PROPTOSIS OTOLARYNGOLO-GIST'S PERSPECTIVE

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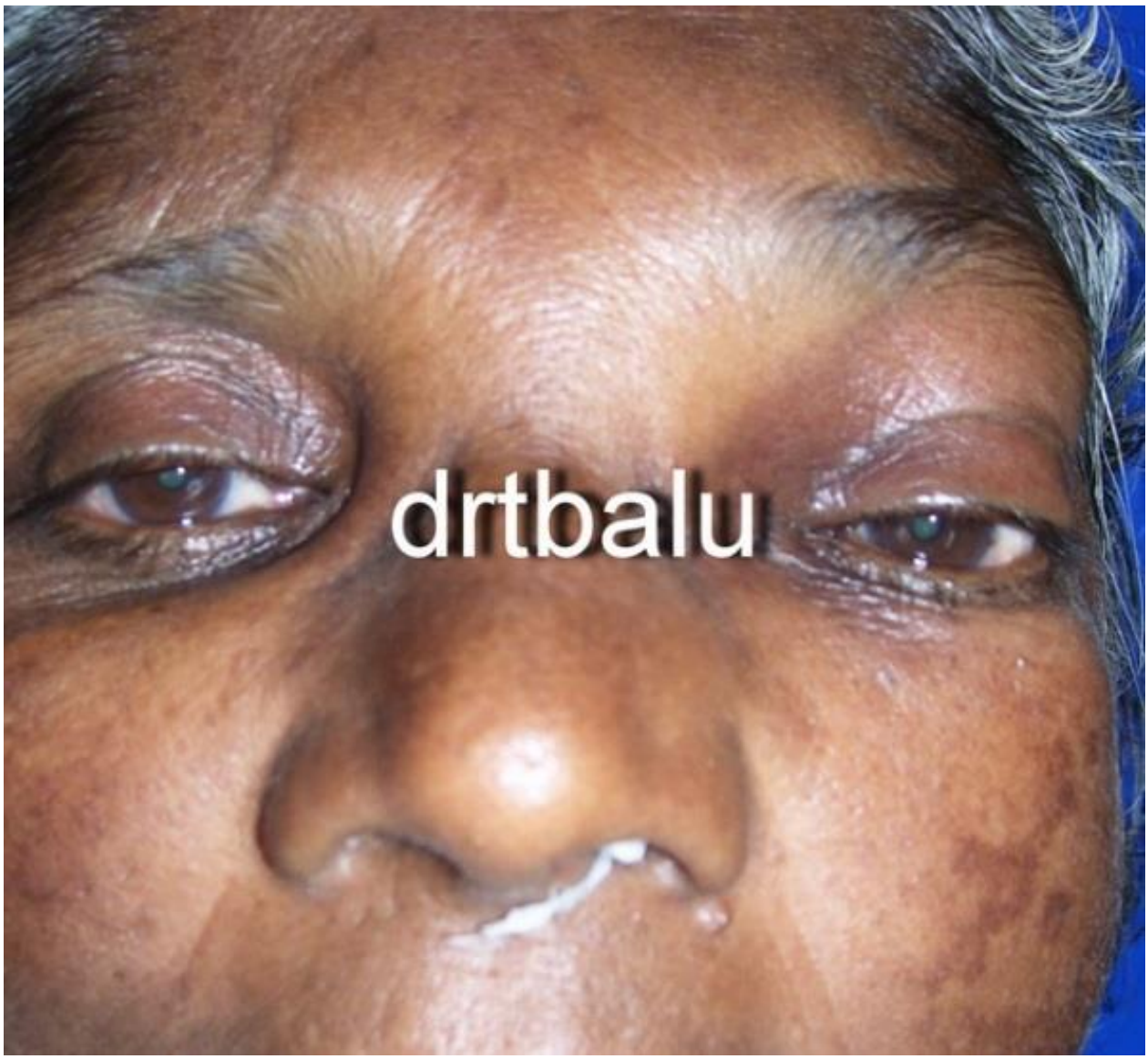

Proptosis Otolaryngologist's Perspective

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Proptosis Otolaryngology causes

Abstract:

Proximity of orbit to nose and paranasal sinuses makes it rather vulnerable to insults due to otolaryngological causes. The medial wall of orbit (Lamina papyracea) is rather paper thin and it happens to form the lateral wall of anterior ethmoid air cells. Infections / tumors involving paranasal sinuses can involve orbit also causing proptosis. This article aims at studying the various causes of proptosis with special emphasis on the various management modalities available.

Introduction:

Proptosis is defined as abnormal protrusion of eyeball. It has in fact been used interchangeably with exophthalmos. Purists consider exophthalmos as proptosis with lid lag (since it is commonly associated with Grave's disease). According to Epstein in Proptosis the globe protrudes out by 18 mm or less and exophthalmos is globe protrusion of more than 18 mm [1]. Henderson prefers reserving the term exophthalmos to proptosis caused by endocrine causes [2]. Proptosis should be differentiated from exorbitism which is caused by decrease in the volume of orbit causing the orbital contents to protrude anteriorly. In exorbitism proptosis is always anterior.

Anatomy of orbit and its relationships with paranasal sinuses:

Orbit is related to paranasal sinuses in two ways:

1. Anatomically by its location and by
2. Venous drainage (They both share the same venous drainage).

The paranasal sinuses surround the orbit from 11 o clock position superiorly to 6 o clock position inferiorly.

The orbit is pyramidal shaped formed by several bones. The superior wall of the orbit is shared by the floor of the frontal sinus, the floor of the orbit is shared by the roof of the maxillary sinus, and the medial wall of the orbit is shared by the lateral wall of ethmoidal sinus. These shared bones are really thin enabling infections to travel from either direction. The medial wall of the orbit is so thin that it is termed as lamina papyracea.

