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Editorial

The Avatar Phenomenon

Dear Friends,

Many Ophthalmologists must have given the ‘out of this world’ 3D movie ‘Avatar’ recently in theaters, a miss. They cannot be blamed for thinking that James Cameron had made it only for the young ‘Sci-fi’ fan.

But the truth is that he had made it as a wake-up call for the ophthalmologist. In ophthalmology if the last century was the period for ‘par excellence’ uniconal vision: from aphakia to IOLS to LASIK and beyond, this century promises to go deeper: In real 3-D depth. Entertainment and movies have shown that they can provide television and art in 3D and it becomes imperative for us, the ophthalmologist to ensure their not one child misses the beauty of the 3D world whether the real one or the artificially created ones.

Having seen so many one eyed individuals function comfortably in their surrounding and having taken our own binocular depth perception for granted, it takes a movie like Avatar to show us how different the 3D is to the 2D: like cheese is from chalk.

Movies like these remind us that we must look at our patient’s both eyes together and not separately. They remind us that great vision in either eye is not the best,

Superb binocular vision is!
Appreciating the Avatar phenomenon is!

Dr. Rohit Saxena

The editorial board of the DJO solicits article from its readers on all aspects of ophthalmology. Major heads under which article are published by the DJO are given in the introduction to authors. We also welcome to article comments and advise on how to improve the DJO.

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ROLE OF TEAR OSMOLARITY IN DRY EYE

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The first ever published association of dry eye with the occurrence of salty tears was described by Galen in the second century.\(^1\) Three centuries later, the concept of tear salinity was based on the taste of overflowing teardrops. The basis of the future concept of osmolarity originated in 1748, when a French priest and scientist Nollet\(^2\) observed the fluid flow through a membrane divided by fluids of different osmolarity. However, the most common method of measuring osmotic pressure in clinics was introduced by Raoult, who in 1882, wrote that the freezing point of water solution depends on the osmolarity.\(^3\)

Currently, the concept and importance of tear osmolarity in dry eye has been re-introduced by the International Dry Eye Workshop (DEWS), 2007\(^4\) that defined dry eye as a multifactorial disease of the tears and ocular surface that results in symptoms of discomfort, visual disturbance, and tear film instability with potential damage to the ocular surface. It is accompanied by increased osmolarity of tear film and inflammation of ocular surface.

Ocular surface is a sensitive part of the eye, where its constituent components are closely linked together and help in maintenance of adequate homeostasis in the region, in which the tear film must maintain the health of the cornea and conjunctival epithelia, and at the same time contribute to the normal physiology of the stroma. The decrease in the production of tears and/or compositional qualitative changes in the tear film, as well as increased evaporation of the tear film favour the phenomenon of hyperosmolarity.

The toxicity of normal tears is mainly determined by its content of cations (sodium, potassium, calcium, magnesium, iron and copper) and anions (chlorides, bicarbonate and phosphates). The most abundant component is sodium chloride. A fraction of toxicity is produced by macromolecules, such as proteins or sugars. Early measurements of tear osmolarity did not directly measure the toxicity, but only expressed the content of salts and macromolecules. Thus, Frerichs (1841) reported that tear has 0.94 to 1.3% solids, including salts, albumin, mucus, fats and desquamated epithelial cells.\(^5\) Later, the total toxicity was expressed by its equivalence to a determined percentage of NaCl water solution. More recently, most authors express tonicity according to the quantity of moles per litre of solution (osmolarity).

The site of tear sampling is of importance in all measurements of tear osmolarity. Various authors have specified the part of the lacrimal sea from which they took the tear sample: inferior conjunctival fornix, tear meniscus, or cisterna lacrimalis. Tear has been taken with absorbent paper or washed cotton and pipette. Mishima et al introduced very thin pipettes to collect small quantities of tear to avoid irritative stimuli.\(^6\) Gilbard et al introduced an L-shaped micropipette under femto-biomicroscopy to avoid production of reflex hyperscretion, taking volumes of 0.4-0.1 microlitre and modifying the technique of storing the sample. The latest instrument for measuring tear osmolarimetry is the Ocusense system, which is able to measure tear sample of only 0.2 microlitre.\(^7\)

Circadian changes in tear osmolarity have also been studied. Mishima et al, in rabbits and Mandell et al, in humans, observed that the cornea is 4% thicker when the eyes are closed than they are in the normal awake state, and they assumed this was because tear film evaporation with an open eye produces a hypertonic film. Uniacke et al reported that tear osmolality is found to be low in mid-morning, high following lunch and throughout the afternoon, and declines in the evening. Benjamin et al found trends during day towards hypertonicity. Terry et al found that during waking, the tear film is hypertonic, with an osmolarity of 310 mOsm/kg and 285 mOsm/kg during sleep, producing corneal edema on awakening.

In 1952, Balik was the first to suspect that the damage of the ocular surface in sicca syndromes could be related to tear hyperosmolarity, but when he found no significant differences in tear NaCl concentration in normal subjects and keratoconjunctivitis sicca (KCS) patients, he concluded that NaCl did not play a role in the pathophysiology of KCS. Mastmam et al demonstrated for the first time that a relationship existed between osmolarity and dry eye. Mishim et al confirmed the tear hyperosmolarity in patients with...
Role of Tear Osmolarity In Dry Eye

Fig 1: Tear film structure: a modern view

Fig 2: Ocular surface homeostasis in a healthy eye

Fig 3: TearLab Osmolarity System intended to measure the osmolarity of human tears to aid in the diagnosis of dry eye

Fig 4: Clinical picture of a patient with dry eye syndrome

Fig 5: Microphotograph showing squamous metaplasia of epithelial cells on conjunctival impression cytology in a case of severe dry eye

Fig 6: Rose Bengal stains dead, devitalized epithelial cells and mucus pink in the inter-palpebral area in dry eye syndrome

Fig 7: Tear break up time: Slit lamp photograph showing break up of tear film as evidenced by black spots where the fluorescein stained tear film has ruptured
Role of Tear Osmolarity In Dry Eye

KCS when they found that normal subjects had a tear osmolarity of 289 mOsm/l in the conjunctival fornix and 304 mOsm/l in the lid margin, while patients with KCS had osmolarity of 329mOsm/l. Gilbard et al confirmed the findings of Mishima et al by using samples of only 0.1-0.4ul.

The designated cut-off or limit between normal osmolarity and hyperosmolarity varies according to the authors. The Pisa criteria of dry eye defines mild hyperosmolarity as about 320 mOsm/l, moderate as 330mOsm/l and severe about 340mOsm/l.8

Today it is accepted that there is a relationship between tear hyperosmolarity and sicca syndrome. Farris et al has taken it as gold standard in diagnosing sicca syndrome and osmolarity of over 312 mosm/l has KCS a sensitivity of 90% and a specificity of 95%.9 Laboratory studies have made it possible to demonstrate that only a 1 % increase in the osmolarity of tear film is able to provoke epithelial lesions and alter the normal flow of liquids to the stroma. The causes of tear hyperosmolarity in dry eye were summarized by Gilbard,10 as follows:

1) Lacrimal gland primarily reduces its secretion, and increases its salinity;
2) Lacrimal sea has a decreased volume, but maintains the same evaporation rate;
3) Lacrimal sea reduces its flow and turn over, but maintains a similar evaporation rate;
4) Physiochemical protectors against evaporation decrease.

The hyperosmolarity11 causes epithelial injury, complete disappearance of cell surface epithelial layers, decreased cytoplasmic density and accumulation of mucus cells and secretions leading to epithelial desquamation and ocular surface damage. This phenomenon is generally evident between 15 to 30 days of the tear film osmolar change. Thus, the hyperosmolarity triggers a series of pathophysiological mechanisms with clear feedback including effects that in turn enhance each other.

Tear hyperosmolarity is regarded as the central mechanism causing ocular surface inflammation, damage, and symptoms, and the initiation of compensatory events in dry eye. Tear hyperosmolarity arises as a result of water evaporation from the exposed ocular surface, in situations of a low aqueous tear flow, or as a result of excessive evaporation, or a combination of these events. Rapid tear film thinning may be hypothesized as a risk factor for tear hyperosmolarity. Hyperosmolarity stimulates a cascade of inflammatory events in the epithelial surface cells, involving MAP kinases and NFKB signalling pathways and the generation of inflammatory cytokines (IL-1, TNF-alpha) and MMPs (MMP9), which arise from or activate inflammatory cells at the ocular surface. There is evidence that these inflammatory events lead to apoptotic death of surface epithelial cells, including goblet cells; thus, goblet cell loss may be seen to be directly related to the effects of chronic inflammation. The increase in the osmolarity of the tear film in dry eye is taken as a stress response by the ocular surface and this triggers the process of inflammation and immunological phenomena such as the presentation of autoantígenos that enhance the inflammatory process.

Studies of inflammatory markers such as NF-Kβ that migrates to the cytoplasm during the inflammatory process, and are directly related to the phenomenon of hyperosmolarity12. The nuclear translocation of the NF-Kβ is directly proportional to the osmolarity of tear film. Berra A et al12 compared the nuclear translocation NF-Kβ in healthy patients, postmenopausal women and patients with Sjögren’s syndrome and linked them with the osmolarity of tear film and conjunctival impression cytology of these patients. Patients of severe dry eye demonstrated high values of the film osmolarity (< 400 mOsm/L) and large nuclear translocation of the factor NF-expression and severe squamous metaplasia.

The effect of tear hyperosmolarity on the ocular surface is evident. Massat (1889) wrote that evaporation of the tear film provokes hyperosmolarity, which can produce epithelial damage and even corneal ulcers, and that nerve terminal endings floating in the tear film induce blinking to recover normal osmolarity. Other processes in which hyperosmolarity may play a role-directly or indirectly-are alteration of corneo-conjunctival epithelium, loss of micropilae, cell membrane disruption, increased Rose-Bengal staining and decreased tear break up time, and decreased desquamation. A link between tear instability and hyperosmolarity has also been established that may in turn produce inflammation and trigger inflammatory neurons.13

Without a doubt, dry eye is currently the most commonly diagnosed clinical condition by ophthalmologists. A clear and concise knowledge of its cascading pathophysiology and its early diagnosis, will enable us to manage this condition effectively. In this regard, assessing the tear osmolarity these patients would not only provide an etiological diagnosis of the disease, but also gives us a powerful diagnostic tool for evaluation of the disease, with values that are directly proportional with the severity of the clinical picture of dry eye, and is always present in these patients.
REFERENCES
5. Frerichs FT, in Wagner R. Handworter Physiologie 1846; 3:617. (German)

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Dissociated vertical deviation (DVD) is a poorly understood eye motility disorder of unexplained etiology. DVD is a form of cyclovertical deviation which is known by various terms like, alternating hypertropia, double hypertropia, dissociated vertical divergence and alternating sursumduction amongst others. Bielschowsky provided the first comprehensive description and clinical analysis of DVD. In DVD, either eye elevates when the fellow eye is fixating. Lancaster and Swan called it alternating sursumduction, emphasizing the monocular nature of the movement.

Clinical features:
The characteristic feature seen in DVD is the spontaneous drifting of either eye upward when the patient is tired or after covering one eye. After the cover is removed from the elevated eye, it slowly drifts downward to settle in the primary position. Other features of DVD include:
- Excycloduction of the elevated eye.
- Incycloduction of the fixating eye.
- Sometimes only excycloduction is seen under cover, when it is called as Dissociated Torsional Deviation (DTD).
- Latent nystagmus seen in nearly half the patients
- Head posture: anomalous head posture is reported in about 30% of patients. Anomalous head posture decreases the magnitude of alternating hyperphoria. Chin depression can also be seen.
- DVD can occur with overaction of inferior oblique as well as superior oblique muscles.
- It is rarely seen in infants, but usually presents at 2-5 years of age.
- DVD is usually bilateral and asymmetrical. Unilateral cases are commonly associated with deep amblyopia and sensory heterotropia.
- DVD is associated with infantile esotropia/ exotropia, and less commonly with Duane’s retraction syndrome.

Diagnostic tests:
When the eye is covered, it elevates and excycloducts; upon removal of cover, the elevated eye returns to the midline slowly along with incycloduction. These torsional movements can be observed by looking into the conjunctival vessels.
- Translucent occluder test (Spielmann’s) figure 1a-c – updrift of the eye behind the occluder can be seen and its downward slow drifting back is observed after removing the occluder.
- Graded density filter bar (Bielschowsky’s) test – as the density of the filter bar is increased, the eye drifts up and as the density of the filter bar is decreased, the eye comes down. This is called Bielschowsky’s phenomenon. figure 2
- Red filter test – this is a dissociating test. The eye behind the filter drifts up and patient appreciates diplopia, with the red image being lower. The amount of separation between the images is used to measure the amplitude of elevation of each eye.

Quantitative assessment of DVD can be made by using base-down prisms, held under the occluder in front of the nonfixating eye until the downward fixation movement of that eye is neutralized.
Aetiology:
Numerous theories have been proposed to explain this intriguing anomaly. Elastic preponderance of the elevator or the depressor muscles, paretic factors especially bilateral paresis of the depressor muscles and imbalances between the amount of innervation originating from each vestibular organ have been proposed earlier. Abnormal visual pathway routing similar to albinism or abnormal, intermittent & alternate excitation of subcortical centers could be responsible. Recent investigations also agree with Bielschowsky’s vertical vergence signal theory on DVD.7,8 Spielmann assumed that DVD is caused by an imbalance of binocular stimulation9. But it does not account for DVD in patients with otherwise normal binocular functions. Several investigators have opined that the vertical vergence movements must be predominantly mediated by oblique muscles. Aetiology of DVD still remains obscure, but Bielschowsky’s explanation of DVD as a vertical vergence eye movement appears more convincing.

