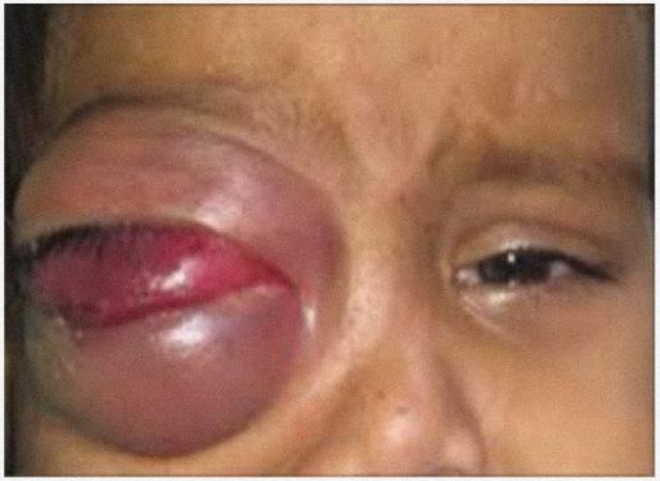
# PROPTOSIS IN CHILDREN



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**FRCS, MRCOphth**
**Fellowship Orbit/Oculoplastics**
**Fellowship Refractive surgery**

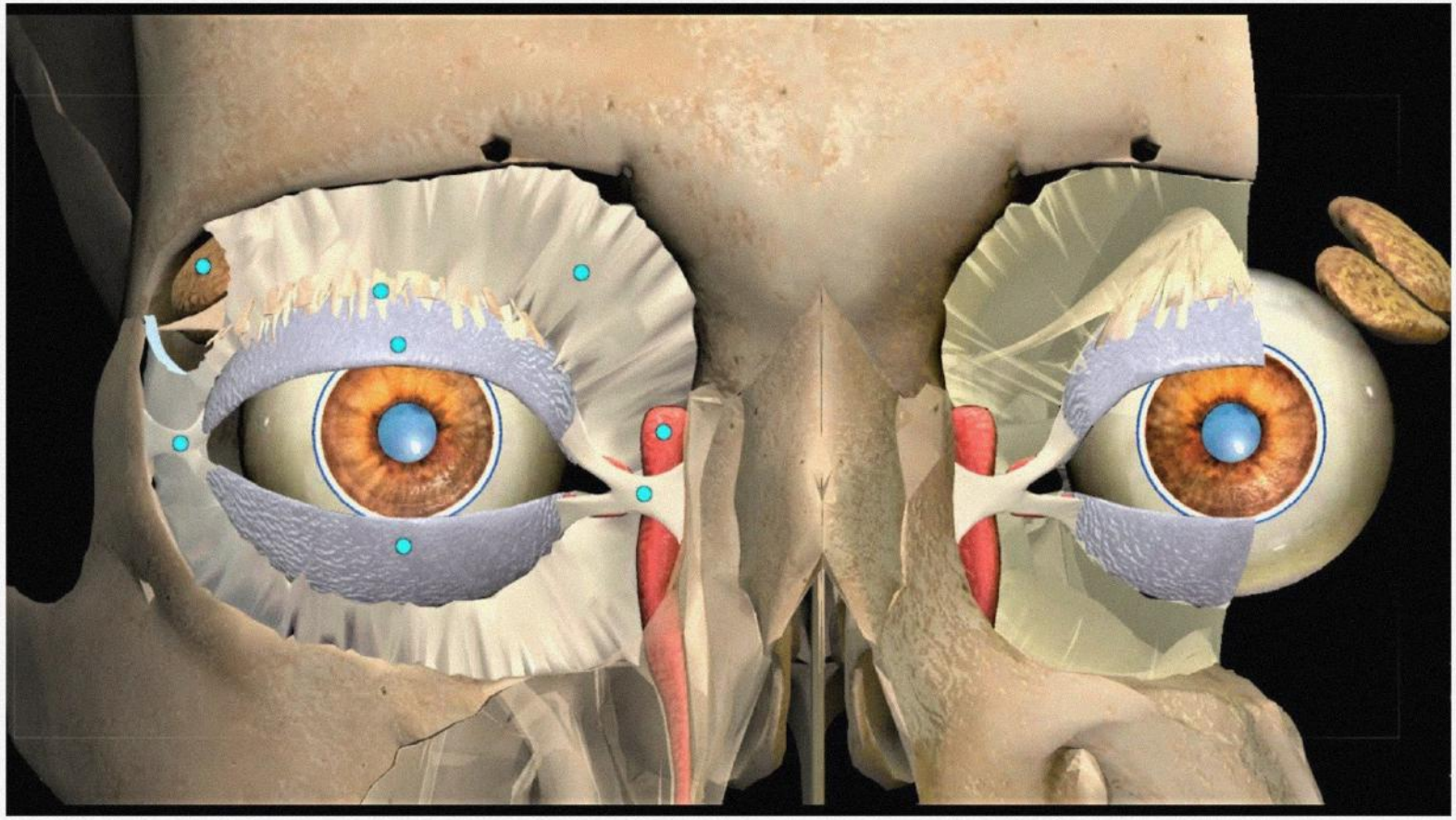
# CAUSES

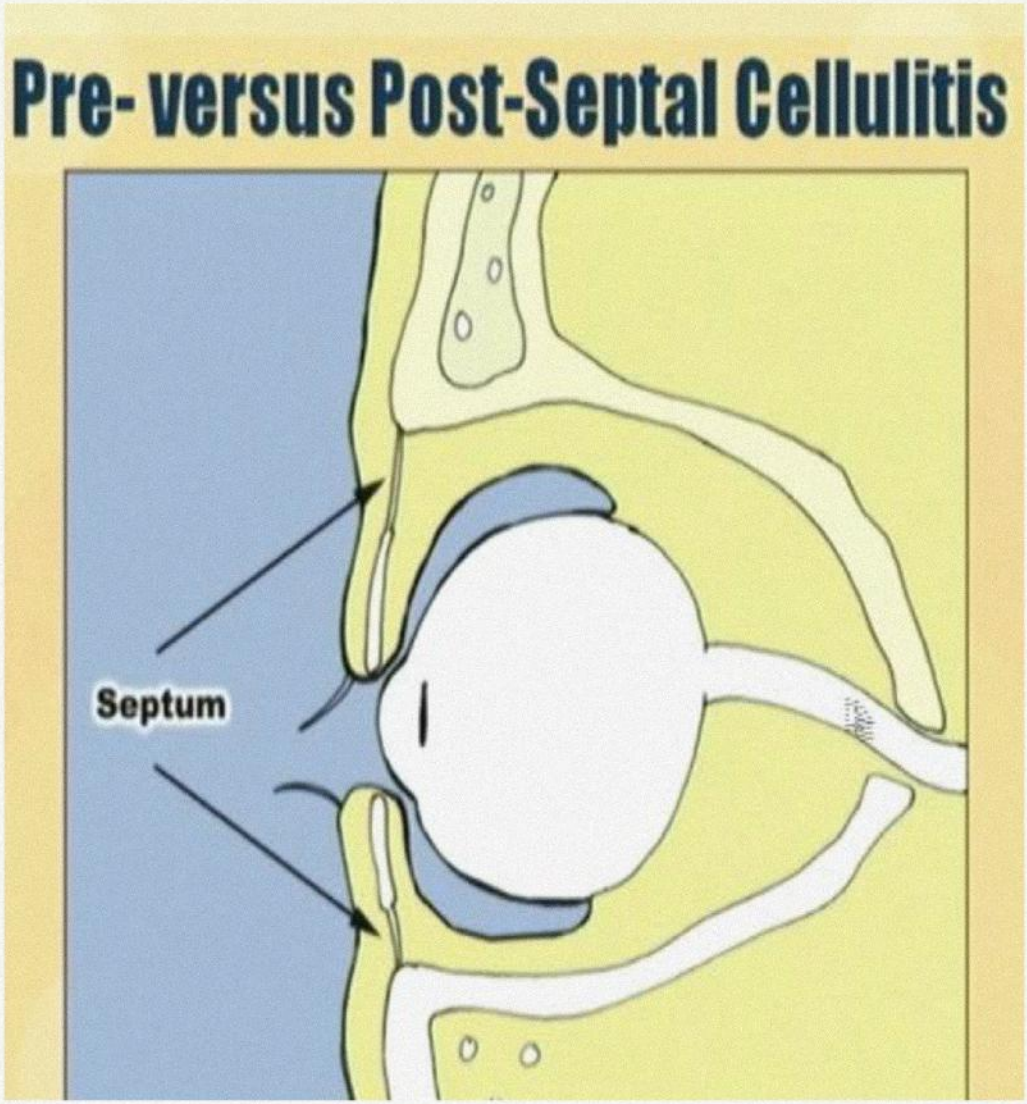
- Orbital cellulitis
- Dermoid cyst
- Capillary hemangioma
- Optic nerve glioma
- Rhabdomyosarcoma
- Retinoblastoma
- Leukemia
- Lymphangioma
- Metastasis
  - Metastatic neuroblastoma
  - Ewing's sarcoma

| BIRTH-4YEARS | 4-10 YEARS | OVER 10 YEARS |
|---|---|---|
| Orbital cellulitis | Lyphangioma | Orbital cellulitis |
| Dermoid cyst | Rhabdomyosarcoma | NSOI |
| Capillary haemangioma | Leukemia | Thyroid ophthalmopathy |
| Lymphangioma | Optic nerve glioma | Dermoid cyst |
| Structural abnormality | Langerhan cell histiocytosis | Leukemia |
| Metastatic (neuroblastoma) | NSOI | Fibrous dysplasia |
| Rhabdomyosarcoma | Orbital cellulitis | |
| Langerhan cell histiocytosis | | |
| Teratoma | | |
| Optic nerve glioma | | |

# Orbital Septum

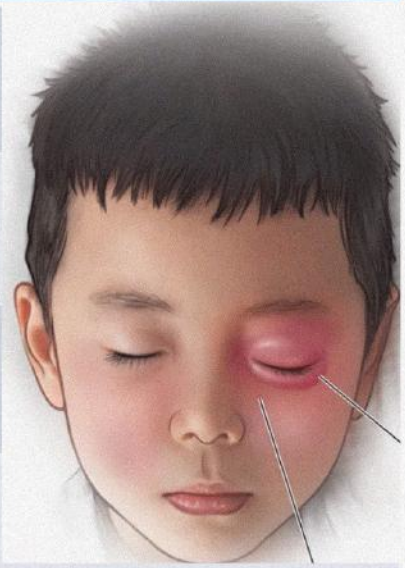
- *The orbit is separated from the soft tissue of the eyelid by the orbital septum*
- *A facial plane that is continuous with the periosteum of the facial bones*
- *It inserts into the tarsal plate of the upper and lower eyelids*
- *It is a barrier which prevents the spread of infection from the eyelids posteriorly to the orbit*

