

# Orbital Disorders

## 429 Ophthalmology Team Notes - F2

Sources: Dr. Adel AlSuhibani's lecture, Toronto Notes 2012, Clinical Ophthalmology - A Systemic Approach 6th ed, OphthoBook 2nd ed. **NOTE:** these notes follow the outline of the lecture. Key points about some of the individual conditions were added as a quick revision/introduction. All pictures from the original lecture are included.

By: AlBatool AlAmmari

## **OBJECTIVES**

### **COURSE OBJECTIVES:**

#### **1. Orbital Disorders:**

##### **Orbital Cellulitis**

- Etiology
- Management

##### **Inflammatory Orbital Disorders**

##### **Trauma and Orbital Tumors**

### **LECTURE OUTLINES:**

#### **1. Orbital Anatomy**

Bones  
Orbital compartments  
Blood supply  
Sinuses  
Annulus of Zinn

#### **2. Evaluation and Orbital Diseases Presentations**

#### **3. Common Orbital Disorders**

##### **Infections:**

Orbital cellulitis  
Pre-septal cellulitis

##### **Inflammation:**

Grave's Disease  
Idiopathic Orbital Inflammatory Disease

##### **Masses and Cysts:**

Lymphoproliferative Disorders  
Rhabdomyosarcoma  
Dermoid Cysts

#### **4. Facial Trauma and Fractures**

#### **5. Imaging Techniques in orbital disorders**

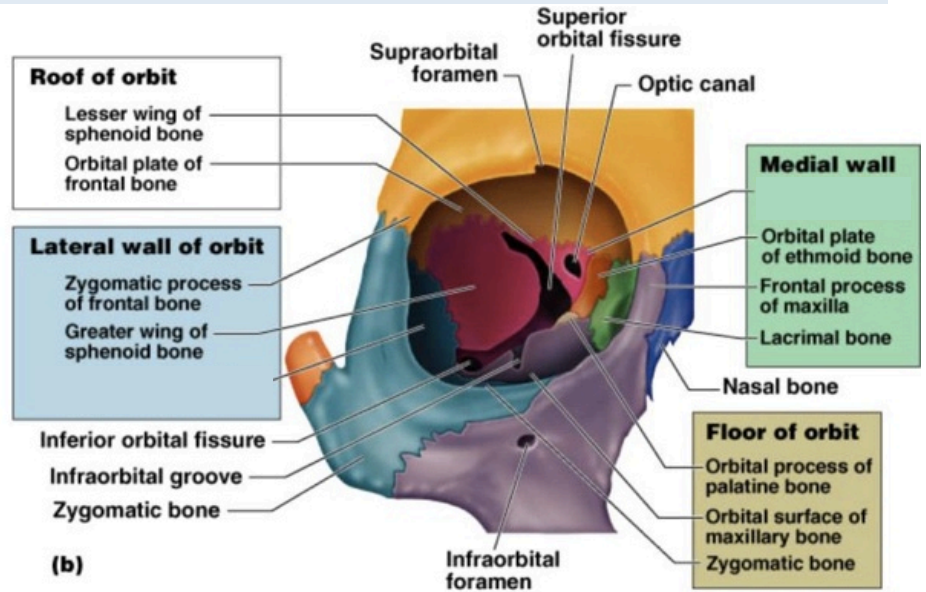
# INTRODUCTION

## ANATOMY:

### BONES:

The orbit is a pear-shaped cavity, the stalk of which is the optic canal.

1. **The roof** consists of two bones: the **lesser wing of the sphenoid** and the **orbital plate of the frontal**. It is located subjacent to the anterior cranial fossa and frontal sinus. [EXTRA: A defect in the orbital roof may cause pulsatile proptosis as a result of transmission of CSF.]



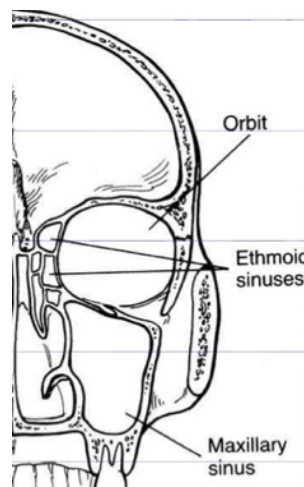
2. **The lateral wall** also consists of two bones: **greater wing of the sphenoid** and **zygomatic**. The anterior half of the globe is vulnerable to lateral trauma since it protrudes beyond the lateral orbital margin.
3. **The floor** consists of three bones: **zygomatic**, **maxillary** and **palatine**. The posteromedial portion of the maxillary bone is relatively weak and may be involved in a 'blowout' fracture.
4. **The medial wall** consists of three bones: **maxillary**, **lacrimal**, and **ethmoid**. The lamina papyracea, which forms part of the medial wall that separates the orbit from ethmoidal sinuses, is paper-thin and perforated. Orbital cellulitis is therefore frequently secondary to ethmoidal sinusitis.

