General Information

Delhi Journal of Ophthalmology (DJO), once called Visiscan, is a quarterly journal brought out by the Delhi Ophthalmological Society. The journal aims at providing a platform to its readers for free exchange of ideas and information in accordance with the rules laid out for such publication. The DJO aims to become an easily readable referenced journal which will provide the specialists with up to date data and the residents with articles providing expert opinions supported with references.

Contribution Methodology

Delhi Journal of Ophthalmology (DJO) is a quarterly journal. Author/Authors must have made significant contribution in carrying out the work and it should be original. It should be accompanied by a letter of transmittal. The article can be sent by email to the Editor or the hard copy posted. Articles received will be sent to reviewers whose comments will be emailed to the author(s) within 4-6 weeks. The identity of the authors and the reviewers will not be revealed to each other by the editorial team. The contributors shall be responsible for statements in his/her/their work including the changes made during editing. Detailed instructions to the contributors and for advertisements are included at the end of journal. Request for reprints or any queries should be addressed to the Editors office by email or post.

Editorial Process

The DJO has Dr Rohit Saxena as its Editor who is assisted by a team of renowned ophthalmologists and an illustrious editorial board. The reviewers, who are leaders in their respective fields, form the back bone of the journal by setting standards for the published work.

Editorial Office

Dr Rohit Saxena, Room No. 479, Dr R.P. Centre for Ophthalmic Sciences, AIIMS, New Delhi-110029
Ph +91-011-26593182, Email : editordjo@gmail.com

Published by: Dr Rohit Saxena, Editor DJO, on behalf of Delhi Ophthalmological Society, Delhi

Editorial Assistant: Varun Kumar
Editorial

5. Impossible is nothing ........................................ Rohit Saxena

Major Review

7. Blepharophimosis
   Saurabh Kamal, Goel Ruchi, Kumar Sushil, Tiwari Bhawna, Dulgach Pratima

15. Accomodative Esotropia
   Deepali Garg Mathur

19. Role of Radiotherapy in Management
   Shraddha Puranik, Prashant Nathani, Sanjay Kumar Mishra

25. Eyelid trauma
   Prashant Yadav, Neelam Pushker, M.S. Bajaj, M. Chandra, B. Chawla, D. Shrey, P. Bhimseria, K. Gupta ,
   K. Rishi, R. Meel

34. High Risk Corneal Grafting
   M Vanathi, Kanchan Chawhan, A Panda, Tushar Agarwal

Preferred Practice Patterns

40. Preparation Of Instruments And O.T.
   Uday Gajiwala, Rajesh Patel, Parin Shah, Rohan Chariwala, Viren Patel

Cases Report

47. Lenticonus Posterior
   Ojaswita Singh, Rashi Shyam, A.K.Gupta

48. Cholesterol Granuloma of the Orbit
   Mandeep S. Bajaj, Seema Kashyap, Neelam Pushker, Vidushi Sharma, R. Balasubramanya,
   Rachna Meel, Anoop Kishore Gupta

50. Cancer associated retinopathy
   Shahana Mazumdar, Navneet Mehotra

54. Mesectodermal Leiomyoma Of Ciliary Body
   Sabia Rashid, M. Q. Keng, M. Farooq, A. R. Khan

History of Ophthalmology

56. Doctor, Your Eponyms Are Showing
   Sashwat Ray

Instruments Scan

59. Role of Radio Imaging in Ophthalmology
   Satya Karna, Varun Gogia

Instruction to Authors

65. Instruction to Authors
Dear Friends,

We meet again through the pages of our third issue and hope that we continue to fulfill our aim of providing knowledge and information in a clear and lucid manner while making reading the journal fun and insightful for all. The entire editorial board is working overtime to bring forth this journal for you and we will welcome all comments and observations that are aimed at enhancing the quality of the journal.

In the beginning of the 1900’s a crisis had engulfed physicists, who felt that all laws that needed to be discovered had already been done and all we needed to know to understand the world around us was there and only minor issues needed to be cleared. Then came Albert Einstein with Quantum Mechanics, and the rest is history.

A similar crisis perhaps had hit ophthalmologists and cataract surgeon who at the peak of ICCE had breached the 2 minute barrier for cataract surgery and thought nothing more is there to cataract surgery. But it took just an idea to change our world. Today ICCE and aphakia is an option only for the incompetent. With CTRs, Cionni Rings and glued IOLs, aphakic glasses are obsolete. Perhaps we will soon have a “quantum Phaco”, something that will make current pheco incisions seem too large.

Each day in our life is an achievement and we must cherish it as such. Every little thought, every little modification is a step forward towards the ultimate goal of change. Newer techniques have to come, newer ideas have to germinate and what was unheard of; has to become law. Every time the “abstract or new” becomes “conventional or gold standard”, we have but a few minutes to admire the new before we start another journey towards a new summit, a new peak, to follow a new ideas, a new dream.

The Delhi Journal of Ophthalmology salutes those men and women who against all odds, defied what was law to follow their dreams, to give us what we take as granted today.

Those who thought big and worked to make it possible because….

Impossible is nothing

Dr Rohit Saxena

This is the third issue of Delhi Journal Of Ophthalmology in this term July 2009 - June 2010.