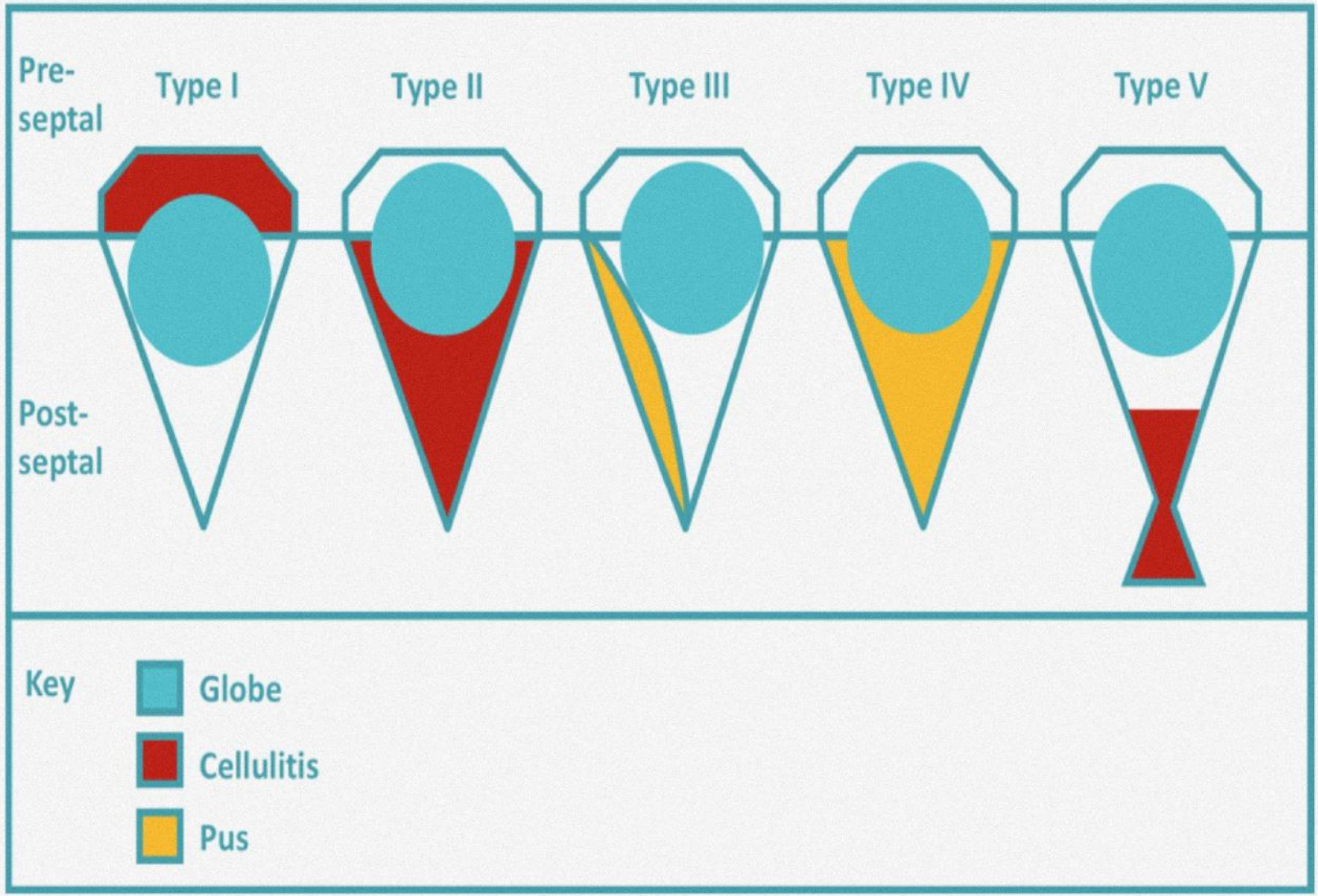


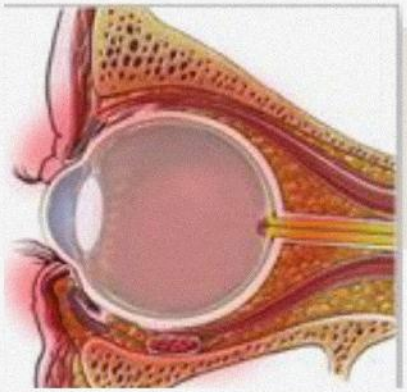


# CHANDLER CLASSIFICATION

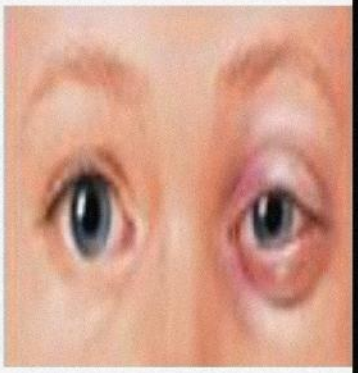
| | |
|---|---|
| GROUP 1 | Preseptal cellulitis |
| GROUP 2 | Orbital cellulitis |
| GROUP 3 | Sub periosteal abscess |
| GROUP 4 | Orbital abscess |
| GROUP 5 | Cavernous sinus thrombosis |




Pre-
septal
Post-
septal
Type I
Type II
Type III
Type IV
Type V
Key
Globe
Cellulitis
Pus



# PRE-SEPTAL CELLULITIS

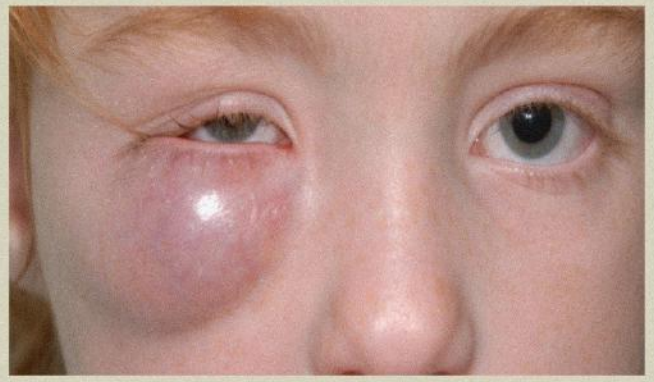


- Infective /inflammatory process anterior to the orbital septum
- Eyelid is swollen and can be non tender
- The VA and EOM are normal and there is no chemosis

## ETIOLOGY

- Bacteria (staph aureus,strep pneumo,anaerobes)
- Local spread from an adjacent sinusitis or dacryocystitis, from an external ocular infection, or following trauma to the eyelid

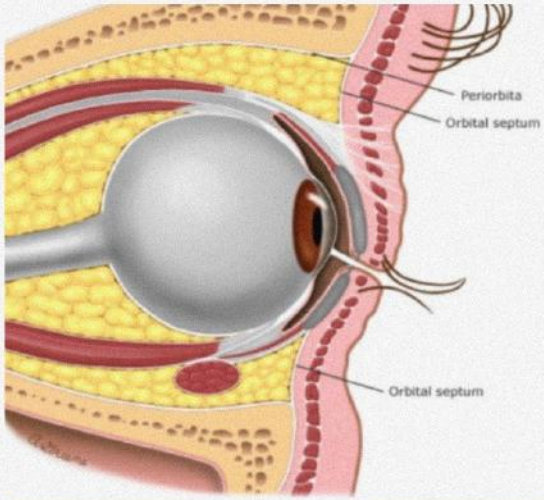
# Management



- **Antibiotics**

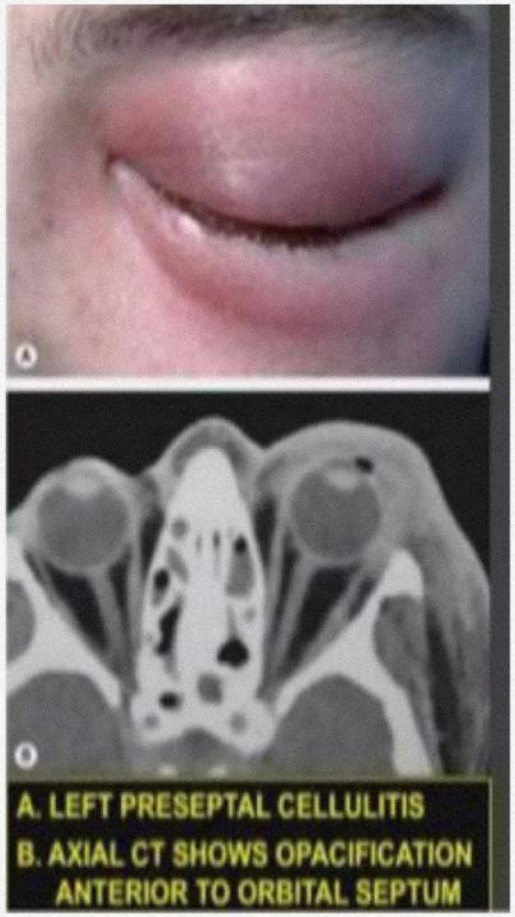
  Amoxicillinclavulanate,Cefuroxime,Gatifloxacin,Moxifloxacin, Levofloxacin
- **Pain killers**
- **Supportive therapy**

**Infection can spread as Orbital cellulitis which is vision and life threatening**

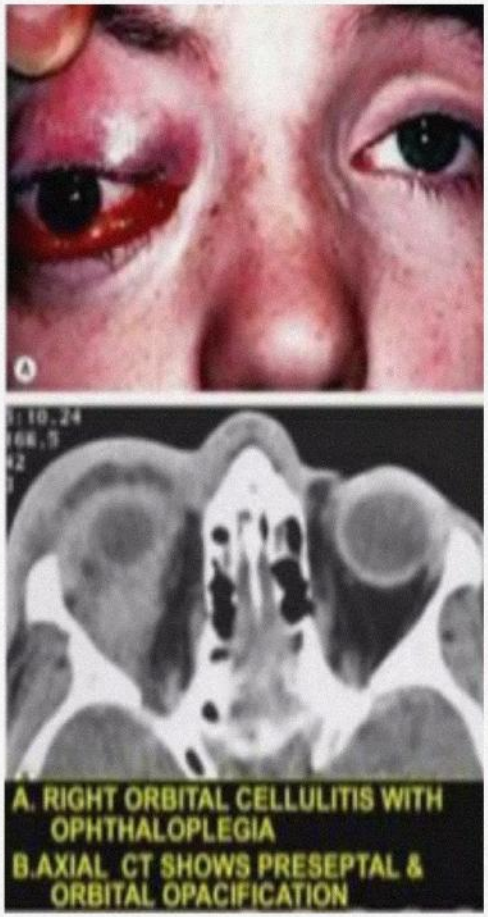



| Preseptal Cellulitis | Orbital Cellulitis |
|---|---|
| ✅ Unlikely to have complications | ❌ Can progress to loss of vision, brain abscess |
| Due to external source | Associated with paranasal sinusitis |
| Staph aureus<br>Strep pneumo + other Strep<br>Anaerobes | Same micro as preseptal cellulitis plus fungal, mycobacterial |

| Clinical Feature | Preseptal Cellulitis | Orbital Cellulitis |
|---|---|---|
| Eyelid swelling with or without erythema | Yes | Yes |
| Eye pain, tenderness | +/- | Yes, can have deep eye pain |
| Pain with EOM | No | Yes |
| Proptosis | No | Usually, can be subtle |
| Ophthalmoplegia, diplopia | No | May be present |
| Vision impairment | No | May be present |
| Chemosis | Rare | May be present |
| Fever | May be present | Usually |
| Leukocytosis | May be present | May be present |



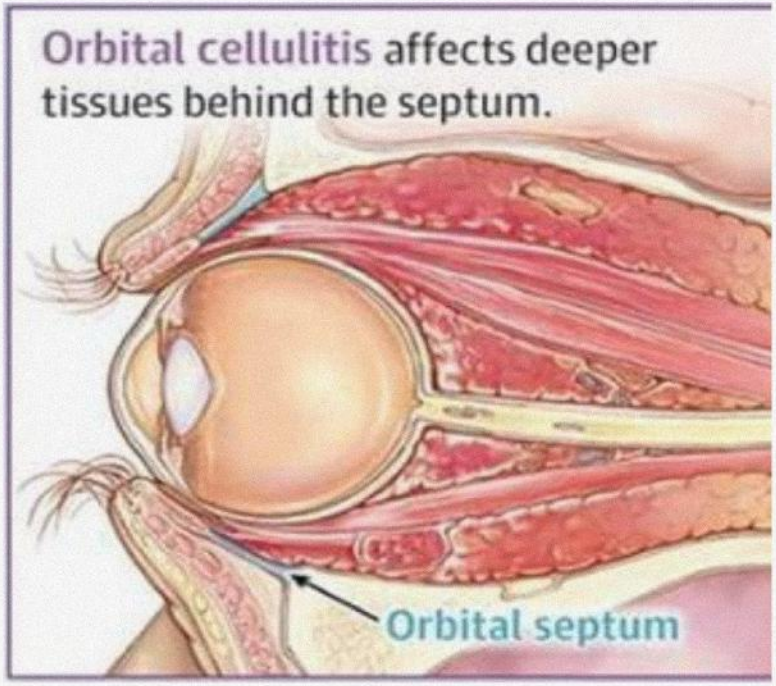
A. LEFT PRESEPTAL CELLULITIS
B. AXIAL CT SHOWS OPACIFICATION ANTERIOR TO ORBITAL SEPTUM



A. RIGHT ORBITAL CELLULITIS WITH OPHTHALOPLEGIA
B.AXIAL CT SHOWS PRESEPTAL & ORBITAL OPACIFICATION

# ORBITAL CELLULITIS

- Infection of the eye tissue behind the orbital septum




# CAUSES OF ORBITAL CELLULITIS

**Extension from periorbital structures**

- Paranasal sinuses (sinusitis)
- Face and eyelids, infection of
- Lacrimal sac (dacryocystitis)
- Dental (odontogenic infection)

**Exogenous causes**

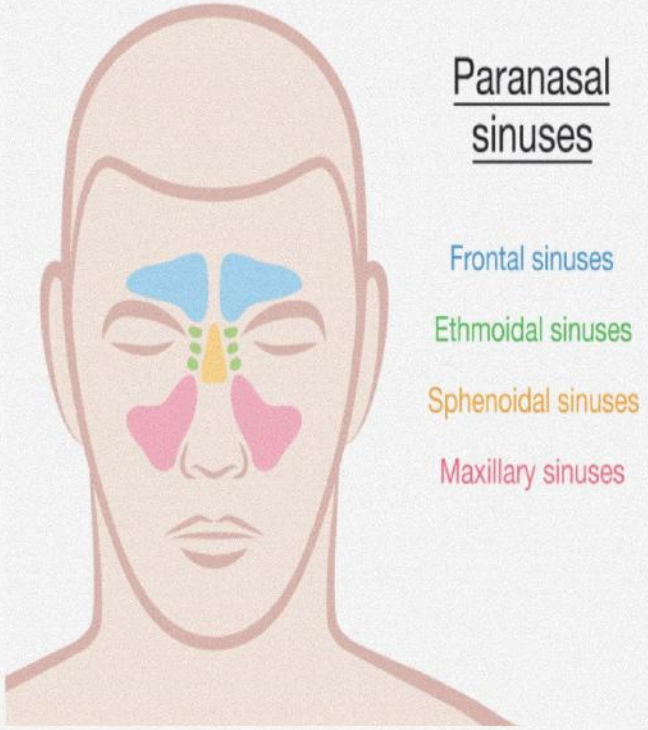
- Trauma (rule out foreign bodies)
- Surgery (after any orbital or periorbital surgery)

