

# OUTLINE

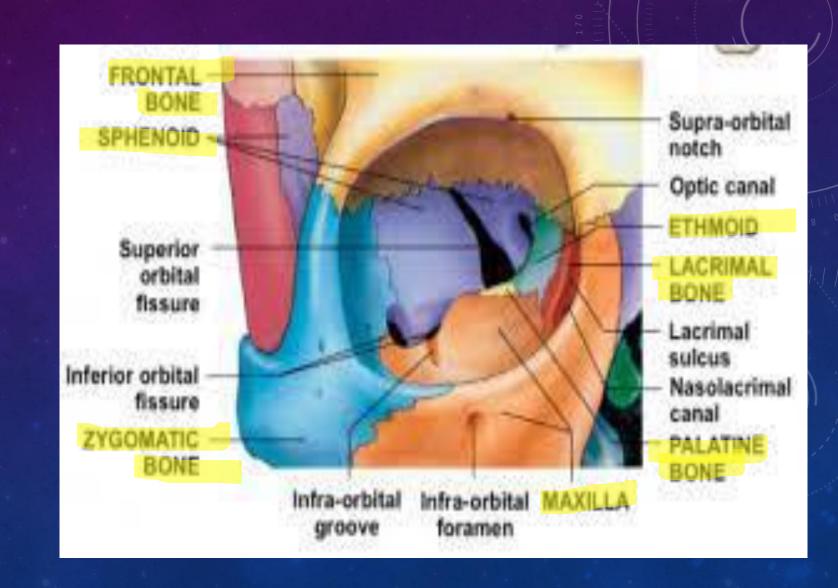
- Definitions
- Anatomy of the orbit.
- Proptosis.
- Thyroid eye disease.
- Diplopia.
- D.dx of orbital diseases.
- Orbital tumors.

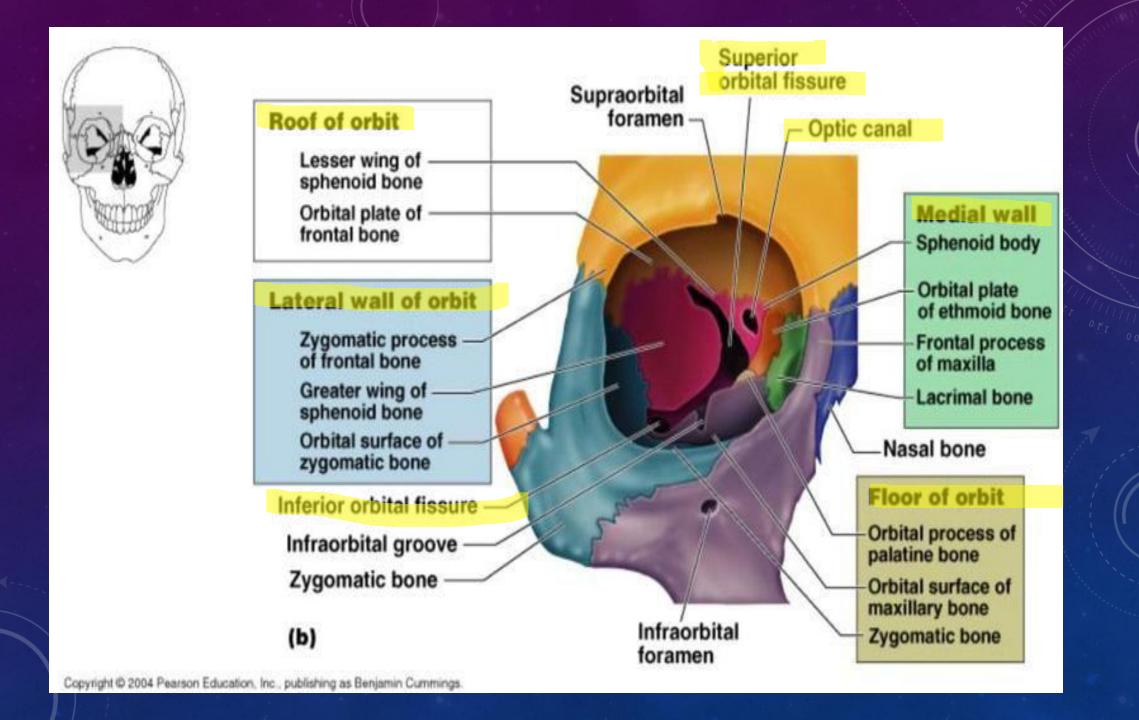
# **DEFINITION**;

# **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 (very small, very crowded area).
- Surrounded by Important nearby structures (Brain, sinuses)

- The orbit consists of **7** bones:
- 1. Frontal bone.
- 2. Ethmoid bone.
- 3. Lacrimal bone.
- 4. Sphenoid bone.
- 5. Maxillary bone.
- 6. Palatine bone.
- 7. Zygomatic bone.



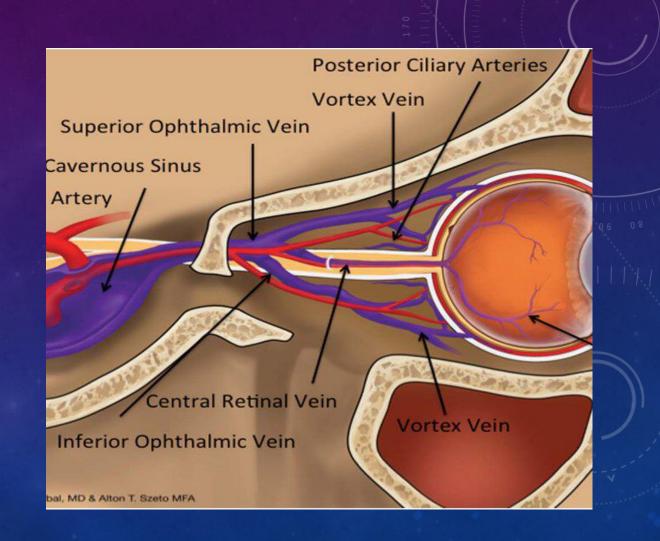


The major nerves and vessels enter the orbit through:

- ✓ Optic canal;
- At the posterior apex of the orbit.
- Transmits optic nerve.
- ✓ The superior and inferior orbital fissures;
- Transmits blood vessels and cranial nerves from the brain to the orbit and face (and from face, sensations and venous drainage back to the brain).