Anatomical uniqueness of orbit:

1. It is a closed space
2. It is devoid of lymphatics
3. Intraorbital pressure may increase to a limit of breaking point
4. Main constituents of orbit are muscle and fat
5. Extra ocular muscles are fine voluntary muscles with excessive interstitial tissue

Peculiarities of venous drainage in this area: The veins draining this area are peculiar in the following aspects:

1. The whole venous system in this area is devoid of valves, consequently a two way pathway of infection between the orbit, nasal cavity and paranasal sinuses become a reality.

2. The superior ophthalmic vein connects the facial veins to the cavernous sinus thus causing infections from face to spread to the cavernous sinus.

3. One branch of the inferior ophthalmic vein connects the orbit with that of the veins of pterygoid plexus and the other branch connects the orbit with that of the cavernous sinus.

The approximate volume of the orbital cavity is about 30 ml. Since the contents of the orbit are within a rigid confines of the orbital walls any disease process within the orbit or adjacent region has a tendency to displace the orbital contents forwards, this displacement of the globe is known as the proptosis. Other symptoms of globe displacement include diplopia, and visual loss.

Exophthalmos due to Graves' disease Proptosis due to frontoethmoidal mucocele

The shape of the orbit resembles a four sided pyramid to begin with but as one goes posterior it becomes three sided towards the apex. The rim of orbit in an adult measures about 40mm horizontally and 35 mm vertically. The medial walls of orbit are roughly parallel and are about 25 mm apart in an adult. The lateral walls of orbit angles about 90 degrees from each other.

Osteology of orbit:

The orbital rim is more or less spiral with its two ends overlapping medially on either side of lacrimal fossa. The inferior orbital rim is formed by the maxillary bone medially and zygomatic bone laterally.
The zygomatic bone forms the lateral orbital rim, while the frontal bone forms the superior orbital rim. The superior rim is commonly indented by a small notch known as the supra orbital notch. This notch is invariably present at the junction of medial and lateral 1/3. The supra orbital nerve and artery pass through this notch to reach the forehead.

The medial portion of the orbital rim is formed by the frontal process of maxilla and the maxillary portion of the frontal bones. A depression known as the lacrimal fossa is formed in the infero medial orbital rim. This fossa is formed by the maxillary and lacrimal bones. This lacrimal fossa is bounded by two projections of bones i.e. the anterior lacrimal crest of maxillary bone and the

posterior lacrimal crest of lacrimal bone. This fossa houses the nasolacrimal sac. This fossa opens in to the naso lacrimal canal through which the naso lacrimal duct traverses.

The naso lacrimal duct is 3 - 4 mm in diameter, courses in an infero lateral and slightly posterior direction towards the inferior turbinate under which it opens into the inferior meatus. This duct is roughly 12mm long. All the walls of the lacrimal duct except its medial wall is formed by the maxillary bone. The medial wall is formed by the lateral nasal wall inferiorly and the descending process of lacrimal bone superiorly.

In the frontal process of maxilla just anterior to the lacrimal fossa a fine groove known as the sutura longitudinalis imperfecta of Weber. This suture runs parallel to the anterior lacrimal crest. Small branches of infraorbital artery pass through this groove to supply the nasal mucosa. The presence of these vessels should be anticipated in any lacrimal sac surgery to avoid unnecessary troublesome bleeding.

Embryology of orbit: The walls of the orbit formed by 7 bones, are embryologically derived from neural crest cells. Ossification of the orbit is complete at birth except at its apex. Except the lesser wing of sphenoid which is cartilaginous the other bones develop by intramembranous ossification.

The roof of the orbit is mostly formed by the frontal bone, only the posterior 1.5 cms of the roof is formed by the lesser wing of the sphenoid bone. The optic foramen through which the optic nerve traverses is located in the lesser wing of the sphenoid bone. The optic nerve enters the orbit at an angulation of 45 degrees.

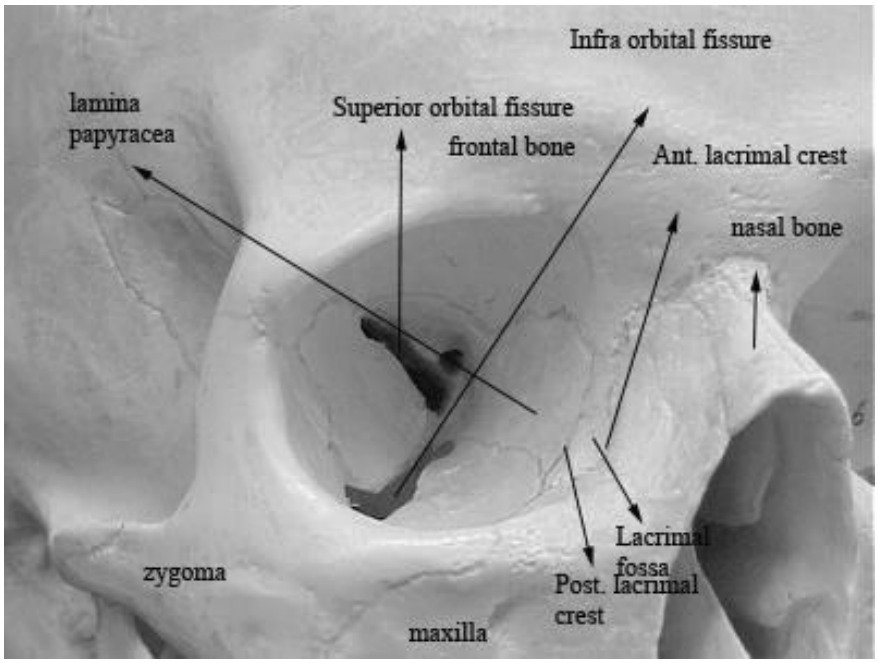
The lacrimal gland fossa is located in the lateral portion of the orbital roof, while the trochlear fossa is located in the antero medial portion of the orbital roof.