**Endogenous causes**

- Bacteremia with septic embolization

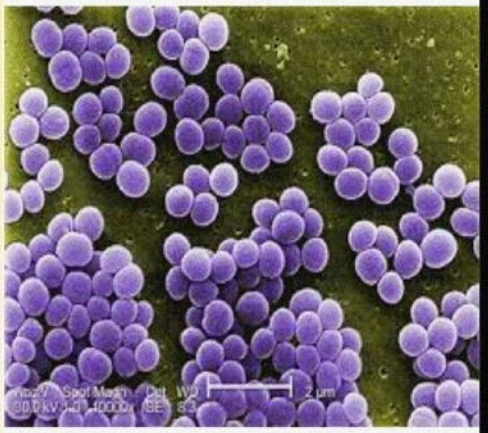
**Intraorbital causes**

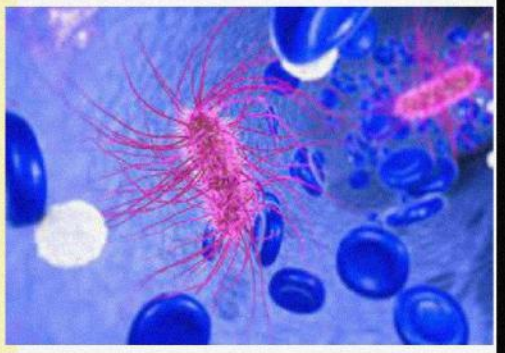
- Endophthalmitis
- Dacryoadenitis



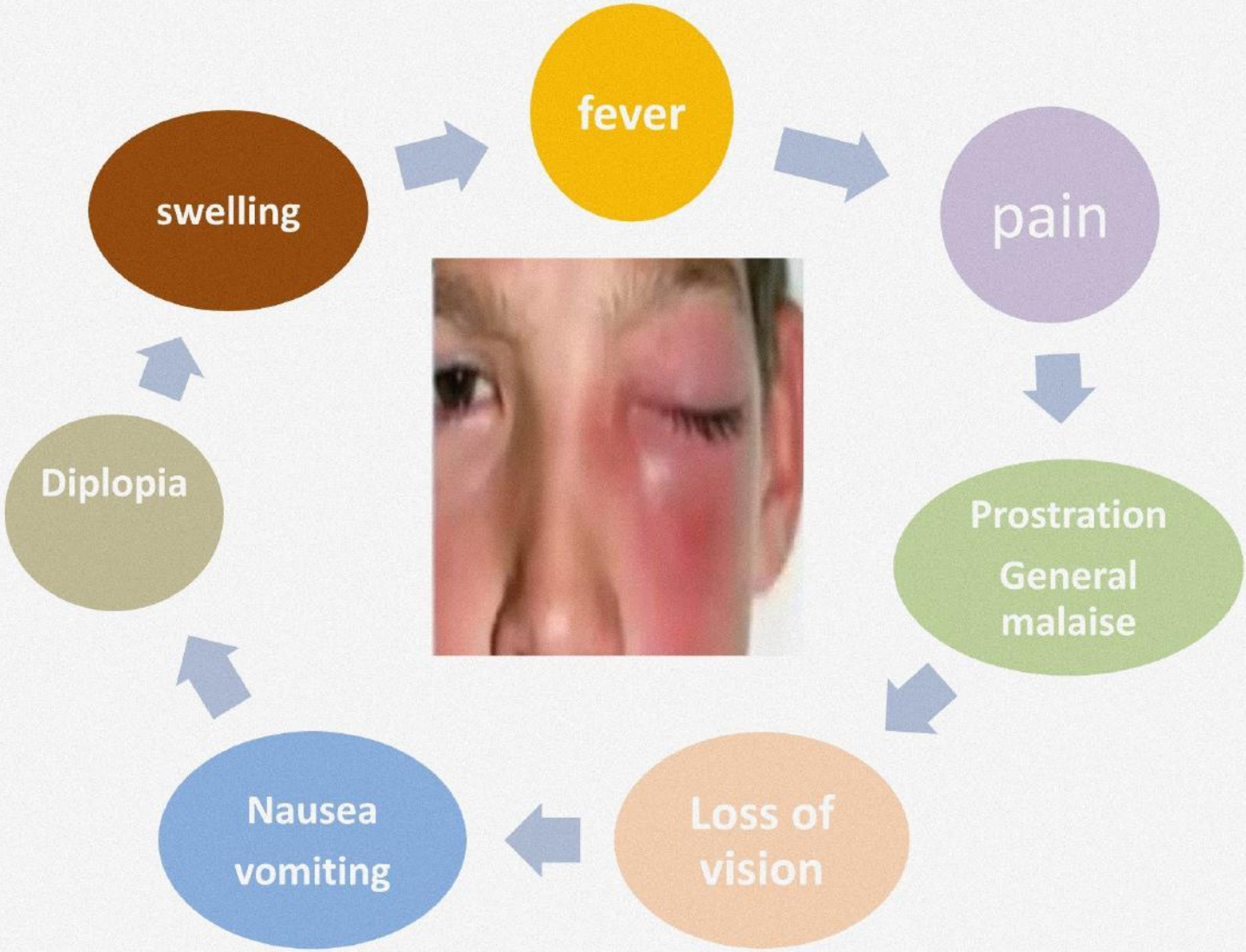
## Bacterial isolates from orbital cellulitis (abscesses), ocular surface or blood

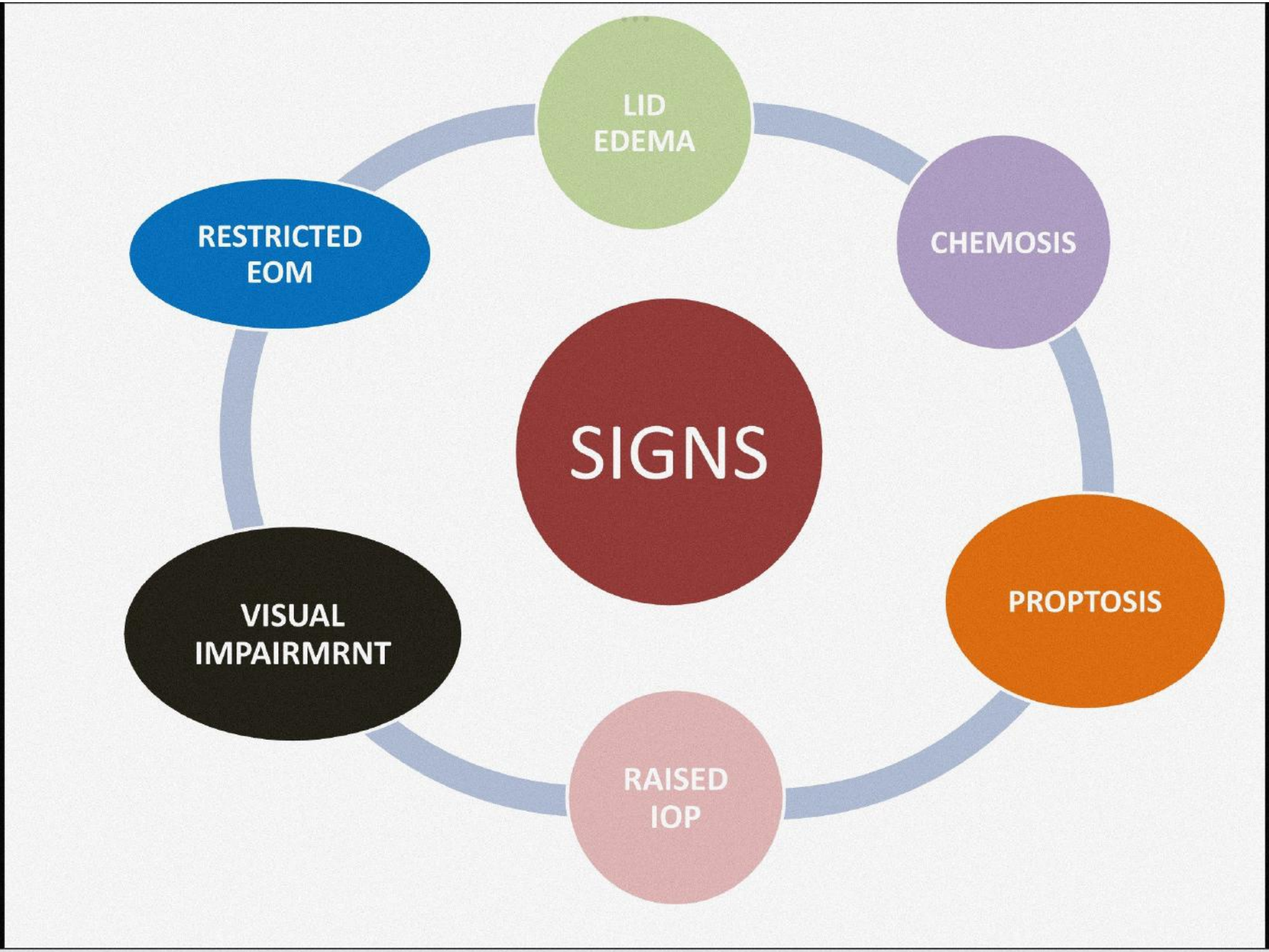
1. *Staphlococcus aureus*, *Staphlococcus epidermidis*
2. *Streptococci pneumoniae*, *Streptococci pyogenes*, *Streptococci sangunis*, *Streptococci fecalis*, *Streptococci mitis*
3. Diphtheroids
4. *Haemophilus influenza*
5. *Escherchia coli*
6. *Moraxella catarrhalis*
7. *Neiserria* sp
8. *Bacillus thuringiensis*
9. *Arcanobacterium haemolyticum*
10. *Pseudomonas aeroginosa*
11. *Pasturella multocida*
12. Atypical Mycobacteria and *Mycobaterium tuberculosis*
13. *Fusobacterium necrophorum*





# SYMPTOMS





## Differential diagnosis of orbital cellulitis in children

(A) Idiopathic nonsupurative inflammation of the orbit
  (i) Nonspecific orbital inflammatory disease
  (ii) Inflammatory thyroid eye disease (rarely seen in children)
  (iii) Wegners granulomatosis
  (iv) Sarcoid related inflammatory disease

(B) Benign and neoplastic disease
  (i) retinoblastoma and its treatment
  (ii) Lymphoma
  (iii) Lymphangioma
  (iv) Eosinophilic granuloma (histiocytosis)
  (v) Rhabdomyosarcoma
  (vi) Leukaemic deposits
  (vii) Dermoid cyst

(C) Systemic disease
  (i) Kawasaki disease
  (ii) Sickle cell anaemia

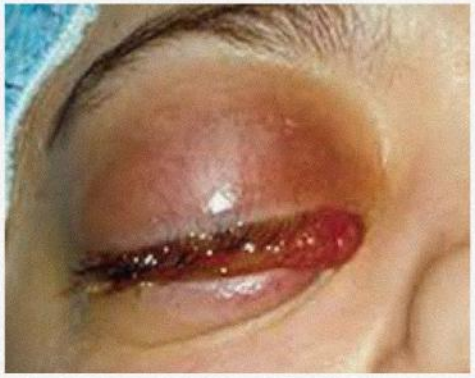
# PROPTOSIS

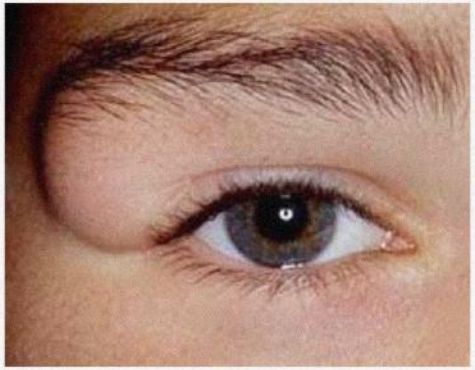
Lid edema
Sinus disease
Restricted motility
Pain/tender
Lid trauma
fever

**ORBITAL CELLULITIS**

Little or no inflammation
Orbital lesion
Young child/adult
Slow growing
Located near bone sutures