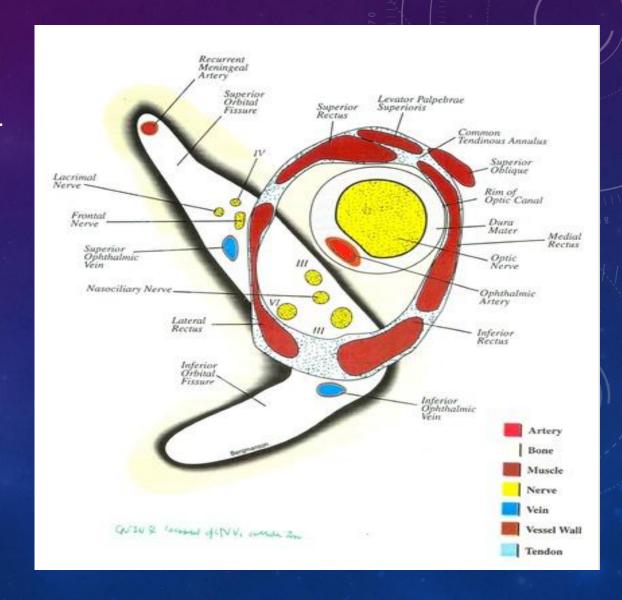
#### The optic canal:

- > 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.



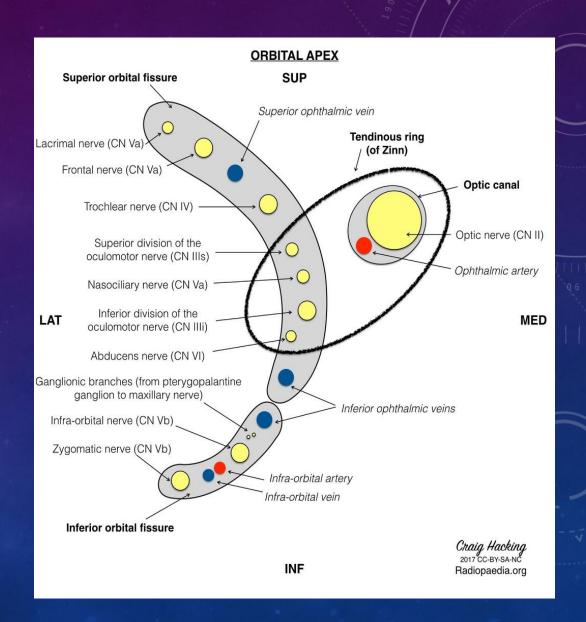
## • The Superior orbital fissure:

- 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.



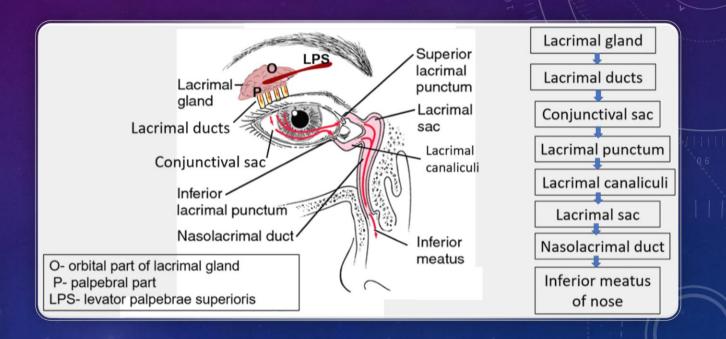
## • The Inferior orbital fissure:

- 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.



## •The lacrimal gland:

- •Lies anteriorly in the superolateral aspect of the orbit.
- •Lacrimal sac lies On the anterior part of the medial wall.

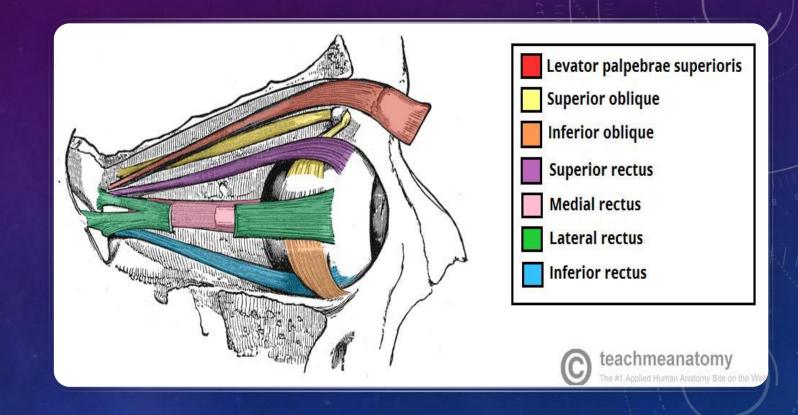


## • Functions of the Orbit?

- Protection of the globe.
- Provides attachment of the extraocular 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,

- <u>Proptosis (exophthalmos)</u> is bulging of the eye anteriorly by a space-occupying lesion (forward protrusion of the globe).
- It can be <u>bilateral</u> (Graves' disease) or <u>unilateral</u> (orbital tumors, infections).



# CAUSES OF PROPTOSIS

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



#### • 1. *Infectious*;

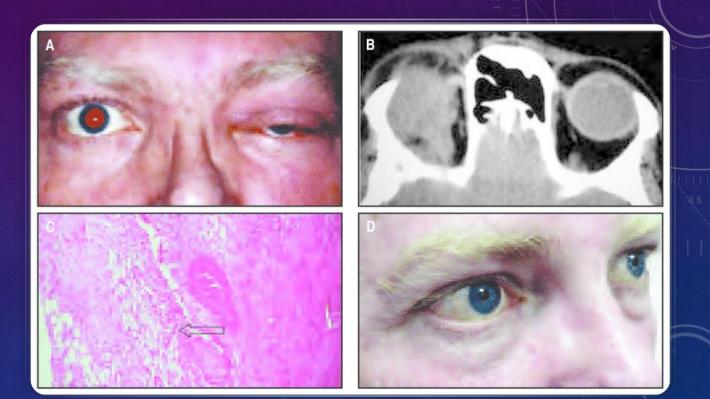
- Orbital cellulitis (Most common cause for unilateral Proptosis in children).
- Mucormycosis (Fungal infection).
- Proptosis is off Rapid onset & painful.

