	**S**ight loss (optic nerve involvement)

nmol/l; FT3L 1.3–2.7 nmol/l) because our pri

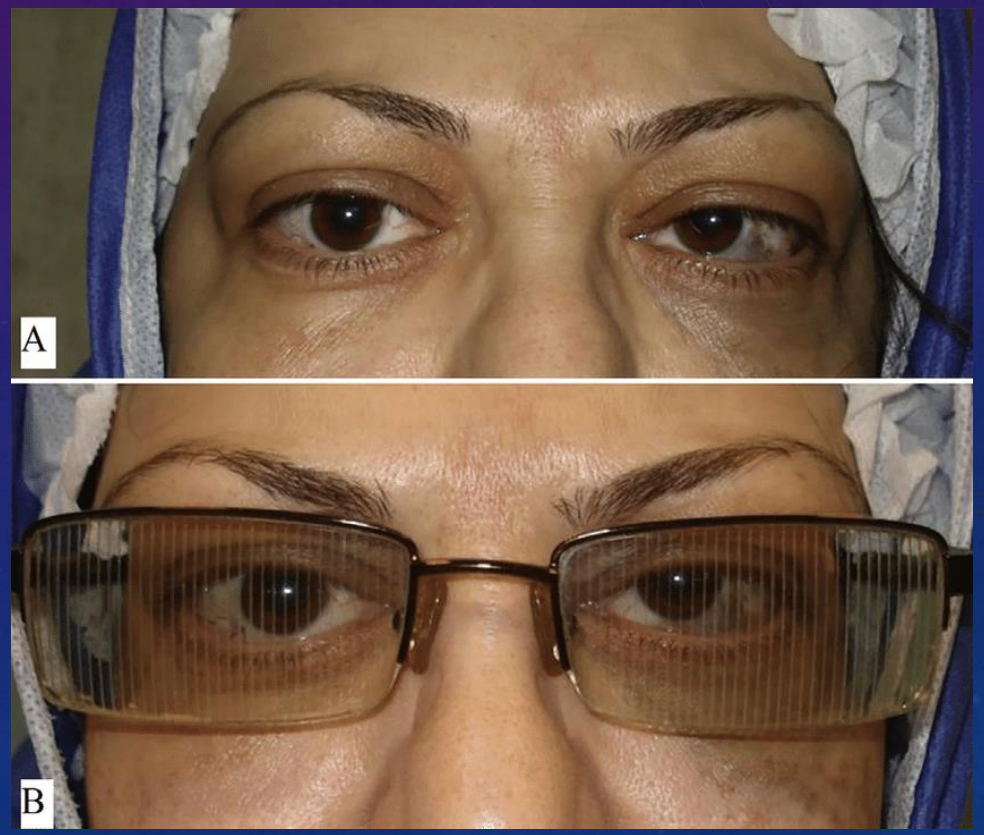
- ***Treatment aims?*** + to preserve vision

1. Improve eye movement problems.
2. Improve the cosmetic appearance.

- ***How?***

لا تغيير على شكل العين بل يكون التغيير على مستوى زاوية النظر / الرؤية

1. Prisms (Diplopia)
2. Corrective surgeries (extraocular muscles and upper eyelid).



DIPLOPIA

DEFINITION

- Diplopia or double vision is the simultaneous perception of two images of a single object that may be displaced horizontally, vertically, or diagonally (i.e. both vertically and horizontally) in relation to each other.

Diplopia vs blurred vision:
Diplopia: a single object is seen in duplicate (two images of a single object/ there is a space between these two images)
Blurred vision: a single image seen by one eye appears unclear (if we feel we're seeing two images of the same object they won't be separated by any spaces and this is what differentiates blurred vision from double vision)

CAUSES

1. Muscles involvement as
   in myosytis and dysthyroid diseases.

2. Nerve supply involvement.

3. Or the junction in between

TYPES

1. monocular diplopia is usually the result of a refractive error

2. binocular diplopia is commonly due to ((More common)) impaired extraocular muscle function, nerve, or N-M junction

Telling them apart can be done by carrying out the cover – uncover test.

Diplopia

Diplopia still present

Diplopia absent

MONOCULAR

BINOCULAR

non neurological

Eyes do not move in sync

likely to be refractive problem

most common cause is astigmatism (abnormal curvature of cornea)

likely pathologies

- cranial nerve palsies
- extraocular muscles dysfunction
- neuromuscular junction problems (myasthenia gravis)

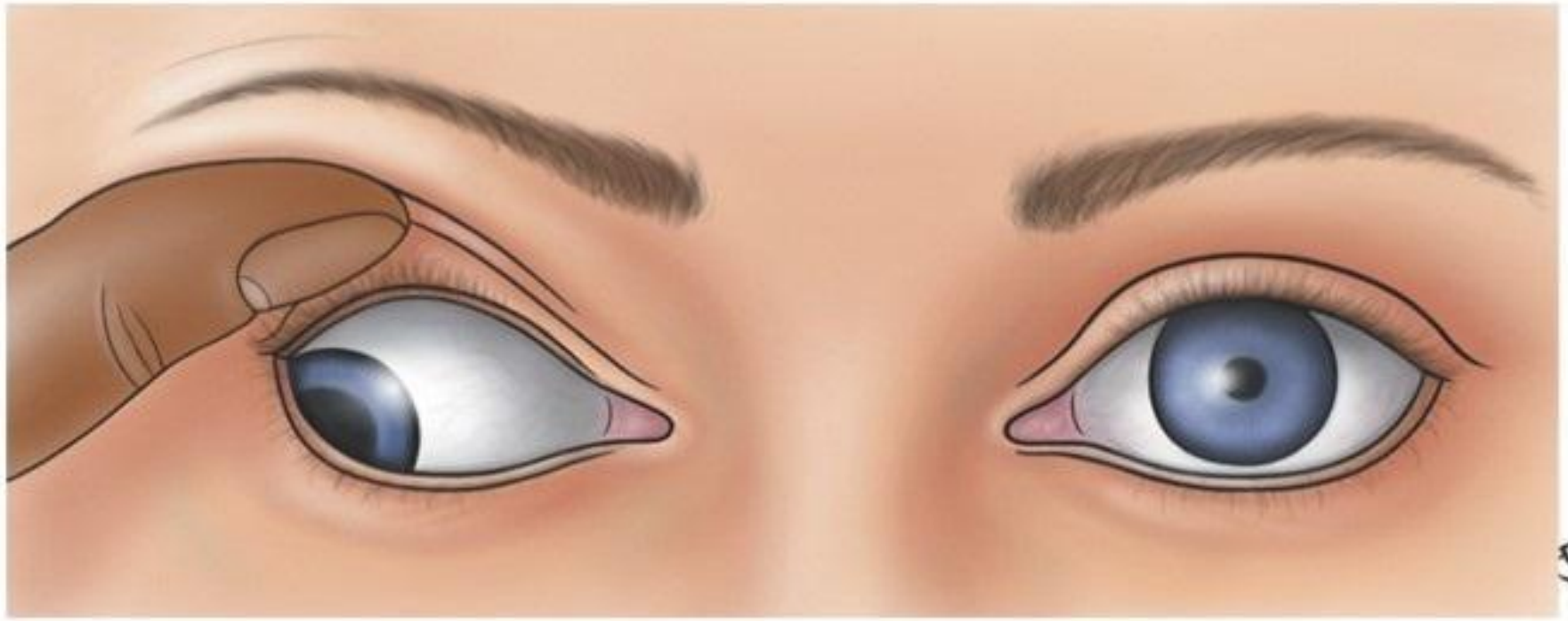
HISTORY TAKING IN DIPLOPIA

1. Pain :

This is essential to ask about in order to rule out a third nerve palsy resulting from a posterior communicating artery aneurysm, which is a neurological emergency.

2. **onset**: the acute onset is more suggestive of a vascular event but is not very specific, the gradual progression or diplopia that has changed the pattern is more indicative of a compressive lesion.

e.g: thyroidopathy

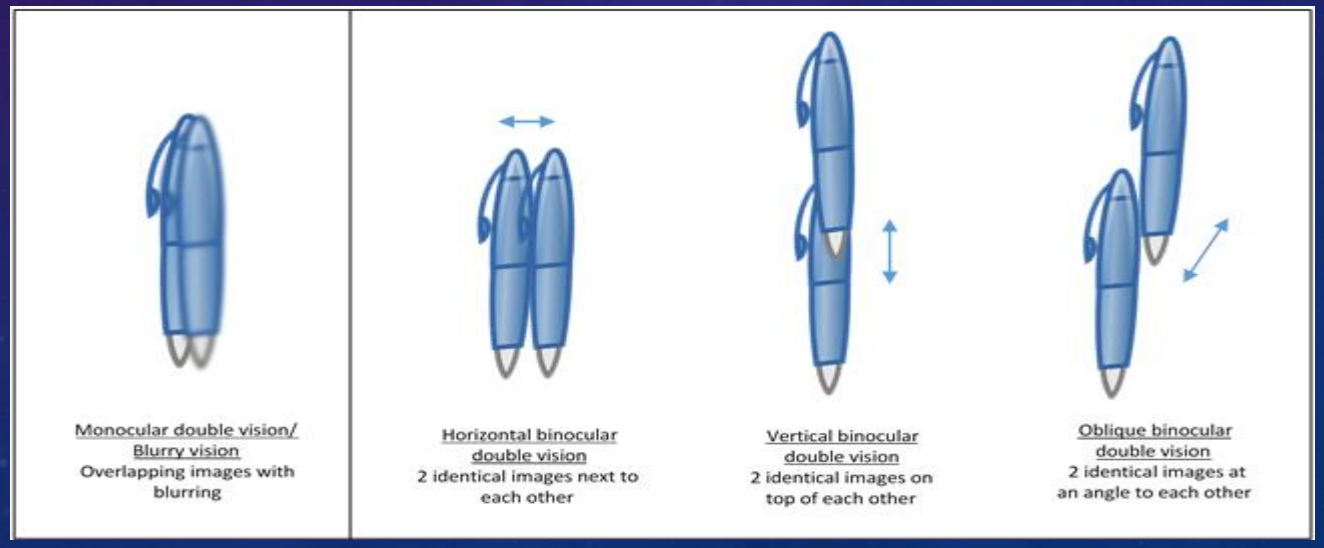


Right eye: Downward and outward gaze, dilated pupil, eyelid manually elevated due to ptosis

Left: Normal

3. Image alignment :

- Is the diplopia horizontal, vertical or oblique?
- Horizontal diplopia with no vertical separation is due to impaired function of lateral rectus and/or medial rectus. This can also commonly occur due to a sixth nerve palsy.
- Vertical or tilted diplopia can be indicative of a fourth nerve palsy. Vertical diplopia can appear in thyroid eye disease (inferior rectus muscle is most commonly affected), orbital floor fractures.