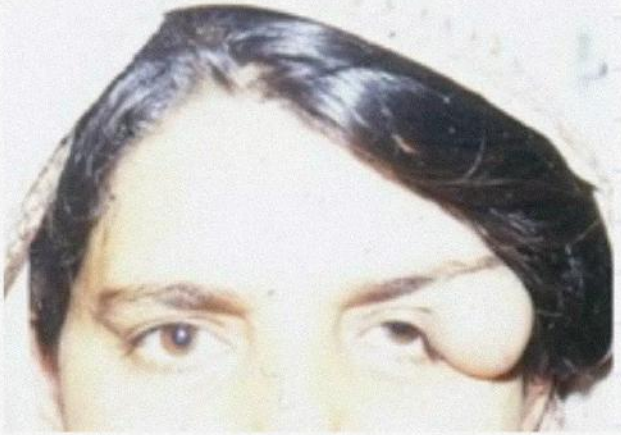
**DERMOID CYST**

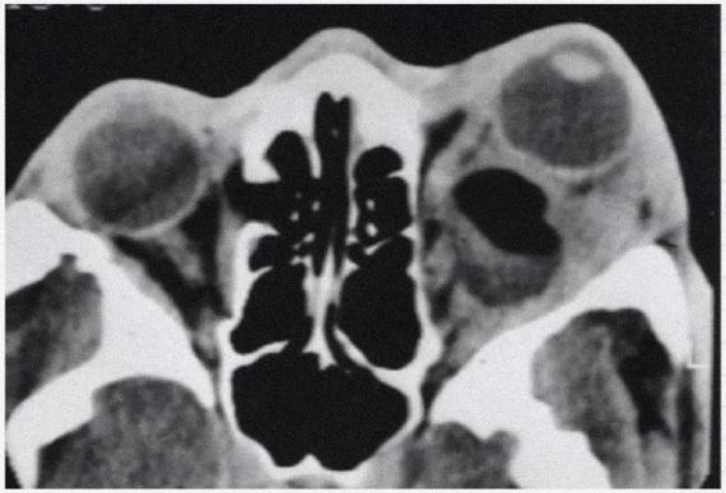




# Dermoid Cyst

- A choristoma found adjacent to suture lines
- Slow growing,nontender mass usually superotemporal mostly in children
- Consist of keratinized stratified squamous epithelium, blood vessels,collagen,sebaceous and sweat glands and hair follicles
- Inflammation if ruptures
- Excision should be in toto





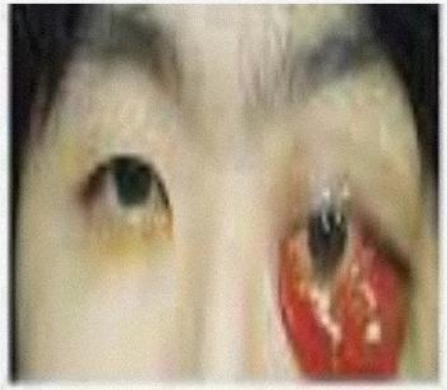
# PROPTOSIS

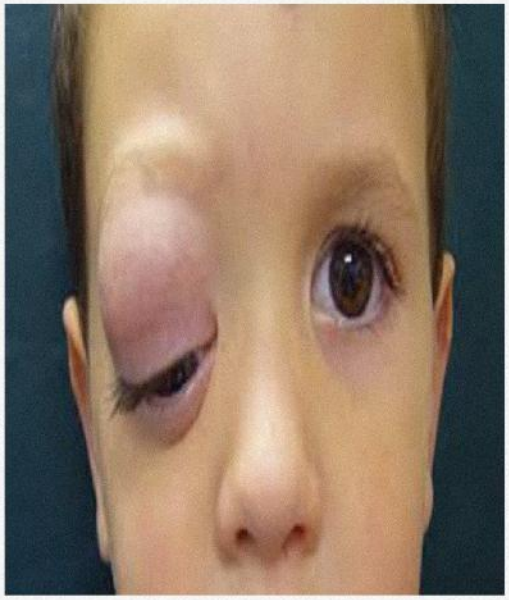
Age ≥5 years
Painful
Upper nasal quadrant mass
Few episodes of proptosis
MRI shows fluid levels

**LYMHANGIOMA**

Age 7-8 years
Unilateral
Hx of nose bleeds
Bluish color lids
Painful
Lymph node involvement

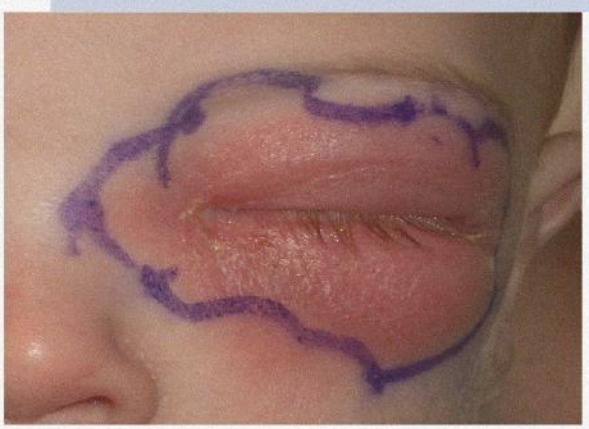
**RHABDOMYOSARCOMA**

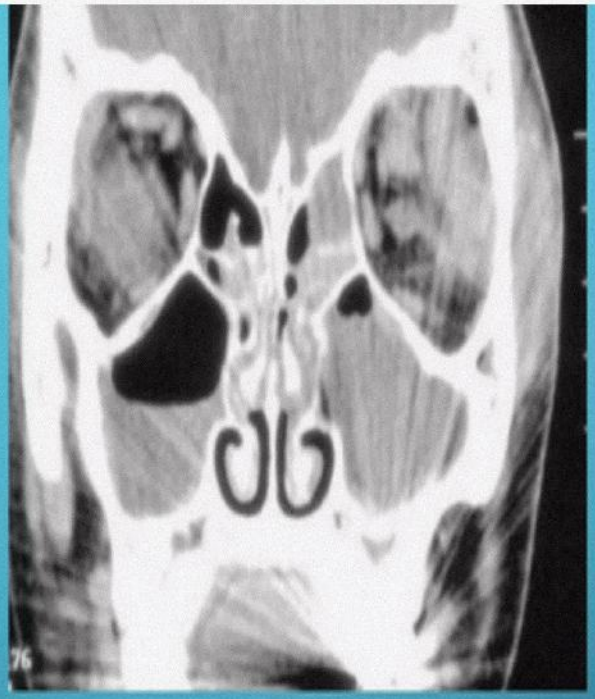




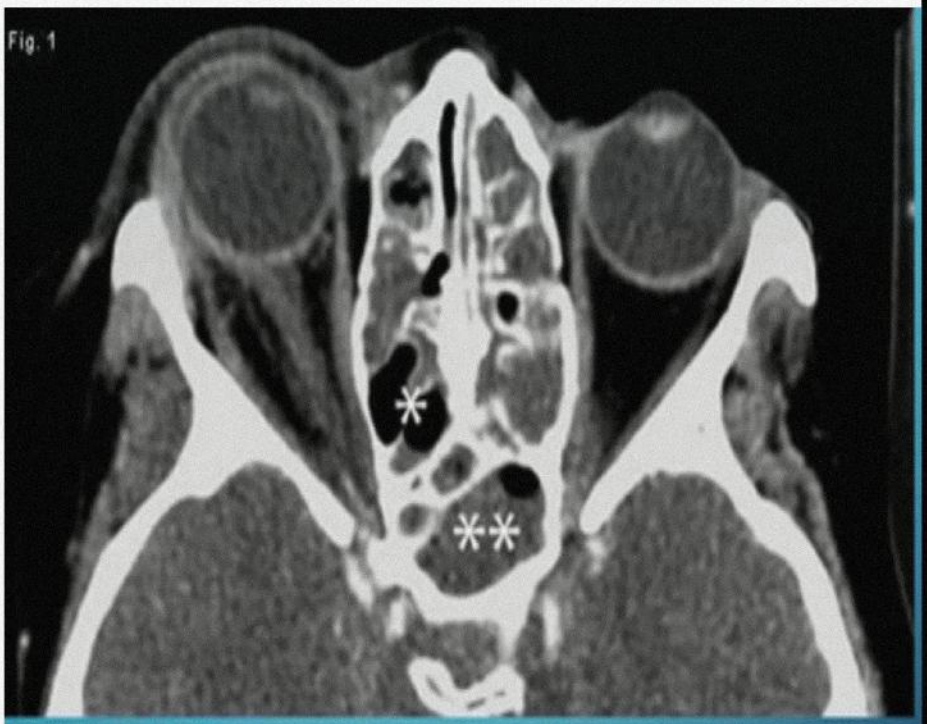
# INVESTIGATIONS/WORK UP

- Full blood count
  Blood cultures
- Nasal swabs
- XRAY PNS
- BSCAN
- CT SCAN
- **MRI**
- LUMBER PUNCTURE



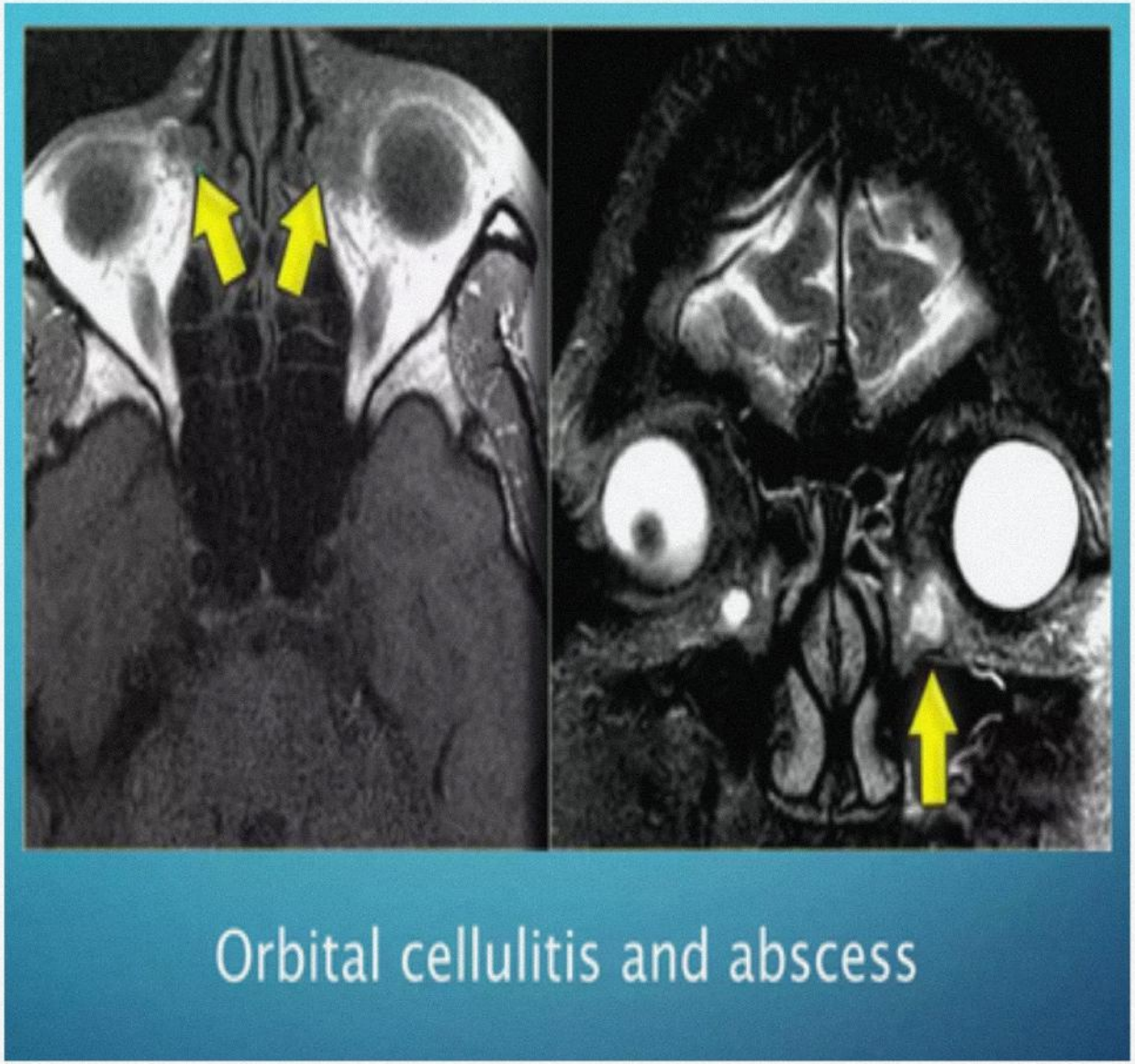


Coronal CT scan in a pediatric patient with sinusitis as well as an orbital and subperiosteal abscess (Left Side)