Differential diagnosis:
DVD should be differentiated from inferior oblique overaction, (Table 1)

<table>
<thead>
<tr>
<th>Features</th>
<th>DVD</th>
<th>IOOA</th>
</tr>
</thead>
<tbody>
<tr>
<td>1) Hypertropia</td>
<td>Same in primary position, adduction &amp; abduction</td>
<td>Maximal in adduction, never in abduction</td>
</tr>
<tr>
<td>2) Recovery movement on uncovering</td>
<td>Slow downward drift</td>
<td>Quick refixation</td>
</tr>
<tr>
<td>3) Superior oblique action</td>
<td>May overact</td>
<td>Usually underaction</td>
</tr>
<tr>
<td>4) V pattern</td>
<td>Absent</td>
<td>Often present</td>
</tr>
<tr>
<td>5) Pseudoparesis of contralateral superior rectus</td>
<td>Absent</td>
<td>Present</td>
</tr>
<tr>
<td>6) Incycloduction on refixation</td>
<td>Present</td>
<td>Absent</td>
</tr>
<tr>
<td>7) Latent nystagmus</td>
<td>Often present</td>
<td>Absent</td>
</tr>
<tr>
<td>8) Bielschowsky’s phenomenon</td>
<td>Often present</td>
<td>Absent</td>
</tr>
<tr>
<td>9) Red filter test</td>
<td>Red image is always lower as eye behind the filter is always higher</td>
<td>Red image is higher or lower on alternation</td>
</tr>
</tbody>
</table>
Dissociated vertical deviation (DVD)

Treatment: Many surgical modalities have been tried with varying success rates.
- Superior rectus recession – unconventional recession of superior rectus by 7 to 9mm is required in bilateral DVD. Asymmetric cases need differential recession.12
- Retroequatorial myopexy of superior rectus (Faden operation) – done along with superior rectus recession may give better results.
- Resection of inferior rectus – is done rarely as a single surgery. It is usually done as second surgery when superior rectus recession fails.
- Anterior transpositioning of inferior obliques – is popular among few surgeons and effective in cases of DVD with inferior oblique overaction.13,14 The antielevation syndrome which is observed after this surgery in some cases can be overcome by bunching the placement of inferior oblique adjacent to the inferior rectus.

Lim15 has described hypotropic DVD, which is mostly unilateral and commonly is associated with monocular visual deficits or high myopia. Although the nature of the intermittent slow downward ocular deviation is similar to that of hypertropic DVD, it should be considered to be a unique form of the dissociated strabismus complex. This rare condition can be corrected surgically by a large recession or a combined recession-resection of the inferior rectus muscle.

Fixating eye penalization with 1% atropine has been shown by few authors to reduce DVD magnitude and decompensatory phases during follow-up.

References:
Dissociated vertical deviation (DVD)

Is this dissociated deviation

Cover – uncover either eye while watching the eye just uncovered

Uncovered eye moves **down**, fellow eye moves **up** when uncovered

Uncovered eye moves **down**, fellow eye may do same when uncovered but **never moves up**

**Dissociated exodeviation**

Recess LR

Elevation may be the same in adduction primary and abduction

Often associated with latent nystagmus

Eye excycloducts when elevated

Bielschowsky phenomenon

Refixation movement is slow (tonic)

May be latent or manifest

May be associated with A – or V - pattern

Recess SR 7 – 10 mm

**Dissociated vertical deviation**

Elevation may be the same in adduction primary and abduction

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May be associated with A – or V - pattern

Recess SR 7 – 10 mm

**Vertical strabismus**

Red light always seen **above** white light with one eye fixing and **below** white light with other eye fixing

**Red glass test**
MANAGEMENT OF SUPERIOR OBLIQUE PALSY

Archana Gupta Mahajan
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For the strabismologist, superior oblique or fourth nerve (IV N) palsy is the most common isolated cranial nerve weakness affecting motility. Possibly because of a process of selection others, particularly the neuro-ophthalmologist might see VI N palsy more often.

Case:
A twenty one year old male presented to us with a history of one month old head trauma. His chief complaint was intermittent vertical diplopia which got better on tilting his head to the right side. On examination, he had a left hypertropia of twenty prism diopters (pd) increasing on right gaze and left head tilt. Ocular movements showed left Inferior oblique over action and left superior oblique under action. A subjective torsion of 8 degrees was measured on double Maddox rod test and on indirect ophthalmoscopy, left excyclotorsion was seen. A diagnosis of left superior oblique palsy was made.

The question to be answered is - how do we proceed from here? What are the diagnostic tests required and what are the management options? Before determining the right treatment strategy, the etiology and the muscle groups involved need to be determined, as well as the scale of discomfort to the patient. That is:

- Etiology - Congenital or acquired?
- Signs and symptoms – diagnosis and isolation of muscles primarily affected
- Investigations
- Treatment

ETIOLOGY
Etiology is important as among other reasons, identifying a case as congenital helps in avoiding expensive investigations and starting with treatment without delay

- Congenital
- Acquired
  - Traumatic
  - Vascular
  - Tumor – very rare
  - Iatrogenic

CONGENITAL
Unlike other isolated nerve palsies, the most fourth nerve palsies are congenital, incidence varies in different studies; out of 190 cases of superior oblique palsy in a study by Helveston et al, 137 were congenital and 53 were acquired.

Usually there is no history of trauma, or the patient may erroneously date the symptoms to a trivial trauma. The symptoms are mostly longstanding and may be worsening over time due to a decompensation. There is a large head tilt and facial asymmetry confirmed on old family photographs (the family album tomography or “FAT scan”). The face is fuller on the involved side – the ocular torticollis – a hallmark of congenital superior oblique palsy.

Although congenital cases do not complain of diplopia, intermittent vertical diplopia may occur in decompensated congenital palsy. There is no measurable subjective torsion.

A Lax superior oblique tendon confirmed by the superior oblique traction test done at surgery. In case of amblyopia and horizontal strabismus absence of the superior oblique tendon should be suspected.
ACQUIRED
The most common cause of an acquired palsy is traumatic.

TRAUMATIC

Unilateral Palsy
Usually occurs after a trivial acute trauma e.g a whack on the head or a bump against furniture. The patient typically complains of incomitant hypertropia and vertical diplopia but there is no facial asymmetry. The superior oblique traction test is normal at the time of surgery and extorsion is typically demonstrated on the double Maddox rod test less than 104-151 degrees.

Bilateral Palsy
Occurs after more severe trauma; severe closed head trauma – associated with a period of unconsciousness. A patient having subjective complaints of torsion should be suspected of having a bilateral palsy; the objective torsion is more than 10 degrees. On examination, there is an alternating hypertropia on head tilt, a V pattern esotropia, chin down head posture and reversing Bielschowsky head tilt test.

Vascular
Seen in the elderly age group, vascular palsies have an acute onset of a small angle hypertropia not more than 4-6 diopters. Mostly associated with Hypertension and diabetes, these palsies are self-limiting and require a physician referral. Temporary relief can be gained with prisms; these rarely need surgery.

Tumor
A rare cause of superior oblique palsy, systemic disease is mostly confirmed by the time the symptoms of palsy appear; treatment is aimed at the underlying disease and prisms for relief of symptoms.

Latrogenic
Now rarely seen, this association was seen quite commonly in the earlier days after superior oblique tenectomy or after ethmoid sinus surgery with trochlear trauma.

History
As the details have mostly been covered above, the following points sum the history:
- Nature of trauma
- Diplopia
- Head tilt to the opposite side
- Any medical problems
- Associated neurological signs i.e other cranial nerves palsies.

EXAMINATION
- Head tilt to the opposite side and in case of a bilateral palsy, chin depression (V pattern)
- Cover tests
- Prism Bar Cover Test in all positions
- Ocular movements – versions
- Parks 3 step test
- Maddox rod – subjective torsion
- Indirect ophthalmoscopy - objective torsion
- Associated neurological signs

Cover Tests
Are important to demonstrate the presence of hypertropia and the prism bar cover test should be done to quantify the deviation in all the gazes giving an idea of which gaze is affected the most and thus corresponding muscles needing surgery.

Figure from: Helveston Eugene M; Superior oblique palsy – Etiology. The Strabismus Minute; Vol 2, no 15
Ocular Movements
Important determinant of what to operate, versions are an extremely important part of the diagnosis of superior oblique palsy. As in the figure, the patient has a left superior oblique palsy. Here, the most characteristic finding is a left inferior oblique (IO) over action and to a lesser extent, superior oblique (SO) under action (sometimes, this is slight or undetectable). The other eye has an apparent superior oblique over action. There is an ipsilateral superior rectus (SR) contracture (as seen in long standing cases).

Bielchowsky head tilt test/ Parks 3 step test
Helps in confirming the diagnosis by three steps i.e
Is it a left or right hypertropia?
Worsens in dextro or levoversion
Worsens in left or right head tilt
The Bielchowsky head tilt test is considered positive for superior oblique palsy when the vertical deviation increases with the head tilted towards the higher side. If the head tilt test reverses, a bilateral superior oblique palsy is suspected.

Double Maddox Rod test
The details of the procedure of this test are not discussed here. The interpretation:
• Subjective torsion seen only in acquired palsy.
• < 10 degree torsion seen in unilateral traumatic palsy – patient does not complain, seen only on dissociation.
• > 10 degree torsion seen in bilateral palsies, patient complains of tilting.

Indirect ophthalmoscopy
Torsion can be seen objectively on examination with an indirect ophthalmoscope. If the macula is rotated downwards or clockwise in the left eye or counter clockwise in the right eye, so the macula is below a line drawn parallel to the orbit floor and temporal from the lower disc margin, torsion can be inferred.

Once the diagnosis of superior oblique palsy has been made or is suspected, it is necessary to measure the deviation - to quantify the superior oblique palsy and to classify it - then it is possible to arrive at a treatment program

DIAGNOSTICS
Include
• Examination of other cranial nerves
• Hess Charting
• MRI scan

MRI
Most patients with isolated SO palsy do not need an extensive neurological workup. As most palsies are congenital, the characteristic facial asymmetry, a review of old family photographs showing a head tilt (the FAT scan) or eliciting symptoms of a long duration is sufficient to rule out an acute acquired process.

Neuroimaging may however be required in cases of:
– Fresh traumatic
– Acquired palsies not directly linked to trauma
– Non isolated palsies

LOCALISATION
The fourth nerve nucleus is in the rostral part of the mid brain in the tectum. The nerve fibres originate from the dorsal part of the brain stem and decussate. Delicate nerve fibrils then pass through the tentorium and pass into the superior orbital fissure to supply the superior oblique muscle. These delicate fibrils are vulnerable to to and fro movements of the brain during sudden deceleration or violent head trauma.

TREATMENT
Superior oblique palsy seen as an incidental finding in a patient without torticollis or symptoms does not require any treatment.
Prisms have a limited role in the management of superior oblique palsy and their role is limited to:
- Very small comitant deviations, usually vascular
- Non surgical candidates.

Most patients presenting to the ophthalmologist with SO palsy do so because of troubling symptoms and are usually surgical candidates.

**KNAPPS CLASSIFICATION**
Knapp in his landmark paper on classification of superior oblique palsy emphasized the importance of the relative magnitude of hyperdeviation in the various fields of gaze. Although many of the treatment recommendations may not be followed at this point of time, the importance of his classification lies in the basic message that the muscle to be treated should be matched to the gaze in which the deviation is the maximum. To summarize,
- Classified SO palsy into 7 groups depending on gaze primarily affected
- Treatment paradigm based on the main muscle involved
- Table enumerates Knapps recommendations

<table>
<thead>
<tr>
<th>Knapp I</th>
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<tbody>
<tr>
<td>Overaction of Antagonist IO</td>
</tr>
<tr>
<td>surgery: Weaken antagonist IO</td>
</tr>
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<table>
<thead>
<tr>
<th>Knapp II</th>
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<tbody>
<tr>
<td>Underaction of the paretic SO with the deviation greater in its the field of action</td>
</tr>
<tr>
<td>Surgery : SO tuck, IO weakening, or yoke Inferior Rectus (IR) Recession</td>
</tr>
</tbody>
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<thead>
<tr>
<th>Knapp III</th>
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<tbody>
<tr>
<td>Deviation equal in the field of the paretic SO and the antagonist IO</td>
</tr>
<tr>
<td>Surgery:</td>
</tr>
<tr>
<td>&lt;20pd: Weaken the IO</td>
</tr>
<tr>
<td>&gt;20 pd add SO tuck if tendon lax</td>
</tr>
<tr>
<td>Or contralateral IR weakening</td>
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<tr>
<th>Knapp IV</th>
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<tbody>
<tr>
<td>Spread of hyper deviation ‘across the bottom’: tightness of the ipsilateral SR</td>
</tr>
<tr>
<td>Surgery &lt;20pd weaken the IO</td>
</tr>
<tr>
<td>&gt;20 pd add SO tuck if tendon lax Or contralateral IR weaken</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Knapp V</th>
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<tbody>
<tr>
<td>Hyper deviation ‘across the bottom’ - long-standing acquired</td>
</tr>
<tr>
<td>Surgery &lt;20 pd SR rec +SO tuck/ IO rec/ C/L IR rec Contralateral SO WEAKENING as originally recommended by Knapp is a STRICT NO</td>
</tr>
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</table>

<table>
<thead>
<tr>
<th>Knapp VI</th>
</tr>
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<tbody>
<tr>
<td>Bilateral superior oblique palsy</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Knapp VII</th>
</tr>
</thead>
<tbody>
<tr>
<td>Brown syndrome with superior oblique underaction (‘CANINE TOOTH’)</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Surgery:</th>
</tr>
</thead>
<tbody>
<tr>
<td>None -- if eyes are aligned around primary</td>
</tr>
<tr>
<td>Yoke IR Recession -- if ipsilateral hypertropia</td>
</tr>
<tr>
<td>Take down tuck, if caused by a ‘too tight’ tuck</td>
</tr>
<tr>
<td>Free SO restriction if ipsilateral hypotropia</td>
</tr>
</tbody>
</table>

Present day treatment plan incorporates most of these criteria plus adds
- Significance of superior rectus contracture
- Superior oblique laxity
- Quantification of superior oblique laxity
- Management of torsion
**Oblique traction test**

Intraoperative testing of the superior oblique is an indispensable part of evaluating patients with superior oblique palsy. Traction testing is important in identifying not only the lax tendons that need to be tucked but also the normal length tendons that cannot be tucked.

**Technique**

**Step 1:**
Surgeon seated above the head of the supine, anaesthetized patient
Two toothed forceps taken
Limbus grasped at 2 and 8 o clock position in the left eye and the 4 and 10 o clock position in the right eye

**Step 2:**
Eye is rotated up into an elevated, adducted position in the superior nasal quadrant while simultaneously pushing the globe down towards the orbital apex

**Step 3:**
Once the tendon is put on stretch, the eye is moved back and forth (temporally and nasally) while maintaining the tendon taut

**Step 4:**
Step 1 to 3 are repeated in the fellow eye
The relative laxity of the tendon should be compared with the other eye. A subjective grading scale is also available.

This test is more valuable in children; the vigorous maneuver may easily tear the fragile conjunctiva in adults.

**TREATMENT ALGORITHM**

Although most surgeons follow the rule to operate in the muscle involved in the position of maximum deviation, the following is a useful algorithm to determine which muscle to operate.