[NOTE: Despite the fragility of the medial wall, it is well buttressed and rarely fractures. The orbital floor, however, breaks most often during blunt trauma. The maxillary bone fractures downward and the orbital contents can herniate down into the underlying maxillary sinus. This is called a "blowout fracture" and can present with enophthalmia (a sunken-in eyeball)]

### 5. The superior orbital fissure

[EXTRA: it's a slit between the wings of the sphenoid bone through which pass the following important structures from the cranium to the orbit:

- The superior portion contains the lacrimal, frontal [branches of the ophthalmic division of the trigeminal nerve] and trochlear nerves, and the superior ophthalmic vein.
- The inferior portion contains the superior and inferior divisions of the oculomotor nerve, the abducens, the naso-ciliary [branch of the ophthalmic division of the trigeminal nerve] and sympathetic fibers.]



### SINUSES:

Both the maxillary and ethmoidal sinuses are separated from the orbit by thin walls. Thus there's always a chance of orbital invasion with ethmoidal and maxillary sinuses' diseases.

# INTRODUCTION

## ANATOMY:

### MUSCLES (ANNULUS OF ZINN) IMP:

The "Annulus of Zinn," a tendinous band that serves as the insertion point for most of the ocular muscles.

- **Inside the Annulus of Zinn:**

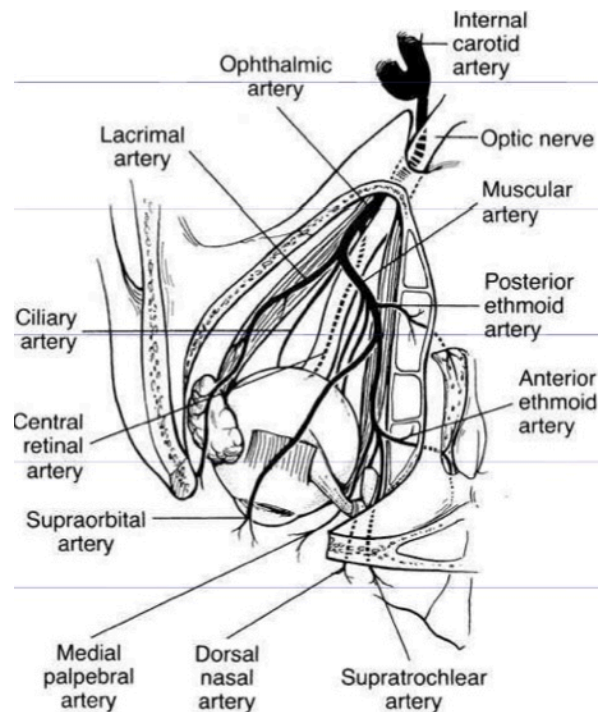
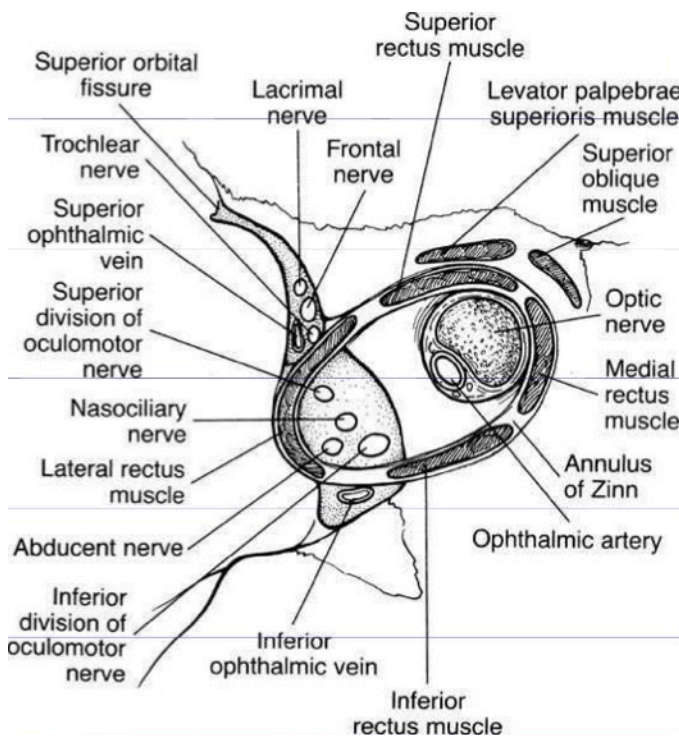
Nerves → Superior and inferior divisions of the oculomotor nerve, nasociliary branch of the ophthalmic division of the trigeminal nerve, the abducent nerve and the optic nerve.

- **Outside the Annulus of Zinn:**

Nerves → Trochlear nerve and the lacrimal and frontal branches of the ophthalmic division of the trigeminal nerve

Veins → the superior and inferior ophthalmic veins

The ophthalmic artery varies, could be inside or outside, but it's usually outside.

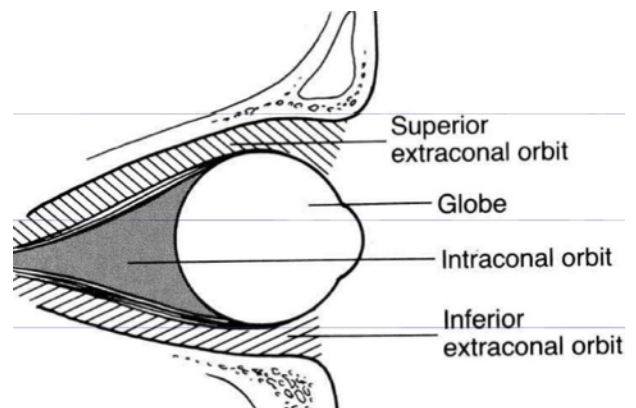


### BLOOD SUPPLY:

All extraocular muscles are supplied by the lateral and medial muscular branches of the ophthalmic artery.

