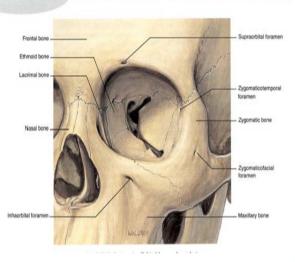
# <u>ORBITAL TUMORS &</u> <u>SURGICAL APPROACHES</u> <u>TO THE ORBIT</u>

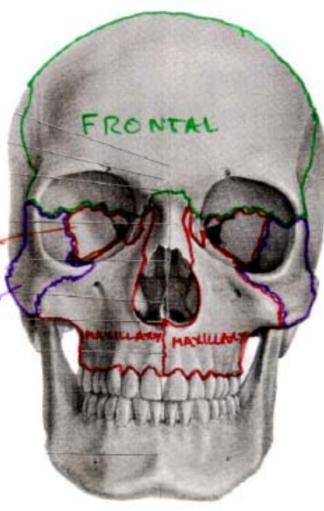
**Dr. Shailesh Jain** 

### I. BONES OF ORBIT



SPHENOID

#### ZYGOMATIC



A. BOUNDARIES 1. ROOF FRONTAL 4. LATERAL WALL ZYGOMATIC SPHENOID

2. FLOOR MAXILLARY

#### 1) OPTIC CANAL- IN BASE

OF LESSER WING OF SPHENOID BONE, CONTAINS OPTIC NERVE & OPHTHALMIC ARTERY

#### 2) SUPERIOR ORBITAL FISSURE BETWEEN GREATER AND LESSER WINGS OF SPHENOID, CONTAINS III, IV, V1, VI, OPTHALMIC VEINS

#### 1) <u>SUPRAORBITAL NOTCH OR</u> <u>FORAMEN</u>-IN FRONTAL BONE CONTAINS SUPRAORBITAL N., A. and V. FROM V1, OPTHALMIC A. and V.

#### 2) INFRAORBITAL FORAMEN- IN MAXILLARY BONE CONTAINS INFRAORBITAL N., A. and V. FROM V2 & MAXILLARY

#### 3) <u>ZYGOMATICO- FACIAL</u> <u>FORAMEN</u>- IN ZYGOMATIC BONE-ZYGOMATICO-ORB. N. ,V2,

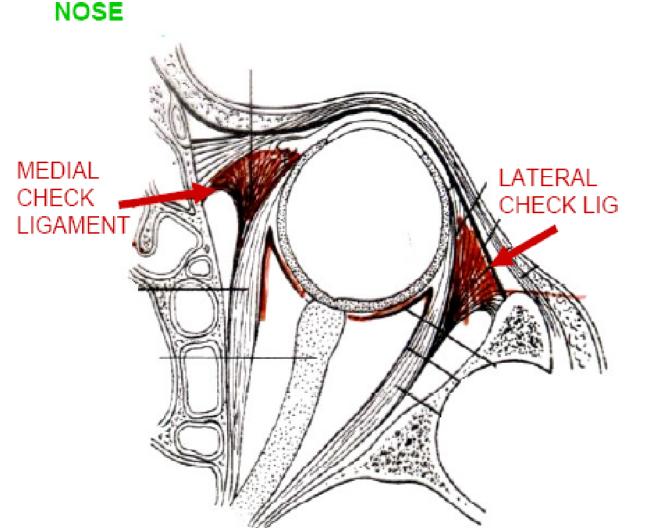
#### 1) INFERIOR ORBITAL FISSURE- SLIT BELOW SUP. ORBITAL FISSURE: BETWEEN SPHENOID & MAXILLARY - CONNECT PTERYGOPALATINE FOSSA AND INFRATEMPORAL FOSSA WITH ORBIT - CONTAINS INFRAORBITAL AND ZYGOMATIC N., A. AND V. (V2, MAXILLARY A.)

#### 2) ANT. & POST. ETHMOIDAL

FORAMINA- BETWEEN ETHMOID & FRONTAL BONES -CONNECT ORBIT & NASAL CAVITIES - CONTAIN ANT & POST ETHMOIDAL N., A. & V. (V1 & OPHTHALIC A.) OPENING OF <u>3) NASOLACRIMAL</u> <u>DUCT</u>- IN MAXILLARY, LACRIMAL BONES & INF. NASAL CONCHA -CONTAINS MEMBRANEOUS NASOLACRIMAL DUCT & TEARS

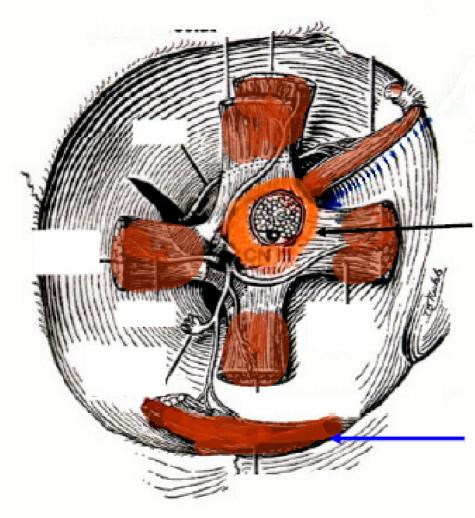
### **OPTIC NERVE SHEATH & FASCIAS**

- The orbit is lined by a periosteal layer, the so called periorbita, which is loosely adherent to the bone.
- Anteriorly, the periorbita blends with the periosteum of the orbital margin and to the orbital septum, while in the posterior region it fuses with the dural sheath of the optic nerve to form a unique sheet, the dura of the optic canal, which is tightly adherent to the bone.
- The orbital septum separates the extraorbital preseptal space from the postseptal orbital space.