**BILATERAL PALSY**

Management is controversial. Some suggested procedures:
- BE Yoke IO weakening
- BE IR weakening
- Harada Ito procedure
- BE Down shift of the MR for V

**References**

1. Helveston Eugene M; Superior oblique palsy – Etiology. The Strabismus Minute; Vol 2, no 15
2. Von Noorden G K, Campos E C. Binocular Vision and ocular motility; theory and management of strabismus. 6; Mosby; 312-320
SUPERIOR OBLIQUE PASLSY MANAGEMENT

TRACTION TEST

Absent Tendon

Markedly Lax

Every one Else (Including Acquired Unilateral)

1) RECESS SR
2) WEAKEN IO

1) TUCK SO TO MATCH 'NORMAL' SIDE
2) WEAKEN IO

INFERIOR OBLIQUE OVERACTION

Yes

No

HT <= 15 PD

Yes

WEAKEN IO

SR RESTRICTION?

Yes

No

1) RECESS SR
2) WEAKEN IO

1) RECESS IR (CONTRALAT)
2) WEAKEN IO

Yes

No

CONTRALAT IR RECESS

IPSILAT SR RECESS

HT <= 15 PD

Yes

No

CONTRALAT IR RECESS

IPSILAT SR RECESS
Management of Corneal Trauma

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Introduction
Trauma to the eye can have various components. It includes trauma to extraocular structures and globe and its intraocular contents. Extraocular injury includes injury to lids, extraocular muscles, orbital margin, orbital floor, face or more serious intracranial injury. Injury to globe includes injury to cornea, sclera, crystalline lens, or the posterior segment. Discussion in this article is limited to corneal and corneo-scleral lacerations.

A corneo-scleral laceration may be partial- or full-thickness injury. A partial-thickness injury does not violate the globe of the eye (abrasion) and may or may not require surgical repair. A full-thickness injury penetrates completely through the cornea, causing a ruptured globe. It may also be associated with lid laceration (Figure 1).

Epidemiology
International epidemiology of penetrating eye injury proves that most people susceptible to trauma are males, between the ages of 20 and 30 years, associated with alcohol ingestion, injury occurring at work, home, during fight and road traffic accident. Be aware of any local activities or traditions that may also predispose the local population to corneal injury like bow arrow injury during Dhussera or fire cracker injury. Cause of injury could be metal fragments, sharp objects, fingernails, air-bag deployment, explosions, blunt force trauma and pellets.

Principles of management of ocular injury:
1. Rule out life threatening injury
2. History
3. Examination
4. Investigation
5. Treatment

1. Rule out life threatening injury to brain, cardiorespiratory system and check vital parameters: Ocular injury may be associated with intracranial injury. Examine the systemic condition by Glasgow Coma Scale assessment, Neurological assessment and CT scan.
2. Patient presenting with acute symptoms may give history of preceding injury. Some patients have insidious injury and later present with secondary infections or complications. Small foreign bodies, digital trauma, or other more subtle sources of damage may not be quickly recalled by the patient. Document information such as the setting of the injury, changes in vision, or sensation of a foreign body in the eye. Foreign body should be suspected and ruled out. Even blunt objects can cause corneal lacerations. All trauma cases are to be treated as medico-legal cases, hence all precautions such as consent, clinical photographs, X rays should be done.

Obtain the patient’s pre-injury vision assessments as well as any history of previous ocular problems or ocular surgery. History of old injury in the same or other eye may be important. Patients usually complain of decrease in vision, watering, photophobia, bleeding and pain. Document pertinent medical history, current medications, allergies, and tetanus immunization status. Determine information regarding the patient’s last oral intake if operative intervention is anticipated.

Examination
It is important to examine both the eyes. Sometimes only the symptomatic eye is given due consideration. The physician must be meticulous in examining the cornea and periorbital structures if there is suspicion of a corneal laceration. Typically, patients who present with this type of injury complain of an
intensely painful, profusely lacrimating eye. The bulbar conjunctiva will be injected with prominent blood vessels.

The first priority in evaluating a corneal injury is to include or exclude a full-thickness injury and the resulting ruptured globe. A full-thickness injury will allow aqueous humor to escape the anterior chamber, which can result in a flat-appearing cornea, air bubbles under the cornea, or an asymmetric pupil secondary to the iris protruding through the corneal defect.

Complete ocular examination including vision, lids, extraocular movements, light reflex, swinging light test, lens clarity, anterior chamber depth, intraocular pressure (not in obvious globe perforation), distant direct reflex, vitreoretinal examination must be done.

**Assess Vision**

Assessment of visual acuity must be performed, taking care not to apply additional pressure to the globe.

- If possible, determine visual acuity prior to examination or treatment, and separately test each eye with corrective lenses. Pinhole testing may help differentiate refractive error from uncorrectable vision when spectacles are not available.
- Use age-appropriate vision testing devices such as the Snellen distance chart or a hand-held vision card.
- Ask the patient to identify typed letters, the clock, or objects on the wall if the patient’s condition prevents formal (standing upright) testing.
- If a patient’s vision is severely limited, determine whether the patient can count fingers, detect movement, or identify light.

**Slit lamp examination**

- Examine the cornea using a slit beam to search for anterior chamber penetration. A shallow anterior chamber, irregularly shaped pupil (teardrop shape), hyphema (blood in the anterior chamber), bubbles in the anterior chamber, or a flat cornea can be signs of corneal perforation.
- Aqueous humor leaking from the anterior chamber can be identified by performing a Seidel test. This test is performed by directly applying fluorescein to the suspected corneal lesion. Visualization of diluted dye under a black light (a positive test) suggests a leak. A negative Seidel test (no dilution of fluorescein) suggests a partial-thickness injury but may be seen in small or spontaneously sealing lesions. One should avoid the temptation to press on the globe to test for a self-sealing injury.
- Be sure to evaluate for a foreign body in the anterior chamber, especially if the patient’s history suggests that the corneal laceration is from a small, high-speed object (such as from hammering metal).

- **Full-thickness corneal lacerations**
  - Do not apply pressure to the globe.
  - Immediately place a protective shield (not a patch) over the affected eye.
  - Instruct the patient to avoid ocular movement because extraocular muscle contraction can cause extrusion of intraocular contents.
  - Full-thickness corneal lacerations often result in a loss of aqueous humor producing a shallow or flat anterior chamber.
  - Prolapse or incarceration of the iris may produce a teardrop distortion of the pupil.
  - Hyphema, or blood in the anterior chamber, can be another sign of anterior chamber penetration. Hyphema may also be seen with blunt, nonpenetrating trauma. (Figure 2)

Once a diagnosis of corneal laceration is made, one must also consider the simultaneous presence of the following:

1. Corneal foreign body
2. Endophthalmitis
3. Intraocular foreign body
4. Posterior chamber injury
5. Retinal injury
6. Orbital injury
7. Lens injury

**Investigations**

**Laboratory Studies**

- No lab studies are useful for detecting the presence of a corneal laceration.

**Imaging Studies**

- Radiography, CT, or MRI may be indicated to locate
Management of Corneal Trauma

intraocular or intraorbital foreign bodies or associated orbital, cranial, or facial trauma. MRI may be of some use if there is suspicion of an organic foreign body (eg, wood, plant matter). However, avoid MRI if there is any possibility of a metallic foreign body in the eye. Ultrasound biomicroscopy may be required to detect foreign body impacted in the ciliary body or iris root. 4,5

Treatment

Prehospital Care
• Cover the patient’s eye with an eye shield or polystyrene/paper cup and avoid any pressure to the globe.
• Instruct the patient to move the eyes as little as possible.
• Administer tetanus toxoid injection, antiemetic and analgesic medication in order to reduce pressure on the globe.

Emergency Department Care
Perform an examination to ascertain the extent of the corneal, anterior chamber, ocular, and associated (eg, facial, cranial) injuries.
• Place a protective eye shield (prefabricated or custom made) on the injured eye. This can be a commercial plastic eye shield or simply a polystyrene/paper cup taped over the eye.
• Administer antiemetics and systemic analgesic medication.
• Tetanus immunization or booster is indicated.
• Consider use of antibiotics including route (topical or intravenously) and frequency.
• In general, topical analgesia and antibiotics should be avoided if a corneal laceration is suspected. Use systemic analgesia and antibiotics. Topical anesthetics may be used, if needed, to facilitate visual acuity testing and the slit lamp examination.

Medication
Recommendations include a combination of a cephalosporin (eg, cefazolin) or vancomycin and an aminoglycoside (eg, gentamicin). In addition, add clindamycin if an intraocular foreign body is present or if vegetable matter has contaminated the wound. The most common organisms identified in posttraumatic endophthalmitis are Staphylococcus epidermidis, bacilli species, streptococci species, and gram-negative species. Fungal endophthalmitis is a relatively rare entity but should be considered in a patient who is recently post-surgical, immunocompromised, unresponsive to antibiotic treatment, or has a history of trauma with vegetable matter.

Antibiotics
Cefazolin: 500 -1000 mg IV q8h; (not to exceed 1 g q6h); Pediatric : 25-50 mg/kg/d IV divided TDS/QID.
Clindamycin: 600-1200 mg/d IV divided BD/QID (not to exceed 4.8 g/d), Pediatric: 20-40 mg/kg/d IV divided bid/qid
Vancomycin: Adult:2 g/d IV divided BD/QID, Pediatric: 30-40 mg/kg/d IV divided BD/TDS

Techniques of repair of Corneal Laceration:

Principles of repair:
Surgical re-approximation of the ocular tissues.
Re-obtain ocular integrity
Avoid surgical damage to lens and iris
Minimal residual astigmatism

a. Lamellar injury;
Examine completely under high slit lamp magnification. Perform Siedel’s test with 2% fluorescein dye and apply gentle pressure if required.
Management options include: pressure patching, bandage contact lens, tissue adhesives6 and suturing may be required in children, unstable deep wounds.

b. Full thickness injury;
CorneoScleral injury:
Then cornea and later the scleral is sutured with exploration of the rest of globe. Scleral should be sutured with 6-0 vicryl and conjunctiva with 8-0 vicryl sutures. 7,8

Corneal injury:
Simple corneal full thickness injury may be tackled with bandage contact lens, tissue adhesives6 or suturing.

Figure 3: Tri-radiate corneal laceration
All corneal lacerations induce flattening of cornea and its repair would lead to astigmatism. Cornea incision should be closed with 10-0 monofilament nylon.\textsuperscript{7,8}

**Following factors need to be considered:**

Compression zone: Each suture produces a zone of compression which is triangular in shape; on either sides of the suture. The zones of compression of subsequent suture should overlap each other. The zone of compression is roughly equal to the length to the suture. Longer and deeper sutures appose deeper stroma, closing any posterior gaping and decrease the propensity for iris or vitreous to adhere to posterior corneal surface. Ideal length of suture should be approx 1.5 mm.

Eversion/Inversion effects: For a single interrupted suture, inverting and everting forces are equal, so there is no tendency toward tissue depression or elevation. A non-radial interrupted suture produces tissue torque.

Suturing should prevent over-riding of tissues, minimize torque and residual astigmatism and obtain water tight closure.

The corneal laceration is considered as a sum up of multiple perpendicular segments. Each laceration segment needs to be sutured. The suture needle exit and entry sites should be equidistant from the incision.

It is important to place the suture at equal depth on both sides of the laceration, approximately 90% depth (Figure 4).

**Order of suture placement:**

Peripheral suture should be placed first to flatten the peripheral cornea and steepen the central cornea. (Figure 5a)

The central cornea is then closed with a gentle appositional suture just sufficient to close the anterior surface.

At the central area, sutures bites should be shorter and no-touch technique of suturing should be used. (Figure 5b)

All suture knots should be buried
After suturing wound leak should be checked.

**Figure 5:** Suturing Technique

- **Figure A**
- **Figure B**

**Corneal injury with iris incarceration:**

Attempt should be made to reposit and retain the iris tissue wherever possible without damaging it further (figure 6). Any devitalized, infected tissue should be excised by gently grasping the iris tissue. No pull should be applied. The iris is cut flush with the Vannas scissors and the cornea sutured accordingly. \textsuperscript{7,8}

**Figure 6: Corneal laceration with iris prolapsed**

**Corneal injury with Vitreous incarceration:**

Any vitreous strand should be cut with manual or automated vitrectomy cutter. The anterior chamber and wound should be free from any vitreous. Vitreous in wound can cause infection, vitreous traction, Cystoid macular edema, Retinal detachment.

**Intraoperative bleeding:**

Use of viscoelastic, epinephrine, cautery, or thrombin may help decrease bleeding.
Corneal injury with Lens involvement:
Cataract extraction along with primary repair should be avoided. Problems encountered are difficult preop assessment, biometry measurement and poor visualization. Second stage cataract surgery gives optimal results. If anterior capsule is breached and lens matter is present in anterior chamber, it should be removed.9

Post operative management:
Analogesics, topical and systemic antibiotics, cycloplegics, oral and topical steroids are given in post operative period.

Follow-up
• Follow-up care is determined at the time of the initial ophthalmologic consultation and will take place in the setting of the patient’s overall condition.
• An isolated partial-thickness corneal laceration may be monitored on an outpatient basis.
• A patient who sustains a corneal laceration as part of other trauma may have to be evaluated in the hospital.

Deterrence/Prevention
• Patients that engage in activities that place their eyes at risk for trauma should be encouraged to wear protective eyewear at all times.

Complications
• Corneal lacerations frequently are complicated by corneal or intraocular foreign bodies, infections, traumatic cataracts, and secondary glaucoma.

References:
**‘A’ AND ‘V’ PATTERN STRABISMUS**

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**Introduction** Pattern strabismus is vertically inconcomitant strabismus, recognized by change in the degree of horizontal deviation in upgaze and downgaze. This entity has been recognized since long time but much significance was not given which resulted in poor surgical outcome in such cases. Due to complex and varied presentation, surgical management of this phenomenon is rather complex.

**Definition**

In A-V pattern strabismus the angle of horizontal strabismus varies significantly while changing the gaze from elevation to depression or vice versa.

- **A pattern** - horizontal strabismus (esotropia or exotropia) whose angle changes more than 10 diopters between 30 degrees upward and 30 degrees downward gaze has an.
- **V pattern** - the variation has to be more than 15 PD when the gaze is changed from depression to elevation.

**Prevalance:**

Due to absence of uniform criteria there is wide variation in the incidence reported. The incidence reported is 12.5% - 87.7%. V esotropia pattern is more common followed in order of frequency by A esotropia, V exotropia and A exotropia.