### ORBITAL COMPARTMENTS:

Superior extraconal, inferior extraconal and intraconal compartments



# CLINICAL SIGNS AND PRESENTATIONS OF ORBITAL DISEASES:

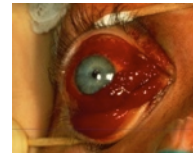
## EVALUATION OF THE ORBIT BY THE 7 P'S:

Pain, Progression, Proptosis, Palpation, Pulsation, Periorbital Changes, Past Medical History

### 1. PAIN:

Pain is a sign of:

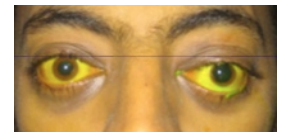
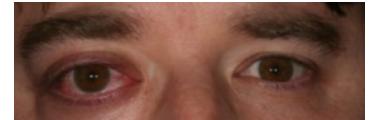
- Infection
- Inflammation
- Acute Hemorrhage
- Malignant Lacrimal Gland Tumor



### 2. PROGRESSION:

- **Minutes to hours** → Hemorrhage
  - Lymphangioma
  - Orbital Venous Varix

[**EXTRA:** Orbital Venous Varix is an uncommon vascular malformation which is composed of enlarged tubular venous channels with direct communication to the systemic venous system]



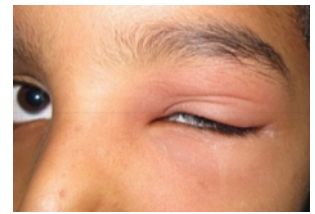
- **Days to weeks** →

In children: Capillary Hemangioma  
Rhabdomyosarcoma  
Retinoblastoma  
Neuroblastoma  
Leukemia



Capillary Hemangioma

In adults: Infection [orbital cellulitis]  
Inflammation [idiopathic orbital inflammatory disease, thrombophlebitis, thyroid orbitopathy, recurrent inflamed dermoid]  
Hemorrhage [orbital hemorrhage, lymphangioma]  
Malignancy [rhabdomyosarcoma, metastatic tumors, granulocytic sarcomas, adenoid cystic carcinoma]  
Fistula [Carotid-Cavernous fistula]  
Trauma or Post-surgical  
Allergy [Allergic eyelid swelling]



Allergic eyelid swelling

- **Months to years** → Dermoid cysts
  - Benign mixed tumors
  - Neurogenic tumors
  - Cavernous hemangioma
  - Lymphoma
  - Fibrous histiocytoma
  - Osteoma
  - Lipoma
  - Glioma
  - Meningioma

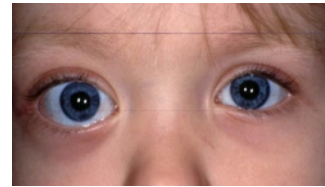
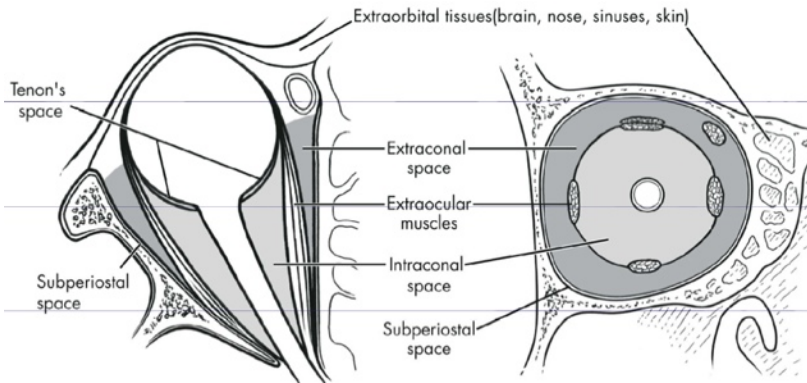


## CLINICAL SIGNS AND PRESENTATIONS OF ORBITAL DISEASES:

### 3. PROPTOSIS:

Proptosis describes an abnormal protrusion of the globe it could be axial, non-axial or pulsatile, present in one or both eyes. Asymmetrical proptosis is best detected by looking down at the patient from above and behind

[**NOTE:** Pseudoproptosis must be ruled out first. Severe proptosis may compromise lid closure with resultant exposure keratopathy, corneal ulceration and infection.]