= TENON'S CAPSULE -THIN MEMBRANE SURROUNDS BACK OF EYE-THICKENINGS -MEDIAL & LATERAL CHECK LIGAMENTS -PREVENT EXCESSIVE ROTATION

### A. ORIGINS OF EXTRAOCULAR MUSCLES



VIEW OF ENUCLEATED ORBIT- EYEBALL REMOVED; MOST MUSCLES TAKE ORIGIN FROM

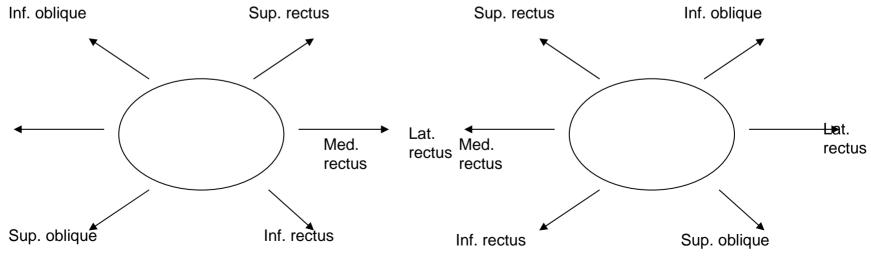
TENDINOUS RING- RING OF CT SURROUNDING OPTIC CANAL & SUPERIOR ORBITAL FISSURE

NOTE: NOT INFERIOR OBLIQUE-FROM FLOOR OF ORBIT

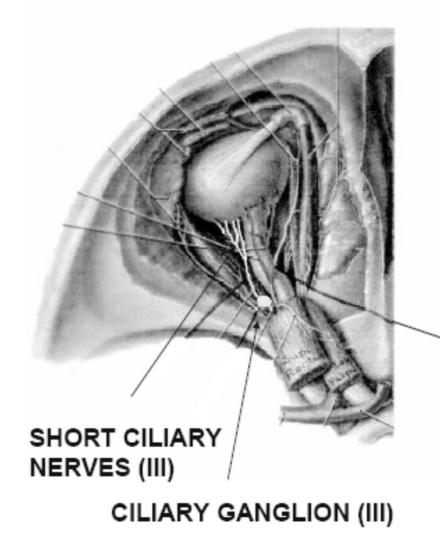
#### - Extraocular muscles

-levator palpebrae superioris muscle – elevates the upper eyelid.

- superior rectus muscle elevates and adducts the eyeball.
- medial rectus muscle adducts the eyeball.
- inferior rectus muscle depresses and adducts the eyeball.
- inferior oblique muscle elevates and abducts the eyeball.
- superior oblique muscle depresses and abducts the eyeball.
- lateral rectus muscle abducts the eyeball.



### VII. CILIARY GANGLION



CILIARY GANGLION-PARASYMPATHETICS OF OCULOMOTOR N (III) INN 1) CILIARY MUSCLES 2) SPHINCTER (CONSTRICTOR) PUPILLAE TRAVEL IN SHORT CILIARY NERVES CONTAIN PARASYMPATHETICS

NOTE: LONG CILIARY NERVES BRANCHES OF V1 (OPTHALMIC) - SENSORY TO CORNEA

- The ciliary ganglion is situated on the inferolateral aspect of the optic nerve and on the medial side of the lateral rectus muscle.
- It receives three branches: the motor (parasympathetic) root from the inferior division of the oculomotor nerve, the sensory root from the nasociliary nerve, and sympathetic fibers from the plexus around the ICA.
- The sympathetic fibers arise in the cervical sympathetic ganglia and pass through the ciliary ganglion without synapsing.
- The short ciliary nerves pass from the ganglion to the globe.

- The arterial supply to the eyeball, orbit, and eyelid comes primarily from the **ophthalmic artery**, which is a branch of **internal carotid artery**.

-The **central retinal artery**, first branch of the ophthalmic artery pierces the optic nerve. It is the only blood supply to the retina.

- The **supraorbital** and **supratrochlear arteries** also branch off the ophthalmic artery, supplying the muscles, skin, and fascia of the forehead.

- The **anterior** and **posterior ethmoidal arteries** supply the ethmoid and frontal sinuses and the nasal mucosa.

- The **lacrimal artery** carries blood to the lacrimal gland and eyelids laterally.

-The **superior ophthalmic vein** drains the eyeball, the superior portion of the orbit, the ethmoid sinuses, and the forehead into the **cavernous sinus**.

- Other veins that drain the orbit are the supratrochlear, supraorbital, angular, and even the maxillary and retromandibular veins.

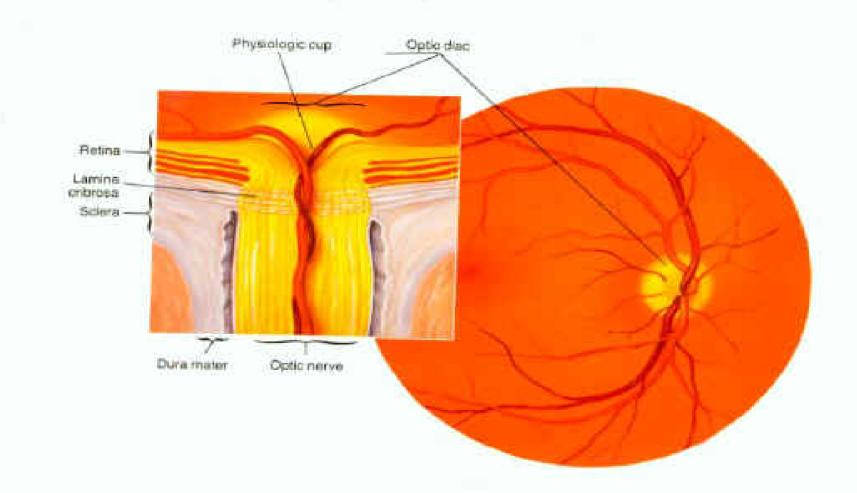
- Orbital tumors arising near the apex of the orbit often compresses the SOV leading to orbital venous congestion.
- Tumors in the anterior part of the middle cranial fossa may compress the exiting ophthalmic veins leading to a mild proptosis.
- Development of a CCF results in the transmission of the arterial pressure into the SOV leading to chemosis & exophthalmos.