Any DOS member who has not received the previous two issue (July - Sept. 2009 and Oct. - Dec. 2009) please contact DOS Secretariat dosrecords@gmail.com, dostimes@airtelmail.in or Editor, DJO editordjo@gmail.com. Some copies have come back due to incorrect addresses, so members are requested to please provide correct address and contact details to DOS Secretariat.
Major Review

BLEPHAROPHIMOSIS – PTOSIS
EPICANTHUS INVERSUS SYNDROME (BPES)

Saurabh Kamal, Goel Ruchi, Kumar Sushil, Tiwari Bhawna, Dulgach Pratima
Guru Nanak Eye Center, Maulana Azad Medical College, New Delhi

Mustarde\(^1\) has categorized congenital eyelid anomalies into three groups: First restricted to soft tissue such as blepharophimosis, epicanthal folds, second includes soft tissue abnormalities associated with bone deformities such as mandibulofacial dysostosis and the third group comprises deformities in which soft tissues may or may not be abnormal but the primary deformity involves the bony orbit such as seen in Apert and Crouzon syndromes. For ophthalmologists, it is important to appreciate that many of these abnormalities are associated with systemic problems.

Blepharophimosis was first described by Dimitry\(^2\) in 1921. The cases in that family were reviewed by Owens et al\(^3\) in 1960. Many authors have documented the primary features of syndrome, namely, blepharophimosis, blepharoptosis, epicanthus inversus, and telecanthus. Kohn and Romano\(^4\) stressed the importance of telecanthus and other associated features. Therefore this syndrome had also been called as Kohn-Romano syndrome.

GENETICS

The frequency of BPES is estimated to be 1 in 50,000.\(^{13}\) A number of authors believe the AD transmission.\(^{3,5-7}\) Townes and Muechler\(^8\) described a family with blepharophimosis, and their proband, a 29-year-old girl, had primary amenorrhea. It is possible that preferential male transmission occurs because of reduced fertility in females.

Three different types are recognized. Type 1 and 2 are inherited in AD pattern.\(^{20}\) Type 1 is characterized by 100% penetrance, male to male transmission, and infertility of females. Type 2 is equally inherited through both sexes, and the penetrance is estimated to be 96.5%. Type 3 resembles type 2 except for having associated hypertelorism. Advanced maternal age, increases the risk for sporadic BPES.\(^{21}\) BPES is caused by mutations in the FOXL2 gene\(^9\) (chromosome 3q23). FOXL2 expression is restricted to developing eyelids of fetal and adult ovarian granulosa cells.\(^{10}\) In previous mutation studies\(^{11}\) intra-genic mutations were found in 70% of patients. Genomic rearrangements encompassing or outside the FOXL2 account for 16% of all molecular defects.\(^{12}\) Large submicroscopic deletions result in mental retardation.\(^{11}\)

MORPHOLOGICAL LANDMARKS OF THE ORBITS

The distance between the orbits varies with age. In clinics, evaluation of the interocular distances is based on the measurement of the following landmarks: Inter pupillary distance (IPD), Inner canthal distance (ICD), Outer inter canthal distances (OCD), and Horizontal palpebral fissure length (PFL). An approximate estimation of normality is to consider that the ICD is equivalent to PFL (Figure 1).

![Figure 1 Morphological Landmarks Of The Orbits](image_url)

The palpebral fissures are 15 ± 2 mm at 32 weeks of gestation, 17 ± 2 mm at birth, 24 ± 3 mm at 2 years of
age, and $27 \pm 3$ mm at the age of 14.\textsuperscript{14,15} ICD and OCD are 16 and 59 mm, respectively, in premature infants; $20 \pm 4$ and $69 \pm 8$ mm in newborn babies; $26 \pm 6$ and $88 \pm 10$ mm at the age of 3; and $31 \pm 5$ and $111 \pm 12$ at the age of 14.\textsuperscript{16}

Interpupillary distance ranges from $40 \pm 2.5$ mm at birth, $48 \pm 2.4$ mm at the age of 3 years and $58 \pm 3.6$ mm at the age of 14 years.\textsuperscript{22} IPD can also be calculated as $(OCD - ICD)/2 + ICD$. The eyelid pathology in dystopia canthorum of Waardenburg produces a short palpebral fissure. This shortness is due to lateral displacement of inner canthi, so the formula would serve as a mean to diagnose hypertelorism. Calculated and measured IPD are the same in the normal eyes and in hypertelorism, but not in Waardenburg's syndrome (Figure 2).

Also ICD is almost half of the IPD.

Alternative indices have been described, such as Farkas canthal index\textsuperscript{17,18} defined by $ICD/OCD \times 10$. It is higher than 42 in hypertelorism and lower than 38 in hypotelorism. It is useful for off-clinic analysis of photographs.

According to the normal range in the Chinese population, the ratio between the ICD and HPFL was approximately $1 - 1.2$.\textsuperscript{22} Depending on this ratio, the BPES can be graded into:

- Mild – 1.3 – 1.5
- Moderate – 1.5 – 1.8
- Severe – > 1.8

The surgical results can be regarded as good if, the ratio can be reduced to less than 1.3

**CLINICAL FEATURES**

BPES is recognized as tetrad of blepharoptosis, blepharophimosis, epicanthus inversus and telecanthus.\textsuperscript{4} (Figure 3)

Type 1 (classic BPES) have ptosis, telecanthus and epicanthus inversus as prominent features. Patients with type 2 have ptosis, telecanthus and lateral ectropion. Type 3 resembles type 2 except for having associated hypertelorism.

Blepharoptosis is usually bilateral, symmetric and severe, demonstrating poor levator action. These patients have hypoplasia of tarsal plate with absence of the eyelid fold. Vertical brow width and its convex arch, is increased from constant utilization of the frontalis.

Blepharophimosis denotes diminution of horizontal fissure from a normal 25-30 to 18-22 mm. Epicanthus inversus fold originate in the lower lid and sweep superiorly and medially over the canthus. This may diminish the normal canthal depression.