Axial



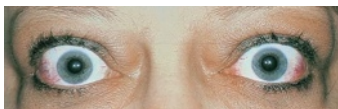
Non-Axial

### TYPES OF PROPTOSIS:

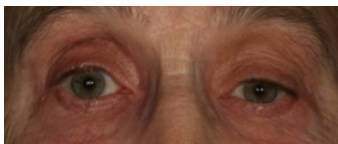
- Axial** arises from intra-muscular or intra-conal pathologies
  - Non-axial** arises from extra-muscular or extra-conal pathologies
  - Pulsatile** seen in Neurofibromatosis Type-1
- d. **Unilateral** → Primary Orbital Neoplasms
- e. **Bilateral** → - Inflammatory diseases [Thyroid disease, idiopathic orbital inflammatory disease or sarcoidosis involving the lacrimal glands]
- Infection [Orbital abscess or cellulitis]
  - Vascular diseases [Orbital hemorrhage, lymphangioma, C-C fistula, orbital varices-proptosis with Valsalva, or vasculitis due to giant-cell arthritis, Wegener granulomatosis, polyarteritis nodosa and systemic lupus erythematosus]
  - Tumors [**Benign**: cavernous hemangioma, lymphangioma  
**Malignant**: adenoid cystic carcinoma, lymphoma, glioma  
**Contiguous**: sinus, intracranial nasopharynx, skin  
**Metastatic**: lymphoma, leukemia, neuroblastoma  
**Rhabdomyosarcoma**]

### DIFFERENTIATION BETWEEN TRUE AND PSEUDO PROPTOSIS:

#### **Clinically:**



- In pseudo-proptosis the lid is retracted while the globe is normal and not protruded.



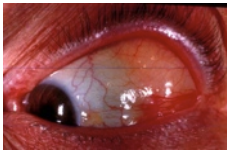
- In some cases the effected eye is herniated inside (enophthalmos) and the normal eye looks pushed out like in floor fractured.

- Proptosis is measured using **Hertel exophthalmometer**.

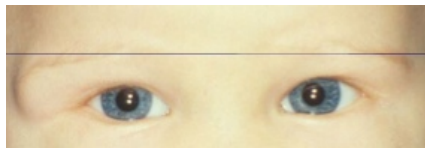
## CLINICAL SIGNS AND PRESENTATIONS OF ORBITAL DISEASES:

### 4. PALPATION:

[**NOTE:** To determine the site, size, shape, consistency, composition and edges of a mass or a cyst. Also to discover its relation to the skin and surrounding tissue and whether it's mobile or not.]



Fat  
Herniation



Dermoid  
Cyst

### 5. PULSATION:

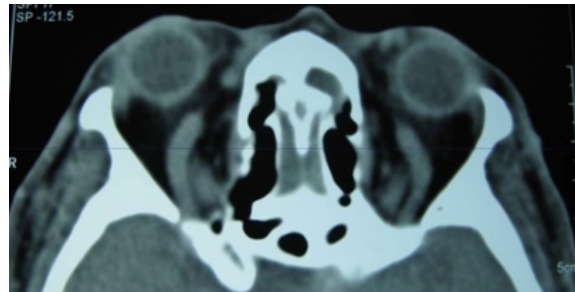
Is caused either by an arteriovenous communication or a defect in the orbital roof.

**With bruit** → Cavernous-Carotid fistula  
Orbital arteriovenous fistula  
Dural arteriovenous (a-v) fistula

**Without bruit** → Meningoencephaloceles  
Neurofibromatosis  
Orbital roof defect (after surgical removal of orbital roof, sphenoid wing dysplasia)



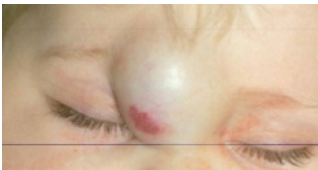
Patient with C-C fistula



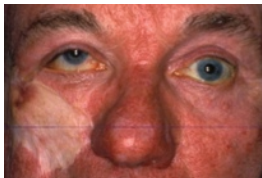
Superior ophthalmic vein enlargement  
in C-C fistula

### 6. PERI-ORBITAL CHANGES:

**Swelling** in relation to the medial canthal area:



If the swelling **above** the medial canthal area brain lesions must be ruled out.



If it's **below** it naso-lacrimal sac pathologies are suspected.

### 7. PAST MEDICAL HISTORY

## ORBITAL DISORDERS

### INFECTIONS **IMP**:

#### Orbital Cellulitis:

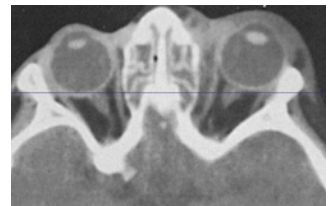
Bacterial orbital cellulitis is a life-threatening infection of the soft tissues behind the orbital septum. It can occur at any age but is more common in children.

[**NOTE**: The most prevalent causative organisms are *S.pneumoniae*, *S.aureus*, *S.pyogenes* and *H.influenzae*.]

**Pathogenesis**: in 90% of cases it's sinus-related, most commonly ethmoidal.

#### Diagnosis:

1. Presentation is with a rapid progression of severe malaise, fever, pain and visual impairment.
2. Signs Unilateral, tender, warm and red periorbital oedema.  
Proptosis, which is often obscured by lid swelling  
Painful ophthalmoplegia.  
Optic nerve dysfunction seen as [pupils and visual field impairment]  
[**NOTE**: Pain, rapid progression, proptosis and periorbital changes]
3. CT shows pre-septal and orbital opacification



**Complications**: There's a high risk of morbidity and mortality as it progresses orbital abscess, brain abscess and cavernous sinus thrombosis if left untreated.