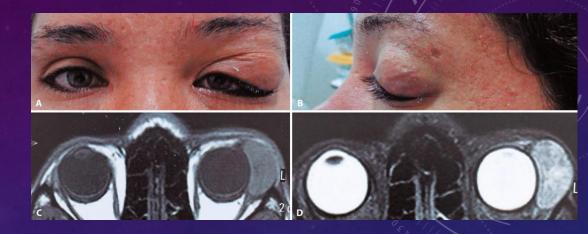


Figure 2 – This extremely irritable infant with red, swollen eyelids and a fever was found to have orbital cellulitis based on the findings of a CT scan of the orbits, which revealed inflammatory changes in the postseptal area.

#### • 2. Inflammatory;

- Thyroidopathy (M.C.C for proptosis in adults is Graves' disease).
- Orbital inflammatory syndrome (orbital pseudomotor, benign orbital inflammation, sarcoidosis).





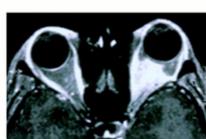
#### • 3. **Neoplastic:**

- Lacrimal gland tumors. (Eye displaced to one side; lesion outside muscle cone {Extra-conal})
- > Lymphoma.
- > Leukemia.
- Meningioma. (proptosis is forward in direction; {intra-conal lesion})
- Optic nerve Glioma.
- Ossifying fibroma.
- Metastatic. (breast in women, lung and prostate in men, GI, kidney)
- \*\*in children, rapidly developing Proptosis; rhabdomyosarcoma.









#### 4. Vasculitis;

Wegner granulomatosis.

➤ Churg-Strauss syndrome. (Eosinophilic granulomatosis with polyangiitis (EGPA)) is a very rare autoimmune condition characterized by necrotizing vasculitis of small- and medium-sized systemic blood vessels.





Orbital varix (venous malformation)

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

- Orbital arteriovenous malformation
- Carotid-cavernous sinus fistula



- > Traumatic/ iatrogenic orbital hemorrhage proptosis in acute phase.
- Orbital fractures.
- > Facial fractures.

## • 7. Pseudoproptosis;

- Buphthalmos.
- > Contralateral enophthalmos.
- > Ipsilateral lid retraction.
- > Axial myopia.
- > Contralateral blepharoptosis.





# The orbit

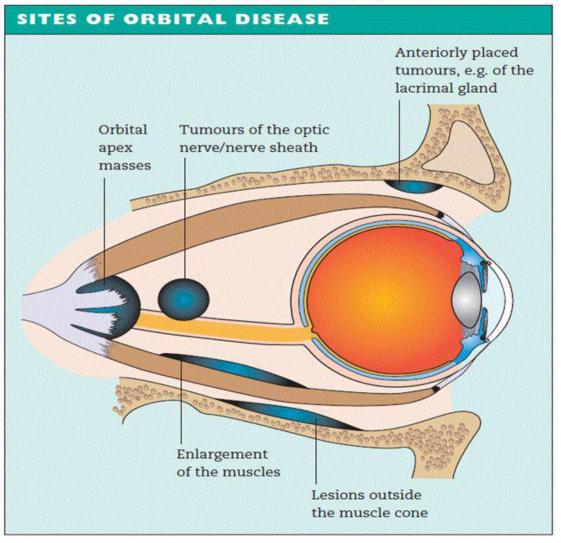
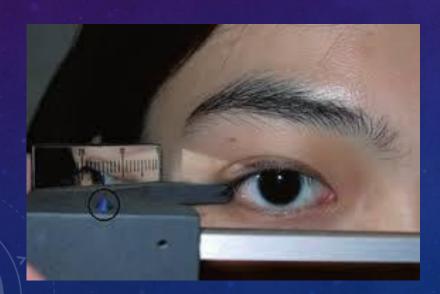
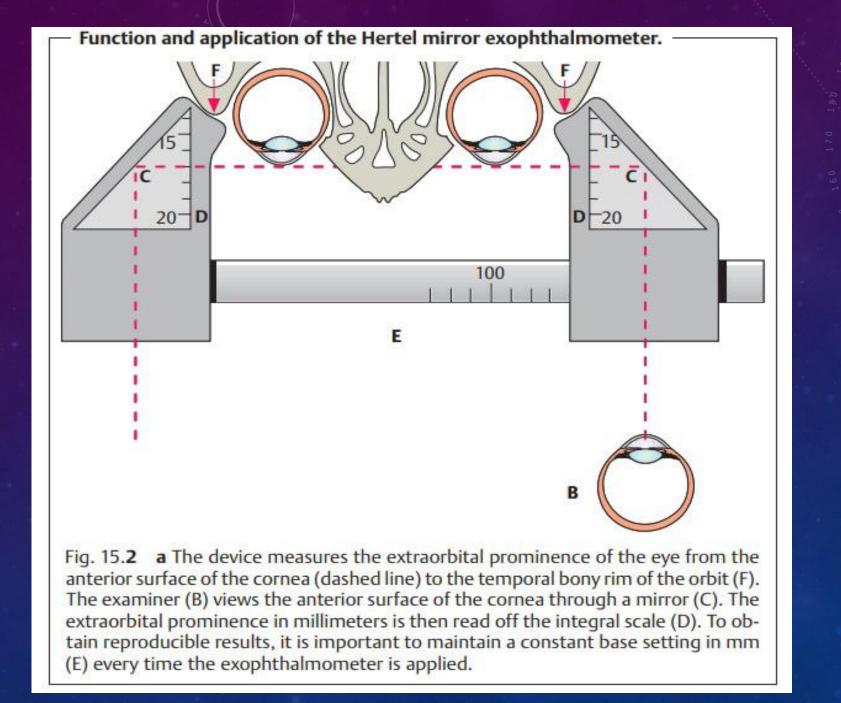