Type 1 (classic BPES) have ptosis, telecanthus and epicanthus inversus as prominent features. Patients with type 2 have ptosis, telecanthus and lateral ectropion. Type 3 resembles type 2 except for having associated hypertelorism. Type 1 (classic BPES) have ptosis, telecanthus and epicanthus inversus as prominent features. Patients with type 2 have ptosis, telecanthus and lateral ectropion. Type 3 resembles type 2 except for having associated hypertelorism. BPES is recognized as tetrad of blepharoptosis, blepharophimosis, epicanthus inversus and telecanthus.\textsuperscript{4} (Figure 3)

Blepharoptosis is usually bilateral, symmetric and severe, demonstrating poor levator action. These patients have hypoplasia of tarsal plate with absence of the eyelid fold. Vertical brow width and its convex arch, is increased from constant utilization of the frontalis.
definition is based on bony landmarks and the CT scan is useful. In most cases, the angle between the orbits is increased. False hypertelorism can be due to flat nasal bridge, epicanthic folds, exotropia, widely spaced eyebrows, narrow palpebral fissures and dystopia canthorum.

Additional lid features includes upper eyelid margin’s characteristics S-shape and the lower eyelid margin’s downward concavity, particularly laterally producing ectropion. Trichiasis also occurs. Lacrimal system may be affected. The lower punctum is displaced laterally, while the upper punctum medially. Other variations includes: posterior ectopia of lower punctum, aplasia of upper punctum, stenosis and elongation of canaliculi, and punctual reduplication. 18% incidence of nasolacrimal drainage problems have also been noticed.26 Additional features include nystagmus, microphthalmos, microcornea, strabismus, underaction of superior and inferior rectus and disc colobomas.

Facial features include broad and flat nasal bridge with bony deficiency at the supraorbital rim. This in addition to epicanthus may give the young patient a Down syndrome-like appearance. However, the intellectual development is usually normal, although familial mental subnormality is described. Mental retardation also occurs, especially in sporadic cases, in which extraocular anomalies also may be found. 19 The palate may be high arched and the ears low set and cupped with overhanging helix.

Another key feature of BPES is gonadal dysgenesis or premature ovarian failure in females. There are two main possibilities22 - Germ cells may be depleted at an unusually rapid rate from the postnatal ovary or primary error may occur during ovarian morphogenesis.

The incidence of strabismus and refractive error has also been studied.24 In this series 20% had manifest strabismus, out of which 70% had esotropia, 25% exotropia and 5% had hypertropia. 34% patients had a significant refractive error, out of which 30% patients had anisometropic hypermetropia and 34% had anisometropic myopia. Bilateral amblyopia is found in ~ 56% of patients.25

OTHER SYNDROMES WITH SHORTENING OF PALPEBRAL FISSURE23

- Campomelic dysplasia – bowed tibia, hypoplastic scapula, flat facies.
- DiGeorge sequence – defects involving fourth brachial arch and derivatives of third and fourth pharyngeal pouch.
- Dubowitz Syndrome – peculiar facies, infantile eczema, small stature, microcephaly
- Fetal alcohol Syndrome – prenatal onset growth deficiency, microcephaly.
- Oculodentodigital Syndrome – microphthalmos, enamel hypoplasia, campodactyly of fifth finger
- Toriello-Carey Syndrome – agenesis/hypoplasia of corpus callosum
- William Syndrome – prominent lips, hoarse voice, cardiac anomalies.
- Waardenburg’s Syndrome (Dystopia canthorum) – medial ankyloblepharon, telecanthus, lateralization of puncti, heterochromia iridis, deafness.
- Trisomy 18 – clenched hand, short sternum, low arch, dermal ridge on fingertips.
- Young-Simpson syndrome - heart defect, hypothyroidism, mental retardation

MANAGEMENT

Surgical correction has traditionally been deferred until the preschool years to allow for growth of nasal bridge, which helps to reduce the epicanthal folds and allows the tissues to enlarge enough to make surgical correction easier.

In a study to determine the optimal age for surgery and incidence of amblyopia, patients with coexistent strabismus had 64% incidence of amblyopia compared to 24% for those without strabismus.26 Patients with severe ptosis had lower rates of amblyopia than those with moderate ptosis but had their ptosis corrected at a median age of 2 years compared to 5 years for later. The authors concluded that BPES has high rate of amblyopia and co-existent strabismus doubles its risk. Ptosis, alone causes mild to moderate amblyopia. Patients with severe ptosis should be corrected before 3 years and all other patients before 5 years of age.

Classically Mustarde31 had staged the correction of the various deformities in the following manner:
1. Correction of epicanthus and telecanthus.
2. Correction of ptosis.
3. Correction, if necessary, of vertical skin shortage.

So far, most of the surgical techniques are two-staged procedures, consisting of Mustarde’s medial canthoplasting followed by fascia lata sling after 1 year. Only a few reports recommend one-stage treatment.27-30 In one-stage procedure, the vertical and horizontal lengths pull against each other. This results in the
Development of excessively strong traction in different directions on the palpebral fissure. There is high risk in developing loosening of the medial canthopexy and poor elevation of upper lid. About 1 week to 10 days after surgery, vertical fissure widens again as the swelling decreases.\textsuperscript{28,32}

**STAGEWISE CORRECTION OF BLEPHAROPHIMOSIS SYNDROME**

**Stage 1 Correction of Epicanthus and Telecanthus**
The basic problem to be addressed is the lateral displacement of the soft tissue at medial canthus, with lengthening of MCT. The surgery consists of the reorganization of the three soft tissue elements – skin and the underlying fascia, subcutaneous tissues and muscle, and MCT (Table 1).

Medial canthal skin is thicker and contains more glandular elements; consequently more scarring. Small dog ears and excessive skin tension, also promote scarring. During soft tissue excision, canalicul damage is prevented by inserting lacrimal probes.

In some there may be lateral displacement of the anterior lacrimal crest and clinically no hypertelorism. The solution to the problem is to perform transnasal wiring.