Gives indication of the affected muscle;
(1) Horizontal binocular diplopia : horizontal muscles (medial rectus & lateral rectus)
(2) Vertical binocular diplopia: vertical muscles (superior rectus & inferior rectus)
(3) Oblique binocular diplopia: oblique muscles (superior oblique & inferior oblique)

4. Constant or intermittent

- the characteristic timing of the diplopia is important as intermittent diplopia which is worse at the end of the day can suggest myasthenia gravis

5. Direction of gaze

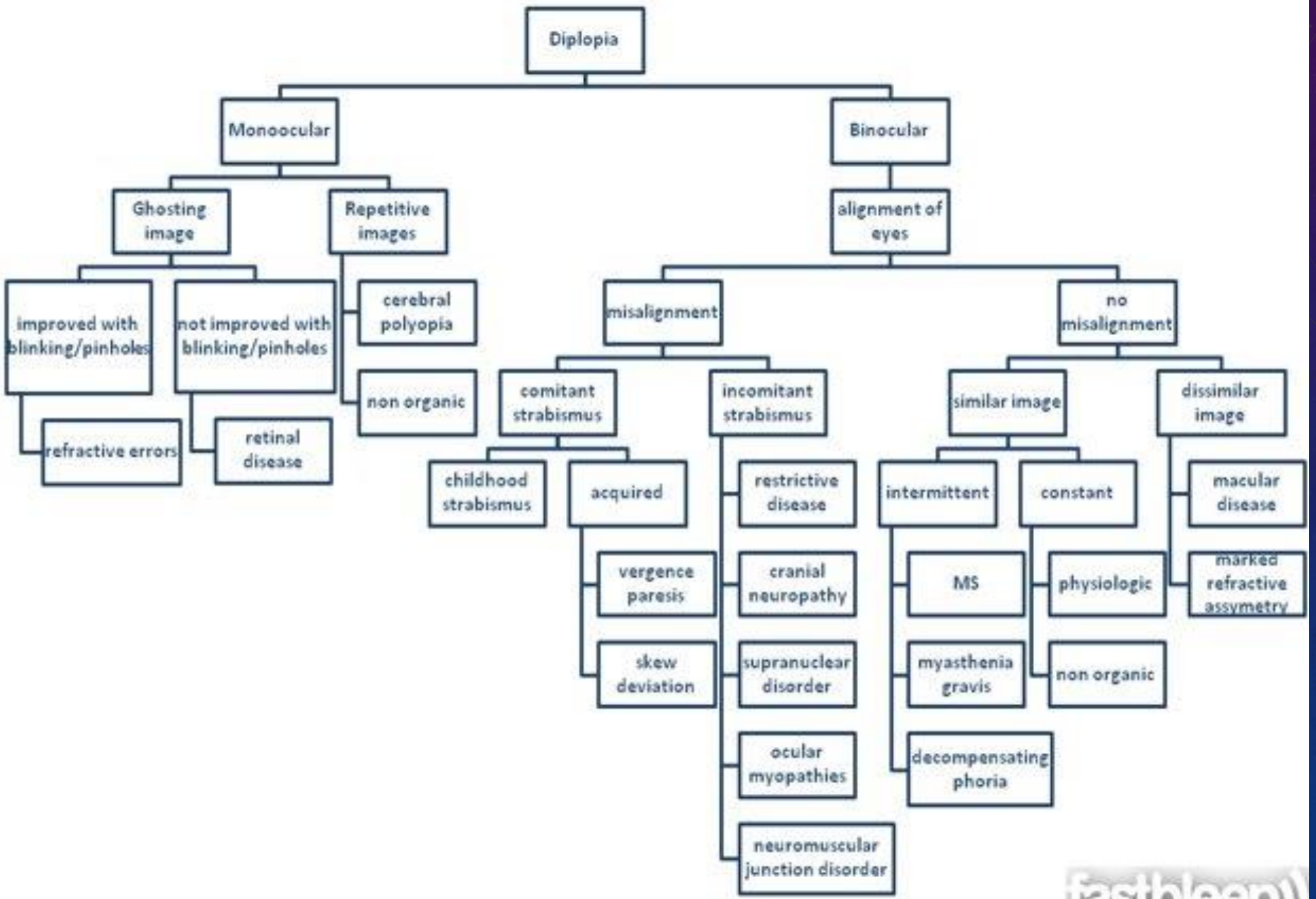
- This can be used to help locate the EOM involved and help to identify whether it is due to a paralytic or restrictive cause.
- Paralytic cause - diplopia when looking in direction of paralytic muscle.eg: Worsening of vertical diplopia in downgaze implicates either a trochlear palsy or inferior rectus weakness, while a worsening in upgaze will occur in complete third nerve palsies since there is weakness of both the superior rectus and inferior oblique
- Restrictive cause - diplopia when looking away from restrictive muscle (normal saccadic velocity.) e.g: thyroidopathy

- 5. **Exacerbating or relieving factors -** monocular diplopia can be improved by blinking if due to astigmatism or dry eyes, however there is no change if the diplopia is caused by macular disease or cataract.

- **6. Trauma -** It is important to enquire about any recent trauma. Any eye or head injury could lead to diplopia through various mechanisms.

- **Past medical history -** This is an integral part of assessing diplopia as any childhood strabismus, past prism correction via glasses and past ocular surgeries can lead to the development of subsequent diplopia.

- **Other medical conditions -** It is important to remember the bigger picture and therefore other systemic diseases should be asked about such as **diabetes**, **hypertension**, **temporal arteritis** and **thyroid disease**. Neurological symptoms should also be asked about such as **weakness**, **paraesthesia**, **blurred vision**, **loss of hearing**, **balance, dysphagia** and **headaches**


Diplopia
Monoocular
Binocular
Ghosting image
Repetitive images
alignment of eyes
improved with blinking/pinholes
not improved with blinking/pinholes
cerebral polyopia
non organic
misalignment
no misalignment
refractive errors
retinal disease
comitant strabismus
incomitant strabismus
similar image
dissimilar image
childhood strabismus
acquired
restrictive disease
intermittent
constant
macular disease
vergence paresis
cranial neuropathy
MS
physiologic
marked refractive assymetry
skew deviation
supranuclear disorder
myasthenia gravis
non organic
ocular myopathies
decompensating phoria
neuromuscular junction disorder

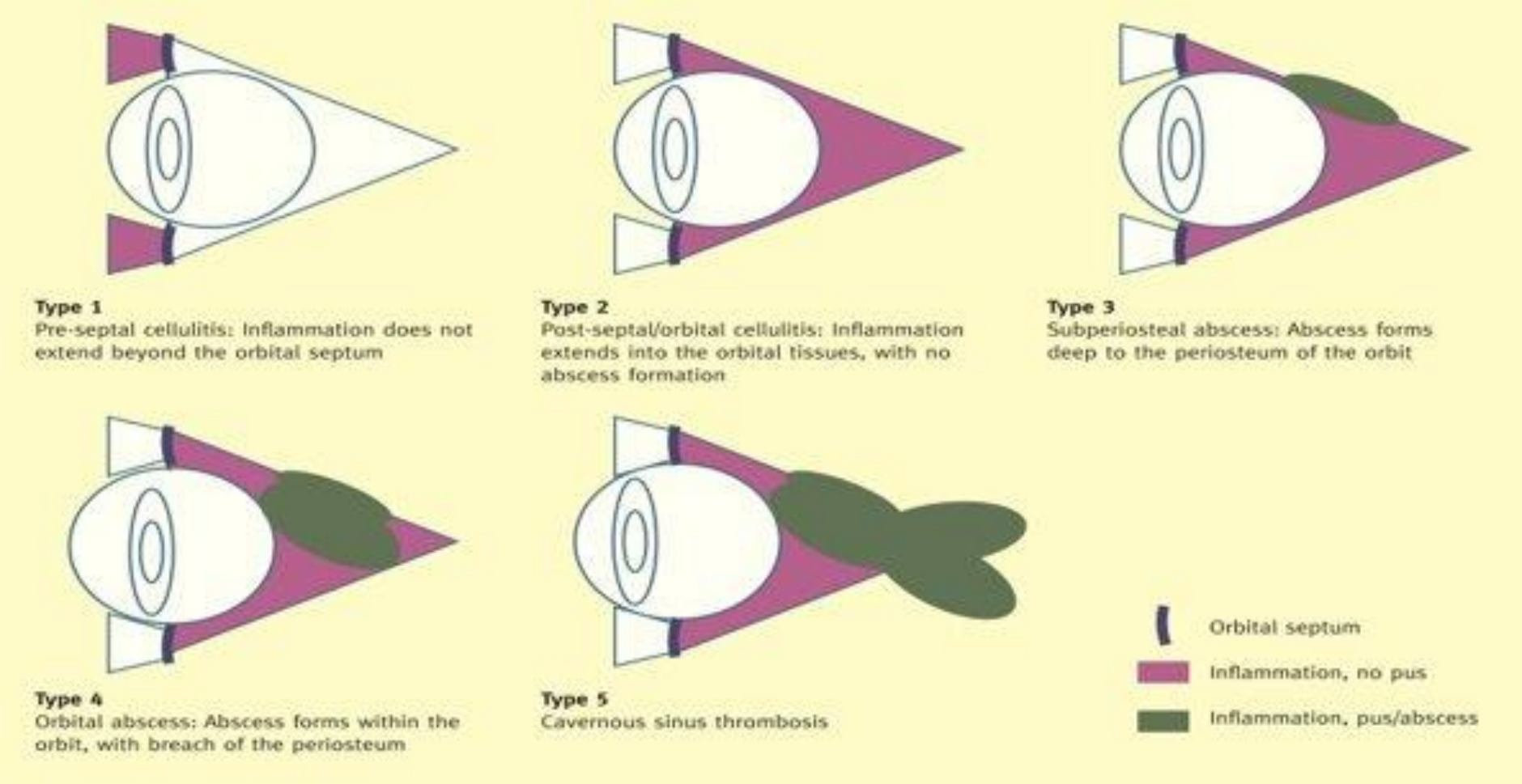
DIFFERENTIAL DIAGNOSIS OF ORBITAL DISEASES