The medial wall of the orbit is formed from anterior to posterior by:

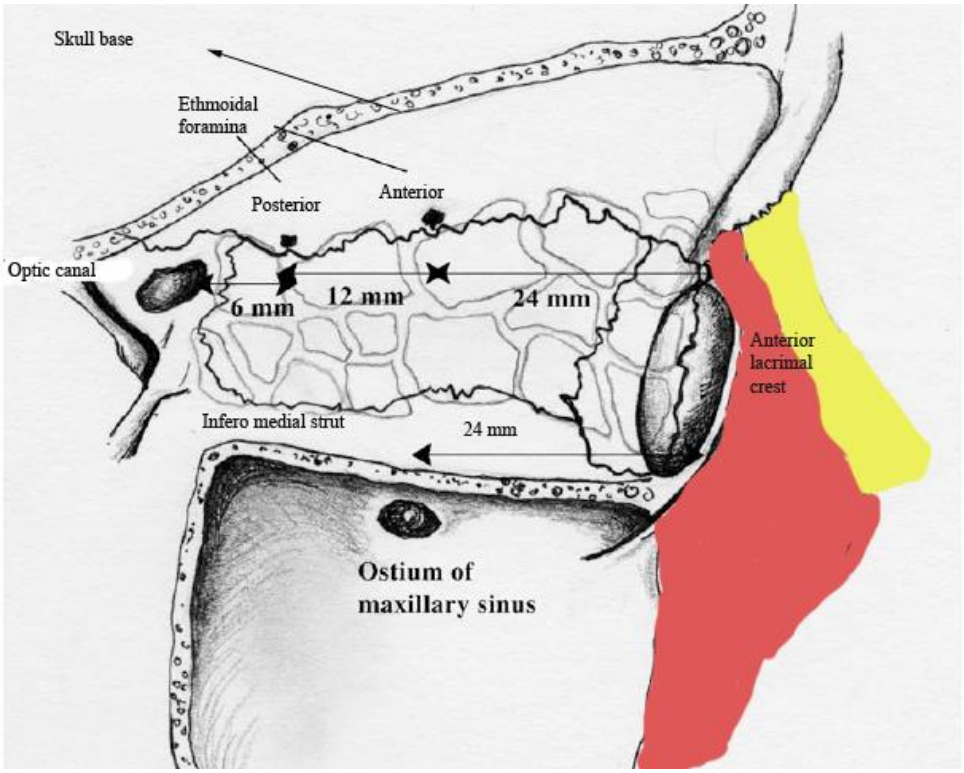
1. frontal process of maxilla
2. Lacrimal bone
3. Ethmoid bone
4. Lesser wing of sphenoid bone

The thinnest portion of the medial wall is the lamina papyracea which separates the ethmoidal sinuses from the orbit. It is one of the components of ethmoid bone. Infections from ethmoidal sinus can easily breach this paper thin bone and affect the orbital contents. The medial wall of the orbit is thicker posterior where the sphenoid bone is present and anteriorly where the posterior lacrimal crest is present.

The fronto ethmoidal suture line marks the approximate level of ethmoidal sinus roof, hence any dissection above this line may expose the cranial cavity. The anterior and posterior ethmoidal foramina through which branches of ophthalmic artery (anterior and posterior ethmoidal arteries) and branches of naso ciliary nerve passes are present in this suture. The anterior ethmoidal foramen is located at a distance of 24 mm from the anterior lacrimal crest, while the posterior ethmoidal foramen is located at a distance of 36mm from the anterior lacrimal crest.



Bones forming orbit shown here



Lateral view of skull showing the posterior relations of orbit

A vertical suture that runs between the anterior and posterior lacrimal crests is the anastomotic area between the maxillary and the lacrimal bone. If this suture is located more anteriorly it indicates a predominance of lacrimal bone, while a more posteriorly placed suture line indicates a predominance of maxillary bone in the anastomotic relationship. The lacrimal bone at the level of lacrimal fossa is pretty thin (106 micrometer). This bone can be easily penetrated during dacryocystorhinostomy surgery. If the maxillary component is predominant it becomes difficult to perform the osteotomy in this area to access the sac because the maxillary bone is pretty thick. Hence lacrimal bone predominance makes it easy to expose the sac during dacryocystorhinostomy.

The floor of the orbit is the shortest of all its walls and is bordered laterally by infra orbital fissure. Medially the floor is bounded by the maxillo ethmoidal strut. The floor of the orbit is almost entirely formed by the orbital plate of maxilla, palatine contributes to a small portion of the floor posteriorly. Zygoma also makes a small contribution to it anterolaterally. The infra orbital groove becomes a canal anteriorly, through this groove passes the infra orbital nerve and artery. The floor of the orbit medial to the infra orbital groove is thin because of the expansion of the maxillary sinus. With the growth of facial bones the infra orbital foramen migrates to about 6-10mm below the infra orbital rim.

The lateral wall of the orbit is formed mainly by the greater wing of sphenoid bone with contributions from zygoma and zygomatic process of frontal bone anteriorly. The recurrent meningeal branch of middle meningeal artery may be seen coursing through a foramen in the suture line between the frontal and sphenoid bones. This artery forms a anastomosis between the external and internal carotid arterial systems. Roughly 4 - 5 mm behind the lateral orbital rim and 1 cm inferior to the fronto zygomatic suture is the lateral tubercle of Whitnall. The following structures gets attached to this tubercle.

1. Lateral canthal tendon
2. Lateral rectus check ligament
3. Suspensory ligament of lower eyelid (Lockwood's ligament).
4. Orbital septum
5. Lacrimal gland fascia.

The frontal process of zygomatic bone and the zygomatic process of frontal bone are thick and they protect the globe from lateral trauma. Just behind this facial buttress area the posterior zygomatic bone and the orbital plate of greater wing of sphenoid are thinner thus making the zygomatico sphenoid suture a convenient land mark for lateral orbitotomy. The zygomatico facial and zygomatico temporal nerves and vessels pass through the lateral wall of the orbit to reach the cheek and temporal regions. Posteriorly the lateral wall thickens and meets the temporal bone which forms the lateral wall of the cranial cavity. When lateral orbitotomy is being done only 12 - 13 mm separate the posterior aspect of lateral orbitotomy to that of the middle cranial fossa. This distance could still be shorter in females.