# Spaces of the Retrobulbar Orbit

- Intraconal space:
  - Contains CN II, ophthalmic artery, superior division of CN III, nasociliary nerve (V1), inferior division of CN III, and CN VI.
- Extraconal space:
  - Contains ophthalmic vein, lacrimal nerve (V1), CN IV and frontal nerve (V1).

## Optic nerve

- Consists of 1 million axons that arise from the retina.
- Leaves the eye through the sclera optic canal into the cranium.



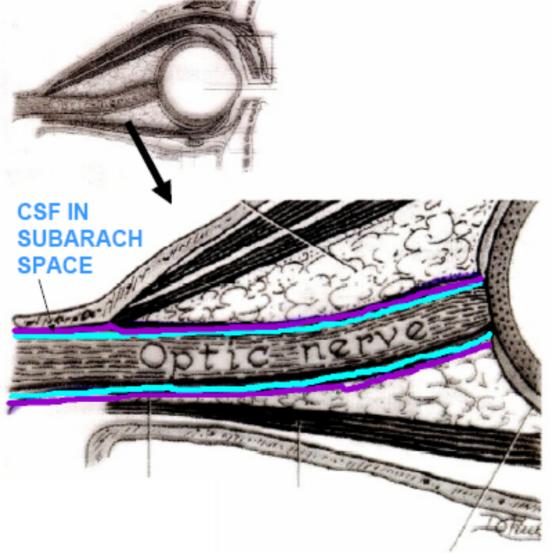
### **OPTIC NERVE**

- The optic nerve can be divided into an intraocular, an intraorbital, a canalicular, and an intracranial segment.
- The intraorbital optic nerve has a length of about 4–4.5 cm, and its course is slightly tortuous.
- The entire intraorbital optic nerve is embedded in meningeal and arachnoidal sheaths in continuation with the respective intracranial layers.

### Cont....

- In the orbit, this sheath divides into two layers, one of which remains the outer sheath, blending with the sclera of the eyeball, the other becoming the orbital periosteum (periorbita).
- A tube-shaped subarachnoid space extending between the intermediate and the inner arachnoid layer communicates with the intracranial subarachnoid space.

#### SUBARACHNOID SPACE EXTENDS TO BACK OF EYE



NOTE:

DURA & SUBARACHNOID SPACE (CSF) EXTEND AROUND OPTIC NERVE; CSF CAN EFFECT VISION; DIAGNOSE BY EYE EXAM

### Cont....

- The length of the optic canal is about 5 mm, its width about 3–4 mm,
- The length of the intracranial prechiasmatic optic nerve is about 10 mm.
- The intradural segment of the ICA starts a little more proximally than the origin of the ophthalmic artery.
- At the intracranial end of the optic canal, the ophthalmic artery is located inferolaterally to the optic nerve inside the subarachnoid space but at the orbital end of the optic canal, the ophthalmic artery exits the dura laterally.

# **Classification on basis of Origin**

Orbital tumors can be classified based on origin:

1) Primary lesions, which originate from the orbit itself;

2) Secondary lesions, which extend to the orbit from neighboring structures and include such lesions as intracranial tumors and tumors of the paranasal sinuses that, by contiguity, extend to involve the orbit; and

3) Metastatic tumors, lesions reach the orbit via hematogenous or lymphatic spread.

## **Classification (Histology basis)**

- A. Benign - Malignant
- B Epithelial: Cylindrinoma, adenocarcinoma, carcinoma (secondary)
  - Mesenchymal: fibroma, lipoma, rhabdomyoma, osteoma, sarcoma
  - Vascular: hemangioma, lymphangioma
  - Neurogenic: glioma, schwannoma, neurofibroma, meningioma
  - Lymphatic and hematogenic: lymphoma, lymphosarcoma, lymphoid hamartoma, plasmocytoma
  - Developmental: teratoma, dermoid cyst
  - Pigment: melanoblastoma

# **Classification (Surgery basis)**

- 1) Those that are wholly with in the orbit not involving the apex,
- 2) Those that involve the orbital apex,
- 3) Those that have significant intracranial extension, &
- 4) Those that have significant extension into the paranasal sinuses.

## **CLINICAL FEATURES**

- INTRACONAL TUMORS :
  - Early visual loss
  - Impairment of mobility
  - Axial Proptosis (Anterior Displacement of globe)
- <u>EXTRACONAL TUMORS</u> :

- Early nonaxial proptosis

 INTRACANALICULAR / APICAL TUMORS : - Early painful loss of vision (accompanied by optic disk edema)

## <u>PROPTOSIS</u>

- Frequently painless proptosis.
- Painful invasion of the orbital walls,
  - involvement of the trigeminal n.
  - Inflammatory, Traumatic or H'agic
  - Exposure keratitis (late stage)
  - + in orbital apex tumor & tumor of cavernous

sinus

• Proptosis is usually of insidious onset except in c/o inflammatory process or CCF.