**ETIOPATHOGENESIS:**

<table>
<thead>
<tr>
<th></th>
<th>V</th>
<th>A</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Esotropia</td>
<td>171 (41%)</td>
<td>105 (25%)</td>
<td>276 (65.5%)</td>
</tr>
<tr>
<td>Exotropia</td>
<td>97 (23%)</td>
<td>48 (11%)</td>
<td>145 (34.5%)</td>
</tr>
<tr>
<td>Total</td>
<td>268 (64%)</td>
<td>153 (38%)</td>
<td>421 (100%)</td>
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Table showing distribution of pattern strabismus

Various etiologies have been proposed but there is lack of consensus among the authors. The presumed etiopathogenesis is due to

1. **Oblique muscle dysfunction**

Torsional imbalance as a result of sagittalization of oblique muscles leading to secondary overaction or underaction of the corresponding cyclovertical muscles. This is the most widely accepted theory. Overaction of inferior obliques can cause a V pattern by increasing its abduction action in elevation and conversely superior oblique overaction can cause A pattern.

2. **Horizontal muscle dysfunction:**

Anomalous insertion of horizontal recti, medial recti higher and lateral recti lower than normal, causing V pattern and reverse in A pattern have been reported. For example underaction of both lateral recti leads to a weakening of divergence on looking up, causing A pattern esotropia and whereas overaction of the lateral recti causes V pattern esotropia.

3. **Vertical muscle dysfunction:**

Primary anomalies in the function of the vertical muscles has also been proposed. The tertiary action of the vertical recti is adduction. If superior rectus is underacting then in upgaze then adduction will also be diminished resulting in divergence in upgaze.
‘A’ And ‘V’ Pattern Strabismus

4. Anatomical anomalies of orbits, e.g., mongoloid or antimongoloid features and torsion of orbits

5. Anomalies of muscle insertions

6. Anomalies of muscle pulley

EXAMINATION:

Symptoms
Asthenopia and diplopia are the usual complaints. The increase in deviation in looking down is tolerated well as it does not interfere with the routine functions like reading or other near work. The increase in the deviation in downgaze may cause visual discomfort.

Work up
The usual assessment routine to any case having problems of ocular motility to be carried out in all the cases. Angle of deviation should be measured in at least 3 vertical directions as follows:

1) In 30 degrees elevation
2) In primary position (PP)
3) In 30 degrees depression

MANAGEMENT:
Management includes treatment of refractive error and amblyopia if present. Surgical management is done when indicated. The indications for the surgery are
1. Deviation manifesting 50 % of time or more and/or
2. Presence of sensory problems, e.g., suppression, defective binocular functions
3. When it poses a cosmetic problem
4. Requirement of normal binocular functions and ocular motility for certain professions

Aims of surgery:
(a) To achieve orthotropia in more practical and useful positions of gaze, i.e., in primary position and infraversion (looking directly downwards). The latter is the position in which most of the daily activities like walking, eating, reading and writing etc. are performed.
(b) To obtain comfortable binocular single vision (also in primary position and looking down).
(c) To correct abnormal (compensatory) head posture. This will automatically result if above two have been achieved.

Planning the line of management of pattern strabismus
- Exact measurements of the pattern (the change in the angle of deviation) from direct elevation (25-30 degrees) to direct depression (25-30 degrees) must be taken.
- Actions of the extraocular muscles should be investigated thoroughly through examination of ocular motility so that an overaction or underaction of a muscle is detected. Success of surgery may depend on this. Unless these problems are corrected response to treatment may not be as anticipated
- The results of the surgery may also be unpredictable because of anomalies of the muscle pulleys and abnormalities of the orbits.
- Oblique muscles should only be operated on if they show a significant degree of dysfunction (underaction or overaction).
- Vertical muscles should only be selected for surgery if other options like surgery on horizontal or oblique muscles have failed or are contraindicated
- In the absence of significant dysfunction of oblique muscles, surgery should be confined to the horizontal recti (vertical displacement with recession / resection according to the need).
- The action of a rectus is weakened in the direction it’s insertion is transposed, e.g., if the insertion of medial rectus is displaced downwards as is required in V pattern strabismus, adduction is weakened in depression
- Medial rectus insertion is always transposed towards the apex of A or V to abolish the convergence of the arms of the letters.
- Lateral rectus insertion is always displaced towards the base of A or V to straighten the divergent arms of the letters.
- As regards the surgery on oblique muscles it should be kept in mind that weakening of an overacting oblique muscle gives better results than does strengthening of an underacting oblique muscle.
- While planning surgery of multiple muscles specially recti one should remember that there is a possibility of anterior segment ischemia. Oblique muscles do not contribute to the circulation of the anterior segment.
deviations should be corrected in one sitting.
Choice of surgical procedures Surgical treatment on horizontal recti, vertical recti and oblique muscles has been advocated according to the muscle involved.
(1) Horizontal muscle surgery with vertical displacement of insertions and/or slanting the insertion of horizontal recti
(2) Oblique muscle surgery, in cases of significant oblique muscle dysfunction, e.g., inferior oblique overaction in V-pattern and superior oblique overaction in A-pattern.
Surgery of horizontal recti in the form of recession and resection procedures along with their upward or downward displacement. 7-10 Whenever the oblique muscle dysfunction is incriminated11,12 oblique muscles are operated. Vertical recti have also been operated upon to correct this pattern.13

1) HORIZONTAL MUSCLE SURGERY: combined with vertical transposition of their insertion. The Principle of the surgery is to weakened the action of the muscle in the direction in which its insertion is shifted. Eg: if the insertion of the lateral rectus is shifted downwards, its action i.e abduction will be weakened in downgaze.
V- pattern: lateral rectus insertion shifted upwards and medial rectus insertion downwards
A- pattern: medial rectus insertion shifted upward and lateral rectus insertion downward.
The amount of shift depends on the difference in the angle of deviation in midline vertical gaze. But usually a moderate amount of shift i.e half the length of the muscle insertion is done.

- Slanting the newly transposed muscle insertion for an added effect in case of a larger pattern so that selective weakening can be achieved in elevation or depression as required. eg., A pattern ET- The medial rectus has to be weakened more in up-gaze than in down-gaze to abolish the convergence of the arms of A. This can be achieved by transposing its insertion upwards and slanting the new insertion so that its upper end is further away from limbus than the lower end. Putting the upper end further back further weakens the adduction action of the medial rectus in elevation.

2) OBLIQUE MUSCLE SURGERY: done in adjunct to the horizontal muscle recession or resection surgery as indicated. It should be done only when there is a presence of oblique muscle overaction
V- pattern: weakening of overacting inferior oblique muscle
A- pattern: weakening of overacting superior oblique muscle
Inferior Oblique weakening can be done by:
- Myectomy - corrects about 20 prism dioptres of pattern
- Recession of the muscle insertion depending upon the amount of vertical incomitance
- Anterior transposition if associated with DVD
- Superior Oblique weakening
- Posterior tenectomy of the superior oblique (PTSO) for mild overaction (15 prism dioptres)
- Tenectomy for moderate overaction (20 prism dioptres)
- Translational recession for 25 prism dioptres of A pattern

COMPLICATIONS OF A-V PATTERN STRABISMUS SURGERY:
1) Iatrogenic vertical and torsional strabismus can occur when a normal oblique muscle is operated upon
2) Vertical and torsional diplopia
3) Superior oblique palsy
4) Consecutive overaction of the direct antagonist muscle
5) Compensatory head posture following the above complications
6) Anterior segment ischemia following vertical recti surgery
7) Persistence of oblique muscle overaction

V pattern exotropia

V pattern esotropia

A primary or secondary overaction of inferior oblique can cause a V pattern by increasing its abduction action in elevation.

A Et

C) Conversely, weakness of inferior oblique or overaction of superior oblique may cause A pattern strabismus by weakening abduction in elevation and strengthening it in depression.
SUMMARY:
Treatment of pattern strabismus is more complicated and difficult than the horizontal comitant strabismus. The aim of the surgery is to reduce the pattern and to achieve fusion in primary and downgazes. The results of surgery are rather unpredictable, especially in case of combined surgery (recti and oblique). Hence, careful planning of surgery is to be done, most important is to determine preoperatively whether there is an abnormality in the action of the oblique muscles.

References:
CURRENT MANAGEMENT OF RETINOPATHY OF PREMATURITY

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INTRODUCTION

Retinopathy of Prematurity (ROP) is a vasoproliferative disorder of the retina affecting premature babies. As more advanced neonatal centers continue to save smaller premature babies, the incidence of ROP is on the rise, and has become an important cause of childhood blindness across the world.

However, in developing countries there is a major lack of awareness about ROP among ophthalmologists and neonatologists, due to which a large number of these children are not screened and left untreated; to be later detected with advanced ROP, when prognosis becomes poor. As awareness is spreading about ROP, more centers have started establishing ROP screening programs to detect and treat this blinding disease in a timely manner and thereby reduce childhood blindness.

The international classification of retinopathy of prematurity (ICROP) group has clearly classified ROP into zones and stages to enable both beginners and experts managing ROP to communicate easily among themselves. Definitive ROP screening guidelines have been established to detect treatable ROP in a timely manner.

The American screening guidelines recommend screening babies <1500g birth weight or <32wks GA or higher in case of unstable babies considered high risk to develop ROP by neonatologist. They suggest the first screening be done at 31wks PCA or 4wks after birth, whichever is later. Indian expert group suggests screening babies <1500g birth weight or <34-35wks GA or higher in case of unstable babies considered high risk to develop ROP by neonatologist. They recommend first screening be done at 31wks PCA or 4wks after birth, whichever is earlier (even at 2-3 weeks in VLBW babies).

TREATMENT OF ROP

Timely screening and prompt treatment is the key to success in ROP. If treated in time, ROP mostly regresses uneventfully and has good visual outcome. The current treatment strategy is guided by The Early Treatment For Retinopathy Of Prematurity Cooperative Group (ETROP) study. ETROP group divides these eyes into 2 categories –

- Type 1 ROP - defined as
  - zone I, any stage ROP with plus disease or
  - zone I, stage 3 ROP without plus disease or
  - zone II, stage 2 or 3 ROP with plus disease.
- Type 2 ROP - defined as
  - zone I, stage 1 or 2 ROP without plus disease or
  - zone II, stage 3 ROP without plus disease.

Type I ROP requires prompt treatment, while Type 2 ROP is regularly followed up and considered for treatment only if it progresses to Type 1 or threshold ROP.

It is essential here to be aware of the newly defined terminology called Aggressive Posterior ROP (AP-ROP) which signifies a rapidly progressive, severe form of ROP in zone I or zone 2 posterior, which is characterized by prominent plus disease, ill-defined retinopathy with flat neovascularization, deceptively featureless junction between vascularized and non vascularized retina, and can rapidly progress to stage 5 ROP. It is essential to be aware of AP-ROP, and treat it promptly when present.

Retcam assisted fluorescein angiography in ROP has emerged as a useful investigative modality which helps to distinguish vascular from avascular retina, identify causes of persisting plus disease, identify skip areas of treatment and detect early neovascularization in AP-ROP enabling timely treatment of these cases. Another interesting area of development has been the computerized analysis of vascular tortuosity. This helps in a better understanding of plus disease and detects changes suggestive of severity or progression of ROP.

Laser Photocoagulation

Laser Photocoagulation has become the treatment of choice for treatable ROP when indicated. The basic principle of laser treatment is the ablation...
of the avascular retina to remove the ischemic stimulus stimulating neovascularization. Laser photocoagulation in eyes of premature babies is done using a laser indirect ophthalmoscopic delivery system with preferably Green or Diode lasers. It is a time intensive procedure and requires trained experts to perform it.

The treatment aims to laser the entire avascular retina, while sparing the ridge, and completing the procedure in one sitting in both eyes. Since laser treatment is a stressful procedure, it requires regular vitals monitoring by a neonatologist. Regular weekly follow up is required till the disease regresses. If laser skip areas are noted, laser augmentation of these patches may be needed. Sometimes the ROP may progress to advanced stages and then require surgical intervention.

Cryotherapy

Before laser photocoagulation gained popularity, Cryotherapy was the preferred treatment and was used to ablate the avascular retina. The main disadvantage of Cryotherapy was it required general anesthesia, which carried a high risk in premature babies. Though cryotherapy allowed faster treatment, yet the treatment parameters were less controlled and it led to extensive chemosis, congestion, cataract, pain and prolonged morbidity and was often associated with more iatrogenic ocular complications. However, cryotherapy is still used at some centers because cryo machine is much more economical compared to the laser equipment, but laser is the treatment of choice now.

Scleral Buckling / Lens sparing vitrectomy

Advanced ROP progressing to Stage IV and beyond requires surgical intervention, and the results and prognosis becomes poorer with disease progression. ROP progression is often due to lack of screening, delayed treatment or progression despite laser, especially in cases of AP-ROP.

Scleral buckling is performed in stages 4A and 4B with the aim to relieve vitreoretinal traction and help in absorption of subretinal fluid. A #240 encircling band / buckle is passed (2.5mm wide) and anchored to the sclera with anchoring sutures. This encirclage requires removal after a few months to allow the eye to grow normally. Scleral buckling has led to favorable results and is widely used.

With advent of modern vitreoretinal surgical techniques, lens sparing pars plana vitrectomy helps to relieve the vitreoretinal traction better internally, and is proving more effective than scleral buckling in selected cases with good results. Now three port 25-gauge transconjunctival sutureless vitrectomy is also emerging as an effective technique to attach the retina in patients with stage 4 retinal detachment in ROP.

Vitreoretinal surgery

Advanced stage 5 ROP requires vitreoretinal surgery by experienced vitreoretinal surgeons and is available only at few tertiary centers. Surgery basically involves lensectomy, dissection and separation of retrolental membranes, removal of vitreoretinal traction from center to periphery to allow the retina to settle. The aim of surgery is an attempt to attain ambulatory vision for the child so they can perform their activities of daily living.

Various diagnostic modalities like ultrasonography can help assess the retinal funnel configuration in the anterior and posterior parts. Open-open funnel configurations have the best surgical outcome, while narrow-narrow configurations fare poorly. UBM has also been found to have a useful role in detecting anterior surgical space and determining the site of instrument entry.

However, in spite of best surgical efforts, the anatomical and visual outcomes are relatively poor. The poor prognosis and bad surgical outcomes are often complicated by associated ocular disorders like glaucoma, uveitis, posterior synechiae etc.
Anti-VEGF drugs

Often cases of aggressive posterior ROP or those with delayed presentation / treatment may continue to progress, and there has been a need for an adjunct to laser which can help reduce the vascularity and control progression of ROP.