#### Treatment:

1. Hospital admission with frequent ophthalmic and otolaryngological assessment is mandatory.  
Intracranial abscess formation may necessitate drainage.  
[**NOTE**: It is usually necessary to drain the infected sinuses as well as the orbit]
2. Antimicrobial therapy  
[**EXTRA**: involves ceftazidime and metronidazole to cover anaerobes. Therapy should be continued until the patient is afebrile for 4 days.]
3. Monitoring of optic nerve function
4. Surgical intervention

#### Pre-septal Cellulitis:

An infection of the subcutaneous tissues anterior to the orbital septum. Although not an orbital disease, it is included because it must be differentiated from the much less common but more serious orbital cellulitis. Occasionally rapid progression to orbital cellulitis may occur.

#### Diagnosis:

1. Signs: Unilateral, tender and red periorbital oedema. Unlike orbital cellulitis **proptosis and chemosis are absent**; visual acuity, pupillary reactions and ocular motility are unimpaired.
2. CT shows opacification anterior to the orbital septum.

**Treatment**: [**EXTRA**: Treated with antibiotics i.g. oral co-amoxiclav.]

Table 4. Differentiating Between Preseptal and Orbital Cellulitis

Finding	Preseptal Cellulitis	Orbital Cellulitis
Fever	May be present	Present
Lid edema	Moderate to severe	Severe
Chemosis	Absent or mild	Marked
Proptosis	Absent	Present
Pain on eye movement	Absent	Present
Ocular mobility	Normal	Decreased
Vision	Normal	Diminished ± diplopia
RAPD	Absent	May be seen



## ORBITAL DISORDERS

### INFLAMMATION:

#### Grave's Disease:

Thyroid eye disease (TED) affects patients with Graves disease, and is the **most common cause of unilateral and bilateral proptosis**.

TED may precede, coincide with or follow hyperthyroidism and *bears no relationship to the severity of thyroid dysfunction*. It may vary from being merely a nuisance, to blindness secondary to exposure keratopathy or optic neuropathy and it's not controlled by thyroid ablation.

#### **Pathogenesis:**

Initiation: T-cells recognize thyroid-fibroblast cross-active antigen. T-cells infiltrate tissues and release cytokines (including interferon), interleukin 1- $\alpha$  and transforming-growth factor  $\beta$ .

Propagation: Expression of immunomodulatory proteins is enhanced and fibroblasts increase glycosaminoglycan production.

Histopathology: Glycosaminoglycan and fluids accumulate in the orbital tissue, so the orbital connective tissue volume increases.

Clinical Expression: Periorbital edema, exophthalmos, eyelid retraction, exposure keratopathy, strabismus, compression optic neuropathy which may lead to visual loss.

**Stages:** There are two stages in the development of the disease.

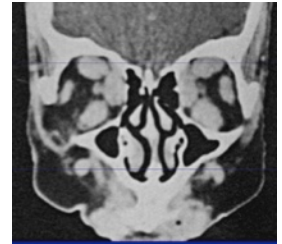
Congestive (active or inflammatory) stage, in which the eyes are red and painful.

[**NOTE**: remit within 3 years and only 10% of patients develop serious long-term ocular problems.]

Fibrotic stage in which the eyes are white, although a painless motility defect may be present.

#### **Treatment:**

- Systemic steroids may be used in rapidly progressive and painful proptosis during the active phase.
- Radiotherapy may be used along with steroids or when steroids are contraindicated or ineffective to suppress the inflammatory process.
- Optic nerve decompression may be indicated



#### Idiopathic Orbital Inflammatory Disease:

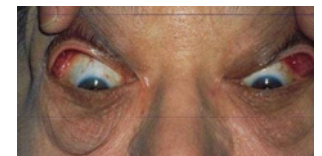
**Previously** referred to as orbital pseudotumour, is an uncommon disorder characterized by non-neoplastic, noninfectious, space-occupying, orbital lesions. The inflammatory process may involve any or all of the orbital soft tissues, resulting in, for example, myositis, optic perineuritis or scleritis.

#### **Diagnosis:**

1. Signs: Unilateral disease is the rule in adults, although in children bilateral involvement may occur, so if it presents bilateral or systemic in an adult think of vasculitis.

[**NOTE**: Pain, rapidly progressive, proptosis, and periorbital changes]

2. CT shows ill-defined orbital opacification and loss of definition of contents



**Treatment:** [**NOTES**: Observation, for relatively mild disease, in anticipation of spontaneous remission.

Biopsy may be required in persistent cases to confirm the diagnosis and to rule out neoplasia. Oral NSAID's are often effective and should precede steroid therapy. Systemic steroids should be administered only after the diagnosis has been confirmed as they may mask other pathology such as infection and Wegener granulomatosis.]

## ORBITAL DISORDERS

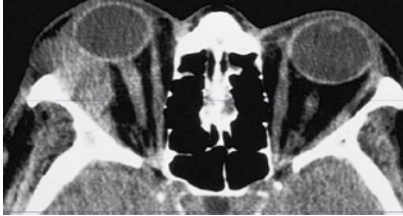
### MASSES AND CYSTS:

#### Lymphoid Hyperplasia and Lymphoma:

Lymphomas of the ocular and benign reactive lymphoid hyperplasia and together constitute 20% of all orbital masses.

##### **Diagnosis:**

1. Presentation: insidious and usually in old age.
2. Signs: Anterior lesions may be palpated, have a **rubbery consistency and a salmon patch appearance**. Any part of the orbit may be affected and 17% of the time involvement is bilateral. 50% of lymphoma are confined to the lacrimal fossa, sparing the orbit.