### **Clinical Presentation**

- Bilateral globe protrusion is more likely in c/o systemic disease such as dysthyroid orbitopathy, as c.f. usual unilateral orbital tumors involvement (except occasionally lymphoma, histiocytoma, or metastatic lesions)
- Distinguish from Pseudoexophthalmos by Grave's d's(I/L), Microphthalmos(C/L) & C/L enophthalmos caused by blow-out orbital #, inflammatory processes

- Vision loss :
- <u>Diplopia</u> :
- <u>Pupillary Size</u> : tumors invade or compress the parasympathetic (third nerve through the ciliary ganglion) or the sympathetic fibers innervating the pupillary dilators e.g. chemodectomas (paragangliomas),
- <u>**Bruits</u>** : Associated with arteriovenous fistulas (more often traumatic).</u>

<u>AGE</u>	<u>COMMON TUMORS</u>
Infants & Neonates	Teratomas, Dermoid cysts and Capillary hemangiomas
Older Childhood	Lymphangiomas, Optic gliomas such as pilocytic astrocytomas
Adults	Lacrimal gland tumors, Metastases, Cavernous hemangiomas, and Neurilemmomas

# Cavernous Hemangioma

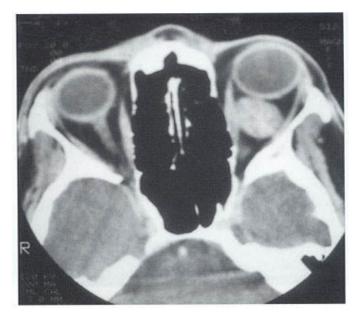
- Cavernous hemangiomas consist of large ectatic (cavernous) spaces lined by flattened endothelial cells and surrounded by a capsule of fibrous tissue.
- Also known as encapsulated venous malformation (good plane of cleavage from adjacent structures)
- Most common vascular lesion & MC primary benign orbital tumor in adults.
- Cavernous hemangiomas do not have a prominent arterial supply or show infiltrative expansion, in contrast to capillary hemangiomas of childhood.
- Cavernous hemangiomas are isolated from the orbital vascular system and thus from the systemic circulation.

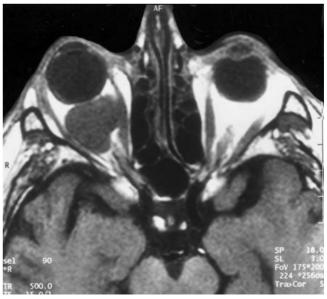
- account for 5%–7% of all orbital tumors.
- Peak incidence middle age (40 years)
- Women > men
- Most tumors are located within intraconal space lateral to the optic nerve
- Slowly progressive painless proptosis occasionally with a mild visual deficit or obscurations
- do not enlarge with Valsalva (d/d orbital varix)

## **Cavernous Hemangioma**

- CT/MRI reveals well-defined oval mass,
- CT : Homogeneous with increased density with variable enhancement on contrast

- MRI isointense to muscle on T1 & hypo on T2 with inhomogenous enhancement
- Calcifications correspond to phleboliths, may be regarded as a pathognomic sign.







• The differential diagnosis should include hemangiopericytoma, fibrous histiocytoma, and neurinoma.

#### • <u>Treatment</u> :

- Periodic observation -- cases where no symptoms are present and the tumor was detected incidentally,
- Complete surgical removal -- when the patient is symptomatic, or the lesion is growing.
- Shrinkage of the tumor by selective coagulation or aspiration may facilitate removal.
- The visual prognosis is excellent in most cases;
- Malignant transformation or local recurrence has not been demonstrated.

# Orbital Meningioma

- 1-2% of all meningiomas
- B/w 5-20% of orbital tumors are meningiomas
- Optic nerve sheath meningiomas account for 10%–33% of orbital meningiomas
- Can be purely intraorbital or with in the sphenoid wing or periorbita with secondary orbital involvement

### Periorbital meningioma

- Arise from the superolateral quadrant of the orbit, usually the SOF,
- May be in continuity with a sphenoid wing meningioma
- More aggressive in children & can extend intracranially via the SOF & to the skull base via the IOF, making total resection difficult

## Optic nerve sheath Meningioma

- Most commonly arise intracranially – Secondarily invade orbit
- 80% of <u>primary</u> intraorbital meningiomas arise from optic n. sheath
- They usually wrap circumferentially around the optic nerve and extend posteriorly into the annulus of Zinn,
- F >> M (middle age)

# Orbital Meningioma

- Exopthalmos (Extradural Tx) 83%
- Decreased VA (Subdural Tx) 38%
- Headache 36
- Diplopia
- Ptosis

36% 21%

20%

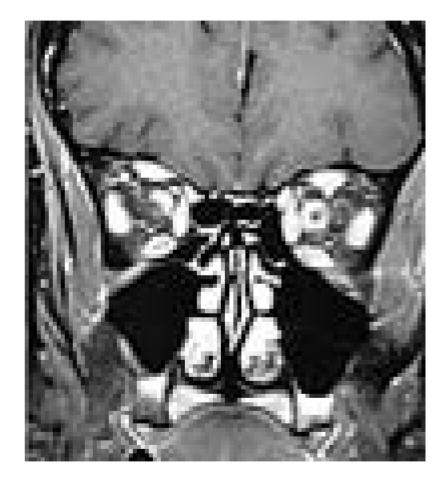
• **Optociliary venous shunts** : preexisting optic disk vessels that enlarge to compensate for chronic retinal venous congestion caused by CRVO --- strongly s/o optic nerve meningioma