DIFFERENTIAL DIAGNOSIS OF ORBITAL DISEASE

- Traumatic orbital disease
- Infective disorders
- Vascular abnormalities
- Orbital tumors

INFECTIVE DISORDERS

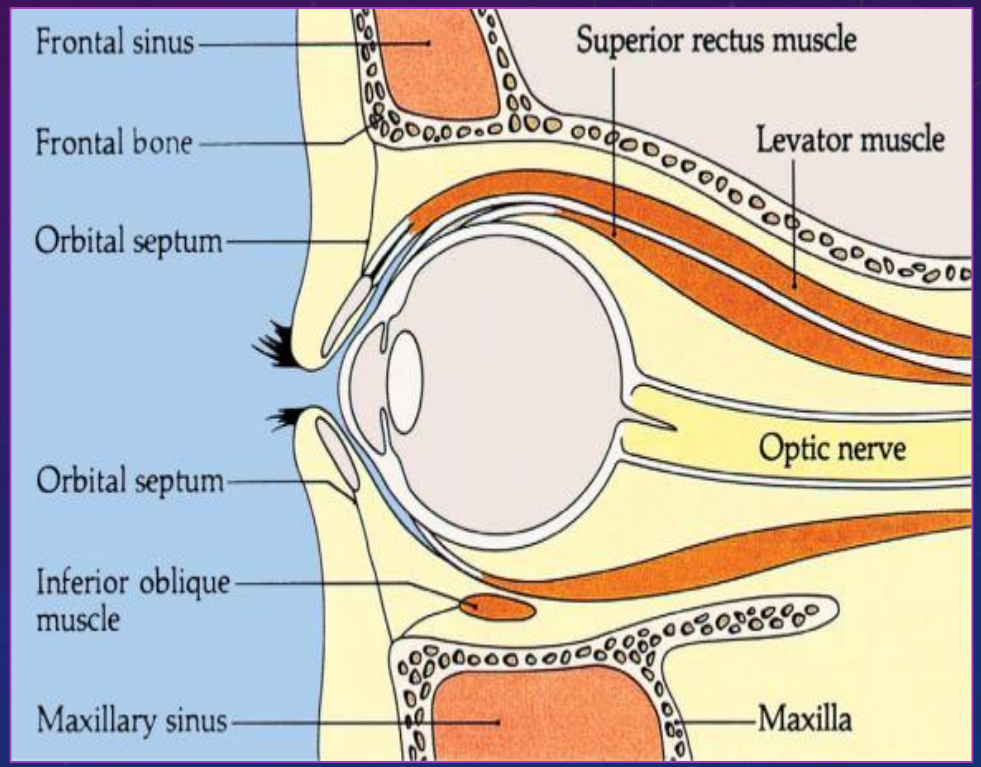
- Classification of orbital infections (Chandler's) :



A - PRESEPTAL CELLULITIS

- Is the infection of soft tissue

anterior to orbital septum (lid structures {skin and muscle})



- Etiology

usually follows periorbital trauma

or dermal infection

- Clinical Features

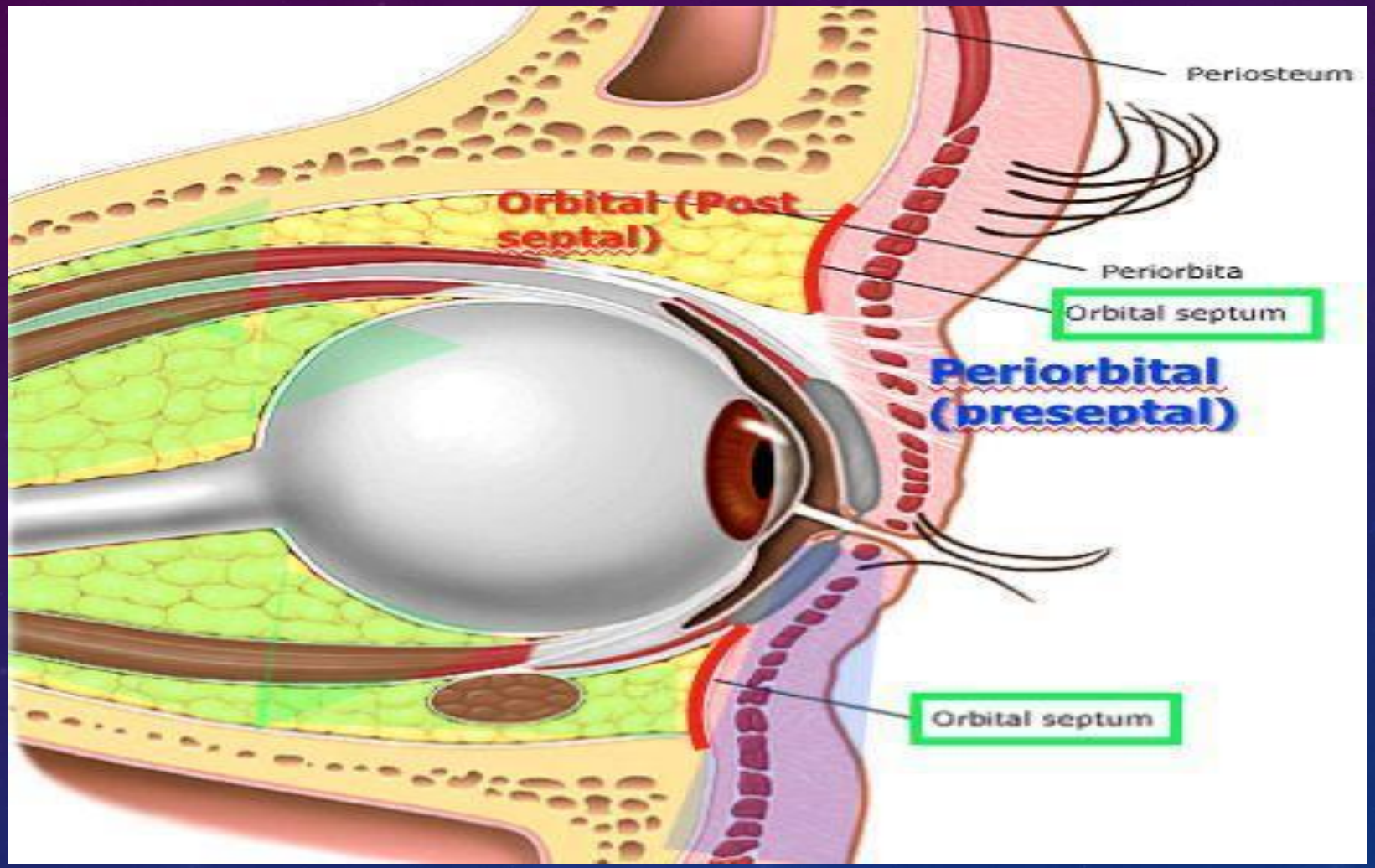
• tender, swollen, and erythematous lids .

• ± low-grade fever .

• Normal visual acuity , pupils , extraocular movements .

• NO exophthalmos .