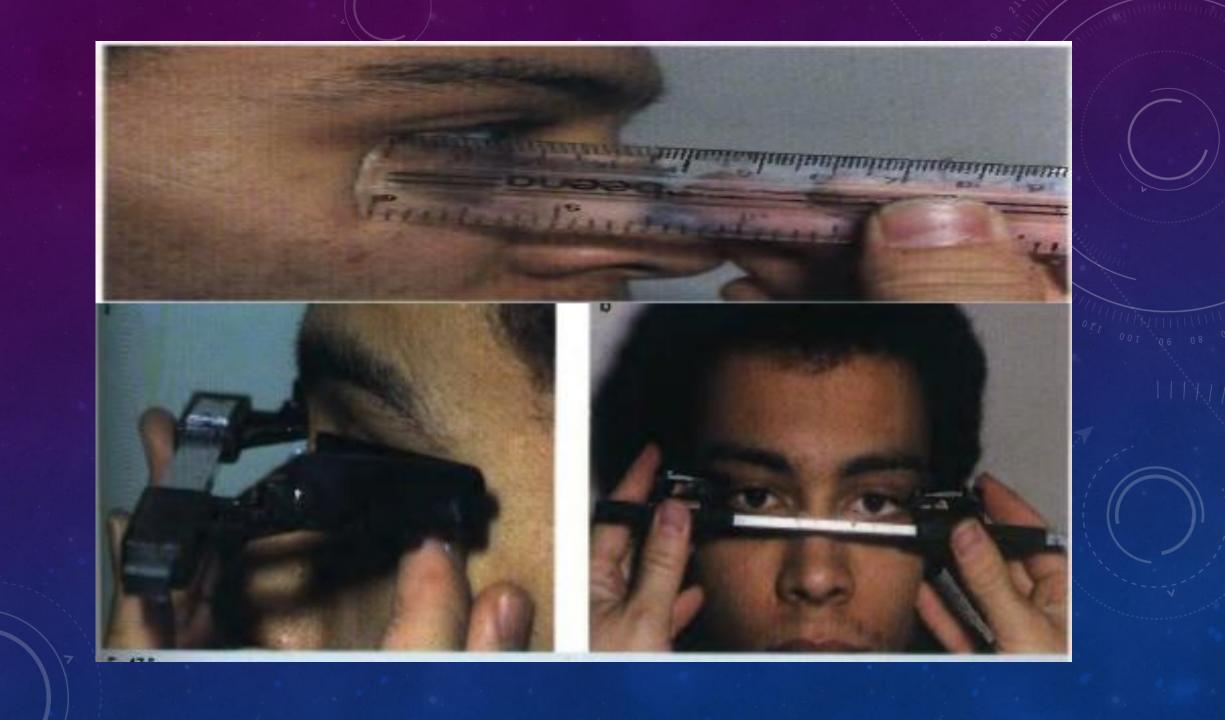
Fig. 4.1 Sites of orbital disease.

- How do we measureProptosis?
- ✓ it can be measured using Hertel exophthalmometer.









# HERTEL EXOPHTHALMOMETER

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.



# THYROID EYE DISEASE (TED)

- The commonest cause of unilateral and bilateral Proptosis in adults and bilateral proptosis in children.
- 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 orbital bones, as swelling continues the eyeball will be pushed forward.
- Particularly in hyperthyroidism (90%), but can happen in hypothyroidism and euthyroidism.



✓ What's the most commonly affected muscle?

The inferior rectus muscle.



# Signs & symptoms of Graves' Ophthalmopathy:

- ✓ Red, painful eye.
- ✓ Periorbital edema.
- ✓ Lid retraction (*sclera visible above or below cornea*)
- ✓ Inflammatory Conjunctivitis.
- ✓ Diplopia.
- ✓ Proptosis.
- ✓ Reduced visual acuity.



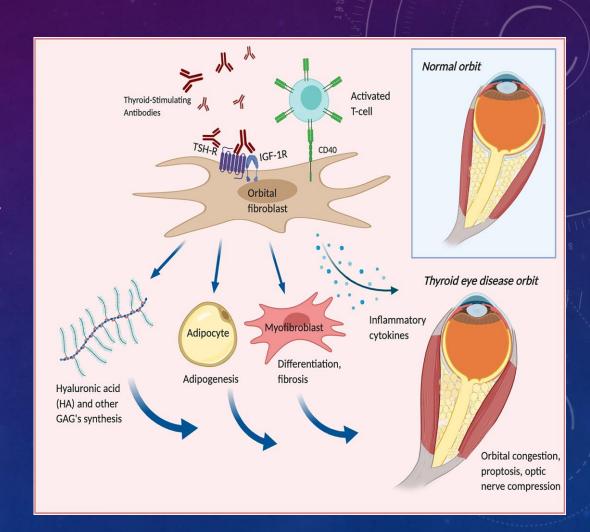
## On physical exam;

- ✓ Proptosis (asymmetrical).
- ✓ Chemosis in conjunctiva.
- ✓ Lid retraction (staring appearance due to Increased sympathetic activity).
- ✓ Lid lag (lid don't follow globe movement on downgaze ).
- √ +/- restricted eye movement (enlarged inferior rectus).



#### 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.



- The initial activation of T cells in Graves' ophthalmopathy is now thought to be initiated by thyrotropin (TSH) receptor antigen. Mechanistic observations, plus the fact that Graves' hyperthyroidism and Graves' ophthalmopathy often occur concomitantly, have led to the suggestion that activation of the immune system must be secondary to an antigen shared by both tissues.
- TSH receptor (TSHR) mRNA and protein can be detected in orbital fibroblasts and adipocytes, and pre-adipocytes from patients with Graves' ophthalmopathy express more TSH receptor mRNA and produce more cyclic adenosine monophosphate (AMP) in response to TSH than do similar cells from normal subjects



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. Exposure Keratopathy: Excessive exposure of the conjunctiva and cornea with chemosis and corneal ulcer; corneal perforation.
- 2. Compressive optic neuropathy: Compression and ischemia of the optic nerve by the thickened muscle leading to compressive optic neuropathy (visual field loss, blindness).
- Treatment of urgent complications?
- 1. Systemic steroids.
- 2. Surgical orbital decompression.

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	No physical signs or symptoms
1	Only signs, no symptoms (eg, upper lid retraction,
	stare, and eyelid lag)
2	Soft tissue involvement (symptoms and signs)
3	Proptosis
4	Extraocular muscle involvement
5	Corneal involvement
6	Sight loss (optic nerve involvement)

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#### **Treatment aims?**

- 1- Protection of the globe
- 2- Improve eye movement problems.
- 3- Improve the cosmetic appearance.
- <u>How?</u>
- 1- Lubrication of the eye.
- 2- Prisms to correct Diplopia.
- 3- Corrective surgeries (orbital decompression, extraocular muscles and upper eyelid).