- Two histological subtypes
  - Meningotheliomatous
  - Psammomatous
- ONSMs appear as a diffuse fusiform or tubular enlargement of the optic nerve.
- CT may demonstrate calcification of the nerve sheath or enlargement of the optic canal
- Axial CT scans reveal "tram tracking," a sign caused by the density difference between the thicker nerve sheath tumor and the residual optic nerve (central lucency)









#### Treatment Algorithm for Orbital Meningiomas

Location	Visual Acuity	Treatment
Mid to anterior	Intact	Observe
Mid to anterior	Loss	Resection
Apical	Intact	Observe
Apical	Progressive loss	Resection/ XRT
Apical	No light perception (or large tumor or IC extension)	Craniotomy

Note: decision for surgery depends on visual status and presence of intracranial extension

### **Optic nerve Gliomas**

- 3<sup>rd</sup> most common in children
- Typically occurs in the first decade of life
- Histologically are (juvenile) pilocytic astrocytoma,
- Benign, slow-growing tumors but a malignant variant occurs in adults (Anaplastic astrocytomas / Glioblastomas)
- 66% of all primary optic nerve tumors,
- 4% of all orbital space-occupying lesions.

- More common than optic nerve sheath meningiomas (4:1) and have a female preponderance
- May occur randomly although often associated with NF type I (up to 50%)
- Bilateral disease pathognomonic for NF1
- Axial proptosis : 94%
- Visual field loss : 63%
- Optic nerve atrophy : 59%

### Optic pathway gliomas classification

Based on their specific location :

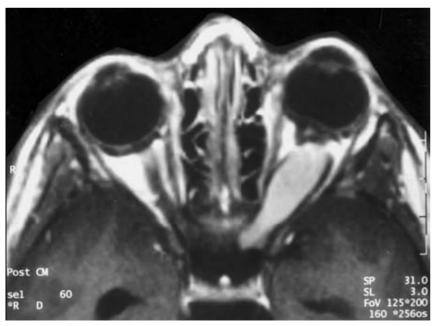
- Type I-A is retrobulbar (intra-orbital);
- Type I-B is retrobulbar (intra-foramina);
- Type II the glioma is in optic tract;
- Type III-A is chiasmal and unilateral;
- Type III-B is chiasmal and bilateral.

The type III (A and B) can involve the hypothalamus.

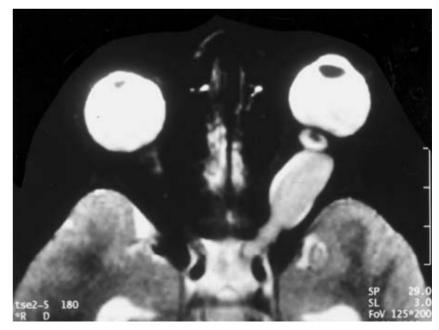
### Imaging

- An enlargement of the optic nerve without calcification, as tubular, fusiform ,or lobulated
- Classically a J-shaped sella;
- Optic foramina views showing an optic foramen > 7.0 mm or a difference of more than 2.0 mm between the left and the right foramina.
- T2WI demonstrate homogeneous high signal intensity of the affected nerve in contrast to the low signal of the C/L unaffected optic nerve









#### Management

- Conservative management can be adopted due to the fact that only 21% of these patients show tumor progression.
- Surgery should be restricted to patients with severe proptosis, a blind painful eye, or when extension along the intracranial portion of the optic nerve threatens the chiasm.
- Surgery involves resection of the optic nerve from the globe to within 2mm of the chiasm through a FT craniotomy.

- <u>Chiasmatic involvement</u>: needle biopsy of the orbital component is occasionally used to confirm the diagnosis if progressive visual loss or significant growth occurs.
- EBRT (50 Gy) is the tumour of choice for chiasmal gliomas.
- Radiotherapy does not appear to alter the ultimate prognosis even in cases where tumor involvement includes the hypothalamus and third ventricle (mortality rate: 28%), since CNS complications of radiotherapy, especially in very young children, might be severe.

- Malignant gliomas (Adults) : lethal <1yr
- FNAB of the orbital component or incisional biopsy by lateral orbitotomy to make the diagnosis
- RT after tissue diagnosis.
- Role of surgical debulking or Chemotherapy is unclear.
- Long-term survival rate and prognosis are excellent when the tumor is confined to the optic nerve.

### Peripheral nerve tumors

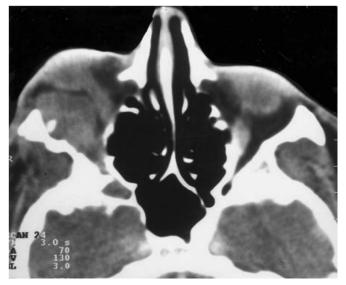
• Constitute 5 - 15 % of the orbital tumors.

• 5 types :

Solitary neurofibroma Diffuse neurofibroma Plexiform neurofibroma Schwannomas Malignant peripheral nerve tumors

### Solitary neurofibroma

- B/w  $3^{rd} 4^{th}$  decade
- Slowly progressive, painless proptosis with minimal or no visual dysfunction
- Typically located in the superolateral orbital quadrant
- Isointense to brain & muscle on T1WI & hyperintense to fat onT2WI with heterogenous enhancement
- Pseudocapsule : easy to dissect
- Prognosis is good
- No need for postoperative RT.