<table>
<thead>
<tr>
<th>Soft Tissue abnormality</th>
<th>Surgical procedure</th>
</tr>
</thead>
<tbody>
<tr>
<td>Skin</td>
<td>Spaeth’s Double Z-plasty</td>
</tr>
<tr>
<td></td>
<td>Verwey’s Y-V medial canthoplasty</td>
</tr>
<tr>
<td></td>
<td>Roveda technique</td>
</tr>
<tr>
<td></td>
<td>Mustarde’s Double Z-plasty</td>
</tr>
<tr>
<td></td>
<td>Kohn’s C-U plasty</td>
</tr>
<tr>
<td>Soft tissue</td>
<td>Excision</td>
</tr>
<tr>
<td>Medial Canthal Tendon</td>
<td>Plication (Bunnell’s technique)</td>
</tr>
<tr>
<td></td>
<td>Resection</td>
</tr>
<tr>
<td></td>
<td>Trans-nasal wiring</td>
</tr>
</tbody>
</table>

*Table 1. Surgical correction of soft tissue elements responsible for Telecanthus*

Earlier lateral canthoplasty was done to extent the fissure. The definity of lateral canthus is lost after such a procedure, and there is tendency for conjunctiva to creep over the lid margin. The lateral support for lid may be lost causing ectropion.\textsuperscript{32}

Operative techniques:

1. **Spaeth’s Double Z-plasty**\textsuperscript{35}

This procedure is used to correct only the Epicanthus (Figure 4). “Z” is marked on each lid with two apices joining over the MCT. Flaps are raised, transposed and sutured.

2. **Verwey’s Y-V medial canthoplasty**\textsuperscript{34}

Horizontally oriented “Y” over the medial canthus with the base of the stem at the desired new position is drawn (Figure 5). Pinching the skin together over the nasal dorsum will help this determination. The arms of “Y” are extended to the supraorbital and infraorbital folds. Skin incision is made and subcutaneous tissue is excised over the MCT. The sutures are used to fix the subcutaneous tissues to the MCT. The flaps are then trimmed and suture into the shape of “V”. It corrects both the Epicanthus and Telecanthus.

Disadvantages include prominent V shaped scar which doesn’t conform to Langer’s line and that only the leading edge of the V is maximally advanced.

3. **Roveda technique**\textsuperscript{34}

An incision is given parallel to and about 5-6 mm medial to the crest of the epicanthal fold (Figure 6). At the midpoint of the curve, “Y” shape incision is given. The arms of the “Y” are extended to a distance of 5-7 mm toward the nose. Thus, a five point junction is formed which should lie 1-2 mm medial to the underlying canthal angle. The two triangular flaps are excised and incision closed in W shape manner.

4. **Mustarde’s Double Z-plasty (Four-flap / Running man / Flying man technique)**\textsuperscript{31}
The intended site of the new canthus is marked, making the proposed ICD one-half of the IPD. By drawing the skin towards the nose and obliterating fold, second point is made and two points are then joined. From the centre of this, two lines are drawn outward at 60 degree, each equal in length to the original line less than 2 mm (Figure 7, 8 ). From these lines, back-cuts towards the nose of the same length are made at 45 degree. Finally, paramarginal extensions along the lids are drawn. The incisions are made, flaps undermined and retracted to expose MCT. The site of new canthus is now cleared of all tissues down to periosteum by blunt dissection. The MCT is then divided and the mattress suture is passed from its medial cut end to the periosteum. The flaps are transposed and trimmed to fit the defect accurately, and sutured.

The considerable suture material placed in small area may accentuate scarring.

6. Five flap technique (combined double “Z” and “V-Y” technique)

Incision lines are mapped out as follows (Figure 10). A point is made on the epicanthal folds medial to the actual site of the present canthus. Another point is made at the desired position of the canthus. A straight line is drawn between these two points on each side. Paramarginal lines are marked on the upper and lower eyelids and connected to the initial point on the epicanthal fold, thus forming a “Y” configuration. A curved line is drawn on the epicanthal fold through the angle of the Y and is equal in length on each side. This line forms the central member of the two “Z” configurations. The final limbs of each are then constructed roughly parallel to the stem of the Y. The angle on the final arm may vary from 45° to 60° depending on the amount of effect necessary. The configuration created is a double Z-plasty formed on the stem of the Y. The MCT shortening is performed. The Y-to-V flap is closed by first placing a suture in the apex of the V with a bite to the bony insertion of MCT to hold the skin as medial and posterior as possible. The Z-plasty flaps are transposed and sutured.

As concluded by the authors, the five-flap technique uses a logical, geometric flap design and provides aesthetically pleasing results. Construction is easily remembered; meticulous measuring, angle plotting, and trimming of flaps are eliminated.
7. Medial canthal tendon shortening

**Bunnell’s suturing technique**

MCT is not cut but is plicated and affixed to the periosteum at its attachment as shown (Figure 11).

**Trans-nasal wiring**

It is required in severe telecanthus with intercanthal displacement of 12 mm or more. After MCT exposure by any above method, an incision is made down to the peristomeum, 5-6 mm medial to the line of anterior lacrimal crest and extending as far up as the frontonasal suture line and down till the level of NLD. The periosteum is stripped with periosteal elevator, so that the lacrimal sac and the attachment of MCT can be displaced laterally. A bony window (12 x 10mm) is cut in the lacrimal fossa involving anterior lacrimal crest. No efforts need to be made to preserve the nasal mucosa. Similar bony window is made on the other side. A 0.3mm stainless steel wire is passed twice through MCT (second time 3mm more medially). The curved awl having a small hole is used to bring wire on to the opposite side. The ends of the wire are twisted and turned upward and posteriorly so as to be away from the lacrimal sac. If one canthus doesn’t advance far enough medially to be symmetrical with the other side, a trans-conjunctival lateral canthotomy can be done.