Superior orbital fissure: is a linear notch between the greater and lesser wings of sphenoid. The superior portion of the fissure is narrower and here the lacrimal, frontal and trochlear nerves passes through outside the annulus of zinn. The annulus of zinn is a ring of fibrous tissue surrounding the optic nerve at its entrance into the apex of orbit. This ring gives origin to the extra ocular muscles. The following structures pass through the superior orbital fissure within the annulus of zinn.

1. Superior and inferior divisions of oculomotor nerve
2. The abducent nerve
3. Naso ciliary branch of ophthalmic branch of trigeminal nerve
4. Major venous orbital drainage exit via the superior orbital fissure to drain into the cavernous sinus.

Medial to the superior orbital fissure is the optic foramen through which the optic nerve passes. This foramen which is present in the lesser wing of sphenoid also conveys the ophthalmic artery. The optic foramen and optic canal are separated from the superior orbital fissure by a bony optic strut. In adults the optic canal is 8 - 10 mm long and 5 - 7 mm wide. The optic foramen is about 6.5 mm in diameter. The optic canal is known to attain its full adult size by the age of 3. The optic foramen on both sides are universally symmetrical. Any variation in size even to the extent of 1mm should be considered as pathological.

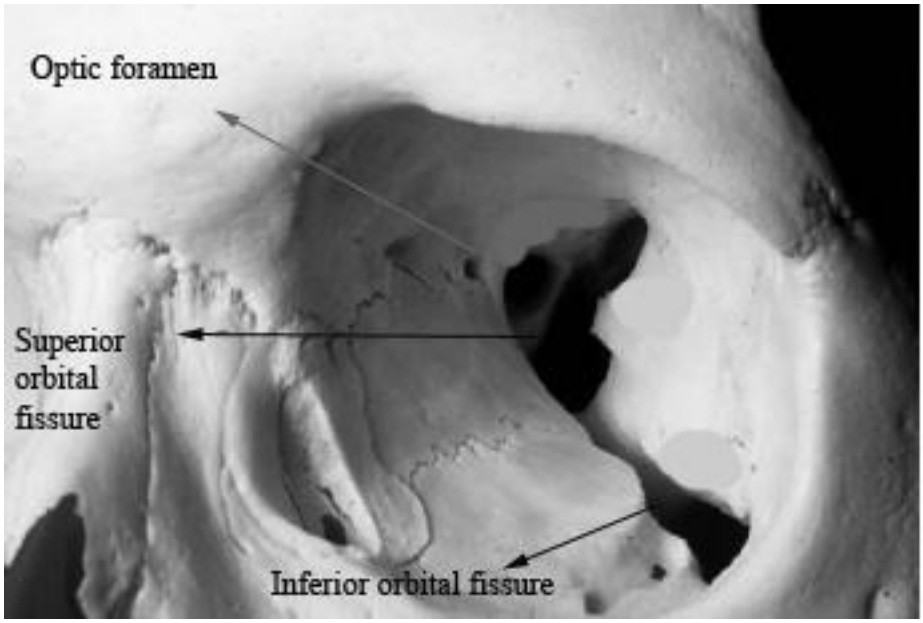


Image showing superior orbital fissure

The inferior orbital fissure lies between the lateral orbital wall and the floor of the orbit. It is about 20 mm long. The following structures pass through this fissure:

1. Maxillary division of trigeminal nerve
2. Zygomatic nerve
3. Branches from the sphenopalatine ganglion
4. Branches of inferior ophthalmic vein leading on to pterygoid plexus.

The maxillary division of trigeminal nerve and the terminal branch of internal maxillary artery enter the infra orbital groove and canal to become the infra orbital nerve and artery. These structures exit through the infra orbital foramen to supply the lower eye lid, cheek, upper lip and upper anterior gingiva.

The roof of the orbit slopes down medially. In fact this slope continues up to fronto ethmoidal suture to form the roof of the ethmoid sinus. This is otherwise known as fovea ethmoidalis.

The anatomical relationship between the anterior ethmoidal air cells and the lacrimal fossa should be borne in mind to avoid confusion between the ethmoid and nasal cavities during dacryocystorhinostomy surgery.

Soft tissues of orbit:

Orbital septum is the anterior soft tissue boundary of the orbit. It acts as a physical barrier against pathogens. This is a thin multi-layered fibrous tissue derived from the mesodermal layer of eyelid. This septum is covered anteriorly by the preseptal orbicularis oculi muscle.

Periorbita: is the periosteal lining of orbital walls. The Periorbita is attached to the suture lines, fissures and foramina of the orbit. Posteriorly the Periorbita is continuous with the optic nerve sheath.

Orbital fat: Adipose tissue present in the orbit has a cushioning effect on the contents of orbit.

The extra ocular muscles of orbit arise from the annulus of zinn and are responsible for the movement of the globe. These muscles are:

lateral and medial rectus
Superior and inferior rectus
Superior and inferior oblique

The lacrimal system:

The main lacrimal gland is located in the supero temporal portion of orbit. It lies in the shallow lacrimal fossa of the frontal bone. The gland is composed of numerous secretory units known as acini which progressively drain in to small and larger ducts. The gland measures 20 mm by 12 mm. A fibrous band incompletely divides the lacrimal gland into two lobes i.e. posterior larger orbital lobe and a smaller anterior palpebral lobe. 2 - 6 ducts from the orbital lobe pass through the palpebral lobe joining with the ducts from the palpebral lobe to form 6 - 12 tubules to empty into the superio lateral conjunctiva. Hence damage to the palpebral lobe may block drainage from the entire gland. About 20 - 40 accessory lacrimal glands of Krause are located in the superior conjunctival fornix, about half this number is located over the lower fornix.
The lacrimal gland is innervated by branches from 5th and 7th cranial nerves, sympathetic supply to lacrimal gland is via the nerves from the superior cervical ganglion. The parasympathetic fibers are supplied via the 6th nerve. Sensory supply is via the branches of trigeminal nerve.