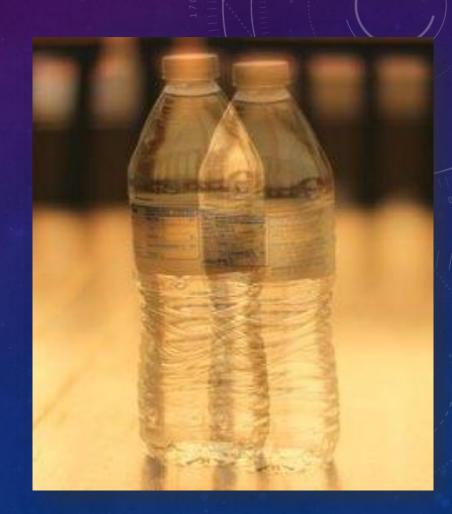
#### Some Causes of Proptosis

Cause	Suggestive Findings	Diagnostic Approach
Graves disease	Eye symptoms: Eye pain, lacrimation, dry eyes, irritation, photophobia, ocular muscle weakness causing diplopia, vision loss caused by optic nerve compression Systemic symptoms: Palpitations, anxiety, increased appetite, weight loss, insomnia, goiter, pretibial myxedema (see <a href="https://example.com/hyperthyroidism"><u>Hyperthyroidism</u>: Symptoms and Signs</a> )	Thyroid function tests
Carotid-cavernous sinus or dural- cavernous sinus fistula	Pulsating proptosis with an orbital bruit	Magnetic resonance angiography
Cavernous sinus thrombosis	Ophthalmoplegia, headache, ptosis, decreased visual acuity, fever	CT or MRI
Congenital glaucoma and unilateral high myopia	Tearing, blepharospasm, redness	Intraocular pressure measurement and funduscopy by ophthalmologist
Orbital cellulitis	Redness, fever, pain, impaired visual acuity, impaired or painful extraocular movements  Usually unilateral	CT or MRI
	obdany dimateral	
Orbital tumors (eg, lymphoma, hemangioma, vascular malformations)	Decreased visual acuity, diplopia, pain	MRI or CT
Retrobulbar hemorrhage	Decreased visual acuity, diplopia, pain, ophthalmoplegia, risk factors	Immediate CT or treatment based on clinical findings
Spheno-orbital meningioma	Pain, headache, visual field defects, ophthalmoplegia	MRI or CT

# 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.



#### **CAUSES**

1. Extraocular muscles involvement as in myositis and dysthyroid diseases.

2. Nerve supply involvement.

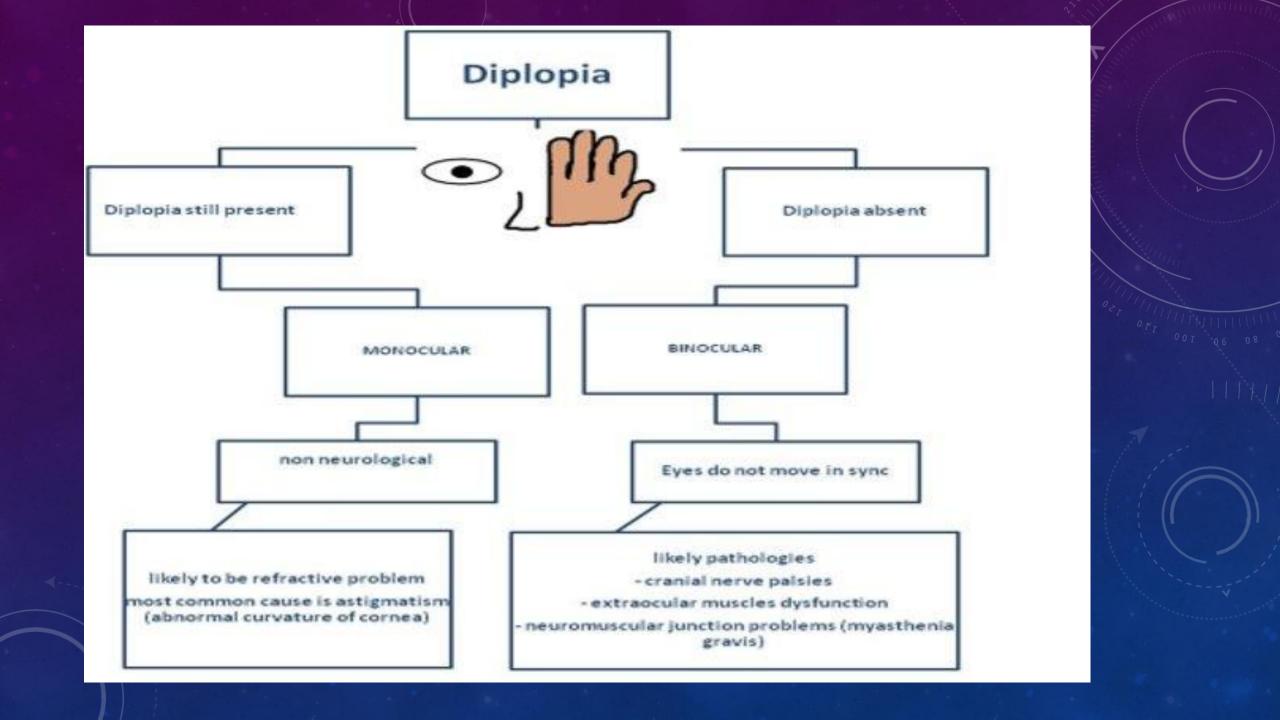
3. Neuro-Muscular junction (N-M junction).