#### Plasma Cells Tumors:

#### Histocytic disorders:

- Macrophages based disorder.

#### Dermoid Cysts:

[**NOTE**: A dermoid cyst is a choristoma derived from displacement of ectoderm to a subcutaneous location along embryonic lines of closure. Dermoids are lined by keratinized stratified squamous epithelium (like skin), have a fibrous wall and contain dermal appendages such as sweat glands, sebaceous glands and hair follicles.]

##### **Diagnosis:**

1. Presentation: in infancy with a painless nodule most commonly **located** at suture lines between the frontal and zygomatic bone.
2. Signs: A firm, round, smooth, non-tender, mobile subcutaneous mass. The posterior margins are easily palpable denoting lack of deeper origin or extension.

[**NOTE**: Painless, slowly progressing palpable mass with no periorbital changes]

**Treatment**: is by excision. [**NOTE**: taking care not to rupture the lesion, since leaking of keratin into the surrounding tissue results in a severe foreign body reaction.]

#### Rhabdomyosarcoma IMP:

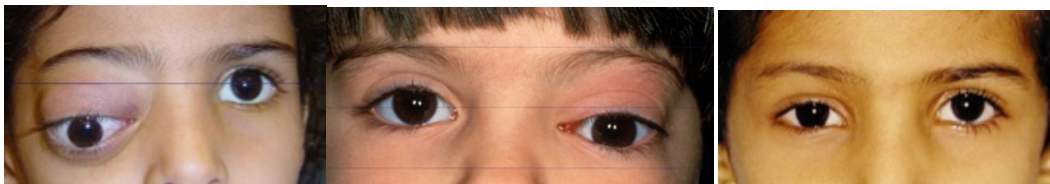
The most common primary orbital malignancy of childhood. [**EXTRA**: The tumor is derived from undifferentiated mesenchymal cell rests, which have the potential to differentiate into striated muscle.]

##### **Diagnosis:**

1. Presentation is in the first decade (average 7-8 years) with rapidly progressive unilateral proptosis.
2. MRI shows a poorly defined mass of homogeneous density, often with adjacent bony destruction.

**Treatment** involves radiotherapy and chemotherapy.

[**NOTE**: Rapidly progressing proptosis in child is an alarming sign and suggestive of malignancy!]



## FACIAL TRAUMA AND FRACTURES:

In facial or eye trauma you must first rule out globe injury and then make sure the orbital walls aren't fractured by asking the patient to move his/her eyes. Limitations of extraocular movement is suggestive of orbital wall fracture.

**TYPES OF FRACTURES:**

- Mid-Facial Fracture
- ZMC Fracture
- Wall and Floor Fracture
- Optic Canal Fractures

### Wall and Floor Fractures:

#### 1. Medial Wall (Lamina Papyracea) Fracture:

Most medial wall orbital fractures are associated with floor fractures.

##### **Diagnosis:**

1. Signs [**NOTES:** Periorbital haematoma and frequently subcutaneous emphysema, which typically develops on blowing the nose. Defective ocular motility involving abduction and adduction, if the medial rectus muscle is caught in the fracture]
2. CT scan will show the extent of damage.

**Treatment:** involves release of the entrapped tissue and repair of the bony defect.

[**EXTRA:** Because of the possibility of forcing infected sinus contents into the orbit, blowing of the nose should be discouraged]

#### 2. Lateral Wall and Orbital Roof Fractures:

Rarely encountered by ophthalmologists. The lateral wall of the orbit is more solid than the other walls, a fracture is usually associated with extensive facial damage.

#### 3. Orbital Floor Fractures:

A 'pure' blow-out fracture of the orbit does not involve the orbital rim, whereas an 'impure' fracture involves the orbital rim and adjacent facial bones.

A blow-out fracture of the orbital floor is typically caused by a striking object which is greater than 5 cm in diameter, such as a fist or tennis ball.

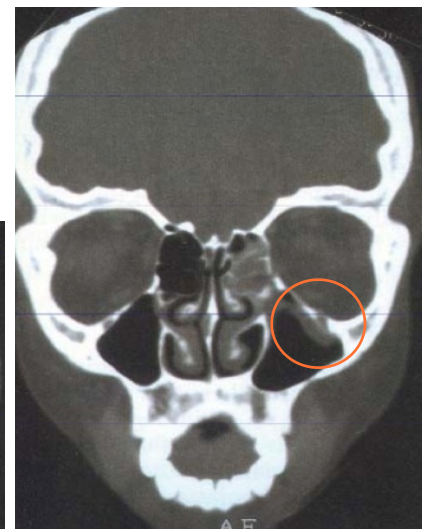
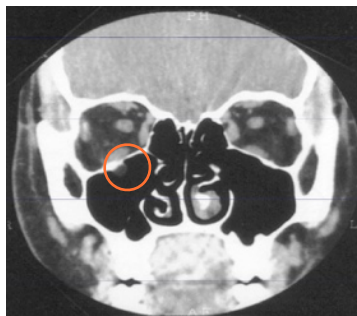


**Classic Signs of Blow-Out**  
Enophthalmos  
Decreased upgaze (IR trapped)  
Cheek anesthetized (infraorbital nerve trapped)