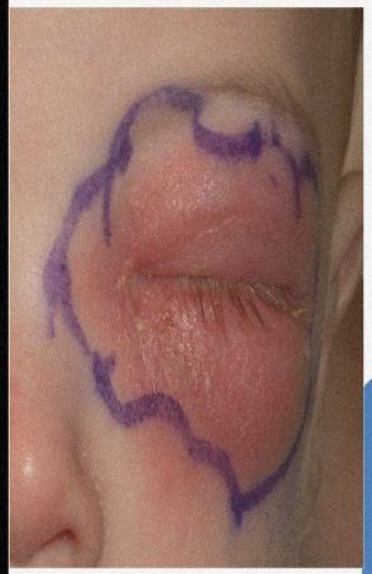

AXIAL VIEW CT scan of the orbit with contrast

- There is Proptosis and Retrobulbar fat stranding.
- Note the mucosal thickening and fluid in the ipsilateral ethmoidal (single asterisk) and sphenoidal sinuses (double asterisk) consistent with acute



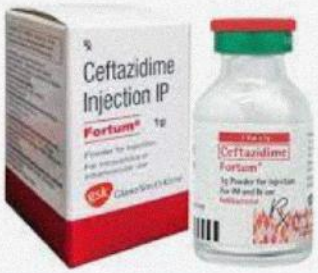
Orbital cellulitis and abscess

# MANAGEMENT



- Hospitalization/ENT opinion
  - Proper work up
    - 4 hourly monitoring of vitals and visual functions/IOP/Pupils
    - Indications for surgical intervention
      - LP for meningeal signs
  - Intensive antibiotic therapy
    - Analgesics
    - Nasal decongestants
    - Anti-inflammatory

# IV ANTIBIOTICS





**Ceftazidime**
**Ceftriaxone**
**Cefotaxime**

**Flucloxacillin**
**Vancomycin**
**Ampicillin -Salbactam**

**Ciprofloxacin/Levoflloxacin**
**metronidazole**

# DOSES

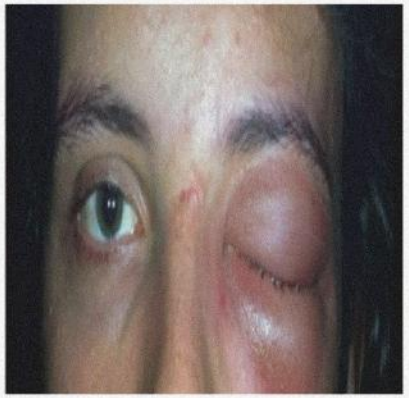
- Ceftazidime (Fortum®)
  - Adults=1-2g/8hr I.V.
  - Pediatric
    - Neonates=30mg/kg IV
    - 1 month-12 years=30-50mg/kg IV
- Ceftriaxone (Rocephin®)
  - Adults=2g/12hr IV
  - Pediatric
    - 20-50mg/kg/day IV
- Cefotaxime (Claforan®)
  - Adults=1-2g/10ml/4hr IV in 3-5 min (max=12g/day)
  - Pediatric
    - 0-1 week=50mg/kg/12hr IV
    - 1-4weeks=50mg/kg/8hr IV
    - 1month-12years=50-80mg/kg/day IV in 4-6 doses

Oral Drugs

- Amoxicillin-clavulanate (Augmentin®)
  - Adults=500mg TDS
  - Children=20-40mg/kg/day 3 divided doses
- Cefaclor (Ceclor®)
  - Adults=500mg TDS
  - Children=20-40mg/kg/day 3 divided doses
- Metronidazole

# INDICATIONS FOR SURGICAL INTERVENTION

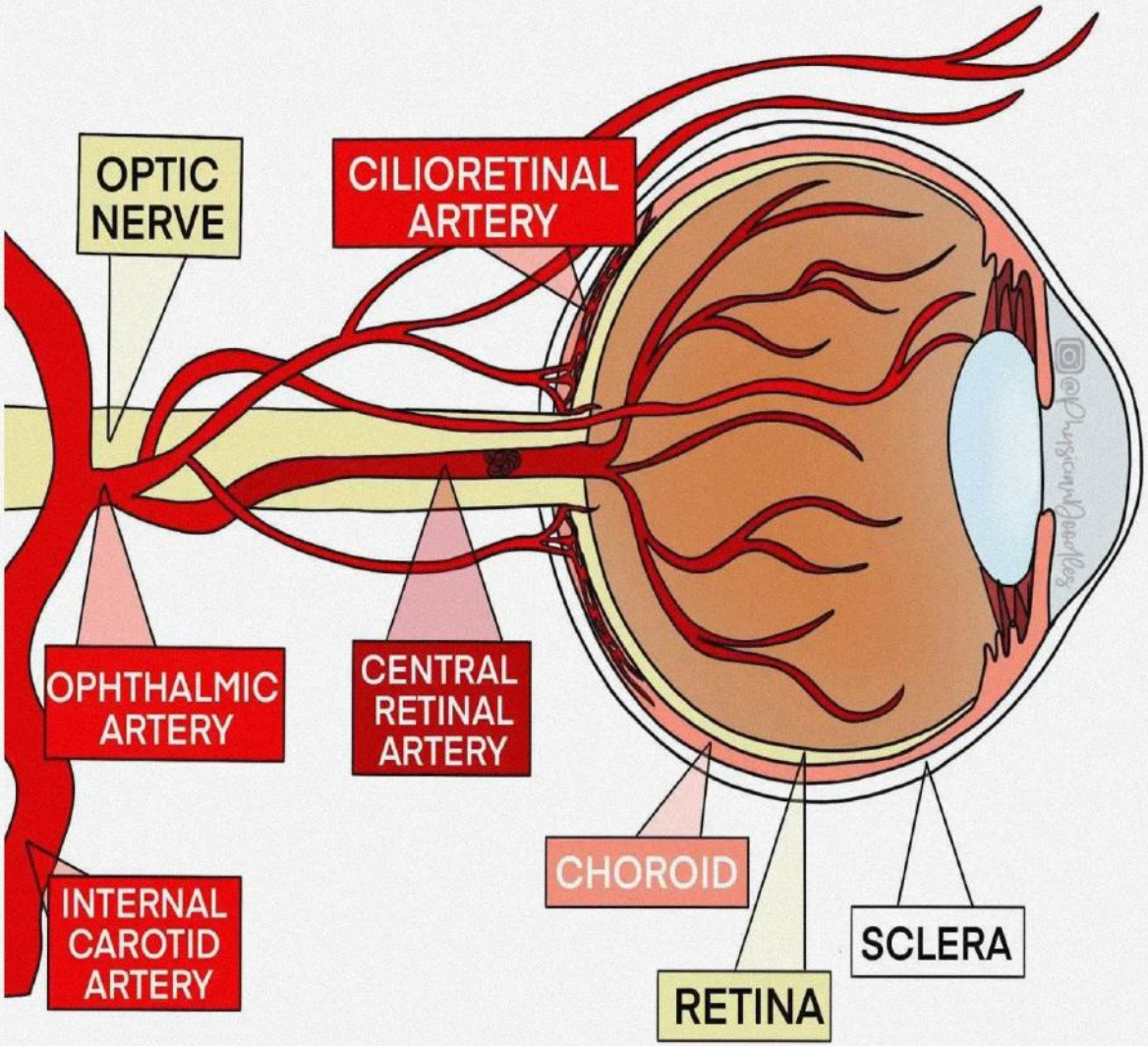
- Suspicion of orbital abscess/foreign body
- Decrease in VA
- EOM defects
- Worsening of proptosis
- Non reduction in abscess size on CT scan after antibiotic therapy

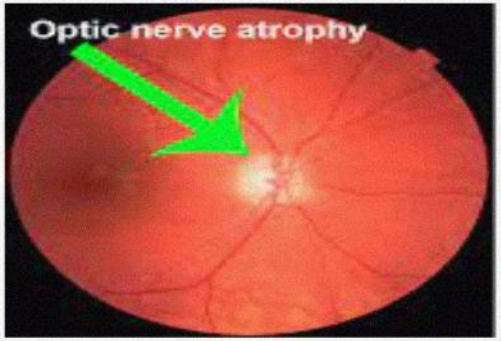


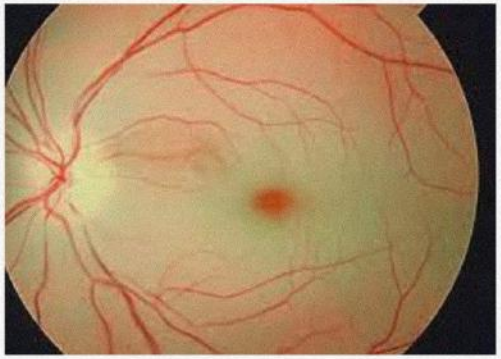
- Canthotomy/Cantholysis in emergency
- Surgical drainage of fluid
- Orbital surgery ± sinusotomy
- For fungal
  - Debridement
  - Exenteration

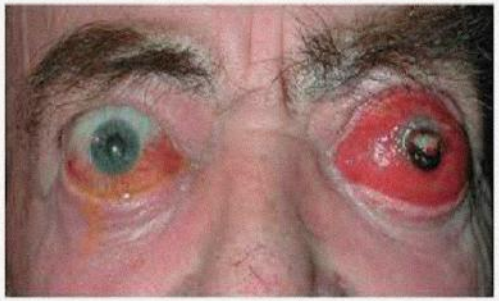
# COMPLICATIONS

- Exposure Keratopathy
- Orbital abscess
- Sub periosteal abscess
- Central retinal artery occlusion/CRVO
- Endophthalmitis
- Increase IOP
- Optic neuropathy
- Cavernous sinus thrombosis
- septicemia
- Hearing loss
- Intracranial spread of infection brain abscess/Meningitis
- Increased mortality

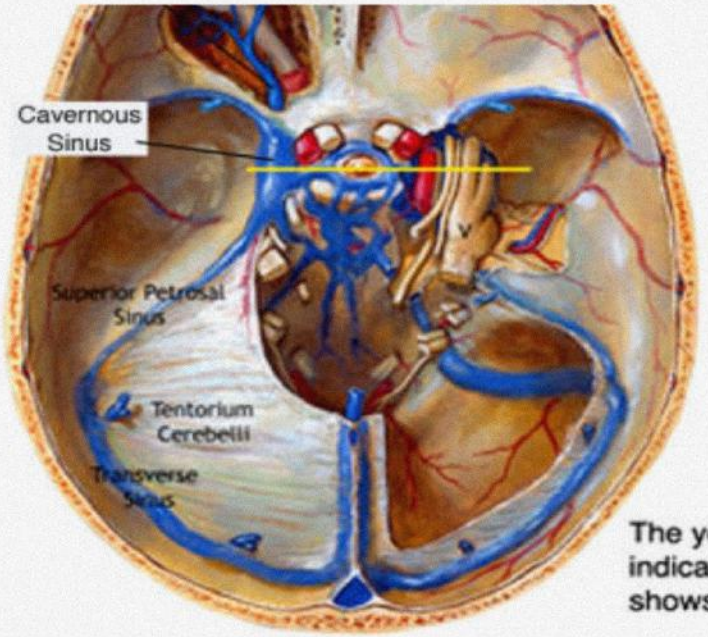

OPTIC NERVE
CILIORETINAL ARTERY
OPHTHALMIC ARTERY
CENTRAL RETINAL ARTERY
INTERNAL CAROTID ARTERY
CHOROID
RETINA
SCLERA

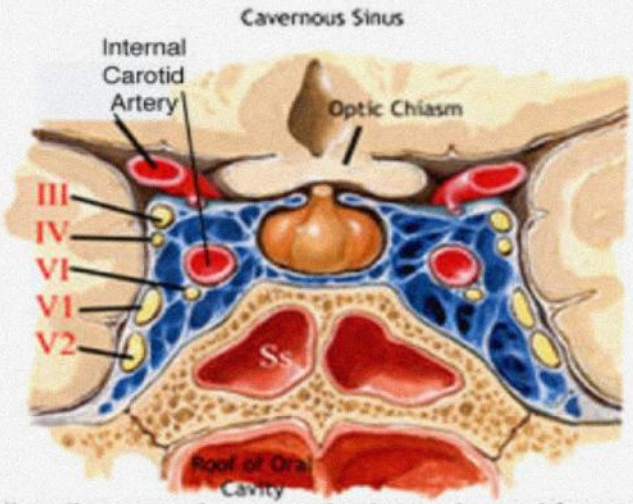

Optic nerve atrophy





# CAVERNOUS SINUS THROMBOSIS




The yellow line crossing over the hypophyseal fossa indicates the plane of section of th image above. It shows the cavernous sinus and its contents.