** It may lead to orbital cellulitis .


Periosteum
Orbital (Post septal)
Periorbita
Orbital septum
Periorbital (preseptal)
Orbital septum

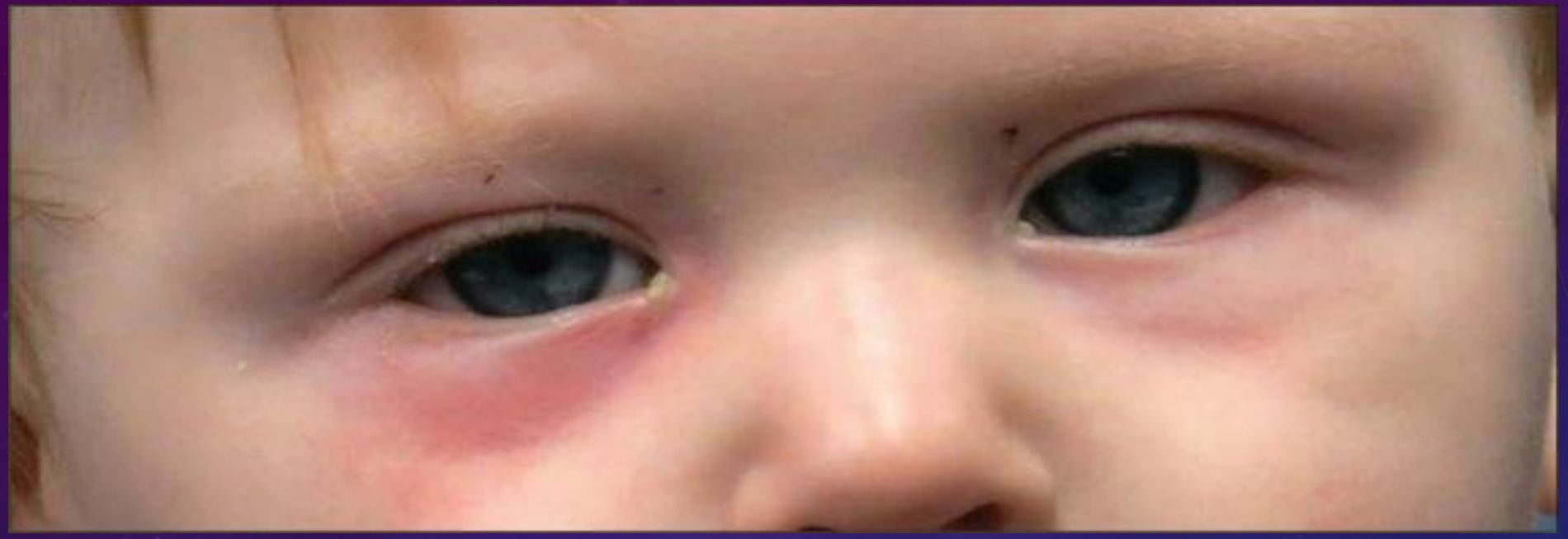
- Treatment

** In adults, topical antibiotics can be as effective**

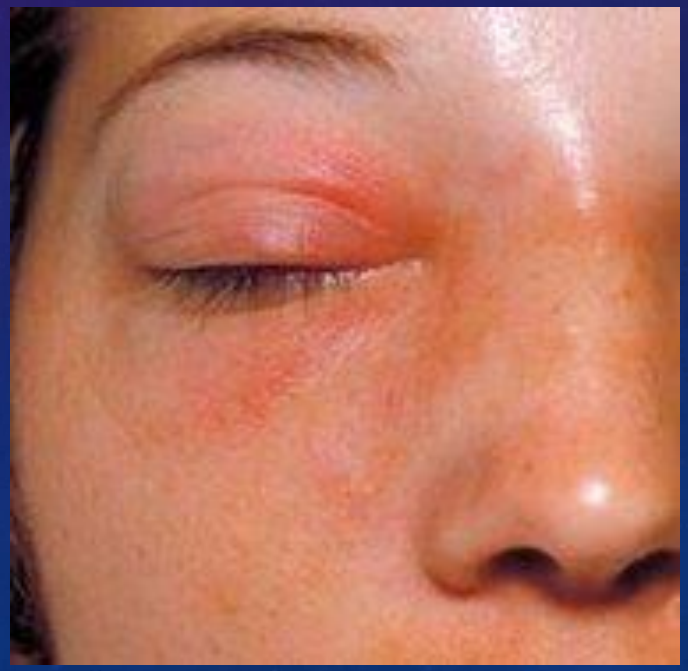
- systemic antibiotics (suspect *H. injluenzae in children ; S. aureus or Streptococcus in adults)*

e.g. amoxicillin-clavulanic acid .

- if severe or child < 1 yr treat as orbital cellulitis .



Preseptal cellulitis



B - ORBITAL CELLULITIS :

- It's an ocular and medical **EMERGENCY!** Defined as an inflammation of orbital contents (fat + muscles) posterior to orbital septum, common in children, elderly, and immunocompromised .

⬅Etiology

(venous drainage)

-usually 2ry° to sinus (ethmoid) / tooth infections or trauma

-Most common microorganisms : Staphylococcus and streptococcus .

- Clinical Features

1. red eye .
2. periorbital inflammation and swelling .
3. pain with and without eye movement .
4. headache and fever .
5. lid erythema, tenderness, and edema with difficulty opening eye .
6. conjunctival injection and chemosis (conjunctival edema) .
7. proptosis, limitation of ocular movements (ophthalmoplegia) .
8. decreased visual acuity (visual loss is possible !) with ± RAPD (Relative afferent pupillary defect).

- **Diagnosis :**

  MRI or CT

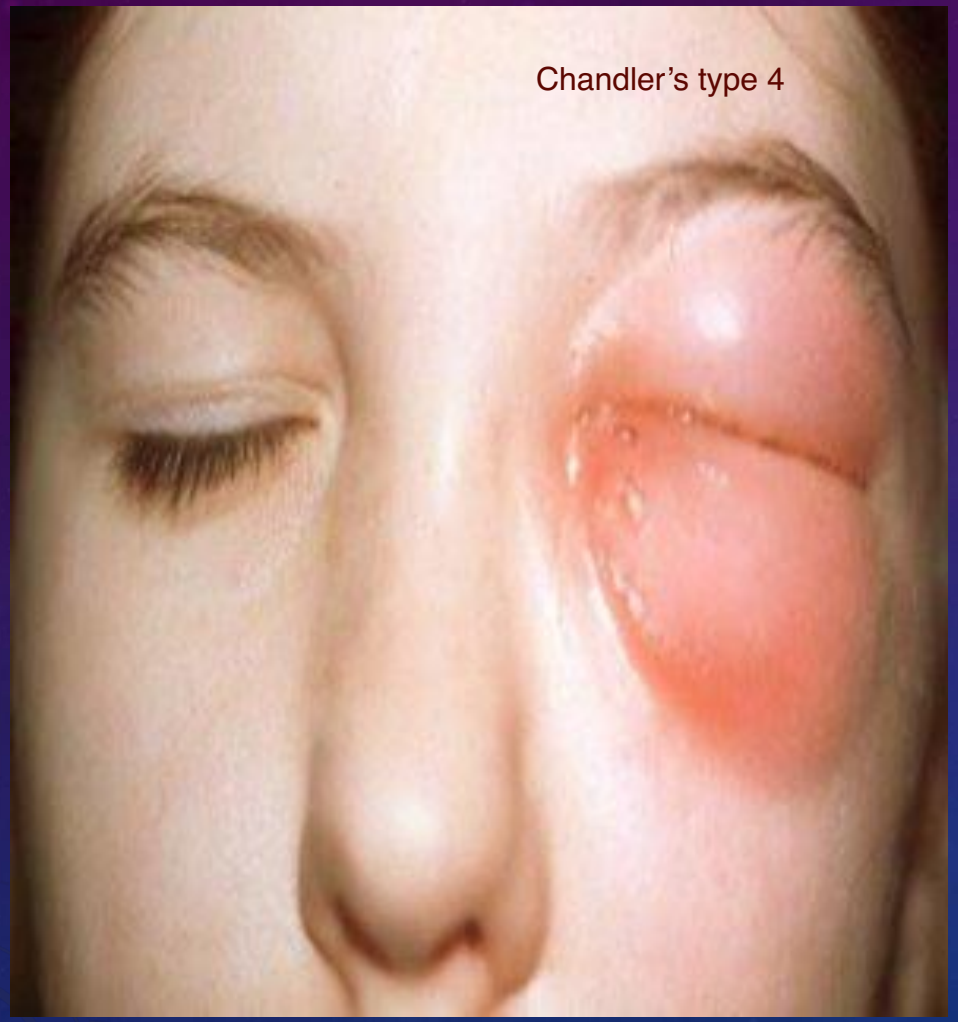
←Treatment :