Due to the success of Anti-VEGF agents like Bevacizumab, Pegabtanib sodium in various neovascular ocular pathologies like wet AMD, diabetic retinopathy, Eales diseases, venous occlusions etc. – many ophthalmologists started trying these agents in ROP with some success. Preliminary studies have demonstrated useful effect of bevacizumab and Pegabtanib in pilot trials since these agents have helped to reduce vascularity, neovascular proliferation and help control the disease process. 12,14

It is important to realize that these drugs are not FDA approved for treatment of ROP, and may have long term side effects following systemic drug absorption which may block normal vasculogenesis of vital organs. Moreover, the complications of repeated intravitreal injections are also of major concern. Thus, there is a need for large scale multi-centric trials to test their efficacy in treatment for ROP. Some notable studies underway investigating into their role include Pan-VEGF Blockade for the
Treatment of Retinopathy of Prematurity (BLOCK ROP) trial in which premature newborn infants with bilateral progressive APROP despite complete peripheral retinal ablation will be given Bevacizumab administered in a single dose; and will assess its safety, efficacy in treatment of ROP. Another notable trial is Bevacizumab Eliminates the Angiogenic Threat of Retinopathy of Prematurity (BEAT-ROP) trial, a prospective, randomized, controlled, multi-centric study where intravitreal Bevacizumab injections versus conventional laser will be compared for vision-threatening ROP (acute stage 3 ROP in zone I or posterior zone II with plus disease).

These study results will give clearer guidelines for the role of anti-VEGF agents in ROP. Till then its usage must be restricted to select progressive APROP cases as an adjunct to laser, after ethics clearance and detailed informed consent.

In conclusion, it is best to run effective screening programs to ensure early detection and timely treatment in children with ROP. As ROP progresses, the treatment outcomes get poor. Therefore, spreading awareness is vital to tackle this preventable cause of childhood blindness.

REFERENCES:
Goldenhar-Gorlin syndrome

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Goldenhar syndrome was first described by Maurice Goldenhar in 1952 in a subject having triad of accessory tragi, mandibular hypoplasia and epibulbar dermoid present since birth. Subsequently it was named as oculo auriculo vertebral dysplasia (OAVD) by R.J. Gorlin (1963) due to the presence of additional vertebral anomalies.

Case report:
A six months old male child was brought to the eye OPD by his parents with the complaints of yellowish white elevated mass in left eye with a gap in the upper lid since birth.

The parents (nonconsanguinous marriage) gave no positive family history of such condition. They had two elder daughters having no abnormality. No history of fever or any other health ailment of the mother during pregnancy period was reported. There was no history of maternal smoking, alcoholism or any toxic drug intake. However the mother had some facial characteristics of prominent and bossed frontal bone, telecanthus, prominent eyes with antimongoloid slant, small sharp nose and pointed chin but no dental abnormality.

On ocular examination of the baby, there was upper lid coloboma of the left eye on medial 1/3rd and a epibulbar dermoid at 3 to 6 O’ clock position (8.5 mm diameter, hemispherical) encroaching up to half of the pupillary area. The dermoid had no superficial hair growth on it. The baby was taking fixation preferentially with his right eye. Ocular examination including pupillary light reaction and fundus examination was normal in both the eyes.

There were three preauricular skin tags on the left side and one on right side. One more skin tag with blind skin fistula was present on left cheek over the line joining angle of the mouth and left tragus.

On general and systemic examination the child showed no abnormality except a bluish grey colored naevus at the nape of the neck.

No abnormality was found on cardiovascular and central nervous system examination. The baby was playful and of normal developmental milestones. With Otorhinolaryngological evaluation, other than preauricular skin tags, there was no sign suggestive of external, middle or internal ear abnormality. Nose and oropharyngeal examinations were normal.

Investigation:
i) X-ray spine showed hemi vertebra of S1 and lumbosacral scoliosis, but cervical spine was absolutely normal.
ii) X-ray mastoid (Schullars view) showed hypoplastic mastoid air cells bilaterally [normal at this age].
iii) USG abdomen found no organomaly.

The clinico-radiological triad of epibulbar dermoid, preauricular skin tags and vertebral anomalies were consistent with the diagnosis of oculo-auriculo-vertebral dysplasia (OAVD), which is a subtype of oculo-auriculo-vertebral spectrum (OAVS) or facio-auricular-vertebral sequence (FAV) or Goldenhar sequence or Goldenhar syndrome. This reported case of OAVD also has unusual features of lid coloboma and congenital scoliosis due to sacral hemi vertebra.

Discussion:
Many authors consider OAVD and Goldenhar syndrome to be synonymous while others believe that the syndrome encompasses two broad subgroups namely OAVD and hemi facial microsomia. This view is based on developmental similarities which are a dysplasia of first and second branchial arches and intervening first pharyngeal pouch and branchial cleft in both the subgroups. That is why Goldenhar syndrome is also known as first and second branchial arch syndrome. The dysplasia is believed to occur either due to the teratogen exposure or a vascular disruption in intrauterine life or maternal diabetes. A new theory put forward for this developmental anomaly states that OAVS occurs as a result of ectodermal non disjunction early in embryonic development during 4th to 8th weeks of intrauterine life with subsequent mesodermal tethering. This theory explains multisystem involvement in OAVS.

The incidence of Goldenhar syndrome has been reported to be between 1:3500 and 1:5600, with a male: female ratio of 3:27. The incidence is
higher, about 1:1000, in children with congenital deafness. It mostly occurs as sporadic case but familial incidences are also noted suggestive of autosomal dominant or recessive inheritance. In addition, some researchers suggest that the disorder may be caused by the interaction of many genes, possibly in combination with environmental factors - multifactorial inheritance. Few other authors have reported its association with trisomy of chromosome 22 and chromosome 9q45. Variable phenotypic expression is the characteristic of this syndrome & the spectrum of phenotypic abnormalities can range from mild to severe even within the same affected family. There are cases of identical (monozygotic) twins in which only one has the syndrome, even though they received the same genetic blueprint; whilst other researchers describe examples of siblings having Goldenhar Syndrome. There are a few families with an affected person having a 50% chance of passing on the syndrome. Family history may include cleft lip or palate, unusually shaped ears, asymmetry of face, small chin, skeletal problems, eye abnormalities, internal organ anomalies or speech and dental problems. The abnormalities are found to be mostly unilateral in 85% of cases and bilateral in 10-33% cases. Kulkarni VV, Shah MD & Parikh AA reported, in Goldenhar syndrome, ocular anomalies especially bilateral dermoids are seen in 60% of the cases, vertebral anomalies in 40% of the cases and ear anomalies also in 40% of the cases.

The various features of Goldenhar syndrome or OAVD:

**a) Ocular:**
Epibulbar dermoids are the commonest abnormality found in about 75% cases. In rare cases they may be bilateral. These are classically located at infero temporal quadrant. These are choriostomas; i.e. Nest of normal tissue at abnormal places. Lipodermoids are present in less than 50% of cases. Dacryocystitis has also been reported. The other rarer ocular associations are upper lid coloboma, microphthalmos, cataract, iris & choroid coloboma, tilted disc, optic nerve hypoplasia, squint, ocular motility problems and anophthalmos.

**b) Auricular:**
Preauricular skin tags, pits/sinuses and accessory auricles are most common features; usually unilateral, but can be bilateral in 33% cases. Other external ear abnormalities range from dysomorphic ear to microtia to anotia (rare, reported by Jaison and Batra). Middle ear may be narrow or atretic with hypoplasia of air cells. Inner ear abnormality includes conductive &/or sensorineural deafness.

c) **Vertebral:**
Involvement of axial skeleton (vertebrae & ribs) has been observed in 24% of patients in the largest series reported by Rollnic et al. Spina bifida is the commonest and least severe of all anomalies. The vertebral abnormalities are known to range from hypoplasia & occipitalisation of Atlas, fusion of posterior element of cervical vertebrae, block vertebra, spina bifida occulta, hemi vertebra, and butterfly vertebrae to complete sacral agenesis. Vertebral anomalies are more commonly seen in cervical region & cervical lordosis has been reported. Congenital scoliosis may occur along with hemi vertebra. Rare case report of kyphosis & scoliosis with hemi vertebra in Goldenhar syndrome has also been reported.

d) **Facio-dental & skull anomalies:**
Hypoplasia of malar bones, zygomatic arch, maxilla and mandible may be present; characteristic of hemi facial microsomia. Some rare associations recorded in the literature are macrostomia, micro-gnathia, high vaulted palate, bifid tongue, cleft palate, malocclusion and other dental anomalies and hydrocephalus.

e) **Other organ anomalies:**
Goldenhar syndrome may present as a complex of craniofacial anomalies associated with vertebral, cardiac, neural, pulmary and genitourinary abnormalities. Although not well reported, up to 70% of cases have urogenital anomalies that include fused kidneys, renal agenesis, vesicoureteric reflux (VUR), pelvi-ureteric junction (PUJ) obstruction, ureteral duplication, multicystic kidney and retrocaval ureter.

The presence of heart disease in such patients accounts for 50% incidence. Tetralogy of Fallot (TOF) and ventricular septal defects (VSD) are the most common cardiovascular anomalies associated with OAVS2. Multiple cardiovascular malformations including Wolff-Parkinson-White (WPW) syndrome, a partial anomalous pulmonary venous connection, patent ductus arteriosus (PDA), an anomalous origin of the coronary arteries, and a right sided descending aorta were revealed by electrocardiography, echocardiography and cardiac catheterization. Goldenhar syndrome is quite rare, but the frequency of cardiovascular malformations in this syndrome is 5-58%. It is necessary to perform a careful evaluation of general malformations, especially cardiovascular malformations.
calcification of falx cerebri, undescended testes, Turner’s syndrome and glaucoma are some rarer reported associations. In approximately 5 to 15 per cent of affected individuals, mild mental retardation may also be present.

A case has been reported with ocular motility disturbances and congenital heart disease as rare association of Goldenhar syndrome. Non axial skeletal involvement in the form of thumb hypoplasia along with Goldenhar-Gorlin syndrome has also been reported.

The syndrome may require to be differentiated from the following conditions:

Other syndromes associated with multiple preauricular tragi include Treacher-Collins syndrome, Wolf-Hirschhorn syndrome, Nager’s acrofacial dysostosis, Wildervanck syndrome (cervico-oculo-acoustic syndrome), Townes-Brocks syndrome and Delleman syndrome. Treacher Collins syndrome is associated with maxillary and mandibular hypoplasia but associated with less common occurrence of eye and ear anomalies. In this syndrome coloboma affect lower eyelids rather than upper eyelids. Ear anomalies may also occur in Pierre Robin syndrome, Moebius syndrome and thalidomide embryopathy. However, these conditions have their own distinguishing features (e.g. marked mandibular hypoplasia in Pierre Robin syndrome, sixth and seventh nerve palsies in Moebius syndrome and phocomelias and amelias in thalidomide embryopathy).

**Prognosis and management:**

Children with Goldenhar Syndrome usually look forward to a long life and normal intelligence. Any systemic anomaly should be promptly identified & managed appropriately.

Special tests such as ultrasound, x-ray, CT scan or MRI may be required in order to obtain complete information about a given case. Hearing aids, Speech therapy, Orthodental treatment and Physiotherapy may be the integral parts of management.

The structural anomalies of the eyes and ears in Goldenhar syndrome can be corrected by plastic surgery. Surgery is usually postponed until the growth of the structure is completed. The epibulbar dermoids should be removed.

**Pre-natal diagnosis:**

Scanning may identify the condition in certain cases where facial or skeletal anomalies are present. Antenatal sonographic detection of OAVS has also been reported by Witters et al and other workers. Pre-natal screening and genetic advice may be offered for future pregnancies.
Consent:
Written consent was taken from the parents of the patient to publish the report and photographs.

References:
A 27 year old male patient presented with sudden onset of pain, redness, photophobia and diminution of vision in left eye since 10 days, no past history of ocular injury.
At presentation visual acuity in right eye was 6/9 and that of left eye was HM +ve; on slit lamp examination corneal ulcer with hypopyon was diagnosed.
On general examination maxillary hypoplasia, parrot beak nose, broad hands with short fingers, acanthosis nigricans were present. X-ray cervical spine, skull, hands and foot showed typical picture of Crouzon syndrome.
Because of rarity of presentation case is being reported.

Key Words: Crouzon Syndrome

Introduction
• It is a part of a group of genetic disorders known as a branchial arch syndromes; more specifically the first branchial (or pharyngeal) arch, which is the precursor of maxilla and mandible. This syndrome is named after Octave Crouzon, a French physician.
• It is also known as craniofacial dysostosis and acrocephalosyndactylyType II.
• It is caused primarily by premature fusion of the coronal and sagittal sutures.
• ICD–10 á Q75.1
1Postgraduate Trainee,

History
A 27-years old male patient presented with sudden onset of pain, redness, photophobia and diminution of vision in Left Eye since 10 days; No Past History of Ocular Injury.

Ocular Examination:

<table>
<thead>
<tr>
<th></th>
<th>RIGHT EYE</th>
<th>LEFT EYE</th>
</tr>
</thead>
<tbody>
<tr>
<td>Visual Acuity</td>
<td>6/9</td>
<td>HM +ve</td>
</tr>
<tr>
<td>Eye Lid</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td>Conjunctiva</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td>Cornea</td>
<td>Normal</td>
<td>Cornea Lesion (Ulcer) 1 x 2 mm, oval with irregular margin in the center of cornea</td>
</tr>
<tr>
<td>Anterior Chamber</td>
<td>Normal</td>
<td>Hypopyon present</td>
</tr>
<tr>
<td>Pupil</td>
<td>NSNR</td>
<td>Cannot be seen</td>
</tr>
<tr>
<td>Fundus</td>
<td>Normal</td>
<td>Cannot be seen</td>
</tr>
<tr>
<td>Vision</td>
<td>6/9</td>
<td>HM +ve</td>
</tr>
</tbody>
</table>

Photograph–2 Parrot Beak Nose with B/L Exophthalmos

Photograph–3 Frog-like Face with Maxillary Hypoplasia and Mandibular Prognathism
Investigations

- Blood Routine Examination:
  - Haemoglobin: 11.3 gm% (78%)
  - Total WBC Count: 7,500/mm3
  - Differential Leukocyte Count: N68E2B0L29M1
- Random Blood Sugar: 75 mg%
- X-rays:
  - X-ray Chest PA View: Normal
  - X-ray Cervical Spine and Lateral View: Normal
  - X-ray of Both Hand AP and Lateral View:
    - Tubular bones of hand are short
    - Distal phalanx of all digits are short

Photograph–4 & 5 Shortening of Distal Phalanx

Photograph–6 X-ray Chest PA View

Photograph–7 X-ray Cervical Spine Lateral View

Photograph–8 X-ray of Both Hand AP View

Photograph–9 X-ray of Both Feet Lateral View
X-ray of Both Feet AP and Lateral View:
- Tubular bones of feet are short
- Distal phalanx of all digits are short

X-ray Skull AP and Lateral View:
- Brachycephaly
- The lateral ridge of bilateral orbits appears elevated giving bilateral “Harrequin” configuration of orbit.
- B/L orbit are shallow with exorbitism
- Profound maxillary hypoplasia
- High arched palate
- Calvaria is thinned out
  with focal lucencies giving “copper seater appearance”
  (suggesting possibly of hydrocephalous)
- B/L Dental malocclusion
- Calcification of B/L stylohyoid ligament

Diagnosis
On the basis of clinical features and x-rays report the case showed typical picture of Crouzon syndrome.