#### **TYPES**

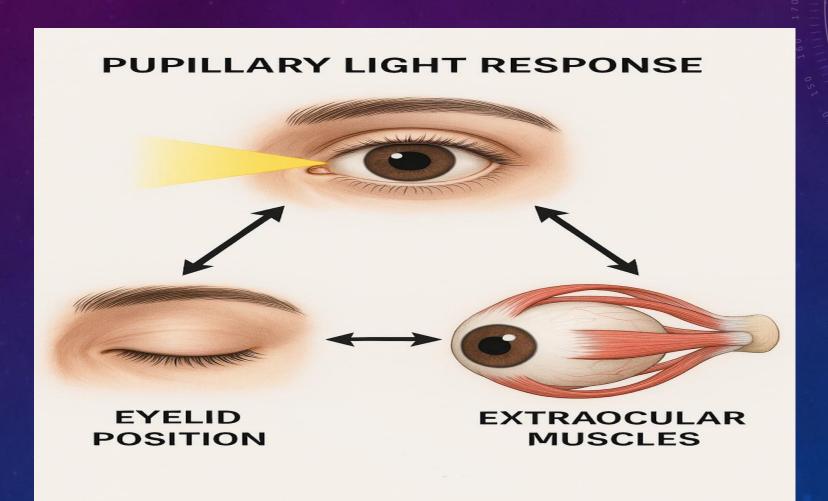
 monocular diplopia (one eye is open) is usually the result of a refractive error

2. binocular diplopia (Both eyes are open) is commonly due to impaired extraocular muscle function, cranial nerve, or N-M junction

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



## THE OCULAR TRIANGLE

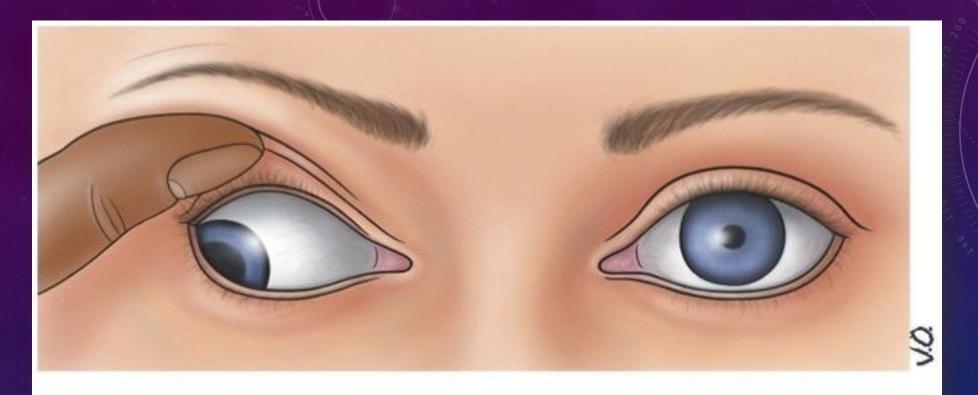


#### HISTORY TAKING IN DIPLOPIA

#### 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 of diplopia is more indicative of a compressive or inflammatory lesion.

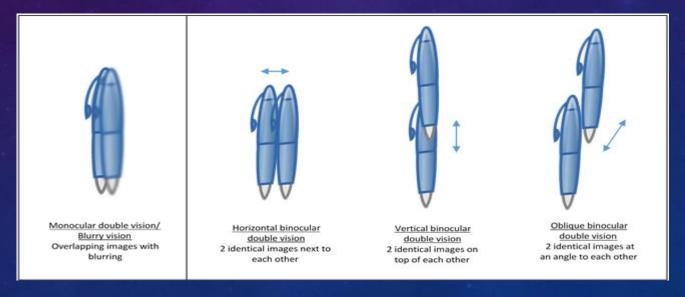


Right eye: Downward and outward gaze, dilated pupil, eyelid manually elevated due to ptosis

Left: Normal

#### 3. Image alignment:

- ◆ Is the diplopia horizontal, vertical or oblique?
- ◆Horizontal diplopia 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), or orbital floor fractures.



#### 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

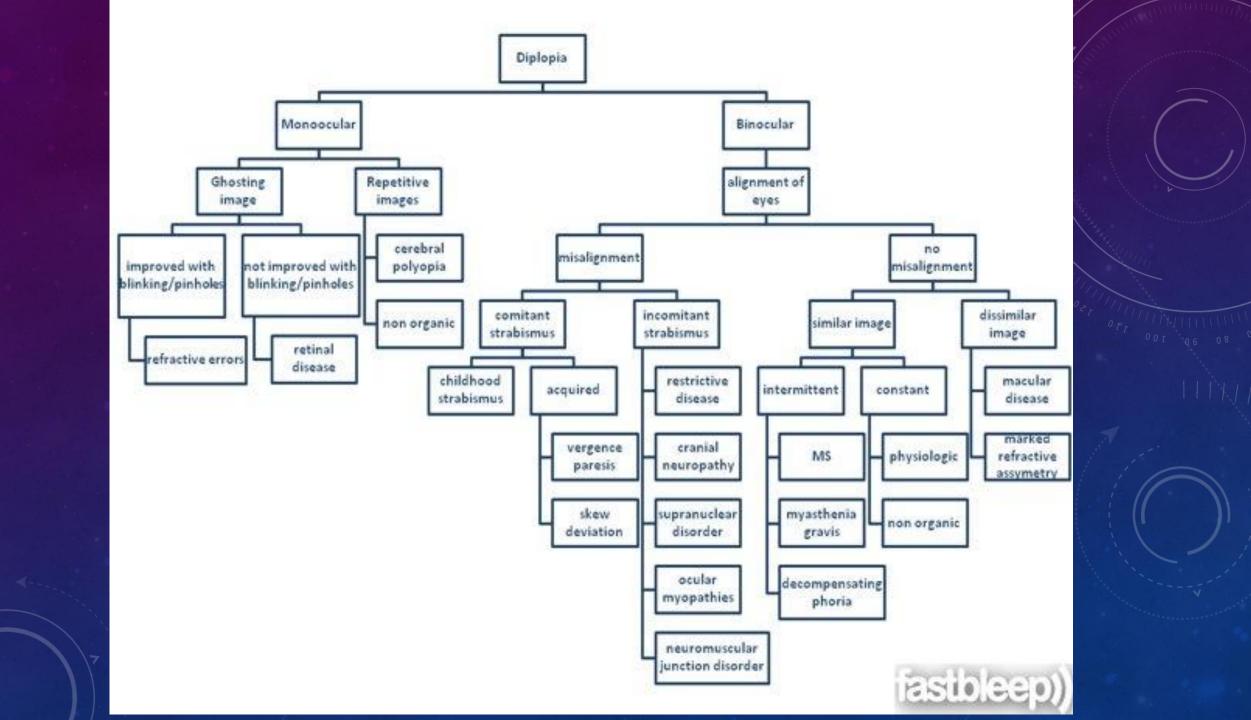
#### 5. Direction of gaze

- 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 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.

• 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 (multiple sclerosis or myasthenia).