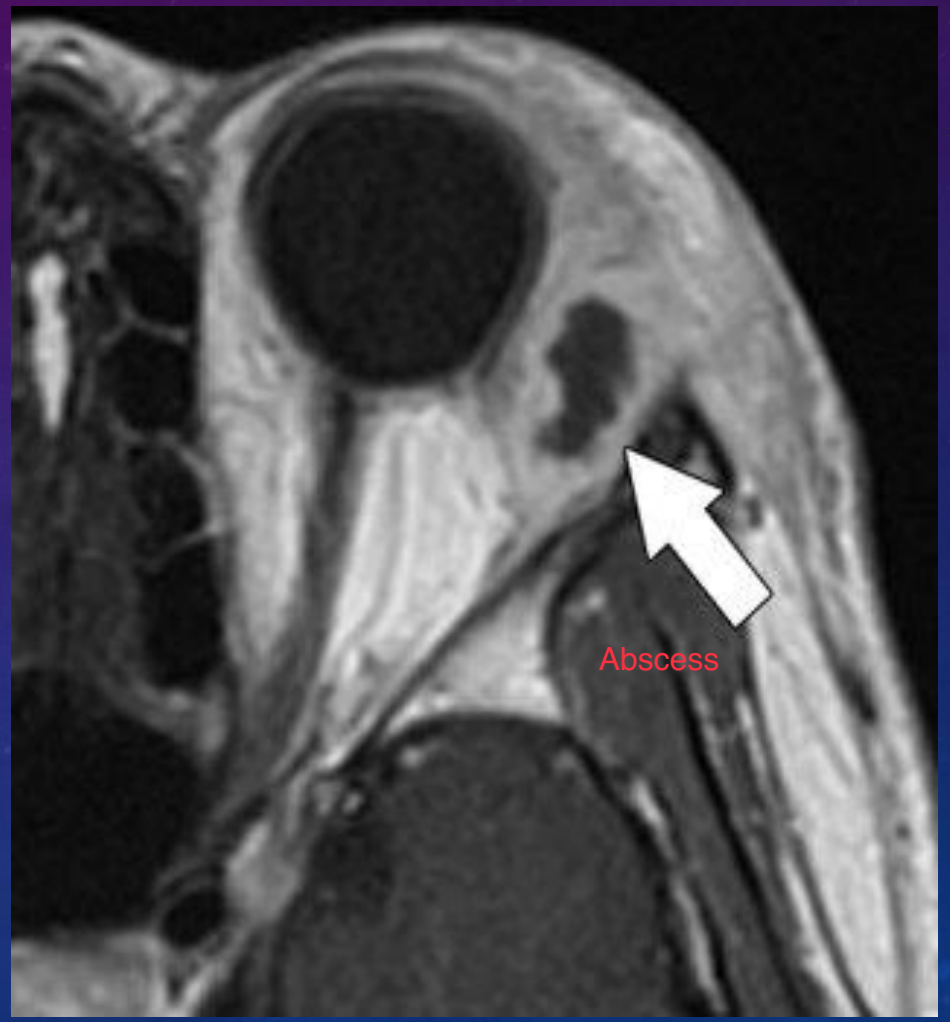
1. Admit the patient, blood cultures, orbital CT, IV antibiotics (ceftriaxone + vancomycin) for 1 week .
2. Surgical drainage of abscess with close follow-up, especially in children .
3. Optic nerve decompression if it's compromised .
   (Endoscopic optic nerve decompression is **a minimally invasive procedure used to relieve some of the pressure on the optic nerve and stabilize or improve vision by removing a portion of the bony optic canal).**

Complications

1. *optic nerve inflammation
2. *cavernous sinus thrombosis
3. *meningitis and brain abscess
4. * possible loss of vision
5. *and in most severe cases death !

Orbital cellulitis





ORBITAL TUMORS

- It can be ***primary*** neoplasm arising from any of the anatomical structures of the orbit.

- Or ***secondary*** orbital invasion from direct extension from contiguous anatomical structures, lymphoproliferative disorders, and hematogenous metastasis.

- **Lacrimal gland tumors:**

Malignant lacrimal gland tumors carry a poor prognosis. Benign tumors still require complete excision to prevent malignant transformation.

- **Optic nerve gliomas:**

may be associated with neurofibromatosis . They are difficult to treat but are often slow - growing and thus may require no intervention.

ADDITIONAL I Optic nerve glioma treatment

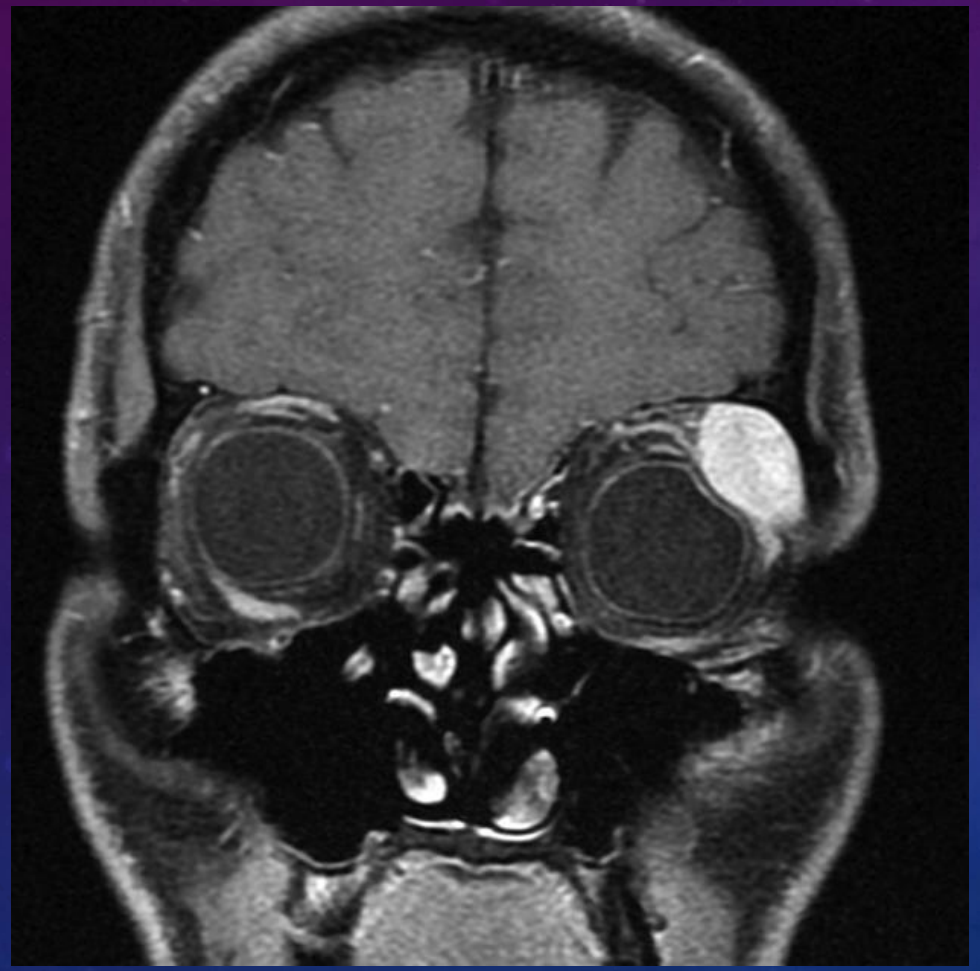
* Treatment of optic nerve gliomas in children remains controversial and is only indicated for those patients with a decline in visual acuity or significant radiologic progression.

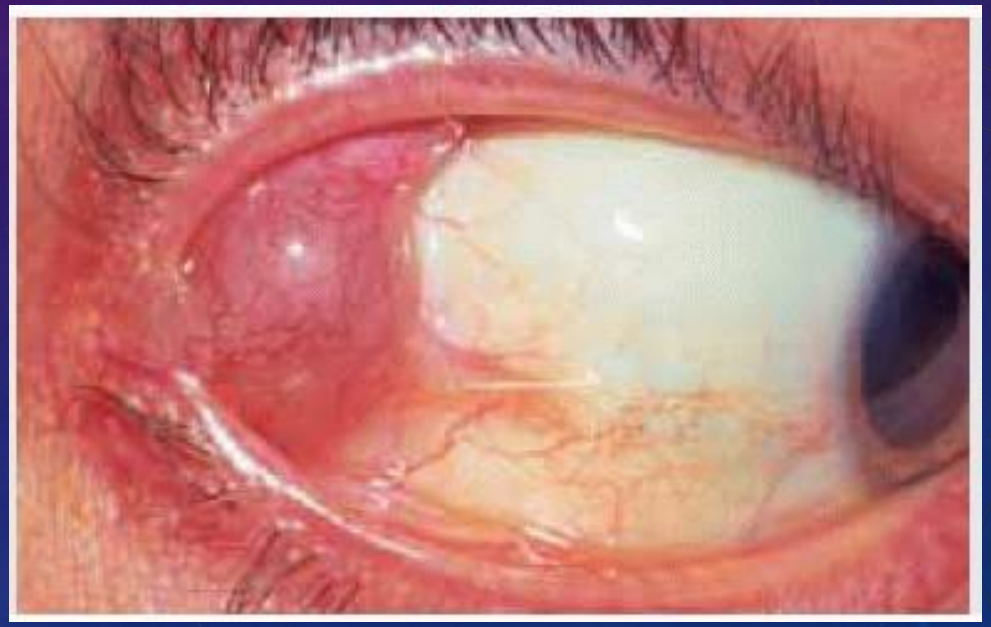
* First-line treatment of optic nerve gliomas remains standard chemotherapy despite modest visual outcomes.

* Molecularly targeted therapies represent a promising new treatment modality for those patients with refractory disease.

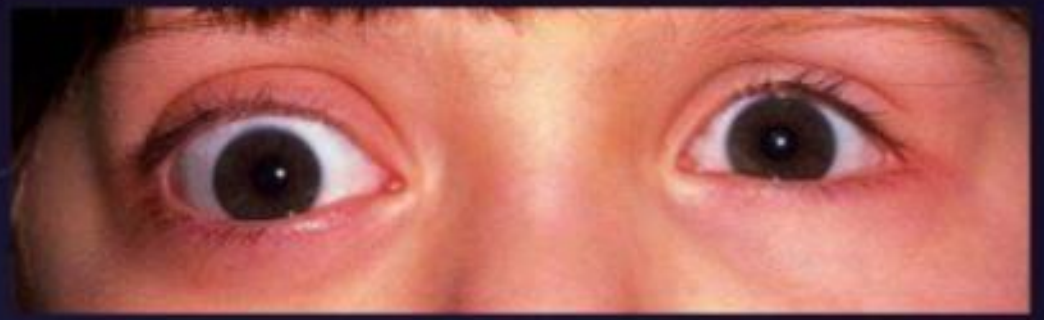
* Although used in the past with good results, radiotherapy (gamma knife radiosurgery) and radical surgical resection are associated with high morbidity and should be reserved as a last resort.