Treatment
Conservative Treatment:
- Antibiotic Eye Drops
- Artificial Tear Eye Drops
- Antibiotic Eye Ointment

Tarsorrhaphy was done in the Left Eye.

Discussion
Crouzon Syndrome is a genetic disorder.
- It is an autosomal dominant disorder.
- Incidence 1: 25,000
- Associated with FGFR2 and FGFR3 gene
- Sex and race not associated

Conclusion
Because of rarity of presentation, case is being reported.

References
(2) Reardon W, Winter RM, Rutland P, Pulleyn LJ, Jones BM, Malcolm S (September 1994). “Mutations in the fibroblast growth factor receptor 2 gene cause Crouzon syndrome”. Nat Genet. 8(1); 98—103; doi 10, 1038/ng 1295—462.
Abstract: Mooren’s Ulcer is an idiopathic, rapidly progressive, painful peripheral ulcerative keratitis with no associated scleritis. It’s a diagnosis of exclusion which means all other diagnosable systemic disorders that could be responsible for the progressive destruction of cornea must be ruled out. The etiology of Mooren’s Ulcer remains uncertain. However, recent studies indicate that it is an autoimmune disease directed against a specific target molecule in the corneal stroma, triggered in genetically susceptible individuals by one of several possible mechanisms.

A 40 years old female presented to us on 25th January 2008 from rural tribal area of Sagbara with bilateral peripheral ulcerative keratitis with perforation and iris prolapse in right eye. She was a known case of diabetes mellitus diagnosed seven years ago. This case report aims to highlight the diagnosis and treatment of Mooren’s ulcer.

Diagnosis of Mooren’s ulcer was made after she underwent extensive medical and laboratory testing to rule out an infectious or systemic cause of corneal melt.

She benefited by right eye patch graft, left eye tissue adhesive with BCL application, Both eyes conjunctival peritomy and immunosuppressive drugs along with control of diabetes.

Case Report:
A 40 year old female from rural tribal area of Sagbara with low socio economical status presented to us on 25th January 2008 with complaint of decreased vision, redness, watering, ocular pain and photophobia in both eyes for last 2 months more so in right eye for last one week. She was having diabetes mellitus for last seven years which was controlled by medication. There was no history of trauma or any associated joint pain.

On examination, best corrected visual acuity in right eye was 6/36 and in left eye, it was 6/24. There was inferior crescent of corneal thinning involving 6 clock hours (3-9 o’clock) with Limbal involvement but no associated scleritis. In right eye, there was a perforation with Iris prolapse at 6-7 o’clock position. In both eyes, there was overhanging edge infiltrated with white cells and spreading towards centrally and circumferentially. It was positive for fluorescein stain indicating overlying epithelial defect. The pupil was peaked inferotemporally in right eye due to perforation and in left eye due to impending
perforation. There were no signs of secondary infection and/or Iritis on examination. Dilated Indirect ophthalmoscopy was normal in both eyes. (No diabetic retinopathy)

She was investigated to rule out systemic disease causing peripheral ulcerative keratitis. RBS was 301 mg% showing uncontrolled diabetes mellitus on the day of presentation.

 Investigations done – Hemogram with ESR, Urine routine and microscopy, VDRL, RA factor, HCV, X-ray chest and Joints, SGPT, ANCA, ANA, HBsAg, Scraping of the ulcer was done, and did not reveal any causative organisms on smear or culture.

She was referred to her physician to control diabetes mellitus as well as to rule out systemic disorders causing peripheral ulcerative keratitis. She was placed on Insulin injections to control DM.

After excluding systemic diseases associated with peripheral ulcerative keratitis, A diagnosis of Bilateral Mooren’s ulcer was made and systemic immunosuppressive therapy was started with oral methotrexate 10 mg once a week and oral systemic steroids at a dose of 1.5 mg / kg / day.

Right eye iris abscission (of prolapsed part) with patch graft (free Hand) was done after controlling diabetes mellitus. Tissue adhesive with Bandage Contact Lens Application with conjunctival peritomy was done in left eye.

Locally she was started on Prednisolone eye drops hourly, Ofloxacin eye drops 4 times a day, timolol maleate eye drops twice a day, 2 % HPMC eye ointment three times a day.

Tissue Adhesive and Bandage Contact Lens were removed after 6 weeks in left eye. It was epithelialized and scarred without any overhanging edge. Right eye patch graft is doing well after 6 months.

Best corrected visual acuity in right eye 6/18 and left eye 6/18 after 6 months follow up.

**Discussion:**
Although the diagnosis of Mooren’s ulceration may
be difficult when a patient first presents with PUK, the clinical appearance is characteristic. However, a thorough medial history, physical examination and appropriate laboratory investigations must be performed to rule out underlying systemic conditions causing PUK, since Mooren’s Ulcer is a diagnosis of exclusion.

Mooren’s ulcer was first described by Bowman in 1849 and then by McKenzie in 1854 as “Chronic serpiginous ulcer of cornea or ulcerus roden”2. Mooren’s name, however, became attached to this rare disorder because of his publication of cases in 1863 and 18673. He was the first to clearly describe this insidious corneal problem and define it as a clinical entity. Nettleship4 summarized the accumulated reported experience with the disorder in a classic article.

Mooren’s ulcer is idiopathic by definition, occurring in complete absence of any diagnosable systematic disorder that could be responsible for progressive destruction of the cornea with no associated scleritis.

Its exact pathophysiology remains uncertain, although a growing body of evidence indicates that it is an autoimmune disease directed against a specific target molecule in the corneal stroma resulting in its destruction by degradative enzymes, which are released primarily by neutrophils attracted into the area by diverse stimuli,12,13 probably triggered in genetically susceptible individual by one or several mechanisms.

Wood and Kaufman having reported9 cases concluded that there were two clinical types of Mooren’s ulcer5. The first limited type, is usually unilateral, with mild to moderate symptoms, generally responds well to medical and surgical treatment. This type is believed to occur in older patients and has become known as typical or benign Mooren’s ulcer. In contrast, the second type is bilateral, with relatively more pain and generally a poor response to therapy in younger patients, became known as atypical or malignant, Mooren’s ulcer. The benign type is bilateral in 25% of patients and the malignant type is bilateral in 75% of Patients.

Keitzman6 published a series of 37 cases of progressive Mooren’s ulcer in Nigeria affecting primarily healthy men between age of 20 and 30 yrs and the clinical course was very rapid. Perforation occurred in 36% of the patients. As a result, the generalized belief has developed that the progressive and relentless atypical form of Mooren’s ulcer has a predilection for young men of African origin.

Lewallen and courtright7, in their published series of Mooren’s ulcer, suggest that younger patients had bilateral disease less frequently than older patients (1.5:1) regardless of race. Although they found that men were 1.6 times more likely to have Mooren’s ulcer than were women.

Different entities have been associated with Mooren’s ulcer, often leading to conjecture that there may be a causal relationship.

An association with helminthiasis has been suggested in Nigeria8, Schanzlin9 speculated that the antigen antibody reaction to helminth toxin deposited in peripheral cornea provoked the inflammation and ulceration. Recently in 2 patients with bilateral Mooren’s ulcers chronic hepatitis with infection C was documented10,11 and they improved after treatment of the hepatitis C with interferon. The authors proposed that molecular mimicry may be involved, with the hepatitis C virus stimulating an autoimmune response to corneal antigens through cross reacting epitopes.

Based on the clinical presentation20 and the low dose anterior segment fluorescein angiographic findings, there seem to be three distinct varieties of Mooren’s ulceration:

1. Unilateral Mooren’s ulceration (UM), characterized by an excessively painful progressive corneal ulceration in one eye in elderly patients, associated with non perfusion of the superficial vascular plexus of the anterior segment.
2. Bilateral aggressive Mooren’s ulceration (BAM), which occurs in young patients, progresses circumferential and only later, centrally in the cornea. Angiography shows vascular leakage and new vessel formation which extends into the base ulcer.
3. Bilateral indolent Mooren’s ulceration (BIM), which usually occurs in middle aged patients presenting with progressive peripheral corneal guttering in both eyes, with little inflammatory response. There is no change from the normal vascular architecture on angiography except an extension of new vessels into the ulcer.

DD:
- Rheumatoid arthritis
- Wegener’s Granulomatosis
- Polyarteritis Nodosa
Other collagen vascular diseases
- Inflammatory bowed disease
- Giant cell Arteritis
- Staphylococcal Marginal Keratitis
- Local infections causes
- Terrien’s degeneration
- Pellucid degeneration
- Ocular Rosacea

**Treatment:**
Today, most experts agree on stepwise approach in management of Mooren’s ulcer.

1. Topical Steroids
2. Conjunctival Resection
3. Systemic immuno suppressive
4. Additional Surgical Procedure
5. Rehabilitation

**Goals:**
To arrest the destructive process and to promote healing and re-epithelialization of the corneal surface.

**Steroids:**
Initially: Intensive topical steroids
Prednisolone acetate 1% eye drop
One hourly

In association with:
Topical Cycloplegics
Prophylactic Antibiotics

If epithelial healing does not occur within 2-3 days, the frequency of topical steroids can be increased to every half hour.

Once healing occurs, the frequency can be reduced and tapered slowly over a period of several months.

Such management, especially in unilateral, benign form has met with good results.
Topical cyclosporine-A therapy (0.5% solution) also found useful in some studies.
Systemic steroids:

**Oral pulse therapy:**
60-100 mg daily of oral Prednisolone is indicated when topical therapy is ineffective after 7-40 days.

When topical steroids may be dangerous because of precariously deep ulcer or infiltrate.
Topical tetracycline may be used for anticollagenolytic properties.

A therapeutic soft contact lens or patching of eye may be beneficial when ulcer is deep.

**Conjunctival Resection:**
- If ulcer progresses despite steroid regimen, conjunctival resection should be performed.
- Under topical or sub conjunctively anesthesia, this consists of conjunctival excision to bare sclera extending at least 2 clock hours to either side of peripheral ulcer and approximately 4 mm posterior to the corneoscleral limbus and parallel to the ulcer.

The rationale of this procedure:
Conjunctiva adjacent to the ulcer contains inflammatory cells that may be producing antibodies against the cornea and cytokines which amplify the inflammation and recruit additional inflammatory cells.

**Multiple resections may be necessary.**

**Systemic immuno suppressive therapy:**

**Indication:**
Bilateral or progressive Mooren’s ulcer that fails to preceding therapeutic attempts will require systemic cytotoxic therapy to bring a half to the progressive corneal destruction.

**Commonly used agents:**
- Cyclophosphamide – 2 mg / kg / day
- Methotrexate (7.5 – 15 mg once a week)
- Azathioprine (2 mg / kg body weight / day)

The degree of fall in white blood cell count is considered as the most reliable indicator of immunosuppressant produced by cyclophosphamide.

Most authors believe that the evidence for the efficacy of systematic immune suppressive chemotherapy for progressive bilateral Mooren’s ulcer is quite strong, and further believe that such treatment should be employed sooner rather than later in the care of such patients, before the corneal destruction, has become too extensive to need for surgery.

Adverse effects of these cytotoxic and immunosuppressive medications, such as anemia,
alopecia, nausea, nephrotoxicity, are likely and it must be administered in close observation of physician.

Additional surgical procedures:
Small perforations / Impending perforation may be treated with application of tissue adhesive – isobutyl cyanoacrylate and placement of a soft contact lens to provide comfort and to prevent dislodging of the glue.

When a perforation is too large for tissue adhesive to seal the leak, some type of patch graft will be necessary. This may range from a small tapered plug of corneal tissue to a penetrating keratoplasty.

Rehabilitation:
Once the active ulceration has stopped and the remaining cornea has been completely opacified, it is possible perform PK on these patients, even in the face of a thinned and vascularized cornea.

Because of the immune systems remarkable memory, surgical attempts at rehabilitation in Mooren’s ulceration, should be done only with concurrent immunosuppressant, even when the active disease has been arrested, or is burnt out because attempts at penetrating keratoplasty often are associated with recurrence and graft failure.

Conclusion:
Bilateral Mooren’s ulcer in young patients can progress rapidly in a circumferential fashion towards the center of the cornea and can present with perforation and Iris prolapse early in a course of disease. Though clinical appearance is characteristics, Mooren’s Ulcer remains a diagnosis of exclusion and systemic diseases associated with PUK must be ruled out. Current treatment options and work up have resulted in a significant improvement in the prognosis of patients with Mooren’s Ulcer. With appropriate management, the eyes can usually be salvaged and visual loss can be minimized. The keys to proper treatment are early diagnosis, judicious use of topical as well as systemic steroids, immunosuppressives, and the use of tissue glue and patch grafting as indicated by the clinical scenario.

References:
The history of glaucoma drainage devices (GDD) is one of triumph of the human imagination spirit and ingenuity. It’s a story of the indomitable scientific spirit of inquiry, and of sheer perseverance and the resilience of the clinicians’ spirit in not succumbing to a disease.

The earliest references of a seton (literally, hair) is a rather scary attempt in 1906, to drain a hypopyon by placing a horse hair through a corneal paracentesis. The same technique was later applied to treat two patients with painful absolute glaucoma.

Anecdotal reports of attempts to shunt aqueous to a variety of unconventional sites, including the vortex veins and the nasolacrimal duct were instrumental in the evolution of the current generation of implants. What these did succeed in doing was to direct the clinician to the fact that the optimal site of drainage was the subconjunctival space.