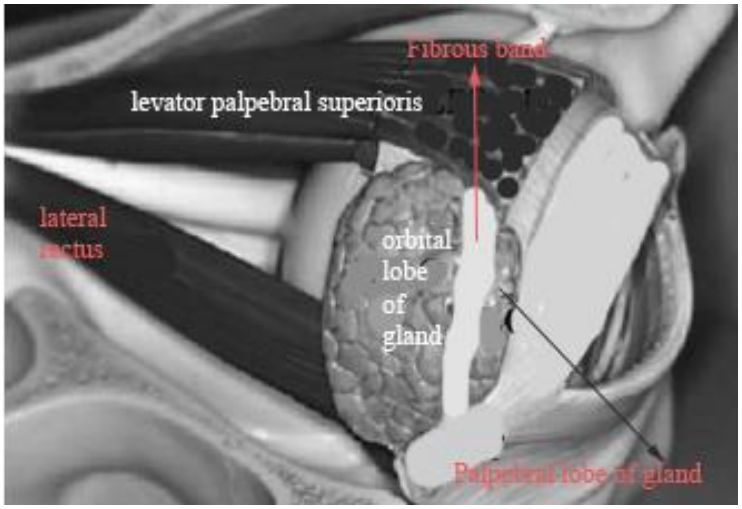


Figure showing lacrimal glands and its relationships

The lacrimal excretory system begins at a 0.3 mm at the medial end of each eyelids known as the punctum. These puncta are directed posteriorly. The punctal opening widens into ampulla, which is perpendicular to the eye lid margin. The ampulla makes a sharp turn to drain into the canaliculi. The canaliculi measures 0.5 - 1mm in diameter and courses parallel to the lid margins. The superior canaliculus is 8 mm long and the inferior canaliculus is 10 mm long. In majority of individuals the superior and inferior canaliculi merge into a common canaliculi before draining into naso lacrimal sac. The opening of common canaliculi into the naso lacrimal sac is known as the common internal punctum. There is a valve at the junction of common canaliculus and lacrimal sac at the common internal punctum level. This is known as the Rosenmuller valve. Another valve known as the valve of Hasner is found at the lower end of the naso lacrimal duct at the level of inferior meatus of nose.
If this Hasner's valve is imperforate in new born infants it causes congenital naso lacrimal obstruction.

The lacrimal sac resides in the lacrimal fossa. It measures about 12 - 15 mm vertically, and 4 - 8 mm antero posteriorly.

The mnemonic VEIN is helpful in remembering the causes of proptosis.

V - Vascular causes

E - Endocrine causes

I - Inflammation and infective causes

N - Neoplastic causes

Vascular causes of proptosis:

Vascular causes of proptosis can be classified into arterial and venous causes. Venous causes are due to the formation of dilated veins known as varices. Patients with these varices give a classic history of positional proptosis (proptosis varying with positions) or proptosis being induced by Val salve maneuver. In patients with long standing varices there is also an associated orbital fat atrophy leading on to a transient stage of enophthalmos. In these patients a valsalva maneuver may reveal proptosis. CT scan performed with jugular venous compression or during a valsalva maneuver may prove diagnostic. Surgical intervention in these patients may prove disastrous, hence observation and treatment of complications is advisable.

In Dural venous sinus fistula the shunt is low flow in type and proptosis is insidious in onset, high index of suspicion is necessary in diagnosing these patients.

Carotid cavernous fistula (high flow shunts) may arise as a result of trauma or spontaneously. These patients have subjective bruits, proptosis, chemosis and vision loss. The conjunctival vessels become arterialized assuming a cock screw pattern. A fistula of spontaneous occurrence has a better chance of spontaneous resolution, but in intractable cases the shunt must be closed with a balloon or carotid artery ligation.

Endocrine causes of proptosis: are the most common cause of exophthalmos. The diagnosis is fairly simple because it is invariable associated with lid signs like lid lag. The major endocrine cause for proptosis is thyrotoxicosis. This condition is also known as Graves' disease.

Characteristic features of endocrine causes of proptosis:

1. Presence of lid lag / lid retraction

2. Presence of temporal flare in the upper eyelid

3. Presence of orbital congestion

CT scan of the orbit show enlarged extra ocular muscles, there may also be a bulging of orbital septum due to protrusion of fat. This is pathognomonic of Grave's disease. TSH estimation show elevated levels in the serum.

Inflammatory causes of proptosis: In inflammatory proptosis the lesion could be either an idiopathic inflammatory orbital pseudo tumor, or due to specific orbital inflammation. Proptosis in these patients appear suddenly and acutely. These patients are invariably toxic and febrile. Myositis of extra ocular muscles may cause pain when eyes are being moved. There may also be associated acute dacryo adenitis. There may also be peri optic neuritis causing blindness. Orbital inflammation, perioptic neuritis and dacryo adenitis are highly responsive to oral prednisolone.

Inflammations involving the paranasal sinuses may involve the orbit causing proptosis. The intervening walls between the medial orbital wall and the ethmoidal sinuses is paper thin (lamina papyracea) which can be easily breached by infections from the ethmoidal sinuses causing spread to the orbit. In proptosis caused by ethmoidal sinus pathology the eye is pushed laterally, whereas proptosis due to maxillary sinus pathology causes deviation of the eye upwards and outwards. In frontal sinus pathology the eye is deviated downwards and outwards. Commonest sinus inflammatory cause for proptosis is the formation of mucoceles in the paranasal sinuses. This commonly occur in the fronto ethmoidal regions.

Neoplastic causes of proptosis: Neoplasms involving orbit may cause proptosis. Here the eye is pushed directly forwards. This type of proptosis is known as axial proptosis. Tumors involving the optic nerve can cause axial proptosis. These patients have pain free disease. The only exception to lack of pain is patients with adenocystic carcinoma of lacrimal gland. These patients have excessive pain because the tumor infiltrates the nerves.