## Clinical

- High **fever**
- **Periorbital edema** and **chemosis** (conjunctival edema)
- **Cranial nerve palsies** (CN VI most common)
- Decreased **visual acuity**

## Dx

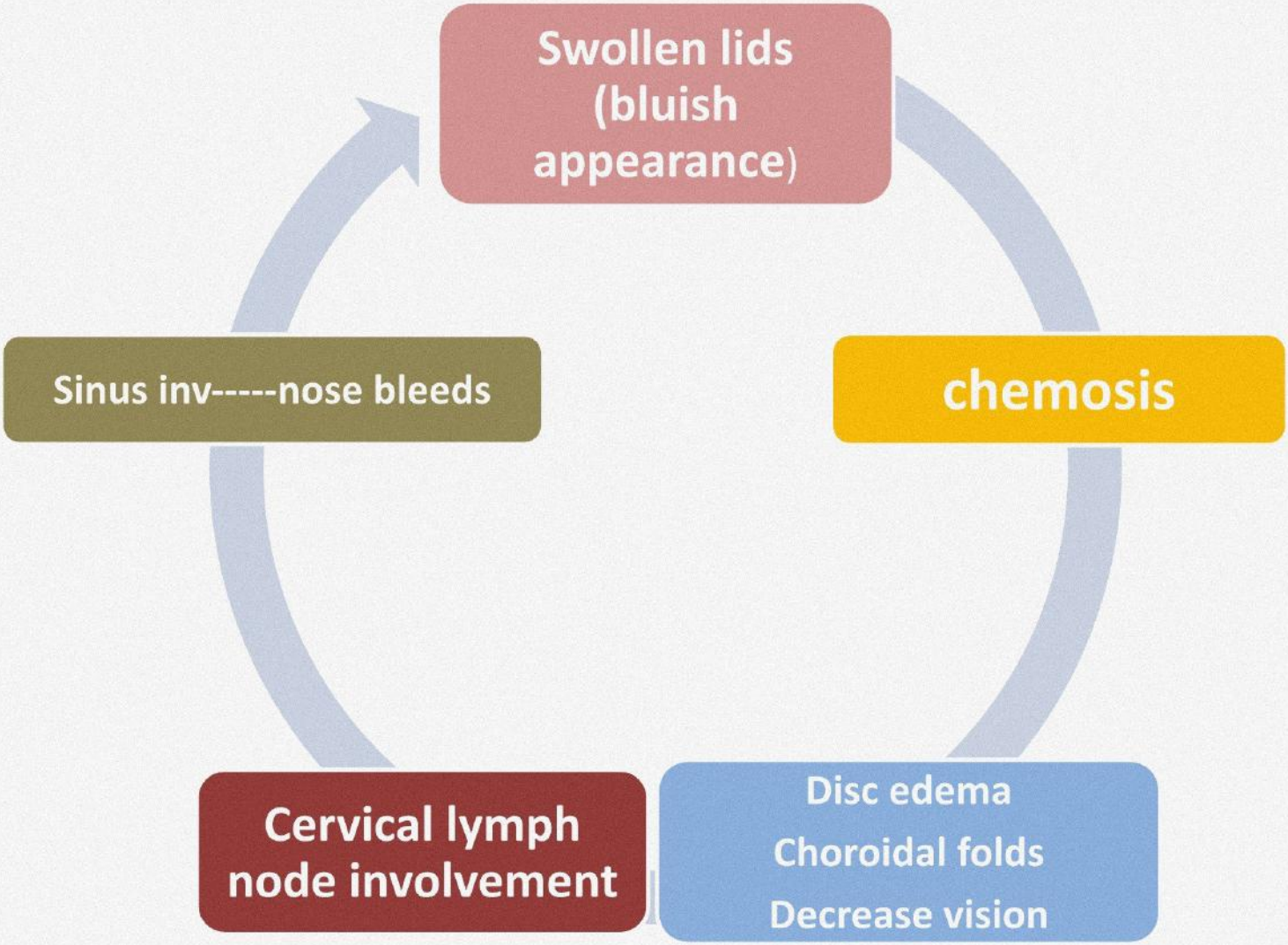
- CT scan
- **MRI**

## Rx

- IV **ABX**
- **Heparin**

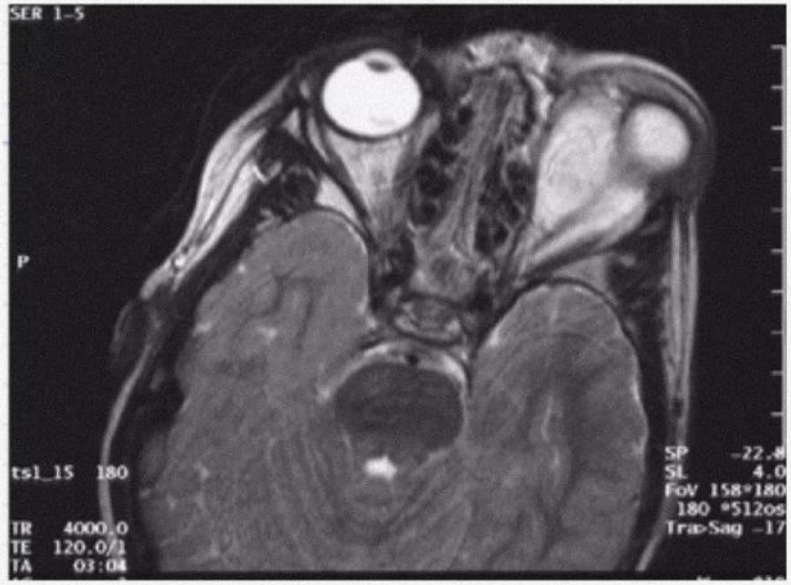
# Embryonal Sarcoma

- Most common primary orbital malignancy of childhood
- Origin……undifferentiated mesenchyme cells
- Called Rhabdomyosarcoma if differentiate into striated muscles
- Usually first decade ……7years
- Boys more than girls
- Rapid onset, unilateral non axial painful proptosis mimics ….orbital cellulitis
- Location….. Superonasal,retrobulbar,superior inferior
- Histology …….Embryonal alveolar and pleomorphic



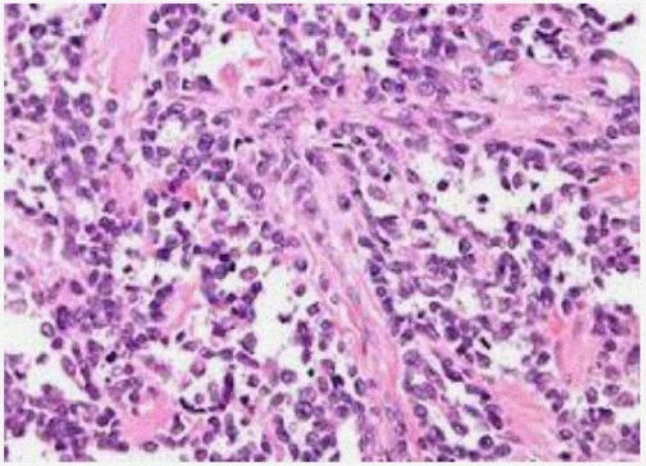
## Work up

- USB,CT,MRI
- Biopsy
- Systemic inv




## Management

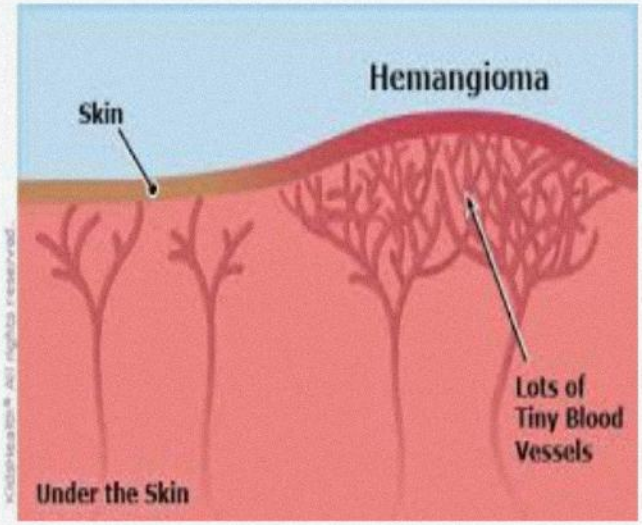
- Refer to pediatric oncologist
- Radiotherapy
- Chemotherapy
- Surgery



| | RHABDOMYOSARCOMA | ORBITAL CELLULITIS |
|---|---|---|
| *Pathology* | Undifferentiated mesenchymal cells, most common primary orbital malignancy of childhood | Infection of soft tissues behind the septum |
| *Demographics* | Boys > girls (avg 7 yrs), | Commonest cause of proptosis in children, adults also affected |
| *Associated* | Cervical L. nodes involved | Usually after trauma, sinus related |
| *Presentation* | Nonaxial, painful, unilateral, rapid onset, swollen bluish lids, nose bleed, Ptosis, chemosis | Painful, lids edema with reddish color, Ptosis, malaise, fever, life threatening, VA impaired, Ophthalmoplegia, |
| *Radiology* | Moderately well-defined homogenous mass, Bony destruction | CT-(orbit, sinuses, brain): diffuse orbital infiltrate, sinus opacity, proptosis<br>MRI for cavernous sinus thrombosis. |
| *Tx* | Radiotherapy, chemotherapy, Exenteration for resistant cases | Medical, surgical |

# CAPILLARY HEMANGIOMA

- Benign endothelial cell neoplasms
- Rapid growth in infancy and involutes later(75%-7years,30-50%-5years)
- Most common orbital tumor of infancy
- Usually located superonasal, brow and eyelid
- Blanches on pressure
- Ptosis and proptosis on posterior extension
- Cutaneous, preseptal,extraconal and intraconal



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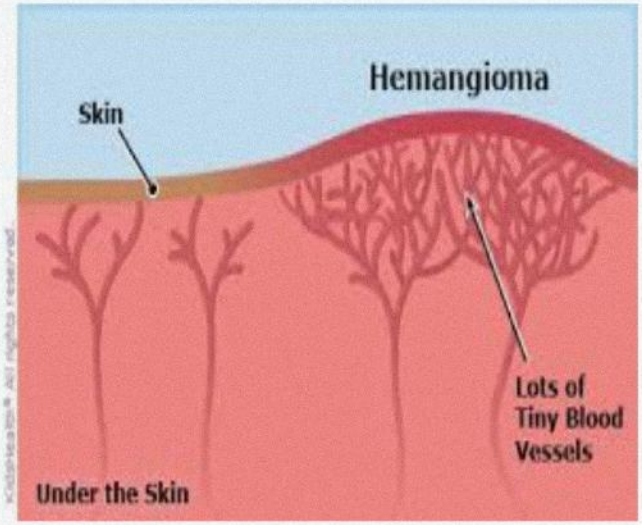
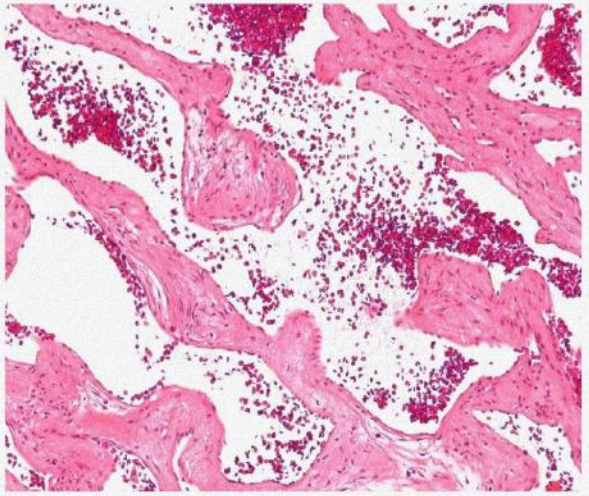
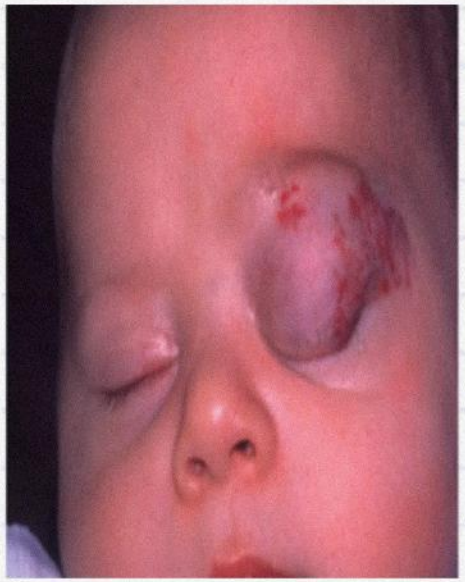
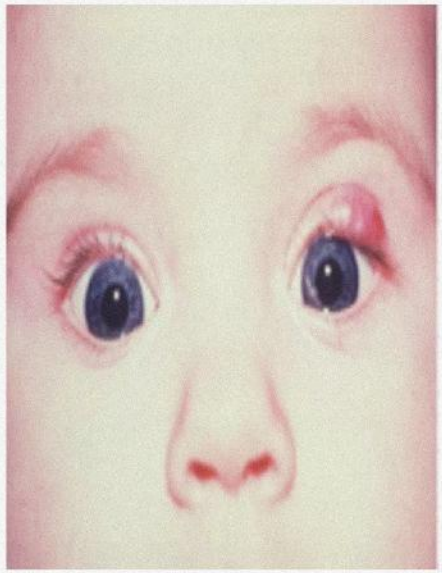