**Generation 1: Translimbal GDDs**

The first translimbal GDD, reported by Zorab in 1912, was a silk thread used as a seton. This was followed by the use of gold, tantalum, magnesium and platinum thread/wire. Results were universally unsatisfactory due to lack of flow control and hypotony together with chronic inflammatory stimulus due to lack of biocompatibility.

Simple translimbal tube devices were similarly unsuccessful, with high rates of early filtration failure. The logic behind the use of these implants was prevention of filtration failure by maintaining patency of the drainage fistula or sclerostomy.

**Generation 2: Anterior Drainage**

It was Molteno’s remarkable observation in 1969 that revolutionized GDDs, heralding a new era in glaucoma surgery. He hypothesised that filtration failure was primarily attributable to subconjunctival fibrosis, with fistula closure occurring as a secondary event.

Molteno therefore launched the concept of tube and plate GDDs, in which aqueous is shunted to a plate device designed to maintain patency of a subconjunctival filtration reservoir in spite of continuing subconjunctival fibrosis. To date the Molteno tube remains the yardstick against which other tube devices are compared.

**Generation 3: Posterior Drainage**

To avoid problems like dellen formation, poor filtration associated with pre-existing anterior conjunctival scarring, and extrusion, posterior placement beneath Tenon’s capsule soon came into vogue. Subsequent tube and plate GDDs share the essential design concept of posterior filtration via a tube in the anterior chamber to a plate element secured beneath Tenon’s capsule. These include the Molteno, Krupin, Baerveldt, Ahmed, and OptiMed GDDs.

**Generation 4: External Resistance mechanisms**

The next landmark innovation is also attributable to the genius of Molteno who recognized that resistance to flow distal to the sclerostomy generally remains low until limited subconjunctival wound healing has occurred and the initially aggressive filtration becomes more limited.

A two stage procedure thus came into vogue, the device was implanted and allowed to encapsulate before insertion of the tube element into the anterior chamber at a second operation after 2–6 weeks.

Variations of a modified single stage procedure are now widely preferred for GDDs with no internal resistance mechanism. These include ligation with an absorbable suture, laser suture lysis, and occlusion with supramid stents or luminal sutures.

**Generation 5: Internal Drainage mechanisms**

A : Dependant on Tissue apposition and fibrosis

The Molteno dual ridge device limits the initial drainage area by dividing the top part of the plate into two separate spaces. Aqueous escapes directly into the channel between two concentric ridges on the plate and must overcome resistance associated with conjunctival tissue apposition. Apposition of the “Bioseal” element of the modified Baerveldt implant to the sclera with absorbable sutures similarly limits initial aqueous escape from beneath the device

B. non-adjustable resistance mechanism

Krupin, Ahmed, and OptiMed GDDs have internal resistance mechanisms and flow characteristics. The
principles of internal resistance of these are slit valves, venturi flow and multiple microtubules respectively.

**Newer Modifications**
The current GDDs have undergone major metamorphosis in terms of the materials used for manufacture of the plates.
The tubes that shunt aqueous from the AC to the plate are almost always made of silicon. The materials used for manufacture of the plates include elastomeric silicone (polydimethylsiloxane), silicone, PMMA, and other hydrophobic polymers.
The tubes placement has also undergone innovations with an alternate site of placement in the posterior chamber, or the vitreous cavity.
The size of the plates of the implant are seen to vary from 134 and 268 sq mm for the single and double plate Molteno to 250, 350, 425, 500 sq mm for Baerveldt’s implants.

**Newer Implants**
SOLX Gold Shunt GMS Plus is a 24 karat gold 3x6 mm rectangle which contains numerous microchannels that bridge the anterior chamber and the suprachoroidal space, thus controlling aqueous outflow.
The express minishunt is a stainless steel device which consists of a Shaft with spur and external plate with a scleral slot. Its size is 400 microns, with an external diameter 27G. It acts by shunting aqueous out of the aqueous into the subcojunctival space.
The T Flux implant is a T shaped poly-MEGMA, hydrophilic acrylic that drains fluid by means of capillarity and osmosis. It has been designed for use with non-penetrating glaucoma surgery.
## Table 1: Landmark Dates in the history of GDD:

<table>
<thead>
<tr>
<th>Year</th>
<th>Event</th>
</tr>
</thead>
<tbody>
<tr>
<td>1907</td>
<td>Rollet and Moreau implanted horse hair head to drain fluid out of anterior chamber</td>
</tr>
<tr>
<td>1912</td>
<td>Silk thread as first translimbal GDD implanted by Zorab</td>
</tr>
<tr>
<td>1959</td>
<td>Epstein unsuccessfully inserted a polythene tube in an attempt to drain fluid, failed because of excessive scar formation near the limbus, seton migration, and conjunctival erosion.</td>
</tr>
<tr>
<td>1969</td>
<td>Molteno introduced the concept of large surface area required to disperse the aqueous (Acrylic tube with thin plate at limbus)</td>
</tr>
<tr>
<td>1976</td>
<td>Molteno implant with a long silicone tube attached to a large end plate placed 9-10 mm posterior to the limbus.</td>
</tr>
<tr>
<td>1976</td>
<td>Krupin introduced the first pressure sensitive unidirectional valve (opening-11mm Hg, closing-9 mm Hg) provided resistance to the flow of aqueous and prevented early postoperative hypotony 1981-Double plate valve by Molteno</td>
</tr>
<tr>
<td>1992</td>
<td>George Baerveldt recognises that increase in the surface area of the end plate(s) results in lower IOPs. Introduced a nonvalved silicone tube attached to a large barium impregnated silicone plate with a surface area of 250 mm², 350 mm², or 500 mm²</td>
</tr>
<tr>
<td>1993</td>
<td>Ahmed introduced AGV</td>
</tr>
<tr>
<td>1997</td>
<td>Introduction of the Helies drainage device which uses an artificial meshwork of PTFE fibers</td>
</tr>
<tr>
<td>1998</td>
<td>Glaucoma Drainage Devices have been implanted in 2,980 patients</td>
</tr>
<tr>
<td>2001</td>
<td>FDA approved the AquaFlow Collagen Glaucoma Drainage Device as an alternative treatment for open-angle glaucoma</td>
</tr>
</tbody>
</table>
References
Pediatric Electrophysiology

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Introduction
Electrophysiology, especially in children, can contribute a significant lot. It is one of the most underutilized diagnostic test in pediatric ophthalmology in our country. The tests are objective, safe, noninvasive, swift and easy to administer. Also, they can give unique insight into the functional integrity of different levels of the visual pathway. It is really advantageous to use visual electrodagnostic tests in preliminary disease stages in young infants. It enhances the chances of successful surgical or clinical intervention and allows genetic counseling when it is most pertinent to young parents. It also helps in emotional acceptance and allows for more easier implementation of educational programs. Despite advances in imaging methods, its application in young children is still questionable.

What does electrophysiology encompass?
The electroretinogram (ERG) indicates the massed retinal responses to the full field luminance stimulation, which reflects the function of the photoreceptors and the inner nuclear layers of the retina, the electro-oculogram (EOG) expresses retinal pigment epithelium (RPE) function and the interaction between the RPE and the (rod) photoreceptors and the visual evoked potential (VEP) reflects optic pathway function beyond the eye to the visual cortex. These tests complement, and supplement, other visual methods of assessment.

Thus, an abnormal ERG may suggest the necessity for metabolic screening and an abnormal VEP in association with a normal ERG can indicate the need for structural imaging studies.

The results of visual electrophysiological tests are usually displayed as graphs of voltage (in microvolts) plotted against time (in milliseconds). These graphs have characteristic waveforms and the constituent positive and negative peaks are quantified by their latency (called implicit time by some workers), relative to the onset of stimulus delivery, and their size (amplitude) relative to the previous peak or an estimated baseline.

ERG (Electroretinogram)
The ERG is the mass electrical response of the retina to luminance stimulation and is recorded using corneal electrodes. The stimuli are delivered by a Ganzfeld (Fig 1) bowl, an integrating sphere enabling uniform whole field illumination. The Ganzfeld provides both flash stimulation and a diffuse background for photopic adaptation. The Krubsfeld is the hand held bowl used for ERG under general anesthesia. The reference electrodes are positioned at the ipsilateral outer canthi if a bipolar contact lens electrode is not used.
of these summed biopotentials is recordable at the front surface of the eye as a series of negative and positive voltage changes (peaks) in the first 200 ms after the light stimulation. These series of peaks are the ERG. The form and timing of the ERG is related to the eye’s state of light adaptation and to the intensity, spatial, chromatic and temporal characteristics of the stimulus.

The flash electroretinogram is generated by a uniform flash of light e.g from the light flashed in a Ganzfeld or by (LEDs) that reflects electrical activity from most of retina.

A pattern ERG (pERG) is recorded when a structured stimuli are used; for example, checkerboards or gratings. Usually, pERGs are recorded using stimuli localized to the macular and paramacular areas. There is no overall change in mean retinal luminance, and light scatter within the eye is minimized.

Multifocal ERG (mERG) is obtained by stimulating the central 300 to 500 of the retina with a contiguous array of flickering hexagons. Multiple hexagons are simultaneously alternated, but the m-sequence determines that no pattern of simultaneous hexagon is repeated twice in the sequence.

There are important clinical advantages in separately assessing the rod and cone activity. Rods can detect single light quanta in dark backgrounds. Rod ERGs are slower responders than cone ERGs, as they are reflecting a longer pathway through the retina. The rod pathway is preferentially activated by dim, short wavelength light stimuli or by a very dim white flashes presented at less than 10/s under darkened (scotopic) laboratory conditions. The cone pathways are preferentially stimulated by high intensity white light and longer wavelength flashes presented under photopic conditions, as well as by fast flash rates delivered above 15/s.

**Origins of waves**

Light stimulation of both rods and cones causes transient and sustained changes in the extracellular ion composition. The movement of these ionic currents through the membrane and interstitial resistance of Muller cells give rise to an associated voltage change and this mass dipolar response is detectable at the front surface of the eye as ERG.

Granit, in 1933, suggested that three processes are involved in generating the flash ERG: process I(P1) is the main contributor to the c wave (EOG), PII to the b wave and P III to the a wave.

The a wave is the main corneal negative wave. The a wave is a reflection of mass hyperpolarisation of photoreceptors. The maximum amplitude of the photocurrent response is determined by the upper limit of dark, or circulating current available. (Figure 2)

![Figure 2 Normal ERG wave](image)

The b wave is a corneal positive component; b wave activity is associated predominantly with the depolarisation of on centre bipolar cells.

![Figure 3 Oscillatory potentials](image)

Oscillatory potentials (OPs) (Figure 3) are a series of high frequency positive wavelets. OPs overlap with the rising phase of b wave. Early wavelets of OPs reflect rod function and “on” pathways and later wavelets are linked with the cone system and “off ” pathway activity.

The c wave is a slow positive going wave, shows variable presence among healthy controls, mainly as it represents the interaction of two retinal potentials with opposite polarity and similar time course.

The d wave is a positive wave that is closely associated
with a reduction in light under photopic conditions. Flicker ERGs elicited by stimuli rates greater than 20/s reflect cone photoreceptor activity. The PERG is typically a biphasic configuration with a positivity at 50ms (P 50) and a negativity at 95 (N95). P 50 indicates macular function and N95 reflects ganglion cell function.

**The machine: parameters & equipment**

**Recording electrodes**

Electrodes that contact the cornea or nearby bulbar conjunctiva are strongly recommended for basic full-field recording. These include contact lens electrodes, conductive fibers and foils, conjunctival loop electrodes and corneal wicks. For most users, contact lens electrodes (Figure 4) will provide the highest amplitude and most stable recordings; such electrodes should be centrally transparent with an optical opening as large as possible, and incorporate a device to hold the lids apart. The corneal surface should be protected during use with a non-irritating and non-allergenic ionic conductive solution that is relatively non-viscous (e.g., no more viscous than 0.5% methyl cellulose).

More viscous solutions can attenuate signal amplitude. Other types of corneal and conjunctival electrodes require more skill to use but may have certain advantages. Users should be aware that signal amplitude is reduced as the point of ocular contact moves away from the corneal apex. Topical anesthesia is necessary for contact lens electrodes but may not be required for other types of corneal and conjunctival electrodes. It is necessary that all electrophysiologists master the technical requirements of their chosen electrode, to ensure good ocular contact, to ensure proper electrode impedance, to ensure that waveforms are comparable to standard ERGs, and to define both normal values and variability (which may be different with different electrodes) for their own lab. Skin electrodes are not generally recommended as active recording electrodes.

Reference electrodes may be incorporated into the contact lens-speculum assembly to make contact with the conjunctiva (‘bipolar electrodes’). This is the most stable configuration electrically. Alternatively, electrodes can be placed near each orbital rim temporally as a reference for the corresponding eye.
A separate skin electrode should be attached to an indifferent point and connected to ground. Typical locations are on the forehead or ear.

**Skin reference electrode characteristics**
The skin should be prepared by cleaning, and a suitable conductive paste or gel used to insure good electrical connection.

**Stimulus duration**
The standard is based on stimuli of duration considerably shorter than the integration time of any photoreceptor. Thus, the light stimulus should consist of flashes having a maximum duration of about 5 ms.

**Clinical Protocol**

**Pupillary dilation**
Pupils should be maximally dilated for all ERG recordings and pupil size should be noted.

**Pre-adaptation to light or dark**
The recording conditions outlined specify 20 min of dark adaptation before recording rod ERGs, and 10 min of light adaptation before recording cone ERGs. If contact lens electrodes are used, the wearing time can be minimized by dark adapting first, and inserting the electrodes under dim red light at the end of the adaptation period. However, care should be used to avoid too bright a red light, and an additional 5 min of dark adaptation may be needed for recovery after lens insertion.

**Fixation**
A fixation point should be incorporated into stimulus domes. A stable eye is important so that eye movements do not alter the optimal corneal electrode position, produce electrical artifacts, or allow blockage of light by the electrode or eyelid.

**Sedation or anesthesia**
Uncompliant children (especially ages 2-6 for whom containment can be difficult) may become compliant with oral sedation or anxiolysis. Medical guidelines should be followed with respect to indications, risks, medical monitoring requirements and the choice of a sedative / relaxant versus general anesthesia. Considering the variability of pediatric records, there will generally be little effect on ERG amplitude or waveform with sedation although full anesthesia may modify the ERG.

**Electrodes**
Contact lens electrodes are applicable to infants and young children, but pediatric sizes will be required with speculum-containing models, and care must be used to minimize corneal and psychological trauma. Special care is required with children to monitor electrode position and compliance in order to avoid artifactual recordings. Burian Allen electrodes are good for children, although they are more preferable in children under anesthesia.