Great care must be taken to avoid injuring the lacrimal sac, common canaliculus or NLD.

**Stage 2 – Correction of ptosis**

It requires frontalis suspension procedure with autogenous fascia lata. In limited number of patients, with less severe ptosis and demonstrable levator action, a maximum resection of levator may prove adequate.

**Stage 3 – Correction of ectropion**

Medial canthopexy, by putting the lids on stretch will reduce moderate ectropion of the lower lid. Residual ectropion should be corrected after 12 – 18 months. Because the orbicularis muscle and orbital septum are short vertically, the latter must be freed from the orbital margin. The lid defect is covered by larger full thickness graft to compensate for the shrinkage. Since the shortage of orbicularis is also there, it results in a gap near the orbital margin which appears somewhat depressed with the passage of time.

**ONE STAGE CORRECTION OF BLEPHAROPHIMOSIS SYNDROME**

The patients with BPES have high rate of amblyopia. Traditional multiple surgeries may prolong the treatment course and most importantly, delay the management of amblyopia. The risk and expenses associated with two separate surgeries are decreased, the hospitalization time is reduced, and subsequent rehabilitation can be initiated in a more timely fashion by one-stage correction.

There have been few reports on the one stage surgical correction of blepharophimosis syndrome. Nakajima et al described good surgical results using medial canthoplasty and levator resection in one stage. Due to tendency towards marked lid lag after sling surgery, they preferred to use levator resection. They concluded that the emotional stress imposed on the child by staged procedure, can be avoided by one stage repair with good results.

Karacaoglan et al performed medial canthoplasty and frontalis suspension in one stage with good results.

Wei-qing et al treated 16 patients with BPES. They first performed the frontalis suspension to overcorrect the blepharoptosis for about 1 mm followed by medial canthoplasty using Mustarde’s technique. When modifying the flaps, they made the third flap accompanying with the double eyelid lines to decrease scar formation. Thus, after suturing the upper lids with 6-0 monofilament, natural double eye lines were achieved. The horizontal fissure of eye increased from 20 to 27 mm on average, the vertical length from 3 to 8 mm and the ICD decreased from 40 to 34 mm on average. Patients had completely closed eyelids without ectropion or exposing keratitis.

Wu et al retrospectively reviewed the 23 patients. They used the ratio of 1 – 1.2 between the ICD and HPFL as normal, and grade BPES into: Mild – 1.3
– 1.5, Moderate – 1.5 – 1.8, Severe - > 1.8. The surgery consisted of lateral canthotomy, medial cantholpasty, transnasal wiring and fascia lata sling or levator resection. In this study, 70% patients had good outcomes. Only 2 patients had the transnasal wiring loosened, attributed to the cheese wiring. 11% patients with HPFL more than 2mm underwent reoperation for ptosis as compared to 100% patients with HPFL less than 2mm. Therefore, regarding the SDWLHQWVZLWKSDOSHEUDO¿VVXUHJUHDWHUWKDQPPRQH stage operation provides acceptable results both in functional and cosmetic improvements.

**Conclusion**

Blepharophimosis - ptosis - epicanthus inversus syndrome (BPES) is a complex of multiple eyelid deformities. It is also associated with ocular, lacrimal, nasal and auricular anomalies. It has a great impact on patient’s functional status and visual development. Patients with severe ptosis especially with co-existent strabismus should have their ptosis corrected before 3 years of age, and all other patients should undergo surgery before 5 years of age. Depending on the levator function, one can choose medial canthoplasty with frontalis suspension using autogenous fascia lata or levator resection. The one-stage surgical correction can be performed when a child is less than 3 years of age with palpebral fissure more than 2mm, otherwise multiple stages of reconstruction are advised as early as possible.

**REFERENCES**

Blepharophimosis – (BPES)

ACCOMODATIVE ESOTROPIA
WHEN TO REMOVE GLASSES

Deepali Garg Mathur
Consultant, Max Eye Care, New Delhi

ABSTRACT: Accomodative Esotropia (AE) is an acquired convergent deviation of the eyes restored to orthotropia at near and distance fixation by optical correction of the underlying hypermetropia. Raab has summarized a number of well known characteristic features of this entity including post infancy onset, initial intermittency of deviation, above normal hypermetropia, elimination or reduction of deviation with glasses and frequent association of anisometropia and amblyopia. The mainstay of treatment is full correction determined by cycloplegic refraction. Details as to how cycloplegia is attained is important, with cyclopentolate being far less effective in causing cycloplegia than atropine. Weaning children with AE off spectacle correction has been known to encourage fusional divergence and emmetropisation. Whether undercorrection of hypermetropia stimulates increased reduction of hypermetropia is controversial. On the other hand, studies have shown that a mild reduction of hypermetropic reduction in AE may lead to larger angles of deviation and precipitate deterioration. Thus the risk of undercorrection may outweigh potential benefits of that approach.

INTRODUCTION:
Accomodative esotropia (AE) is a common entity, accounting for approximately half of all childhood esodeviations. Described by Donders way back in 1864, it is defined as a convergent deviation of the eyes associated with the accommodative effort necessary to overcome the blurred image caused by hypermetropia. It classically presents in the preschool years, 1.5 – 3 years, (though an early onset type has been seen under 1 year too), the deviation being typically eliminated by controlling the accommodative effort with optical correction of the hypermetropia.

On first glance, the treatment seems straightforward and one would expect good results. However, our evidence indicates that AE treatment outcomes are inconsistent and often less than ideal. The results are largely governed by the initial management of hypermetropia while the deviation is still intermittent (How early and how much?), the recognition and treatment of associated abnormal distance-near relationship and amblyopia and patient compliance.

Variations in treatment measures in hypermetropia management (full v/s undercorrection) as well as duration of treatment may account for differences in stereoaucity levels and precipitation of deterioration of control whereby strabismus surgery may be indicated.