Neoplastic lesions involving the paranasal sinuses can also cause proptosis. The common benign tumor involving the sinuses causing proptosis are:

1. Inverted papilloma

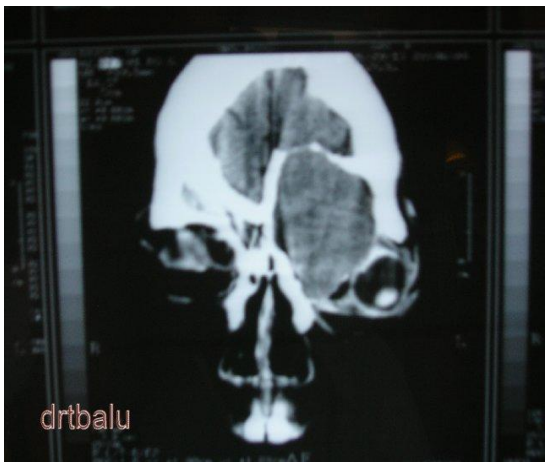
2. Fungal infections involving the paranasal sinuses

3. Mucoceles involving the paranasal sinuses

4. Fibrous dysplasia of the maxilla [5]

5. Osteomas involving the frontal and ethmoidal sinuses [4]

6. Juvenile nasopharyngeal angiofibroma [6]



Coronal CT scan showing proptosis due to frontoethmoidal mucocele

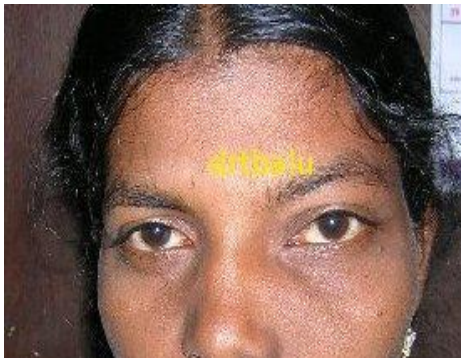


Image showing proptosis due to fibrous dysplasia

Figure showing proptosis due to frontoethmoidal mucocele

Figure showing proptosis due to fungal sinusitis

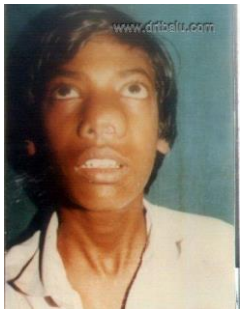
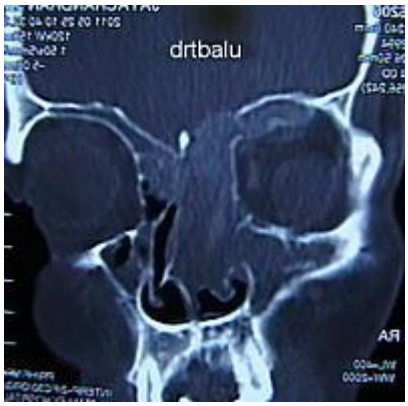


Figure showing proptosis due to JNA



Coronal CT scan Image of nose and sinuses showing proptosis due to fungal sinusitis

Measuring proptosis using exopthalmometer:

Hertel mirror exophthalmometers are used to measure the degree of protrusion of the eyeball. The distance between the lateral orbital rim and corneal apex is used as an index for measuring proptosis. Under normal conditions this distance is roughly 18 mm, there may be individual and racial variations.

Procedure: The examiner is seated in front of the patient at the eye level of the patient. The exopthalmometer is then positioned with the blue arched support at the temporal lateral orbital walls. The instrument is maneuvered using both hands and firmly propped first against the right-hand orbital wall on the temporal side (which should be felt against the lowest part of the support point). The moveable part is then set in such a way that the left-hand orbital wall lies against the lowest part of the arched support. The distance between the lateral orbital walls can then be read from the upper side of the scale; this distance can be noted for future reference. The examiner asks the patient to look straight ahead with eyelids wide open. The examiner measures for proptosis in each eye separately by looking into the mirror (which has a millimeter scale marked on it) with one eye and moving the head horizontally until the red fixations line is at 22mm. The examiner can now determine the position of the corneal apex of the patient from the millimeter reading.

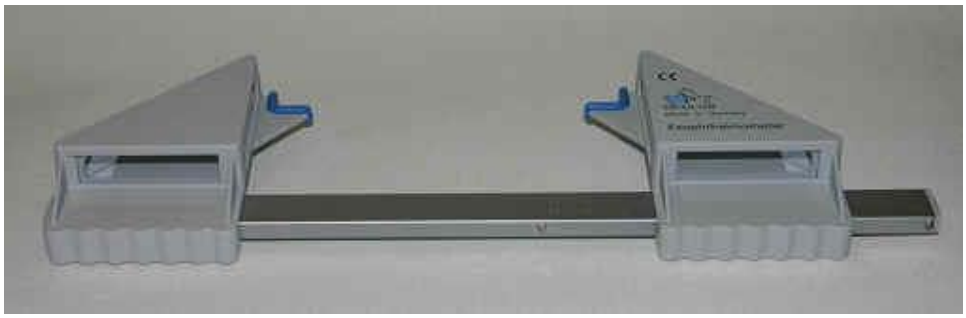


Figure showing Hertel mirror exopthalmometer

Examination of a patient with exophthalmos:

Evaluation and management of proptosis falls within the ambit of various specialities like:

Neurosurgery, ophthalmology, otolaryngology and plastic surgery. Before proceeding any further a patient with proptosis should be evaluated to rule out pseudo proptosis. Careful history taking will help in this process. Previous ocular infection / trauma may cause atrophic / phthisical globe causing the normal eye to appear more prominent. Unilateral prominent eye due to unilateral high myopia can be mistaken for proptosis. Complete ophthalmological examination will reveal the presence of high myopia. Complete examination of nose and paranasal sinuses will help to exclude rhinological causes of proptosis. These patients will have purulent drainage from the sinuses and nasal cavity. In case of doubt imaging will clinch the diagnosis. History of nasal obstruction and epistaxis are the common presentation in patients with rhinological causes of proptosis [7].