# Mortality/Morbidity

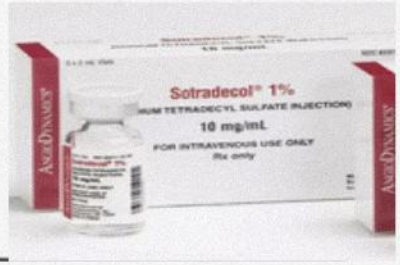
- Kasabach-Merritt syndrome
  - Coagulopathy, thrombocytopenia
  - large visceral / Nasopharyngeal hemangiomas
  - DIC may occur, high output CCF
  - Mortality...30-50%
- Ophthalmic morbidity
  - Space occupying
  - Amblyopia

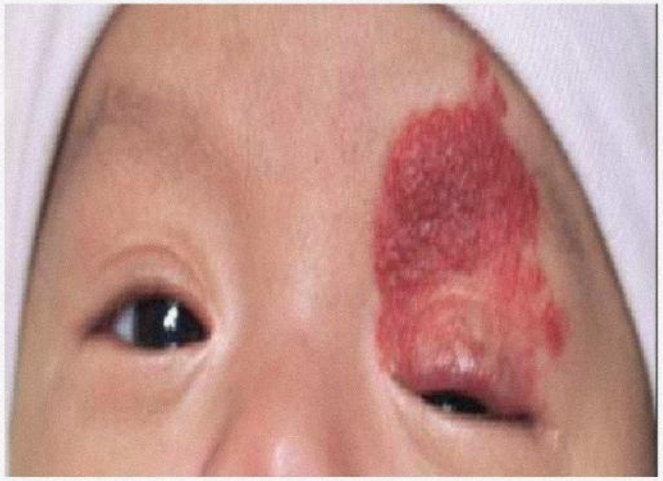


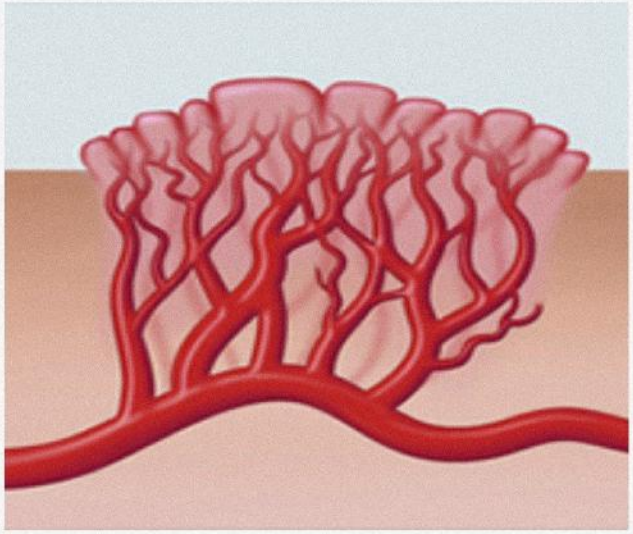


# MANAGEMENT

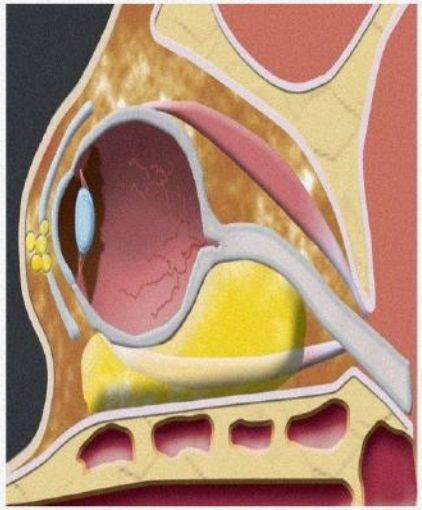
| Medical | Surgical |
|---|---|
| **Topical** | |
| Topical corticosteroids under occlusion | Laser therapy |
| Intralesional corticosteroids | Cryotherapy |
| Becaplermin | Radiotherapy |
| Imiquimod | Surgical excision |
| **Systemic** | Compression |
| Systemic corticosteroids | Embolization |
| Vincristine | Sclerosant injection |
| Interferon-alpha | |
| Bleomycin | |
| Cyclophosphamide | |

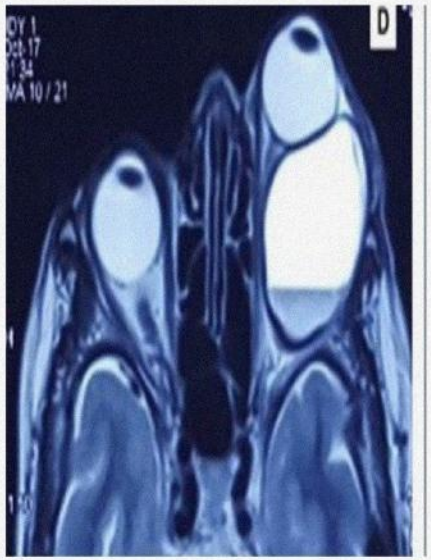


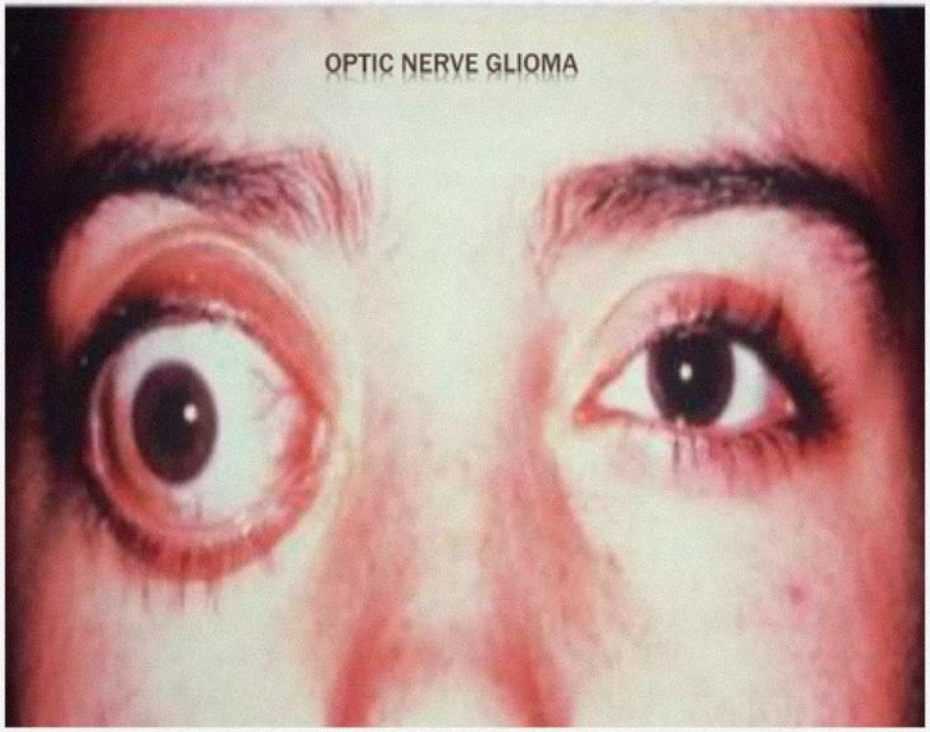


| | CAPILLARY HEMANGIOMA | ORBITAL LYMPHANGIOMA |
|---|---|---|
| *Pathology* | Vascular hemartoma | Isolated vascular hemartoma |
| *Demographics* | Infant, commonest benign orbital tumor in childhood, spontaneous involution | Early childhood |
| *Associated* | Visceral, nasopharyngeal hemangioma, kasabach-merit syndrome | - |
| *Presentation* | Nonaxial, +ve Valsalva, superficial/deep | Nonaxial, -ve Valsalva, acute episodes of spontaneous hemorrhage, may be superficial/deep |
| *Radiology* | Intra/extraconal mass, poorly defined | Low density cyst-like mass, enlargement of bony orbit |
| *Tx* | Observation, surgery | Guarded prognosis, surgery, drainage |





# OPTIC NERVE GLIOMA



# Classification

- **Primary Neural Tumors**

  Optic nerve
  Peripheral nerves

- **Secondary Neural Tumors**

  Metastatic and infiltrative neoplasms
  **Breast cancer**, prostate cancer, melanoma, lung cancer

  ON may be affected by peripheral nerve tumors in the orbit

# OPTIC NERVE GLIOMA

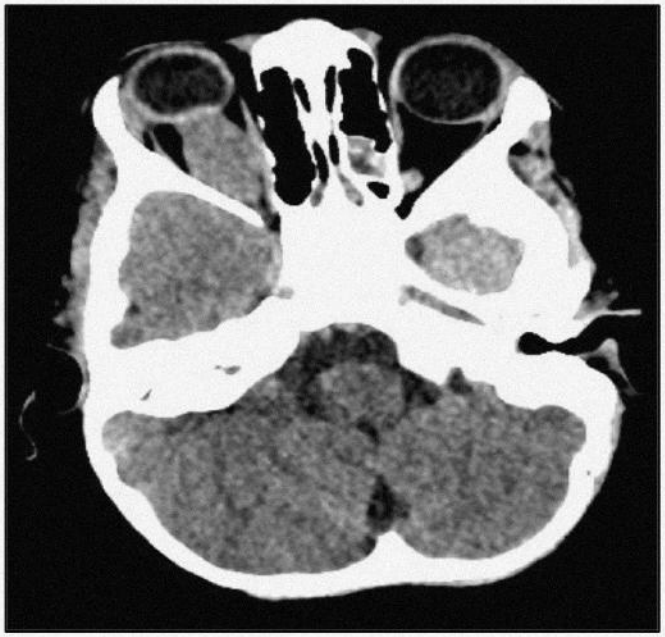
- Most common ON neoplasm, arising from astrocytes within the ON

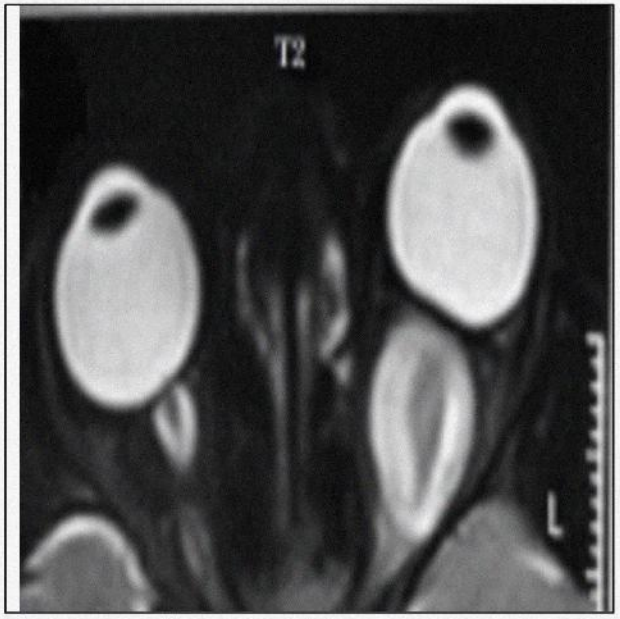
- BENIGN_ common in children (juvenile pilocytic astrocytomas )
- MALIGNANT in adulthood (malignant glioblastoma)

- 90% diagnosed during the first 2 decades of life (median age 5 years)
- Females preponderance (60%)

- 10-70% of patients diagnosed with juvenile pilocytic astrocytoma have associated (NF-1)
- In patients with NF-1, 8-31% are diagnosed with associated ON glioma

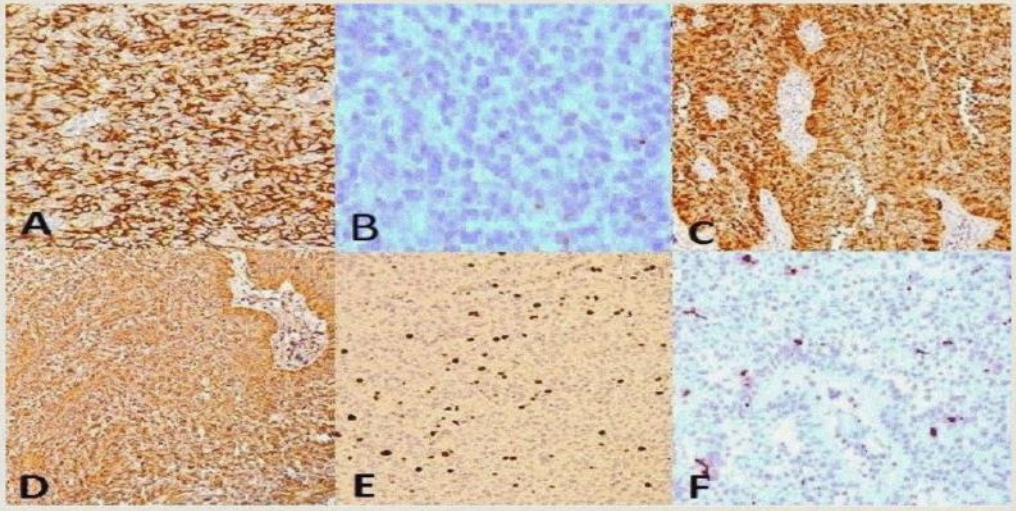
| | NON –NEUROFIBROMATOSIS TYPE 1 | NEUROFIBROMATOSIS TYPE 1 |
|---|---|---|
| Presentation | Visual loss, strabismus,proptosis | Asymptomatic, visual loss |
| Progression | 63% | 12% |
| Visual outcome | Poor | Good |
| Growth rate | Faster- occasionally rapid | Stable, slow growing |
| Location | Discrete, unilateral | Multifocal, diffuse, bilateral |
| Survival | 5 year =83% 10 years =63% | 5 year =93% 10 years =81% |
| Radiographic findings | Fusiform ON enlargement , loss of perineural space | Fusiform ON enlargement , kinking of intraorbital nerve |
| Associated features | None | Café au lait spots, lisch nodules |
| Hydrocephalus | 79% ( radiologically) | Very rare |
| Follow up | Regular imaging | Not routine unless symptomatic |