To add to our consternation many patients of accommodative esotropia do not have bifoveal fusion. Emmetropisation and spontaneous resolution of AE occurs rarely if at all and may take many years. Whether a child with accommodative esotropia will ever “Grow out of glasses” remains a pertinent question, the main outcome measure being resolution or non-resolution of esotropia following weaning and eventual discontinuation of glasses.

CASE PROFILES:
To exemplify the course of AE, the following cases are cited:

CASE I : 4 YRS MALE CHILD FULLY ACCOMODATIVE ESOTROPIA WITH LEFT AMBLYOPIA
A 4 year old male child presented with left convergent squint at the age of 2.5 years. There was an associated left amblyopia. Cycloplegic refraction done under atropine revealed a hypermetropic error of +3.50 D in both eyes. AC/A ratio was normal by gradient method. Child was prescribed the full cycloplegic correction (corrected only for distance) with occlusion therapy. Within 6 months, the child was orthotropic for near and distance with glasses. Visual acuity with Lea Symbols was 3/4.5 in both eyes with good alternation. Stereoacuity was 240 sec/arc.

**CASE II - 11 YRS MALE CHILD WITH FULLY ACCOMODATIVE ESOTROPIA WITH LEFT AMBLYOPIA**

The child presented at 3.5 yrs with a left convergent squint. He was refracted under tropicaeryl and Homatropine variously and groosly undercorrected with +1.50 D in both eyes till 5.5 yrs. Occlusion therapy was however started for left amblyopia. He was advised surgery at the age of 5 years. Refused surgery and presented to us at 5.5 yrs. Presenting BCVA was 6/9 in the right and 6/18 in the left eye. Refraction under atropine revealed +4.0 D in the right eye and +5.0 D in the left eye. In 6 months, he was orthotropic for near and distance with glasses. BCVA improved to 6/6 in the right and 6/6p in the left eye. Stereoacuity remained poor (480 sec/arc).

**CASE III: 6 YRS FEMALE CHILD : ACCOMODATIVE ESOTROPIA WITH HIGH AC/A RATIO**

Diagnosed as accommodative esotropia at the age of 4 yrs. Refraction done under atropine and full refractive correction prescribed. BCVA was 6/6p in both eyes. The child is orthotropic for distance with glasses. Residual near deviation with high AC/A ratio overlooked. Repeat refraction and prescription of bifocals planned. Stereoacuity – 240 sec/arc.
CASE IV: 8 YRS MALE CHILD ACCOMODATIVE ESOTROPIA WITH TREATED HIGH AC/A RATIO

Onset of Accomodative esotropia at 3 years. Initially prescribed constant use + 1.50 D in both eyes. Bifocals with + 3.0 near add given at 4 years. Orthotropic for near and distance on regular follow up. At 7 years near add reduced by 1 D and by 1 D more this year. Maintains good alignment. Stereoacuity- 120 sec/arc.

CASE V: 18 YRS FEMALE CHILD – DETERIORATED ACCOMODATIVE ESOTROPIA

This patient had an onset of accommodative esotropia at about 5 yrs. No treatment initiated till 12 years of age. On examination her esotropia reduced by 15 PD with hyperopic correction of + 2.0 D in both eyes. The BCVA was 6/9 and 6/6p in the right and left eye respectively. Residual esotropia of 25 PD with glasses( non – accommodative element) requiring strabismus surgery. Poor stereopsis.

DISCUSSION:
An understanding of accommodation, convergence and hypermetropia and their interrelationships is essential to the appropriate management of AE. Practically the most important factor is how to manage the hypermetropia.

Our evidence from the case profiles enumerated above bring to light the following facts:
1) Children with fully accommodative esotropia require maximum hypermetropic correction.
2) Refraction under atropine should be carried out especially under 10 years of age.\textsuperscript{13}
3) Check alignment for both distance and near
4) Binocular function should be assessed regularly
5) Detect and treat associated amblyopia
6) Delayed treatment may precipitate deterioration and that is irreversible.
7) Regular follow up is mandatory

Keeping these in focus, we are still faced with the dilemma of when if at all we can wean the child off glasses??
Perusal of literature in this regard has varying and conflicting views. Excessive hypermetropia plays a causative role in at least 50% of all cases of accommodative esotropia. Studies have revealed that hyperopia increases before the onset of esotropia and then decreases after 7-8 yrs. Raab observed a reduction of 0.18 D per year in accommodative esotropes compared with 0.22 D per year in normal hyperopic children. Other studies have indicated that esotropes behave differently from normal hyperopes and are destined to remain more hyperopic than their counterparts. The need for bifocals can however be eliminated by early teens.

The impact of full correction of hypermetropia versus undercorrection has also been under close scrutiny. The stimulus for emmetropisation is blur. So it has been postulated that prescribing the full spectacle correction may prevent emmetropisation. Also speculated is that esotropes may be able to discontinue spectacle use as a result of increased fusional divergence amplitudes, the loss of hyperopia or a reduction in the synkinesis between accommodation and convergence.

However other studies state that accommodative esotropes behave differently from the normal population because of an intrinsic defect in emmetropisation behavior leading to slower loss of hyperopia and are destined to remain that way. How long does the patient have to wear glasses? Most reviews of AE are concerned primarily with its management in childhood. One of the few long range follow ups done by swan on 39 adults (23-46 yrs), found that 38/39 adults diagnosed and treated for AE were still wearing glasses or contact lenses full time. Shipmann et al reported a recurrence of accommodative esotropia in 11 adults between 20-65 yrs of age. In synchronisation, Ewen states that even a small reduction of only 1 D in the full spectacle correction of a fully controlled esotrope can result in a significant increase in the deviation. Also while undercorrection may improve fusional divergence, it may not cause a more rapid decrease in hypermetropia.