# DIFFERENTIAL DIAGNOSIS OF ORBITAL DISEASES

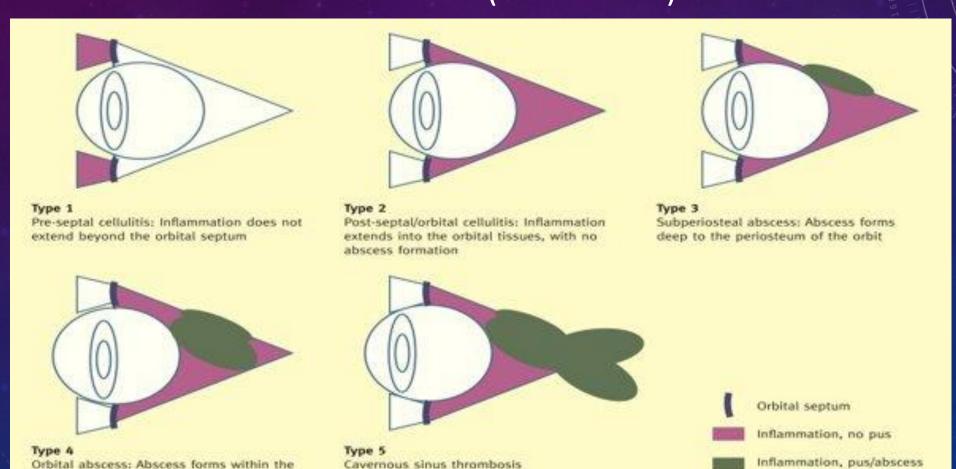
#### **DIFFERENTIAL DIAGNOSIS OF ORBITAL DISEASE**

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

#### INFECTIVE DISORDERS

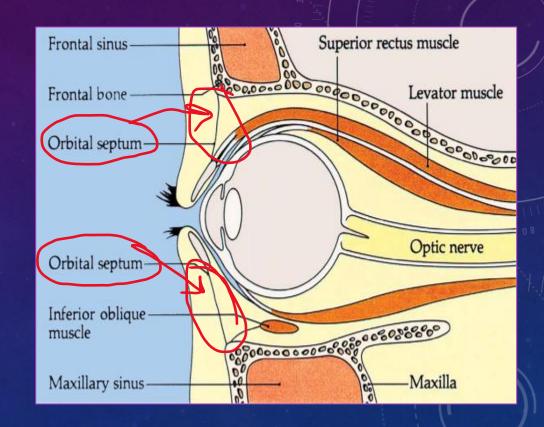
orbit, with breach of the periosteum

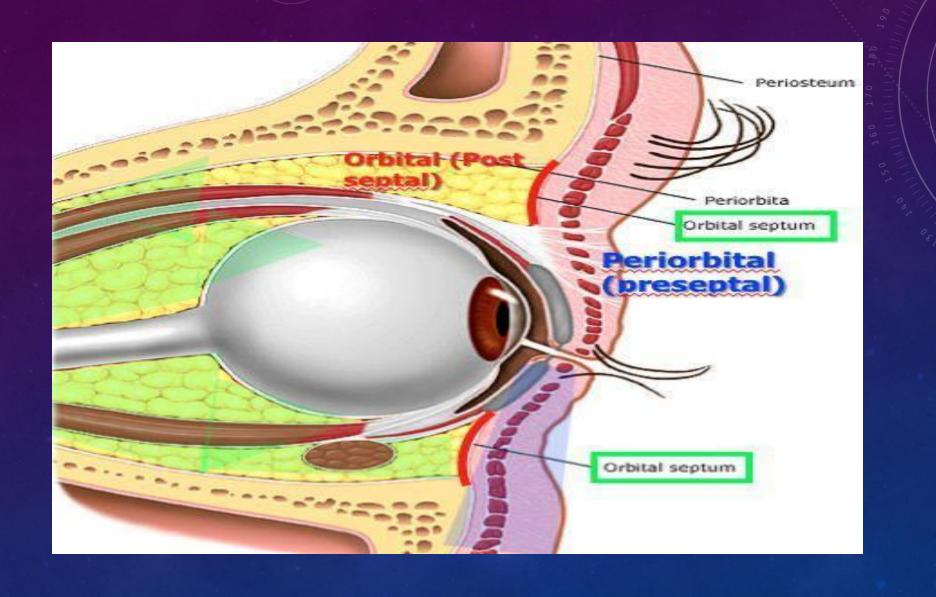
• Classification of orbital infections (Chandler's ):



#### 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 .





• Treatment

topical antibiotic.



• if severe or child < 1 year, 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

- -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).
- decreased visual acuity ( visual loss is possible!) with ± RAPD ( Relative afferent pupillary defect).

## ORBITAL CELLULITIS





#### Diagnosis:

MRI or CT

#### **◆**Treatment:

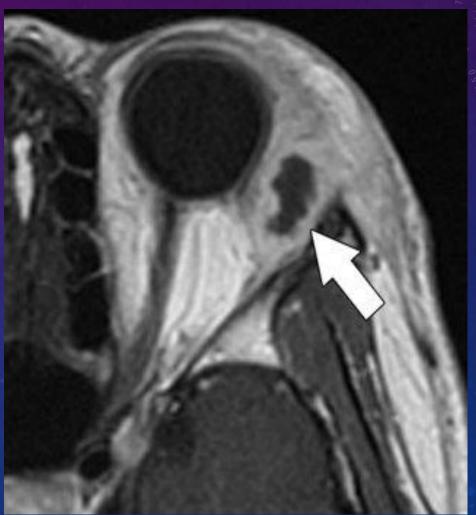
- Admit the patient, blood cultures, orbital CT, IV antibiotics (ceftriaxone + vancomycin) for 1 week.
- 2. Surgical drainage of abscess with close follow-up, especially in children.
- 3. Optic nerve decompression if it's compromised. (Endoscopic optic nerve decompression is a minimally invasive procedure 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

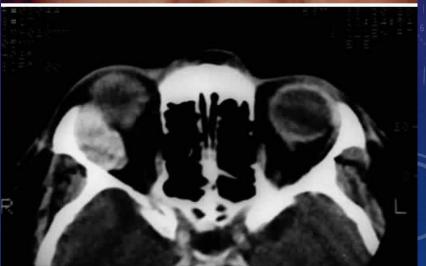
 It can be <u>primary</u> neoplasm arising from any of the anatomical structures of the orbit.