Graves' disease, the most common cause of bilateral proptosis should be ruled out. These patients show clear clinical evidence of thyrotoxicosis like:

1. Bilateral proptosis
2. Lid retraction
3. Lid lag
4. Descent of eyelid cheek complex
5. Gritty sensation in the eye
6. Retrobular pain
7. Tearing
8. Palpitation
9. Sleeplessness
10. Diarrhea
11. Menstrual disturbances (in case of females)

Majority of patients with Graves' disease show evidence of increased levels of T3, T4, free T3, TSH etc. Some of these patients may also be euthyroid with normal hormone levels. Graves's orbitopathy is more common in women. It is 5 times more common in middle aged women than men. Hypertrophy of extra ocular muscles can cause ophthalmoplegia and diplopia.

Complete ophthalmological examination is a must in these patients. Exact amount of global protrusion / proptosis can be measured using exopthalmometer. Optic neuropathy should be ruled out by assessing visual acuity, visual fields and color saturation.

Imaging studies:

CT scan and MRI scan of the orbit may prove beneficial in diagnosing the cause for proptosis. The presence of fat in the orbit serves as an inherent contrast medium for the study. 3 mm cuts are ideal for the study of orbit. Ultrasound studies using either A or B mode may help in rapid diagnosis of the cause for proptosis.

Imaging helps to identify:

12. Presence of sinusitis
13. Septal deviation
14. Hypoplastic maxillary sinuses
15. Presence of inflammatory / mass lesions in the nose and paranasal sinuses

These findings have a valid bearing on surgical management of these patients.

MRI measurement of T2 relaxation time is very useful in identifying extra ocular muscle oedema. Patients with proptosis with increased T2 relaxation time in MRI respond better to anti-inflammatory drugs, where as those with no increase in T2 relaxation time need to undergo orbital decompression to alleviate the symptoms.

Pseudo tumor cerebri is the next common cause for bilateral proptosis. Imaging reveals generalized oedema of soft tissues of orbit and sometimes the brain. There is no hypertrophy of extra ocular muscles.

Management:

Treating the causative factor should always be given priority. Rhinological causes should be treated. Mass lesions involving nose and paranasal sinuses should be removed surgically. Inflammatory lesions of nose and sinuses should be treated with antibiotics. Resistant lesions should be treated by performing endoscopic sinus surgery.

In patients with Graves' disease with hyperthyroidism, the patient should be treated with antithyroid drugs bringing the patient to euthyroid state. This alone could be sufficient in reducing proptosis. In patients with deterioration of visual acuity with Graves' disease Prednisolone in dose of 80-120 mg /day for 2 weeks is administered. Low dose irradiation can be administered in patients who are not fit for orbital decompression and who has not responded to steroid therapy. This treatment involves administration of 200 cGY of fractionated photon radiation over 2 week period. Low dose radiotherapy should be tried only in those patients who refuse surgery / in those who are poor surgical risk. Orbital decompression and orbital fat manipulation is rather difficult following irradiation because of associated fibrosis.
Immune deregulation has been implicated in the pathophysiology of proptosis in Graves's orbitopathy. Immunomodulation using cyclophosphamide and cyclosporine [8] has been attempted with varying degree of success.

Surgical management:

Ultimate goal of any surgical procedure for managing proptosis is enlargement of orbital volume by removing 1 – 4 walls of the orbit combined with incision of periosteum allowing prolapse of orbital fat into nasal cavity and maxillary sinus cavity. Studies reveal that reduction of orbital pressure was maximum when one wall of the orbit was taken down. Incision of orbital periosteum provided only a marginal reduction in Intraorbital pressure [11]. The amount of proptosis reduction is directly proportional to the number of orbital walls taken down for decompression.[9.] Studies reveal that one wall decompression causes 4 mm reduction in proptosis, two wall decompression results in reduction of 6mm in proptosis and 3 wall decompression causes a 10 mm reduction in proptosis. Technically it is possible to obtain 15 mm decompression by removing all 4 walls of the orbit [10]. It should be borne in mind that sudden excessive surgical decompression of orbit can lead to intractable strabismus.

Before embarking on orbital decompression procedure the operating surgeon should be made aware of the risks posed by the procedure which include:

Diplopia
Injury to optic nerve / retina due to prolonged globe retraction
Retrobulbar hematoma (could cause blindness)
Injury to infraorbital nerve
Epistaxis

Indications for surgical decompression of orbit:

Patients in whom steroids fail to improve visual disturbance
In patients who require steroid for maintaining vision on long term basis
Prevention of corneal exposure and the resultant keratitis

Globe prolapse anterior to the eyelids
Cosmesis

In patients with advanced exopthalmosis extensive surgery should be performed even for achieving modest benefits. This should be followed by other surgical procedures aimed at correcting strabismus, lid lengthening procedures because these patients have lid retraction in addition to advanced proptosis.

Following are considered to be indices that indicate a successful procedure:

Reduction of proptosis
Increase in the orbital volume
Reduced orbital pressure
Last but not the least satisfied patient

Superior orbital decompression (Naffziger) [12]:

This surgical procedure involves orbital decompression via superior orbital wall. This procedure was first performed by Howard C Naffziger in 1931 to treat proptosis in a patient with Graves' disease. The entire orbit was unroofed via frontal craniotomy. The roof of the orbit is composed of orbital plate of frontal bone, behind which is the lesser wing of sphenoid. Bone is actually thin in this area, measuring less than 3mm in most areas. It could be thick in the sphenoid area. In this procedure a large amount of orbital bone can be removed. Since craniotomy needs to be done, there is always the possibility of transmitted pulsations from the frontal lobe over the orbit. This can of course be prevented by careful placement of titanium shield.

This procedure is ideally performed with the assistance of Neurosurgeon. It is important to visualize the optic nerve after performing the frontal craniotomy. The roof is removed from just anterior to the optic foramen to the antero superior orbital rim. Care should be taken to leave superior orbital periosteum intact while the bone is being taken down to prevent damage to levator muscle. After exposing superior orbital periosteum in its entirety "H" shaped incision is made on the periosteum to allow fat to prolapse through it. The orbital roof area can be covered using titanium mesh which can be held in place using screws. After completion of surgery cranial flap can be replaced. Temporary tarsorraphy can be considered if there is danger of oedema which could cause worsening of proptosis. Post op steroids can be given for 4 days to allow for cessation of orbital oedema before removing tarsorraphy sutures. The current indication for this procedure is orbital trauma.