##### **Diagnosis:**

1. Presentations: Pain, diplopia
2. Signs: Restricted extra-ocular movement, and infraorbital anesthesia (CN V2), enophthalmos, periorbital ecchymoses
3. CT scan coronal view of orbits [**NOTE:** Shows maxillary soft-tissue densities]

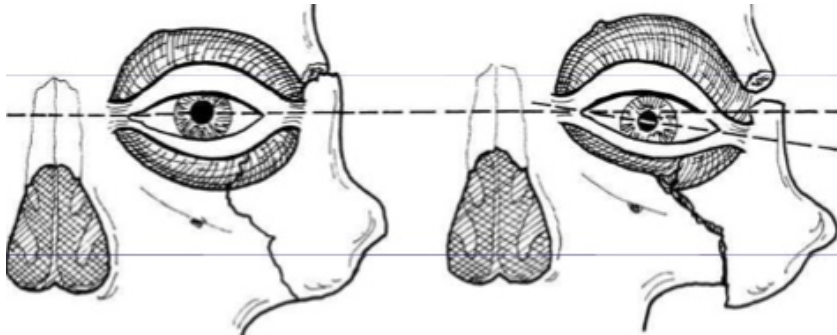
**Treatment:** surgical repair.





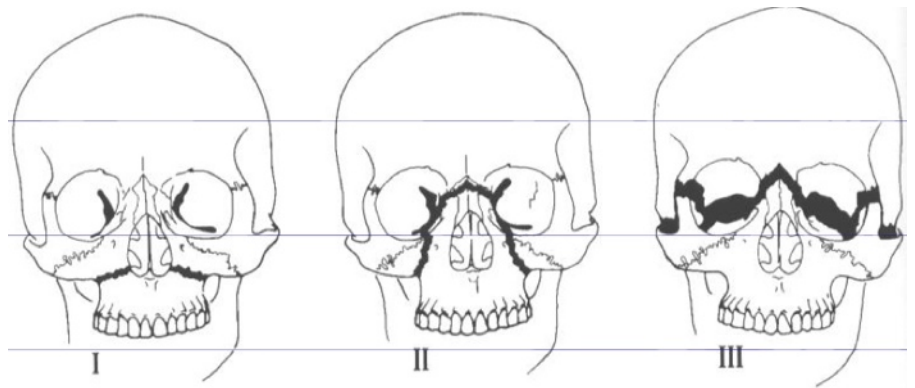
## FACIAL TRAUMA AND FRACTURES:

### Zygomaticomaxillary Complex Fractures:



### Le Fort fractures:

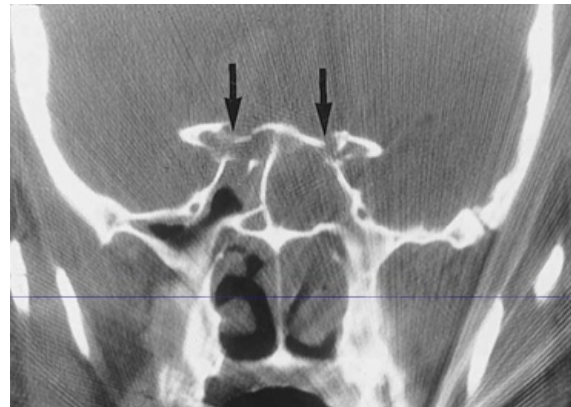
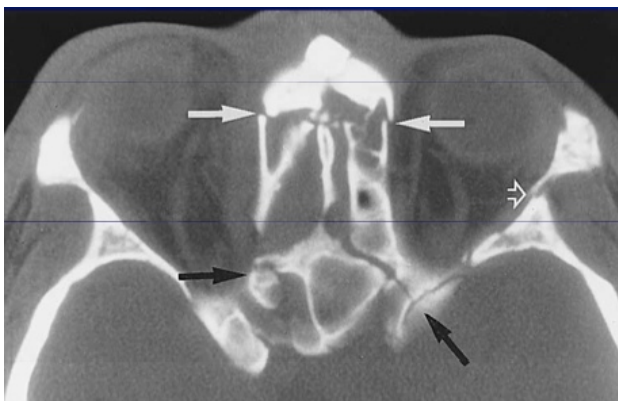
Le Fort fractures are types of facial fractures involving the maxillary bone and surrounding structures in a usually bilateral and either horizontal, pyramidal or transverse way.



### Optic Canal Fractures:

Presents with or without displaced bony fragments. This type of fractures causes traumatic optic neuropathy.

[**EXTRA:** Optic neuropathy presents as sudden visual loss secondary to either blunt or penetrating trauma to the orbit that cannot be explained by other ocular pathology. The damage to the optic nerve is either direct (haemorrhage or compression), shearing (acceleration of the nerve at the optic canal where it is tethered to the dural sheath) or transmission of a shock wave through the orbit.]



## IMAGING TECHNIQUES:

### Imaging Options:

- Plain films
- CT scan
- MRI
- Ultrasound

### Plain Films:

Infrequently used.