**Normal values and measurement**
The ERG matures during infancy, and newborn and infant signals must be interpreted with great caution. Later infantile and young childhood ERGs approach adult waveform and size. Pediatric ERGs should ideally be compared to those from normal subjects of the same age, even though there may be little normative data available. Because movement and poor fixation can make pediatric records variable in amplitude and waveform, several repetitions of each ERG be recorded in order to recognize reproducible waveforms and choose the best examples.

**EOG (Electro oculogram)**
The EOG is a measure of the function of the RPE and the interaction between the RPE and the photoreceptors. The patient makes a fixed 30 degree lateral eye movement during a period of 20 min dark adaptation and then during a 12-15 min period of light adaptation. The eye movements are made every 1-2 seconds for approximately 10 seconds every min. The amplitude of signal recorded between electrodes positioned at medial and lateral canthi reaches a minimum during dark adaptation, the dark trough, and a maximum during light adaptation, the light peak. The development of a normal light peak requires normally functioning photoreceptors in contact with a normally functioning RPE. The EOG is quantified by calculating the size of the light peak in relation to the dark trough as a percentage, the Arden index. A normal EOG light rise is >175% for most laboratories.

**VEP (Visual Evoked potentials)**
The VEP is normally largest in the midline, around 3 to 5 cm above the inion. VEPs are elicited to stimulate a wide central area of the visual field. For clinical purposes, uniform light flashes and checker board pattern stimuli are usually used.

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*Figure 7 Cortical origins of VEP*
VEPs reflect surface activity of cortical gyri and therefore mainly reflect activity of areas of the visual field represented at the surface of gyri. P100 component of the flash VEP arises from cortical activation by the retino geniculate pathway. Flash VEP is used mainly when visual acuity is poor or when cooperation is limited. This is a complex waveform and shows great variability.

Pattern reversal VEP is usually elicited by checks that alternate from black to white and vice versa. Pattern onset VEPs are elicited by the brusque appearance of a pattern usually lasting between 100 and 300 ms. It is used more widely in clinical practice as its waveform is maintained across the life span. In older children, it is useful in identifying the abnormal pathway projection associated with albinism.

How are the waves recorded? Various parameters

**Electrodes**
Standard silver-silver chloride or gold disc surface electrodes are recommended for recording VEPs. The electrodes should be fixed to the scalp and maintained using procedures recommended by the manufacturer.

**Electrode placement**
The scalp electrodes should be placed relative to bony landmarks, in proportion to the size of the head, according to the International 10/20 system. The anterior/posterior midline measurements are based on the distance between the nasion and the inion over the vertex. The active electrode is placed on the scalp over the visual cortex at Oz with the reference electrode at Fz. Commonly used ground electrode positions include the forehead, vertex (Cz), mastoid, earlobe (A1 or A2) or linked earlobes.

**Clinical protocol**

**Preparation of the patient**
Pattern stimuli for VEPs should be presented when the pupils of the eyes are unaltered by mydriatic or miotic drugs. For pattern stimulation, the visual acuity of the patient should be recorded and the patient should be optimally refracted for the viewing distance of the screen. Monocular stimulation should be performed. In infants or some other special populations, when binocular stimulation may be used to assess whether any afferent signals are reaching primary visual cortex. When a flash stimulus is used with monocular stimulation, care should be taken to ensure that no light enters the unstimulated eye.

The pattern onset /offset response shows a greater intersubject variability than the pattern reversal VEP but shows less sensitivity to confounding factors such as poor fixation or eye movements.

**DIAGNOSTIC APPLICATIONS**

**ERG**
The flash ERG plays its most vital role in conditions where ophthalmoscopy shows a normal or questionably normal, fundus and optic disc appearance and yet the child seems to have poor vision.

**Both rods and cones are affected**
In Leber’s congenital amaurosis, both rod and cone ERGs are most usually non-detectable. Concurrent VEP helps as the presence of flash VEP activity indicates cases who are likely to have some degree of rudimentary vision as opposed to complete blindness.

**Predominantly rods are affected**
Rod dysfunction can occur in isolation as in retinitis pigmentosa (RP) or linked with a wide variety of conditions importantly vitamin A deficiency. In the early stage, ERGs show a partial preservation of cone ERG activity and sometimes mixed rod/cone activity attenuation. In early and middle childhood, VEPs to flash and pattern stimulation are commonly of normal size and latency which as the disease progresses P VEP is delayed and reduced in amplitude.

**Predominantly cones are affected**
In children with rod monochromatism (achromatopsia), the rod mediated ERG to dim white or blue flash is usually normal but the white flash cone mediated ERG elicited under photopic conditions and 30 Hz flicker response is not detectable. The VEP is invariably significantly attenuated, degraded and delayed.

**Inner retinal dysfunction**
The normal bright flash mixed rod/cone ERG has a b wave that is 1.5 to 2 fold the size of the a wave. When the b wave amplitude is markedly attenuated or not detectable, the mixed ERG has a larger than average a wave. The negative ERG morphology indicates preserved photoreceptor activity and dysfunction of the inner retina layers, usually affecting the bipolar and Muller cells. The negative ERG is associated with X linked congenital stationary night blindness, X linked retinoschisis, Bull’s eye maculopathy, infantile and juvenile neuronal ceroid lipofuscinosis.

**Visual Evoked potential (VEP)**
ERG and VEPs recorded together can give valuable information in pediatric population regarding the macular and extramacular areas and the quality of
macular pathway function.

**Ocular opacity**
Bright flashes usually penetrate all but the densest ocular opacities, and flash VEPs and ERGs are particularly valuable when ocular opacities prevent adequate visualization of the perimacular fundus.

**Optic nerve disorders**
Monocular stimulation and transoccipital multichannel VEP recording will help to indicate whether a disease process is affecting the optic nerve, chiasm or pathway beyond the chiasm.

Optic Neuritis. In optic nerve demyelination related to optic neuritis with visual involvement, the P VEP is almost invariably markedly delayed to non detectable. Once the acuity has improved P VEP levels rise to moderate levels which is unlike adulthood cases where they maintain significant delays.

Optic nerve hypoplasia. In moderate hypoplasia cases the VEP is variably attenuated probably related to the degree of hypoplasia. In severe cases the VEP are markedly attenuated.

In Optic nerve compression, the PVEP is usually degraded, attenuated and mildly delayed in optic atrophy caused by compressive ischemic or degenerative conditions.

For craniofacial dysostosis, flash and P VEPs can provide an early indication of visual pathway dysfunction in syndromic and non syndromic craniosynostosis.

Optic nerve glioma usually leads to a broadening, attenuation and mild delay of the P VEP from the affected eye.

**Albinism**
The crossed asymmetry VEP distribution in albinism is opposite to that of chiasmal compression or achiasma. In infancy, the “albino” crossed asymmetry can be shown by using a flash stimulation.

**Perinatal hypoxia**
The flash VEP during the acute stage can give an indication of the degree of visual function and eventual outcome.

**Amblyopia: Visual testing?**
Objective estimates of visual acuity can be inferred by noting the smallest pattern size to consistently elicit a P VEP when refractive error is corrected.

A strabismic amblyope may give higher resolution acuity than recognition acuity secondary to spatial distortion. VEP acuity is also a different measure and it is unrealistic to expect a direct relationship between objective and behavioral acuity measures.

However, the P VEP findings do provide a good benchmark and are particularly useful when comparing the two eyes that is the interocular differences in the acuity.

What is pattern and multifocal?

**PERG**
The PERG is the response of central retina to and iso-luminant stimulus, usually a reversing black and white checkerboard. It allows both a measure of central retinal function and and evaluation of retinal ganglion cell function.

The transient PERG has two components: P 50 at approx 50 ms and N95 at 95 ms. Measurement concentrates on the amplitude of P50 from the trough of the early negative N35 component. N 95 is a contrast related component generated in the retinal ganglion cells. (Figure 8)

The P50 component of the PERG is abnormal in disorders of macular function with preservation of N95:P50 ratio. The N95 component (ganglion cell) of the PERG is usually selectively affected if the PERG is abnormal in optic nerve disease but the PERG may be normal.

**Sweep VEP**
P VEP amplitude tends to fall of linearly with spatial frequency near the limit of acuity. The extrapolation of high frequency limb of the spatial tuning function to zero amplitude produces an intercept that correlates with subjective visual resolution; this has been the basis of the sweep VEP techniques.

Sweep techniques can progressively and rapidly test (usually in 8-16 s) a range of spatial frequencies (checks or gratings) to determine a VEP threshold.
Each spatial frequency is presented for a discrete interval (usually for 5-10 s) and the change in pattern size can be continuous or sampled.

**Multifocal ERG (MERG)**
The MERG provides an “ERG visual field” of the electrical response of the retina in different locations extending out to about 250 radius. It can be used to differentiate whether a visual field defect is due to retinal or optic nerve disease.

Since the first order signals are generated from the photoreceptor and bipolar cells in the retina, the standard MERG will pick up diseases of the outer retina and inner retina but will be normal in cases where the ganglion cells or optic nerve is injured.

**ERG and VEP in clinical practice**
When ERG and VEP are used together, they help in finding out whether the nystagmus or a failure of baby to fix and follow is secondary to and dysfunction in the anterior visual pathway or it is otherwise. They can help in finding out the level of the clinical problem in the sense that it determines the level like at photoreceptor level (rods or cones), the inner retinal layer, optic nerve or at the chiasm or postchiasmal pathway.

**Reporting an ERG**
The standards for reporting an ERG (by ISCEV) is to obtain five ERGs (Figure 9):
1. ERG to a weak flash (arising from the rods) in the dark-adapted eye
2. ERG to a strong flash in the dark-adapted eye
3. Oscillatory potentials
4. ERG to a strong flash (arising from the cones) in the light-adapted eye
5. ERGs to a rapidly repeated stimulus (flicker).

The first 3 are the scotopic (dark adapted) responses and the last 2 are the photopic (light adapted) responses.

**Reporting a VEP**
A minimum of two recordings of each VEP condition should be acquired, measured and displayed. Reports of the standardized conditions should specify the stimulus parameter; the field size of the stimulus, the strength (time integrated luminance) of the flash or mean luminance of the pattern, the pattern element size and contrast of pattern stimuli, the frequency of stimulation, the eye tested and the recording parameters; the filter settings and the locations of the positive (i.e., active) and negative (i.e., reference) and indifferent (i.e., ground) electrodes.

In older infants, these tests can help to finding out why the patching is not responding, it becomes an objective indicator of posterior hemisphere dysfunction with visual field loss.

It also helps in older children with complaints of headache or possible functional visual loss.
**Exercises**

**Case 1 (Figure 10)**
The 1st waveform shows response which is less than normal. The 2nd waveform shows a good response for full threshold scotopic waveforms. Oscillatory potential and photopic waveforms are flat. The last waveform which is the 30 Hz flicker response in photopic conditions shows flat response suggestive of a cone dysfunction.

**Case 2 (Figure 11)**

![Standard Electro-Retinogram](image)

First waveform that is scotopic –25dB response, which shows delayed response and low amplitude. However, full threshold scotopic response which shows a negative ERG. The a wave is prominent and almost as big as b wave. Also, the cone responses are also normal with a sharp b wave (cone shaped pointed). This was suggestive of congenital stationary night blindness.

**Case 3 (Figure 12)**

![Kurbsfeld ERG](image)

A 8 year old child presented with complains of reduced vision in both eyes. He was using glasses both eyes -2.5 D cycl 180 improving to 6/24 since 2 years. Stereoacuity was 1800 sec and retina was normal. The child was subjected to ERG and the waveforms show a subnormal response for photopic thresholds. The ERG waveforms of left eye are shown in the picture. The scotopic responses are good. The photopic responses are very small. The 30 Hz flicker revealed a increased latency and a reduced amplitude. The diagnosis of ametropic amblyopia was changed to cone dysfunction.

**Case 4 (Figure 13)**

![Standard Electro-Retinogram](image)

A 5 year old child presented with complains of loss of vision in RE. There was a history of pars plana vitrectomy with lensectomy for traumatic endophthalmitis and child had 3/60 vision. He was referred for amblyopia treatment. An ERG was done and all the waveforms were found to be flat suggestive of nil photoreceptor function and therefore the chances of improvement of vision and amblyopia treatment were severely reduced which was informed to the parents.

**Conclusion**

Electrophysiology is a wonderful diagnostic tool especially for children where the subjective responses are difficult to obtain, are slow and sometimes absent and unreliable.

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2. Sutter E. The interpretation of multifocal binary Kernels. Documenta Ophthalmologica. 2000; 100: 49-75
Aims of the Journal

Delhi Journal of Ophthalmology (DJO) is the quarterly journal published by Delhi Ophthalmological Society. The DJO aims to become an easily readable fully referenced journal which will provide the specialists with up to date data and the residents with articles that give expert opinions that are backed with references. We aim to help the reader by providing in a systematic manner:

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2. Case reports and clinical enigmas
3. Original articles preferably of clinical relevance
4. Articles on diagnostic and surgical techniques
5. Selections, annotated by experts, of the most interesting papers from the great wealth of original publication.
6. New ideas and innovations, new devices and instruments.
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2. Use double spacing, throughout the manuscript including references, tables, and legends.
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4. In the upper right-hand corner, identify each page with a number and a running title.
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6. Other than the title page, do not identify authors else-where in the manuscript.
7. On other pages authors could be identified, if necessary, with their initials in parentheses.
8. Numeric equivalents must precede all percentages, for example: of 100 patients, 30 (30%) had significant visual field loss.
9. For a listing of standard abbreviations consult: Scientific Style and format, 6th Ed. (New York: Cam-bridge University Press: 1994). Abbreviations should be used sparingly and must be preceded by the full form when used for the first time, for example, intraocular pressure (IOP). However, common abbreviations must be used without full forms, for example, mm, mm Hg. Please use right eye and left eye, rather than OD and OS.
10. All hematological and clinical chemistry measurements should be reported in the International Systems of Units (SI). Temperature should be given in degrees Celsius. Length, height, weight and volume should be given in metric units.

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The manuscript (including reference, legend, and tables) must be typed in double spacing on a 21.6 x 27.6cm (8.5”x11”) paper with at least 2.5 cm
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It should contain the manuscript title and each author’s full name with academic degree(s). The abbreviated title (running title) should not exceed 40 characters, including spaces. The department and institutions where the study was performed should be indicated. Sponsoring organization and grant support are to be acknowledged on the title page.

The name and mailing address of author to whom requests or correspondence should be directed must be indicated including the e-mail address.

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