CONCLUSION
1) Full spectacle correction in accommodative esotropia restores alignment, averts amblyopia and maintains stereopsis.
2) The hyperopia reduces but not significantly by early adulthood.
3) Accomodative esotropes have to wear optical correction till adulthood. The prospects of discontinuing glasses remain poor while the need for bifocals can be eliminated by 10-12 years.
4) Weaning of glasses if attempted should be only in cases with good alignment and binocularity but under close scrutiny and follow up for deterioration. Present efforts should be directed to a more cautious and realistic though unfortunately not more encouraging advice to affected individuals.

References
Choroidal melanoma is the most common primary malignant intraocular tumour. Traditional management of choroidal melanoma is enucleation and local resection. Radiotherapy is emerging as an alternative to surgery, salvaging the vision of the patient. The most commonly used mode of radiotherapy is brachytherapy. With Iodine 125 plaque, 50% of the tumours are seen to reduce to 50% of their pretreatment size. External beam irradiation using charged particles such as protons and Helium ions is another option available. Trans-pupillary thermotherapy, Gamma knife therapy and Carbon ion therapy are thought to be the future of treatment of choroidal melanoma.

Choroidal melanoma is the most common primary malignant intraocular tumour and the second most common type of primary malignant melanoma of the body. It occurs in 6 persons per million of the population annually. The mean age of diagnosis is mid 50s. These melanomas occur sporadically. The role of sunlight and other environmental factors in its pathogenesis remains unknown.

In the histopathological examination, three distinct types of cells are recognised: spindle A, spindle B and epitheliod. They may have variable coloration, ranging from darkly pigmented to purely amelanotic. Growth of the tumour can occur silently but with time it may produce a visual field defect, vision loss, cystoid macular edema, exudative retinal detachment, vitreous hemorrhage, cataract or rubiosis iridis. On presentation, patient can have systemic metastases, most commonly in liver.

The management of choroidal melanoma has following options –
1. Observation
2. Local Resection
3. Enucleation
4. Trans-pupillary Thermotherapy
5. Photo-coagulation
6. Radiotherapy
Periodic observation for the growth of the tumour is done with serial fundus photography and USG. Local resection is done in selected cases of small melanomas, in the form of Sclerouveoretinectomy (full thickness eye wall resection)\(^1\) or partial lamellar sclerouveactomy. Enucleation and exenteration are other surgical options for advanced tumours. Photo-coagulation\(^2\), done in past with Xenon laser, gives better tumour control. Nowadays, it is done with Argon Laser which offers fewer complications. Trans-pupillary thermotherapy (TTT) is the modality of treatment that delivers heat to the tumour in infrared range using a modified Diode Laser delivery system. It is preferred over Photo-coagulation for tumours less than 3mm in thickness and are located more than 3mm away from the fovea. TTT is used as a supplement to Plaque therapy. Radiotherapy is most commonly used in the form of Plaque therapy. In the past cobalt-60 plaques were used. These days, Iodine-125 and Ruthenium-106 are used. The other form of radiotherapy is charged particle application. Metastases in liver are treated with hepatic arterial chemo-embolisation with Cis-platin and Polyvinyl sponge.

**Brachytherapy for melanoma**

Brachytherapy is the application of radiation from isotopes over very short distances, in contact with the tissue surface. It was first done by Moore in 1930 with Radon seeds. Now we use Iodine 125 and Ruthenium 106. In Collaborative Ocular Melanoma Study (COMS), the medium sized tumour patients were randomised between enucleation and I- 125 brachytherapy\(^3\). COMS detected equal mortality rates in both the groups up to 12 years of follow up.

Indications of brachytherapy in choroidal melanoma are-
1. Small melanoma with evidence of growth on follow up
2. Medium sized melanomas of choroid and ciliary body in eyes with useful vision
3. Large melanoma ( Up to 16 mm in diameter and 8-10mm in thickness)
4. Unilateral large melanoma
5. Melanoma overhanging the optic disc

Total dose of radiation given is 50- 100 Gy . They gave 100 Gy radiation to the tumour apex at the rate of 50-125cGy per hour.

Iodine 125 is convenient with respect to half life, shielding, tissue penetration and physical form. Gold plaque is used which has 0.4mm thickness with lip around its perimeter. Within the gold plaque, there is sialistic seed carrier insert with evenly spaced troughs that accept I-125 seeds. The carrier is designed so that seeds are adjacent to gold. The prepared loops facilitate anchoring the plaque to the sclera with sutures.

Before placement of plaque, it is very important to measure dimensions of tumour (esp the base) properly either comparing it with the disc diameter or with Indirect ophthalmoscopy and 20D lens with grid. Extra ocular extension less than or equal to 2mm is acceptable for brachytherapy\(^4\). Usual implant duration is 4-5 days. On follow up, I/O, fundus photography and USG should be done. Half of the tumours shrink to 50% of their pre-treatment thickness. On first follow up visit, the tumour actually increases in height due to edema. If any time other than first visit, the tumour increases by 15% or boundary expands by 250 mm, tumour expansion is suspected. In such a case, patient should be followed up every 3 monthly. If additional expansion by 250 mm or increase in thickness by additional 15% is detected, brachytherapy is declared to be a failure.

Plaque tilt after initial accurate placement occurs frequently during brachytherapy and may represent an important cause of local treatment failure. In COMS, 10% of eyes were enucleated because of suspected or documented recurrence\(^5\) after brachytherapy. Factors affecting recurrence are-
- intra operative USG to see placement of plaque
- Size of tumour at the time of surgery
- Proximity of posterior tumour margin with the disc.