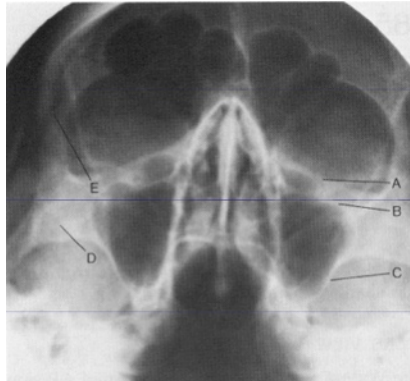
#### Advantages:

- Quick
- Used to rule out foreign bodies

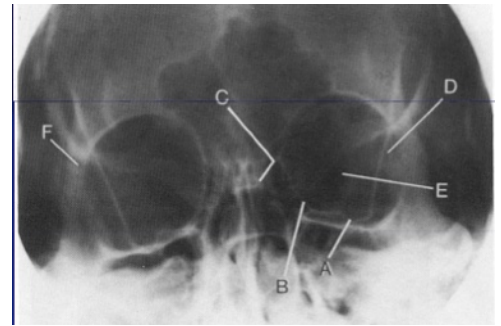
#### Views:



Base View



Waters' view

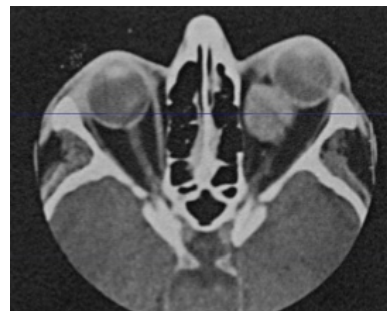


Caldwell's view

### CT-Scans:

#### Advantages:

- Spatial resolution
- Shows the bones
  - Fractures
  - Bone destruction
  - Calcification
- Takes only a few minutes
  - Used in emergencies trauma
- Relatively inexpensive



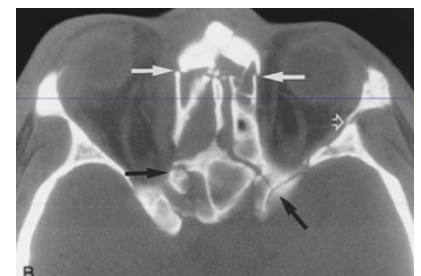
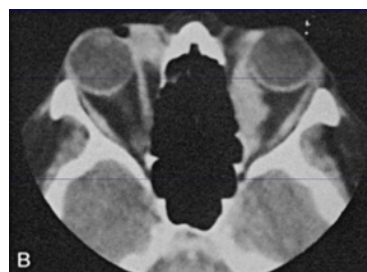
#### Disadvantages:

- Exposes the patient to ionizing radiation: 1-2 cGy
- Soft tissue definition
- Contrast iodinated
  - Contraindicated in allergic patients
- May need MRI anyway [Expensive]



#### Protocols:

- Axial and coronal
- With or without contrast





## IMAGING TECHNIQUES:

### MRI:

#### Advantages:

- Tissue:
  - T1 > anatomy
  - T2 > pathology
- No radiation

#### Disadvantages:

- Magnetic
  - Pacemakers, surgical clips
- Claustrophobia

#### Protocols:

- Axial, coronal or sagittal
- Gadolinium contrast [**ONLY in T1**]
  - Non-iodinated allergies RARE
- Orbital lesions
  - Fat suppression

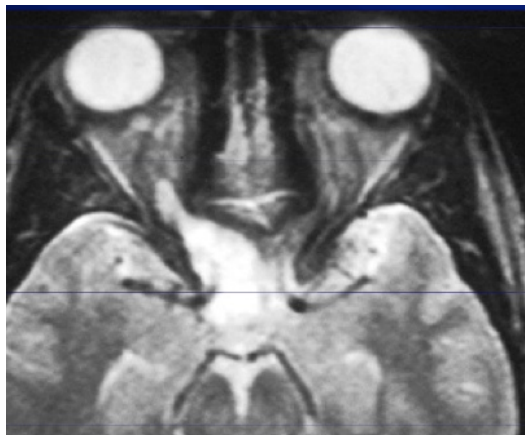
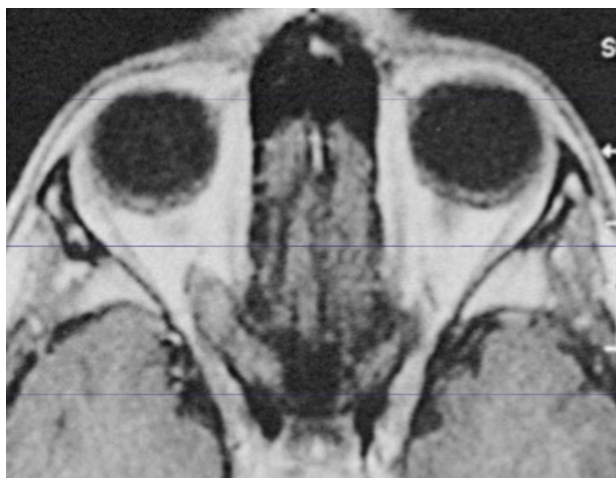
#### Description:

T1 or T2?

Axial, coronal or sagittal?

Contrast?

Lesion?



T2-weighted axial image - vitreous is hyperintense



T1-weighted axial image through the globes and optic nerves - vitreous is hypointense

### Orbital Echography:

- Dynamic
- Less expensive +/-
- Availability variable