Medial orbital decompression (Sewell) [13]:

In this procedure popularized by Sewell the medial wall of orbit is taken down with an intension to increase orbital space. Approach is via Bicoronal incision or conventional anterior ethmoidectomy incision. Bicoronal incision is cosmetically better and medial canthal tendon can be left undisturbed. If anterior ethmoidectomy incision is used medial canthal tendon is tagged and divided. During the procedure, anterior and posterior ethmoidal arteries must be identified and the anterior ethmoidal artery should be secured. A complete ethmoidectomy is performed beginning from lacrimal fossa. Care is taken not to injure optic nerve. Since ethmoidectomy is performed from above there is always danger of damaging lacrimal sac and insertion of the trochlea. After complete ethmoidectomy is performed the medial orbital periosteum is incised allowing the orbital periosteum to gently prolapse into the ethmoid cavity. Injury to medial rectus is avoided during this stage of the procedure.

Inferior decompression (Hisch & Ubaneck):

This procedure crudely put is creation of blow out fracture of inferior orbital wall with sparing of the infraorbital foramen. This procedure is performed using transconjunctival / subciliary incision combined with Caldwell Luc procedure. The entire floor of the orbit is taken down under vision. Initially the bone over the floor of the orbit is thin but as drilling reaches posterior portion of orbit it becomes thicker. 3 cms bone removal in this area is considered to be ideal and safe. Medially the floor can be taken down up to the lacrimal fossa and laterally up to the zygoma. After completion of the bone drilling the Periorbita is incised allowing orbital fat to prolapse into the maxillary antrum. After incising the Periorbita forced duction test should be performed to rule out entrapment of extraocular muscles.

Lateral decompression (Kronlein) [14]:

This actually was the first described technique of orbital decompression in literature. This approach was followed later by Dollinger (1911). Kronlein was aware that lateral wall of orbit was rather thin, so he breached it to remove an orbital tumor from a patient. He was associated with Joseph Lister who was considered to be the father of aseptic surgery. His results were good because he followed Joseph Lister's sterilization techniques using carbolic acid.

Incision used is coronal incision / direct rim incision / lateral extension of subciliary incision. Orbital contents are protected by gently retracting them medially. Periosteum is incised along the lateral rim. It is then elevated on both infratemporal fossa side and the orbital side of the lateral rim. The roof and floor of the orbit are also exposed. Lateral canthus is left intact. Lateral wall of the orbit is drilled out leaving behind the orbital rim. Lateral orbital periosteum is excised allowing prolapse of orbital fat. CSF leak is a common complication of this procedure, because drilling through greater wing of sphenoid can eventually damage Dura in that area causing CSF leak.

Combined medial and inferior decompression (Walsh – Ogura) [15]:

This approach like inferior one involves Caldwell – Luc / Transantral approach. This technique was the most preferred one during 1990's. This procedure managed to achieve 5mm proptosis reduction.

Endoscopic orbital decompression [16]:

Advantages of this technique include:

Skin incision not needed
Morbidity is less
Good access to orbital apex
Can be performed under local anesthesia

This procedure can be performed under local / general anesthesia. Nasal mucosa is decongested using 4% xylocaine mixed with 1 in 50000 units' adrenaline. Eyes are protected with corneal shields during the entire surgical procedure.
30 degree 4 mm nasal endoscope is used for this procedure. Uncinectomy is performed first. Large middle meatal antrostomy is performed first. Infraorbital nerve is identified in the roof of the maxillary sinus. Total ethmoidectomy is performed. Sphenoid ostium is identified and enlarged. The lamina papyracea is skeletonized along its entire length. Position of anterior and posterior ethmoidal arteries should be identified. Middle turbinate can be excised to provide space for future

endo-cleaning procedures. If middle turbinate could be retained than it would prevent prolapsing orbital fat from obstructing sphenoid sinus ostium. While removing lamina papyracea a small portion of it should be left close to the frontal sinus ostium to prevent prolapsing orbital fat from obstructing frontal sinus drainage. A curette is used to breach lamina papyracea in its mid portion. Pieces of lamina should be carefully removed without breaching Periorbita in order to prevent prolapsing fat obscuring endoscopic vision. Lamina papyracea should be removed up to the roof of ethmoid superiorly, anterior face of sphenoid posteriorly, maxillary line (nasolacrimal duct) anteriorly and maxillary antrostomy inferiorly.

Next the inferior wall of the orbit is removed. The orbital floor is removed by fracturing the floor medial to the infraorbital nerve. This can be performed using a spoon curette by downward pressure over the floor of the orbit. It usually fractures medial to the inferior orbital fissure. The Periorbita is breached using a sickle knife allowing orbital fat to prolapse into the ethmoidal cavity and maxillary sinus cavity.

Orbital fat removal:

Removal of fat from the orbit can reduce tension within. Preoperative CT scan will help in identifying fat pockets that need to be removed. Fat pockets can be removed via upper lid skin crease / subciliary / transconjunctival approaches. Removal of 5-6 ml fat will cause a reduction of about 6mm of proptosis.

How to choose appropriate technique?

For mild exophthalmos 2-3 mm any of the above mentioned decompression approaches would suffice. For moderate exophthalmos (5mm) inferior decompression alone would do. For severe exophthalmos (7 mm) 3 wall decompression will have to be performed.

Complications of orbital decompression:

Diplopia – can be avoided by performing balanced decompression of medial and lateral orbital walls. In endoscopic orbital decompression Metson's [17] orbital sling technique can avoid diplopia. In this technique two horizontal incisions are made in the medial wall Periorbita close to the superior and inferior margins of medial rectus muscle. Periorbita above and below this incision can be excised allowing orbital fat to prolapse into the nasal cavity while allowing medial sling to tissue to prevent excessive medial displacement of orbit which could cause diplopia.
Corneal abrasion
Retrobulbar hematoma
Infraorbital nerve injury
Retinal hemorrhage
Orbital cellulitis
Retinal vascular occlusion

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