 Or <u>secondary</u> orbital invasion from direct extension from contiguous anatomical structures, lymphoproliferative disorders, and hematogenous metastasis.

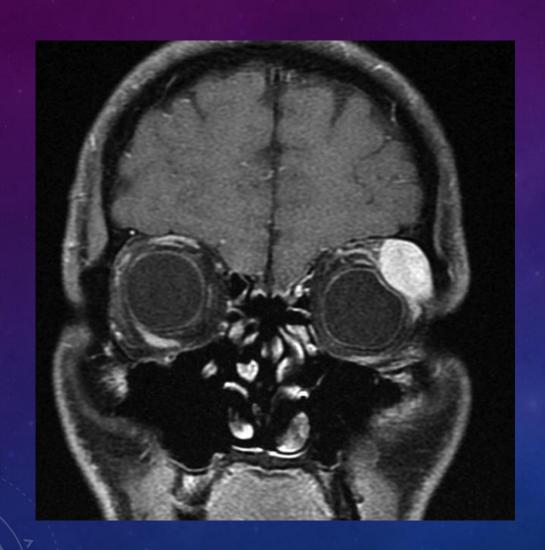
#### Lacrimal gland tumors:

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





## **Lacrimal gland tumors:**



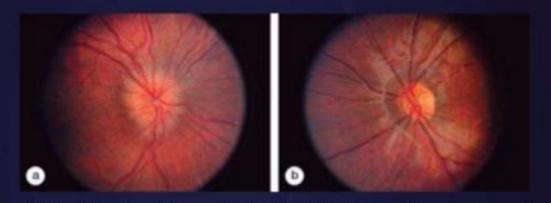
#### Optic nerve gliomas:

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

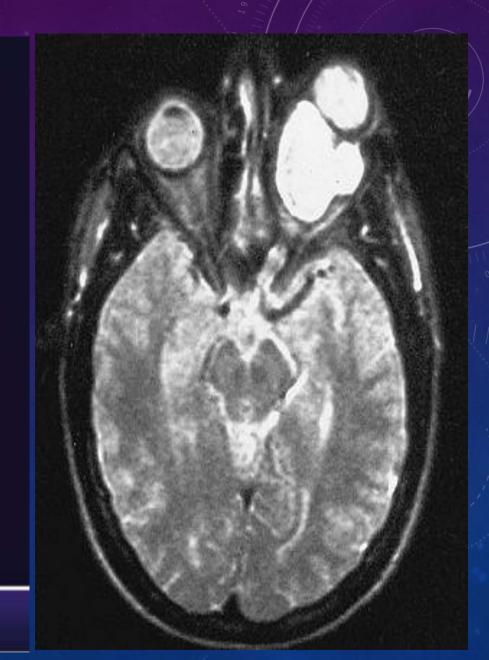
## Optic nerve gliomas:



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.



## Optic nerve sheath Meningiomas

Meningiomas of the optic nerve sheath are <u>rare</u>, and may also be <u>difficult to excise</u>.

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

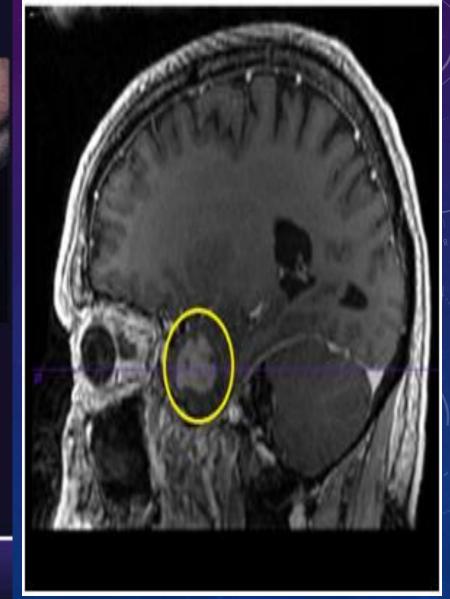
Meningiomas arising from the middle cranial fossa, however, may spread through the optic canal into the orbit.

#### Meningiomas





Face shot of a woman complaining of proptosis of the left eye. Neuroimaging 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.

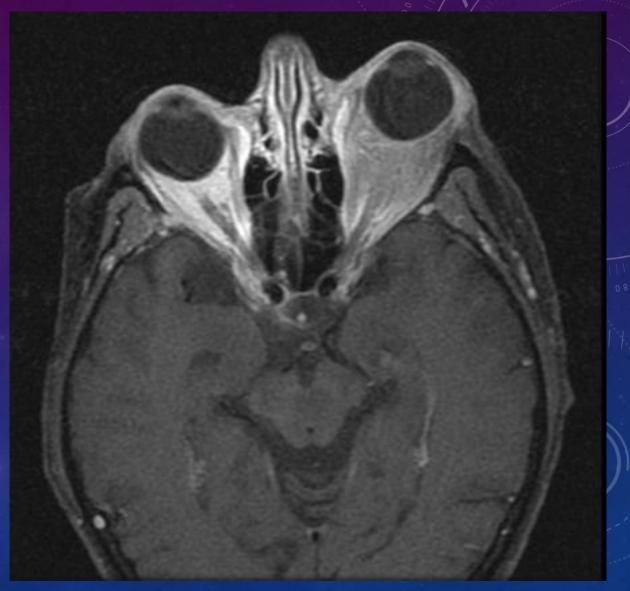


## Lymphomas

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 6-9 years old (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:



Metastasis from other systemic cancers

#### \*Children:

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

#### \*Adults:

Breast, lung, prostate, or GI tract

- Metastatic tumors reach the orbit by hematogenous spread, since the orbit is devoid of lymphatics.
- Metastasis is usually from the breast in women and from the lung in men.
- Proptosis vs. enophthalmos.
- In children, the most common metastatic tumor is Neuroblastoma, which is often associated with spontaneous periocular hemorrhage as the rapidly growing tumor becomes necrotic.
- Many metastatic orbital tumors respond to radiation, some to chemotherapy.
- Small localized tumors that are symptomatic can sometimes be completely or partially excised.
- Neuroblastomas' in children under 11 months have a relatively good prognosis.
- Adults with metastatic tumors in the orbit have a very limited life expectancy.

#### **DERMOID CYSTS**

- These congenital lesions are <u>caused by the continued</u> 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 <u>cosmetic</u> reasons and to <u>avoid traumatic rupture</u>, 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.



# THANK YOU!