Lacrimal gland tumors:

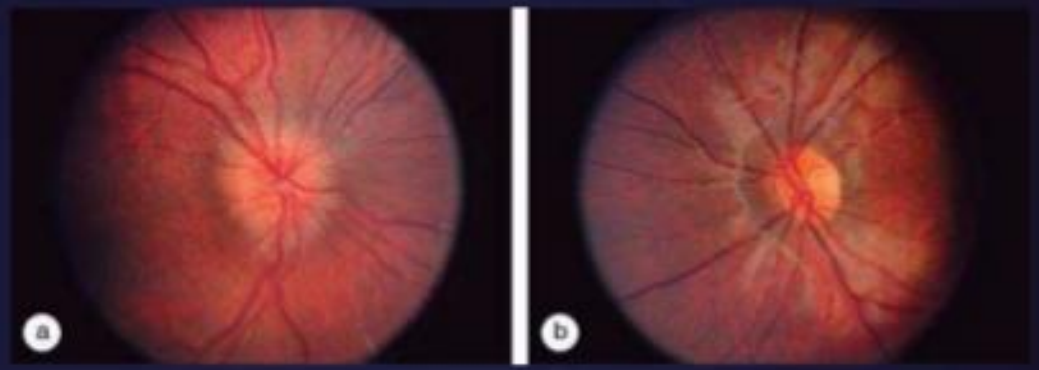




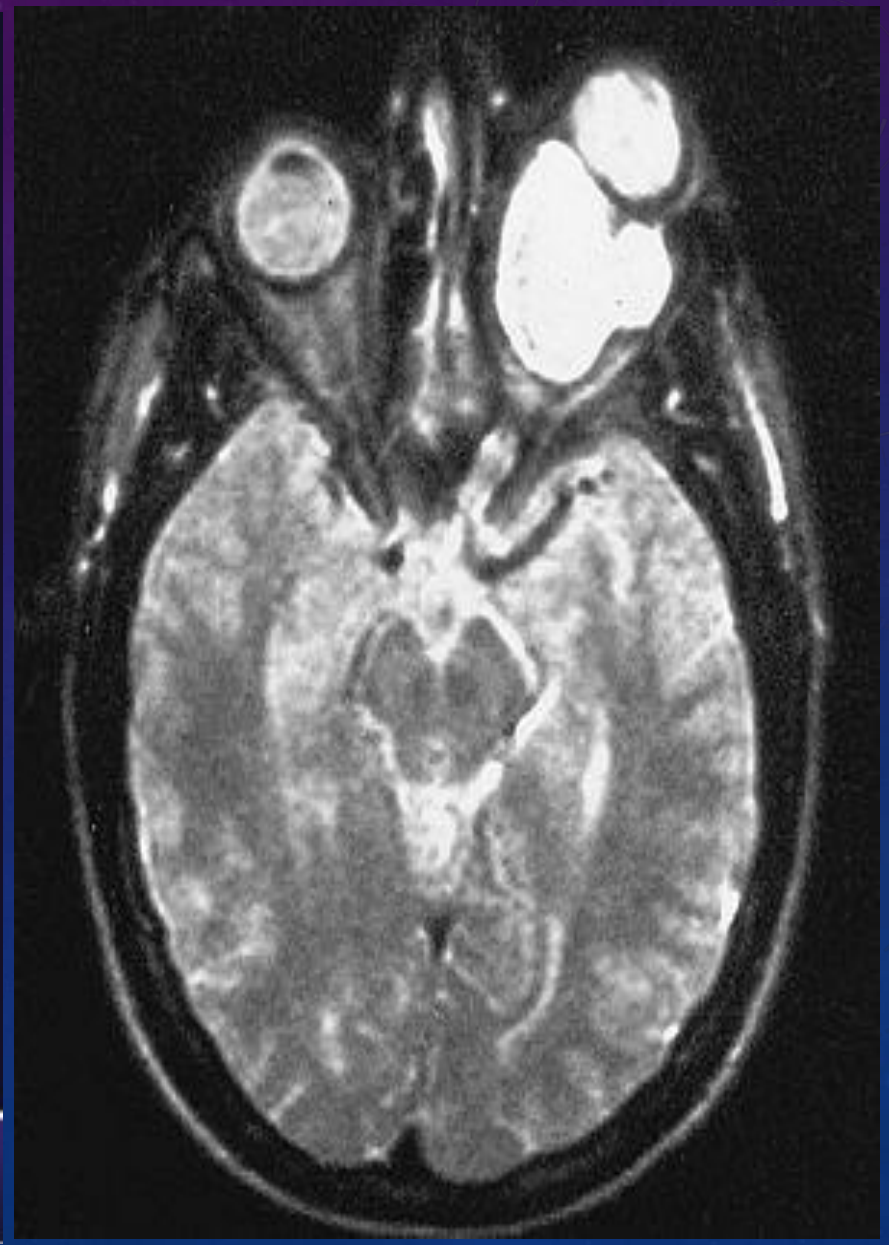
Optic nerve gliomas:



Face photo of a 5-year-old girl who developed noticeable proptosis OD and found to harbor an optic nerve glioma.



(a) The right fundus of a 5-year-old girl with a right optic nerve glioma reveals an optic nerve with mild edema. Her visual acuity was moderately to severely reduced. **(b)** The left nerve was normal.



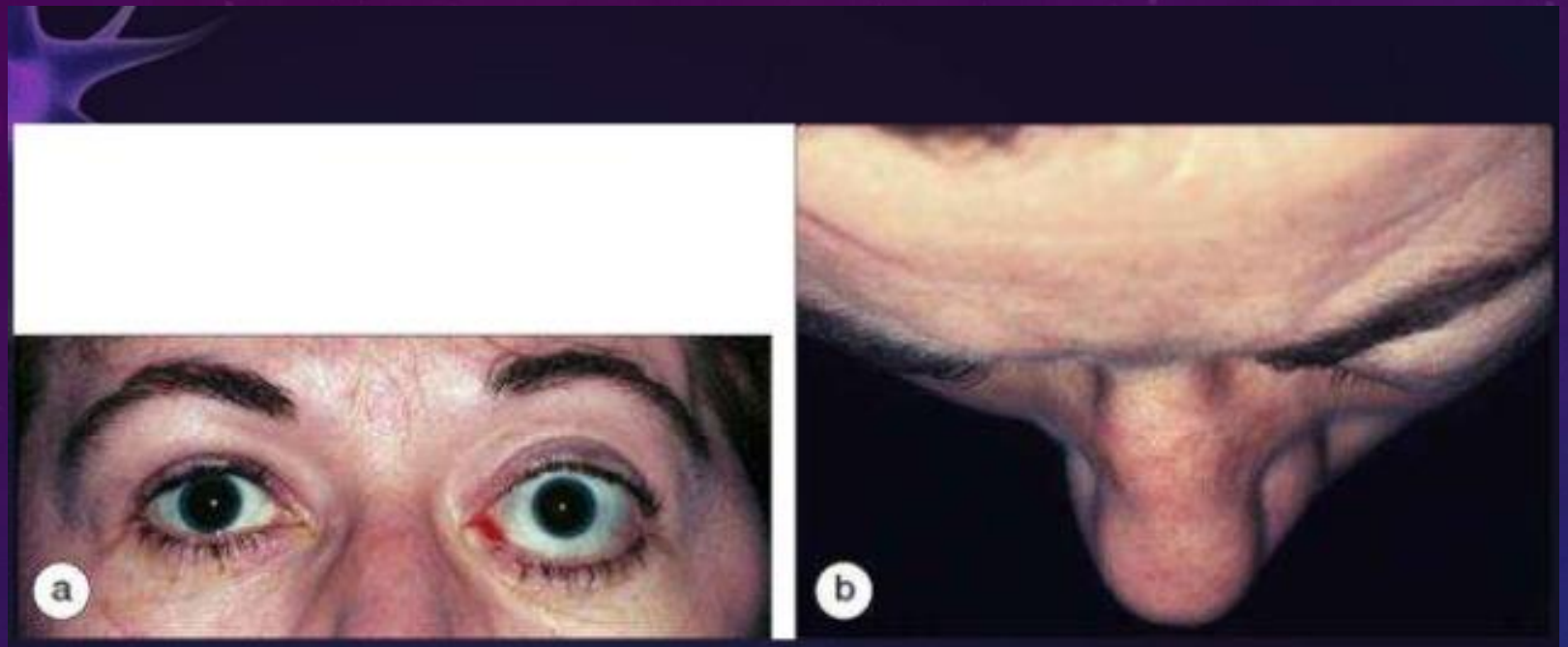
- **Meningiomas**

Meningiomas of the optic nerve sheath are **rare**, and may also be **difficult to excise**.