# Plexiform neurofibroma

- Associated with NF
- Occur mostly in infants & children
- A palpable mass in the eyelid (usually lateral third) with subsequent ptosis & lid hypertrophy
- May spread to forehead or adjacent areas of temple



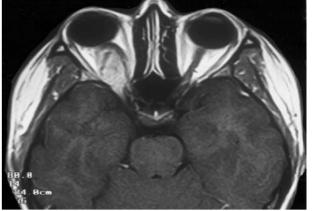
### Diffuse neurofibromas

- Similar to plexiform but more extensive : permeate the orbital tissues, fat, & muscles
- Less frequently associated with NF
- Imaging : Infilterative with irregular borders & enhance with contrast
- Do not have a good dissection plane, making total excision almost impossible & recurrence frequent
- Goal of Sx : Debulk the tumor while preserving normal orbital structures

#### <u>Schwannomas</u>

#### (Neurilemmomas, Neurinoma)

•  $2^{nd} - 5^{th}$  decade, F>M



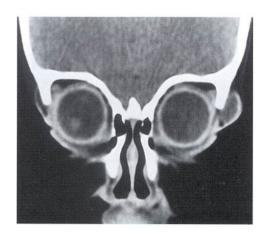
- Usually originate from sensory br. of the trigeminal nerve
- High incidence in patients with NF 2.
- Well encapsulated
- C/f : Proptosis, dysmotility, trigeminal distribution of pain & numbness
- T1WI : Iso- to hypointense signal in relation to the orbital fat with a varying degree of contrast enhancement
- Malignant transformation is rare.

# Pediatric Orbital Tumors

- Differs substantially from adult types
- More often congenital lesions and infectious
- Most common cystic lesions (Dermoids)
- 2<sup>nd</sup> most common vascular lesions
- Most common malignancy rhabdomyosarcoma

# Cystic Lesions – Dermoid Cyst

- 4 6% of orbital tumors
- Most common
- Preschool child
- Superotemporal mass
- Mobile and nontender
- Well circumscribed on CT with rare bony remodeling



### Cystic Lesions – Dermoid Cyst

- Deeper lesions usually show bony abnormality
- May present with proptosis and visual c/o
- Surgical excision at around 1 year of age
- Total excision must be achieved to prevent recurrence

# **Capillary Hemangioma**

- 1/3 diagnosed at birth
- 90% visible by 6 months
- Most common presenting as superficial tumor that develops "strawberry" appearance
- Enlarge with Valsalva
- MC site : Superomedial extraconal quadrant of the orbit,

• CT/MRI show diffusely infiltrating non-encapsulated mass



## **Capillary Hemangioma**

• Usual course

Normal at birth – noticed at one month – enlarge till 1 to 2 years of age – spontaneous involution by age 4 to 8 yr

• Complications – amblyopia or astigmatism, superinfection, ulceration, Kasabach-Merrit

# <u>Capillary Hemangioma -</u> <u>Treatment</u>

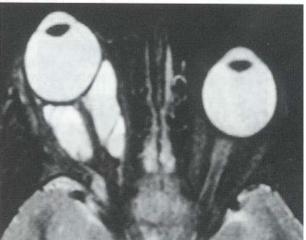
- Indications include any complication
- Medical therapy Steroids (systemic, intralesional) or interferon
- Radiation therapy
- Surgical resection for unresponsive or wellencapsulated lesions

#### Lymphangioma

- Benign congenital venous lymphatic malformations
- Usually identified prior to teenage years
- No spontaneous regression
- Sudden proptosis from hemorrhage into cyst
- No enlargement with Valsalva
- Primarily as an extraconal lesion

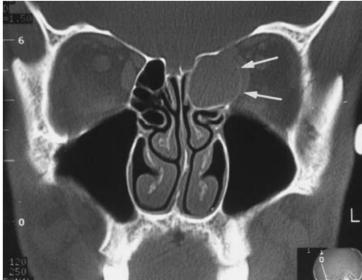
## **Imaging**

- CT/MRI shows multi-compartmental lesion
- "Chocolate-cyst": on MRI may show an inhomogeneous signal due to thrombosis and hemorrhages
- Treatment for significant proptosis, corneal exposure or optic nerve compression
- Debulking and cyst drainage usually
- Complete removal often not possible



#### <u>Mucoceles</u>

- MC result from inflammatory obstruction of the ostium of the affected sinus(primary mucocele).
- Secondary mucoceles are posttraumatic, postoperative, or are seen in neoplastic disorders of the nasal sinuses.
- On CT, a hypointense, expanding mass, originating from a paranasal sinus (*most frequently in the frontoethmoidal sinuses*), characterized by sharp, and thinned remodeling of the bony wall



#### <u>Rhabdomyosarcoma</u>

- Most common malignant tumor in children
- 90% of cases occur before age 16.
- Rapid unilateral proptosis and globe displacement
- Early diagnosis to preserve visual function & to improve the outcome
- CT scan shows irregular margins and often bony destruction
- FNAB / Excisional biopsy for diagnosis if suspected
- Chemotherapy and XRT after biopsy with minimal or no residual (90% 5-yr for localized disease)

# Fibrous Dysplasia

- Most often fibro-osseous tumor
- Occurs in 1<sup>st</sup> two decades
- Replacement of normal bone with immature woven bone
- Polyostotic (Albright's) and monostotic types
- Usually stabilize after puberty
- Conservative treatment is the rule
- Complete resection preferable for significant cosmetic deformity or vision loss

### Metastatic Tumor: Neuroblastoma

- Most frequent in children
- Neuroblastoma accounts for 10% of all childhood malignancies
- Primary: usually adrenal
- Bilateral metastasis with eyelid ecchymoses and proptosis common (50%)
- Survival rate 15%