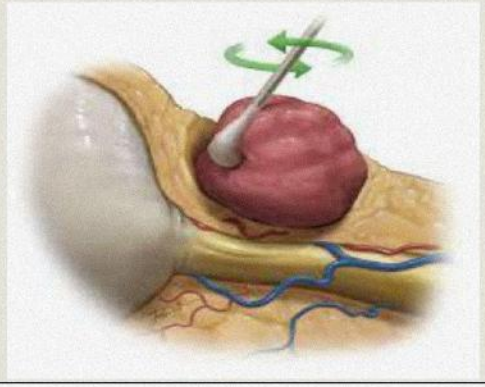
# Cellular morphology
# Immunohistochemistry

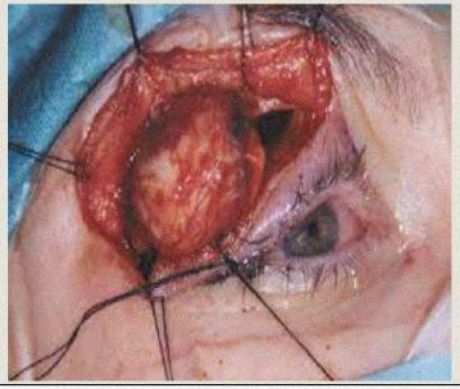
- Meningiomas are positive for EMA and S-100
- Gliomas are positive for GFAP and negative for S-100 and EMA
- Peripheral nerve sheath tumors contain Schwann cells that stain positively for S-100
- Perineural cells may also be positive for EMA
- Granular cell tumors stain positively for S-100 and may be positive for CD68 and leu-7

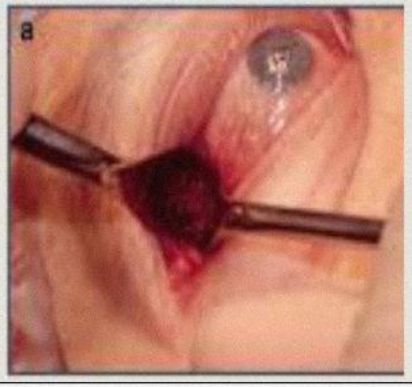


# Treatment Options

- Observation
- Radiotherapy
- Chemotherapy
- Surgical excision





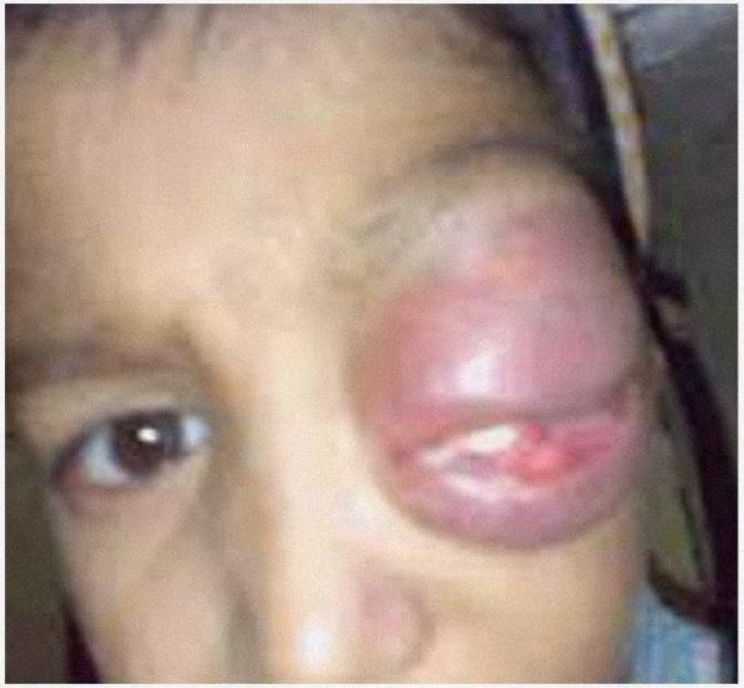


# CHILDHOOD METASTATIC TUMORS

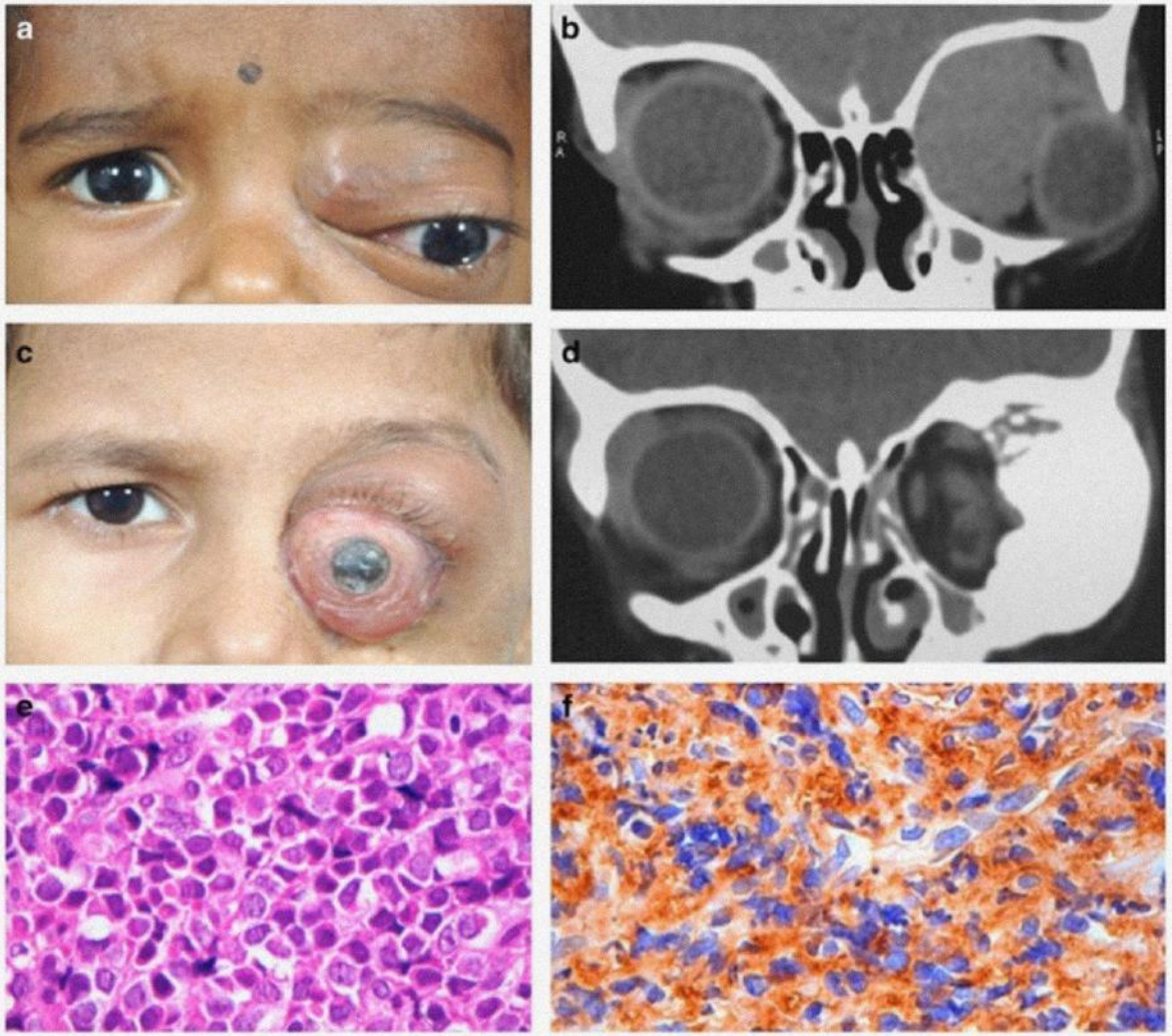
## Neuroblastoma

- Most common childhood cancer.
- Frequent source of orbital metastasis.
- Originates in either adrenal gland or sympathetic ganglion chain in the retroperitoneal mediastinum.
- Metastatic Neuroblastoma in the orbit typically produces proptosis, & periorbital ecchymosis.
- CT – shows evidence of bone destruction.
- With intensive treatment including radio & chemotherapy prognosis is considerably better in infants under the age of 1 yr. than older children.

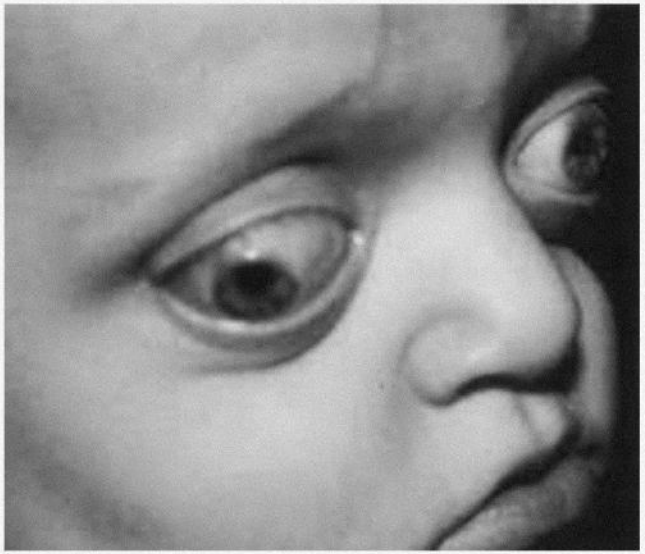
# NEUROBLASTOMA



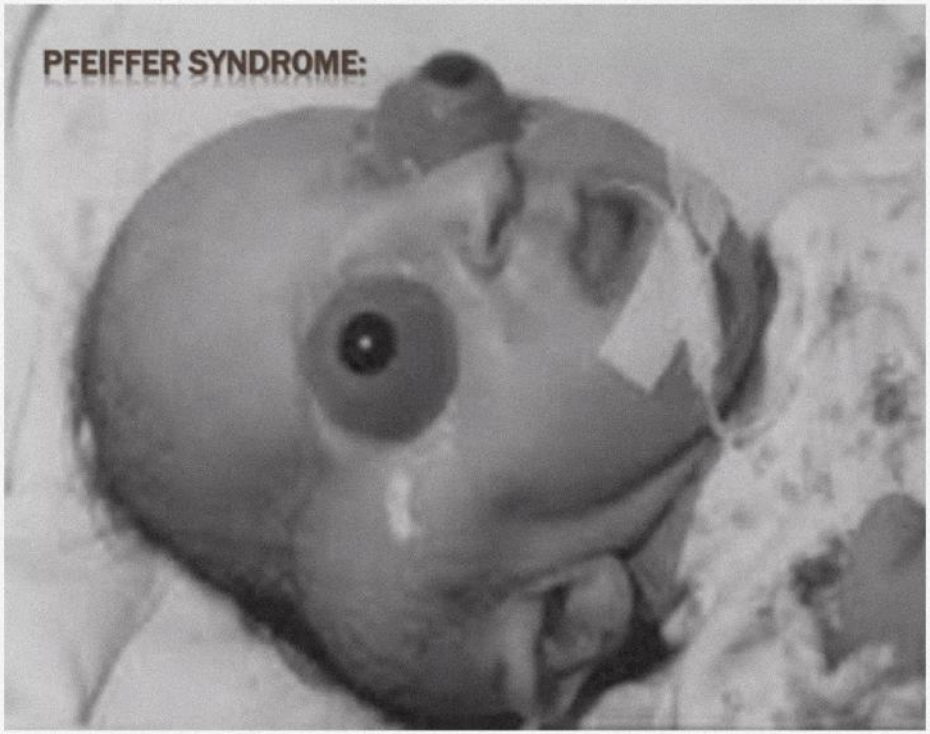
# EWINGS SARCOMA



# DEVELOPMENTAL ANOMALIES



CROUZEN SYNDROME



# CONCLUSION

- General examination, CBC, peripheral smear and local imaging study is mandatory to rule out malignant conditions

- Histopathology aids in the precise diagnosis

***Unilateral proptosis in a child-need for prompt diagnosis***