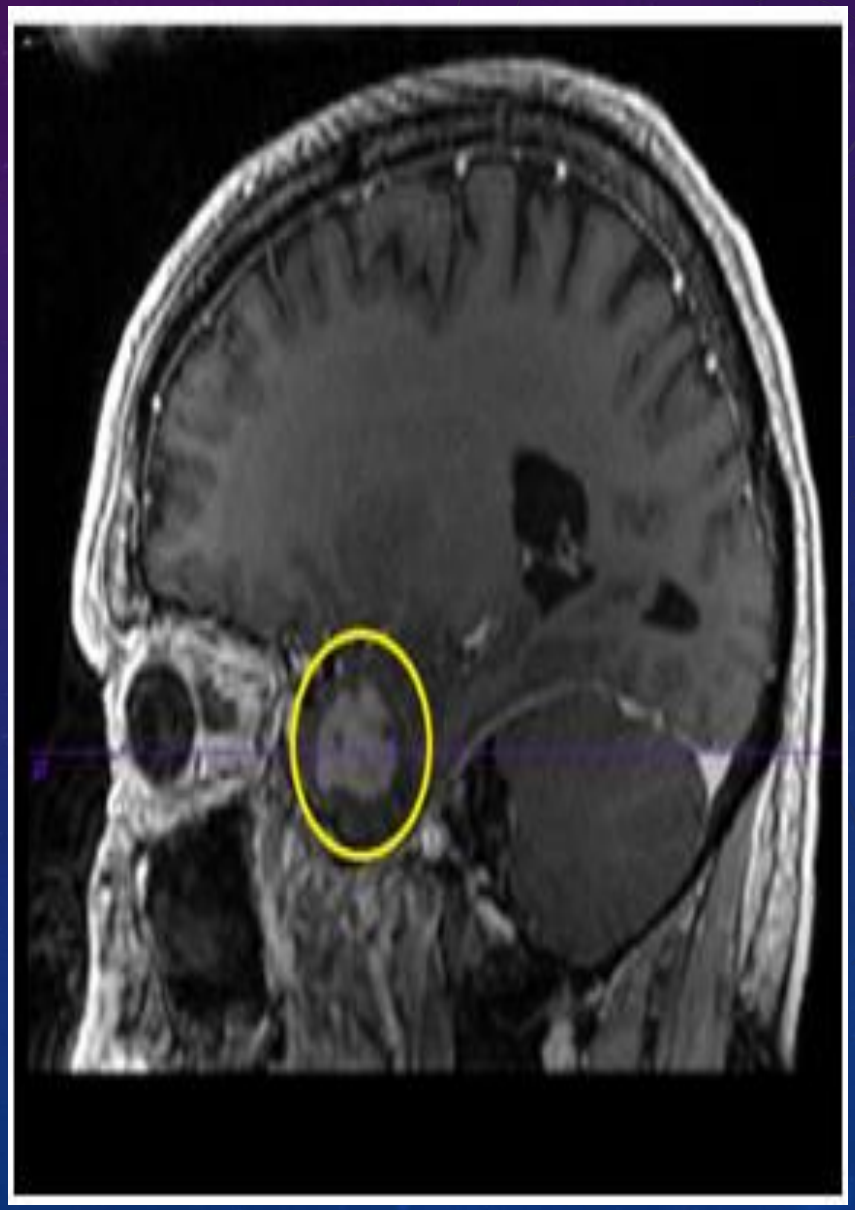
They can be monitored over time and some may benefit from treatment with radiotherapy.

Meningiomas arising from the middle cranial fossa, however, may spread through the optic canal into the orbit.

Meningiomas



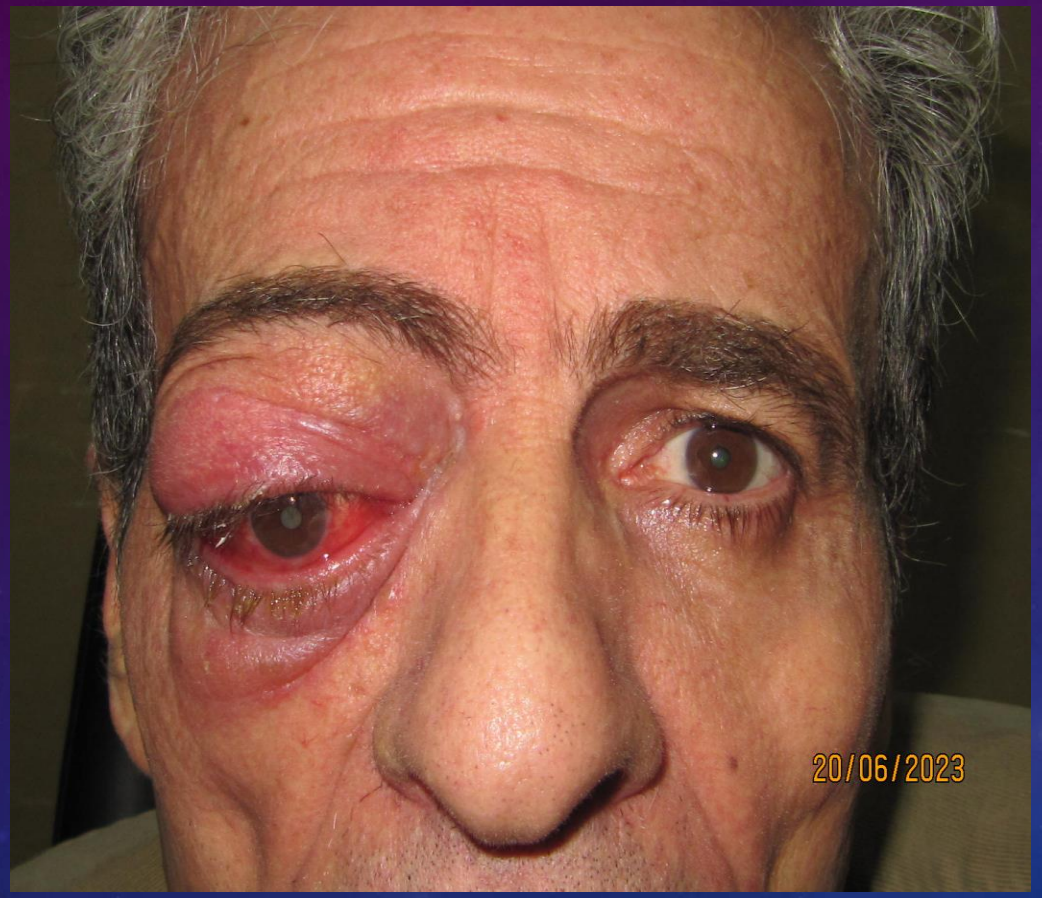
Face shot of a woman complaining of proptosis of the left eye. Neuro-imaging revealed a left sided sphenoid wing meningioma. The left eye shows lid retraction and lagophthalmos with protrusion of the left globe. **(b)** Picture taken from above the patient demonstrating the degree of proptosis visually.

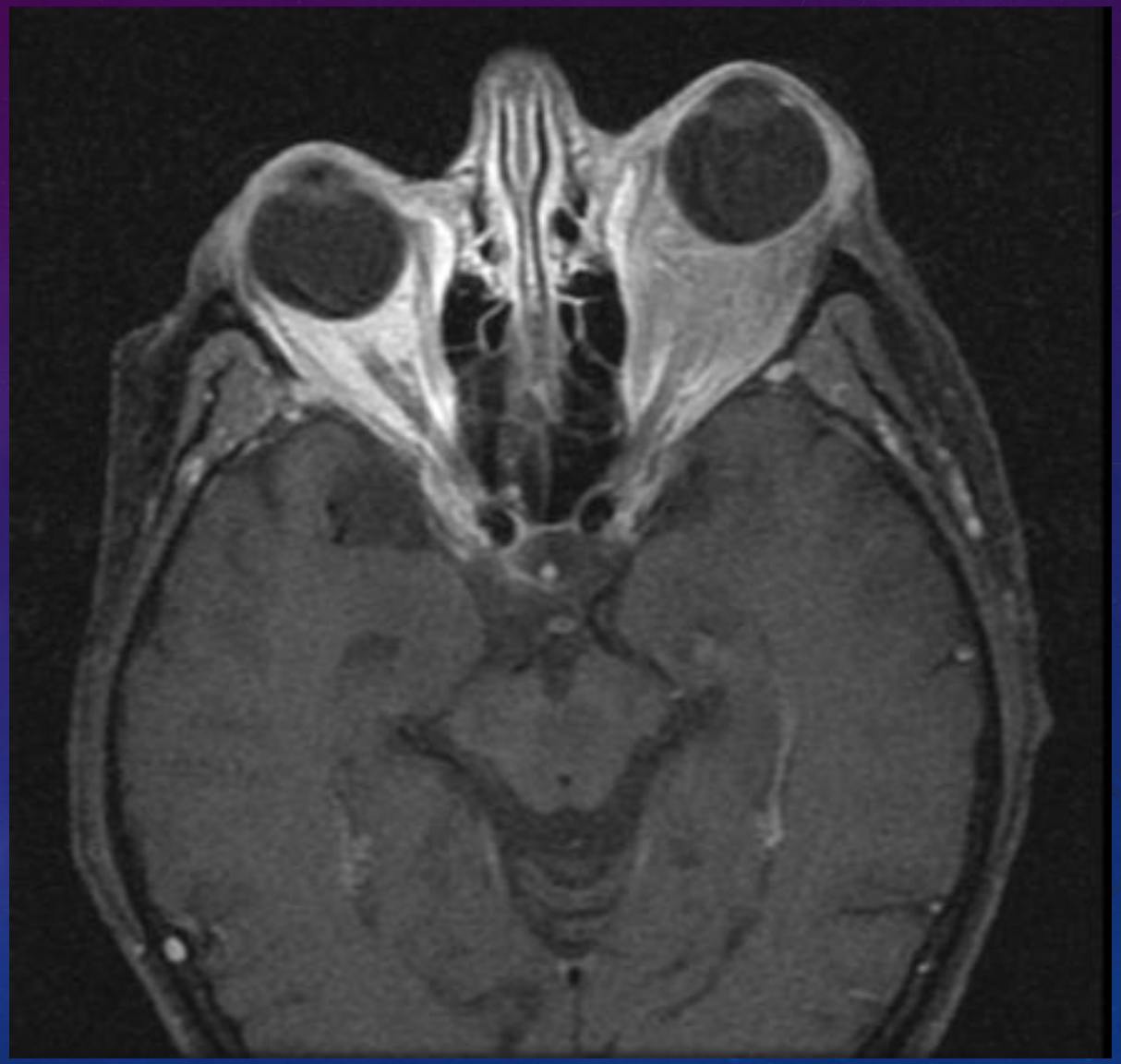


- **Lymphomas**

Treatment requires a full systemic investigation to determine whether the lesion is indicative of widespread disease or whether it is localized to the orbit. In the former case the patient is treated with chemotherapy, in the latter with localized radiotherapy.