### Metastatic Tumors

- 8% of all orbital tumors
- Most common in women breast
- Most common in men prostate & lung
- Symptoms proptosis, diplopia, pain, vision loss
- Presents in 7<sup>th</sup> decade
- FNAB for diagnosis (80%)
- Prognosis is very poor (avg. survival 10 months)
- XRT usual; Chemo and Hormonal occasional

# Surgical approaches to orbit

Divided into 2 groups :

- A) Extracranial approach :
- Lateral orbitotomy
- Anterior orbitotomy
- Anterior medial orbitotomy
- Transethmoidal
  orbitotomy
- Inferior orbitotomy

#### B) Transcranial approach :

- Fronto-orbital
- Fronto temporo-orbital
- Subfrontal
- Pterional

INTRACRANIAL APPROACHES	REGION OPTIMALLY EXPOSED
Subfrontal	Superior & medial orbit
Fronto-orbital	Superior & medial orbit, orbital apex
Pterional	SOF, OC, Apex, Intracranial
Fronto-orbito temporal	Superior & medial orbit, SOF, OC, Apex, Intracranial

EXTRACRANIAL APPROACHES	REGION OPTIMALLY EXPOSED
Lateral	Lateral orbit (Anterior 2/3 <sup>rd</sup> )
Anterior	Anterior half extraconal
Anterior medial	Anterior medial intraconal, Inferior extraconal
Transethmiodal	Medial orbit & optic canal

# Anterior orbitotomy

- Useful for lesions of the anterior two thirds of the orbit.
- Devised by Knapp in 1874 and popularized by Benedict.
- A misnomer because bone removal is often not required.
- Mainly for incision biopsy, if FNAB fails

#### Anterior orbitotomy

- With osteotomy of the superior orbital rim : for larger lesions
- Without osteotomy :

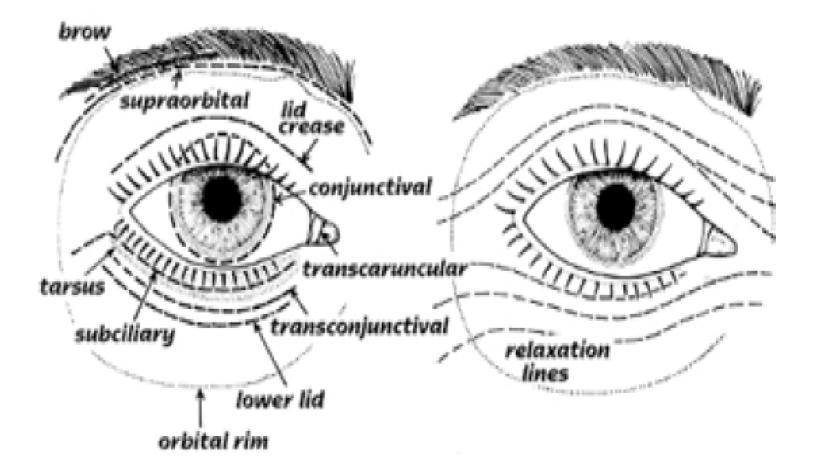
Superior mass :

Eyelid Supraorbital, or Subbrow incision

Inferior mass :

Transconjuctival Subciliary, or Lower eyelid incision

### Incisions for orbital approaches



#### Superior Approach With Superior Osteotomy.

- For Larger extraconal superior orbital lesions
- Incision : a subbrow or a supraorbital with its horizontal extent at least 3 cm.
- The supratrochlear and supraorbital neurovascular bundles are preserved.
- The superior orbital rim is removed which is replaced and secured with titanium miniplates after tumor removal

### Anterior transconjuctival orbitotomy

- Medial conjuctival peritomy done
- MR muscle is detached from the globe
- Orbital fat is retracted from the anterior optic nerve
- ONS fenestration : posterior ciliary N & vessels dissected from the medial surface of the nerve sheath f/b fenestration to allow free egress of the CSF
- MR is reattached to the globe & conjuctiva closed

### Trans-septal anterior orbitotomy

- Orbit being entered via the orbital septum of the upper or lower eyelid
- Upper eyelid : a superior lid crease incision
- Lower eyelid : a direct incision or a subciliary incision
- Skin & muscle flap is elevated to expose the orbital septum, which is then opened.

#### Anterior medial orbitotomy

- The medial approach was described in 1973 by Galbraith and Sullivan to decompress the optic nerve and relieve papilledema.
- Indicated for biopsy, decompression or removal of tumors of the medial intraconal space (area b/w the optic nerve & the MR muscle) such as cavernous hemangiomas, schwannomas, hemangiopericytomas, and isolated neurofibromas.
- This procedure may be used in conjunction with a sinus procedure in cases in which both regions are involved.

# Lateral orbitotomy

- The lateral approach was first proposed by Kronlein in 1889 for a large orbital & temporal fossa dermoid cyst.
- Recently standardized in 1976 by Maroon & Kennerdell.
- The lateral orbitotomy is useful for retrobulbar lesions, and it can be extended for more posterior lesions.
- The procedure involves temporary removal of the lateral wall of the orbit to gain access to the entire lacrimal gland and lateral, superolateral, and inferolateral tumors e.g. pleomorphic adenomas, cavernous hemangiomas.