Lymphomas





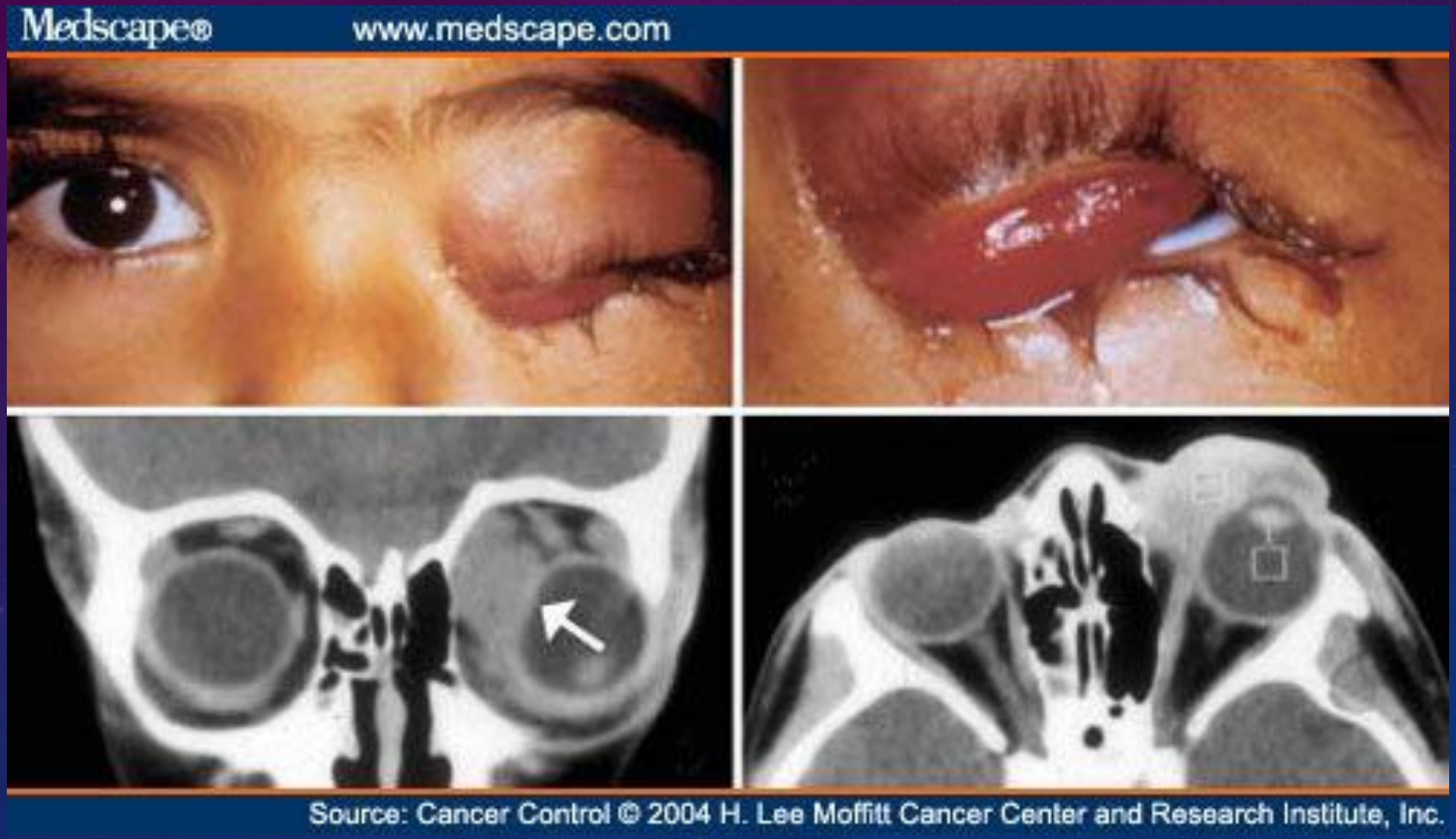
- **Rhabdomyosarcoma**:

It's the commonest **malignant primary** orbital tumor in children (vs. capillary hemangiomas as the most common **benign** orbital tumors affecting children).

Rhabdomyosarcoma is a rapidly growing, malignant tumor of primitive striated muscle. Chemotherapy is effective if the disease is localized to the orbit.

Rhabdomyosarcoma:

Can be confused with orbital cellulitis as it manifests similarly and is rapidly growing. However, there are no signs of inflammation in rhabdomyosarcoma.



- **Metastasis from other systemic cancers**

***Children:** (especially when in the thoracic part of the sympathetic chain)

Most commonly from : Neuroblastomas, Ewing sarcoma, Wilms tumor, and leukemias

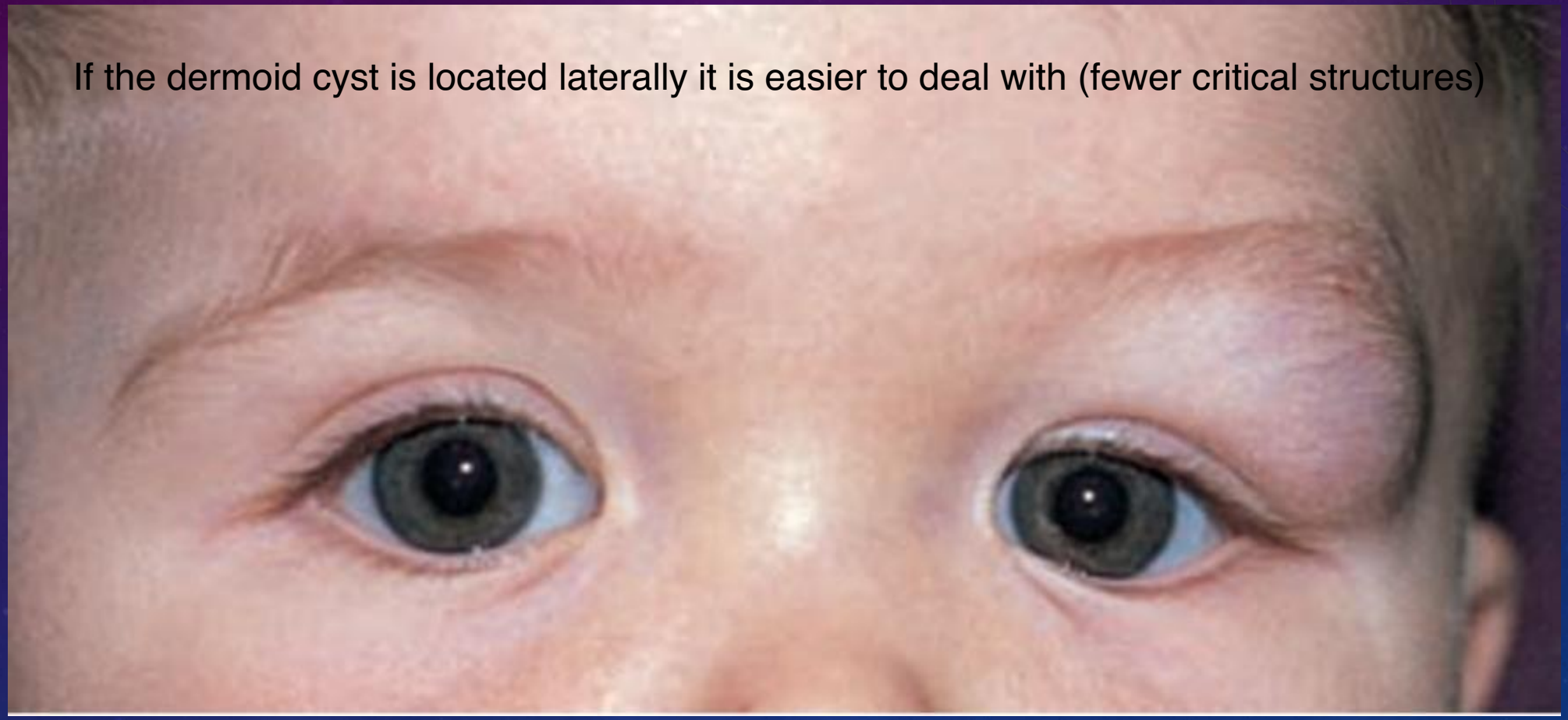
***Adults:**

Breast, lung, prostate, or GI tract

DERMOID CYSTS

- These congenital lesions are **caused by the continued growth of ectodermal tissue beneath the surface**, which may present in the medial or lateral aspect of the superior orbit .
- Excision is usually performed for **cosmetic** reasons and to **avoid traumatic rupture**, which may cause scarring.
- Some may be attached deeply by a stalk, and a CT scan may be necessary before surgery to identify this deeper connection.
-

If the dermoid cyst is located laterally it is easier to deal with (fewer critical structures)



THANK YOU!