### Medial Lateral Orbitotomy

- Large or posteriorly located medial orbital tumors may be resected by combined lateral-medial orbital approach.
- Large, primarily medial intraconal tumors are ideally suited for this approach, particularly large neurofibromas or hemangiomas located deep in the medial orbital compartment.
- The lateral approach is identical except the lateral incision is made directly through the canthus, 3 cm posteriorly
- This is done to allow the globe to be prolapsed into the bone defect laterally so that more room can be obtained medially to extract the tumor

- Orbital Exenteration : for tumors of conjunctiva (SCC / Melanoma) or eyelids (BCC / SCC / Melanoma)
- Orbital Decompression : for thyroid orbitopathy usually with optic neuropathy or extreme proptosis with corneal exposure / inoperable tumors

#### Transcranial approaches

- First report published in 1922 by Dandy
- For tumors located in the orbital apex and/or optic canal, or involving both the orbit and adjacent intracranial areas
- Divided into two types based on whether the orbital rim is or is not elevated in exposing the orbital lesion
- Small frontal or fronto temporal craniotomy, with removal of the orbital roof and/or lateral wall : for limited lesions
- Orbitofrontal or orbitozygomatic approach, with elevation of the orbital rim with the bone flap : for larger lesions

### **Orbitofrontal Craniotomy**

- For lesions involving the optic canal and orbital apex.
- Bicoronal incision
- Keyhole site burr hole which expose periorbita on its lower edge and frontal dura on its upper edge.
- The orbitofrontal bone flap includes the superior rim of the orbit and part of the orbital roof.
- The thin part of the roof of the orbit behind the orbital rim is opened to prevent the fracture across the orbital roof

- **One-piece exposure**: superior rim is elevated with the bone flap,
- **Two-piece exposure** : small frontal bone flap above the supraorbital rim is elevated as the first piece and the superior rim is removed as the second piece.
- In one-piece approach, chances of # of the last segment of the orbital roof between the medial and the lateral margins of the cuts in the roof, which may extend into the ethmoid air cells or cribriform plate.
- Three routes through an orbitofrontal craniotomy can be taken to the orbital contents: medial, lateral, and central.

### The Medial Orbitofrontal Approach

- Directed through the space between the SO muscle, which is retracted medially, and the levator and SR muscles, both of which are retracted laterally.
- It exposes the optic nerve throughout the interval from the globe to the optic canal.
- It is the most direct surgical approach to the apical part of the optic nerve.
- MC selected for tumors of the optic sheath or optic nerve.
- Not suitable for lesions located on the lateral side of the optic nerve or for those involving the SOF and the cavernous sinus.

## The Lateral Orbitofrontal Approach

- Optic nerve is approached between the LR muscle, which is retracted laterally, and the SR and levator muscles, both of which are retracted medially.
- The lateral approach provides a wider working space than the medial or central approach.
- The best of the three orbitofrontal routes for exposing the deep apical area on the lateral side of the optic nerve.
- Combined with an orbitozygomatic craniotomy, it is suitable for lesions along the anterior clinoid process and sphenoid ridge and middle fossa which extend through the superior orbital fissure into the orbit on the lateral side of the optic nerve.

- Two variants of the lateral approach : depends on whether the superior ophthalmic vein is retracted medially or laterally.
- On medial retraction, access to the deep apical area is limited because the superior ophthalmic vein blocks the view.
- On lateral retraction, the orbital septum must be opened, thus risking damage to the cranial nerves that pass through the superior orbital fissure

### The Central Orbitofrontal Approach

- The levator muscle is retracted medially and the SR muscle is retracted laterally
- The least used of the three approaches
- Most direct and shortest way to the midportion of the intraorbital segment of the optic nerve.
- Two variants : choice depends on whether the frontal nerve is retracted medially with the levator muscle or laterally with the superior rectus muscle.
- The approach in which the frontal nerve is retracted medially carries less risk of damaging the frontal nerve
- When frontal nerve is retracted laterally, it provides a wider exposure of the orbital apex

### Orbitozygomatic approach

- A variant of the frontotemporal craniotomy in which a variable amount of the upper and lateral orbital rim and zygomatic arch are elevated as a single piece in continuity with the bone flap or as a second step after elevation of a fronto temporal (pterional) bone flap.
- Provides an excellent exposure of the cavernous sinus and orbital contents and the structures passing through the optic canal and superior orbital fissure

- **One-piece orbitozygomatic approach** :part of the upper and lateral orbital rim, and possibly the zygomatic arch with the pterional flap.
- **Two-piece orbitozygomatic approach** : frontotemporal bone flap is elevated as the initial step and the orbitozygomatic osteotomy is performed as the second step.
- It exposes nearly 180 degrees of the orbital rim, and provides excellent access to the superior and lateral aspects of the orbit.
- This exposure allows access to the optic nerve from the chiasm to the globe, and permits the orbital contents to be exposed from above or laterally.

# **Complications**

- <u>PTOSIS</u> : levator muscle & / or its nerve damage
- <u>DIPLOPIA</u>: EOM damage, ocular motor nerve damage, adhesions of EOM, trochlea damage
- <u>VISUAL LOSS</u> : CRA trauma / occlusion, globe compression, optic nerve trauma / compression (H'age, edema)
- <u>CSF LEAK</u>: inadvertent opening of the paranasal sinuses(post ethmoid) while optic canal deroofing.
- <u>EYELID MALPOSITION</u>: faulty wound closure, adhesions b/w lids & orbital rim
- <u>PUPIL & ACCOMODATION ABNORMALITIES</u> : Posterior ciliary N & vessels damage

- <u>PULSATING PROPTOSIS</u>: due to extensive deroofing of the orbit
- <u>FRONTAL BRANCH OF FACIAL N INJURY</u>: Incision >4cm from the lateral canthal margin in lateral orbitotomy
- <u>OCULAR OR FACIAL SENSORY LOSS</u>: sensory nerve damage (nasociliary N, 1<sup>st</sup>/2<sup>nd</sup> division of trigeminal N)
- <u>CORNEAL ULCERATION</u> : direct corneal trauma, corneal dessication