Local edge recurrences can be controlled with photocoagulation or TTT. Brachytherapy can cause changes in the surrounding retina like atrophy,
hyperplasia, fibrous metaplasia of RPE, retinal gliosis or blood vessel abnormalities. Tumour and radiation effects lead to poor visual acuity in 77\% of eyes. The amount and severity of retinopathy and optic neuropathy after iodine-125 brachytherapy increased through 8 years of follow-up. Assessment of photographs and angiograms has provided reliable estimates that local tumour control was achieved in 90\% of the cases. Tumour and radiation effects led to poor visual acuity in 77\% of eyes. The metastatic rate was 13\% and the mortality rate was 3\%. The 5-year cumulative incidences of cataract, iris neovascularization, and glaucoma are found to be (20) 69\%, 62\%, and 60\% respectively. The 5-year incidences of maculopathy and optic neuropathy are 52\% and 46\%, and those of vitreous hemorrhage and persistent RD were 36\% and 25\%, respectively. Cataract is the earliest complication to appear.

### Charged Particle Irradiation of uveal melanoma

This is done with the help of external beam irradiation using charged particles such as protons and Helium ions. These have minimal scatter and finite, well defined tissue range. The tumours selected are up to 24 mm in diameter and 14 mm in height. Tumours involving macula, disc or both or with small extrascleral extensions are not contraindications for this therapy. Any systemic metastasis or primary malignancy in the other tissues must be ruled out. The conjunctiva is incised, tumour is localised with transillumination and indirect ophthalmoscopy. Extrasceral extension is to be looked for. Edges of the tumour are marked with pencil and four metal rings, 2.5 mm in diameter, are sutured to the sclera to outline the tumour. If the tumour is in contact with optic nerve, rings are placed only anteriorly and laterally while the distance of rings from the posterior margin is estimated from fundus photographs. For tumours extending to ciliary body and iris, rings are placed at the choroidal edges of the tumour and the distance of anterior margins from the rings is measured. A light beam coaxial with the central axis of proton beam is used to position the tumour relative to the beam. Three dimensional treatment planning computer programmes is used. A total dose of 70 cobalt Gy equivalents is delivered in five equal treatments over 7 to 10 days. Positioning of the patient is achieved with a head holder which allows controlled rotation of the head around two mutually perpendicular axes intersecting at a point that is positioned accurately on the axis of collimator. Orientation of patient’s eye is established by voluntary fixation of eye to be treated or the other eye. Lid speculum is used to keep the eye open. A 20x magnification monitoring system is used. A margin of 1.5mm is included in the treatment field around the base of the tumour. Each treatment takes about 1 min. First follow up is to be done 6 weeks after treatment and then 6 monthly. Most of the tumours show regression after the first 6 months of treatment. Disappearance of tumour with a flat remnant scar is considered complete regression and is observed in 15\% of cases. Serous retinal detachment due to the tumours can transiently increase in size after therapy. But it resolves after few months. Post-treatment visual outcome depends on height of the tumour, its proximity in relation to fovea and disc, pre-treatment vision, dose of radiation received at macula, degree of retinal detachment and presence of diabetes. Risk of significant loss of vision (\(<20/200\)) by 10 years after treatment is 16\% in low risk group and 99\% in high risk group. The complications of external beam radiation therapy include intratumour hemorrhage, transient diplopia, lid epithelitis, punctual occlusion,
epithelial keratopathy, radiation vasculopathy, maculopathy and papillopathy. Rubeosis iridis, Neovascular glaucoma are observed in 16% eyes and are associated with large tumour size. Cataract development chances increase with the amount of lens doses and larger tumour height. Local recurrence after therapy is observed in 2 to 5% of the tumours. It can be retreated successfully with repeat proton irradiation or laser photocoagulation. Survival is poor after local recurrences; 10 year survival rates were 72.6% for patients who experienced tumour control and 47.5% for those with re-growth. The 5 year cumulative probability of developing metastasis after charged particle irradiation and enucleation is 20%. The probability of metastatic death varies between 5% for patients with lower risk characteristics and 63% for those with high risk characteristics (large tumour, advanced age, moderate to heavy tumour pigmentation, presence of symptoms, ciliary body tumour origin and green or blue iris colour).

**Laser Treatment**

Argon blue–green laser (wavelengths 488 and 514.5 nm) are used. For TTT, Nd:YAG laser with a wavelength of 1064.5nm is considered these days. But the risk of unwanted tissue disruption and haemorrhage is higher with it. Krypton red (647.1nm) offers deeper penetration in the choroid. Krypton laser offers benefits in treating amelanotic melanomas, but the power generated by it may be insufficient and chances of laser induced haemorrhage are higher. Lasers treat the intra-ocular tumours at a temperature level of more than 75 degree C. Photocoagulation should be restricted as a treatment modality for small choroidal melanomas, not extending to the ciliary body, <3mm in thickness, lateral extension <30 degrees and presence of clear media. Laser can be used in treating the tumours close to macula or disc where radiation therapy can cause severe vision loss, optic neuropathy, radiation retinopathy or macular oedema. During therapy, the tumour is first surrounded by choriotinal scar, separating the mass from its blood supply followed by direct tumour coagulation. Because of the presence of pigmentation, the tumour absorbs more light and doses are accordingly adjusted to avoid explosive burns. Firstly tumour is surrounded by 1 or 2 rows of laser with a spot size of 800 to 1000mm and an exposure time of 1 to 1.5 seconds. After 4 to 5 weeks a second circulating coagulation is performed with treatment of surface of the tumour. Third coagulation is performed after additional 4 to 5 weeks when entire surface of the tumour is photocoagulated again. Treatment is suspended when choroidal mass is converted to an atrophic, flat chorioretinal scar. 7 to 8 sessions can be required to achieve this. Argon laser is used in the first step and krypton laser is used for the next ones. Neovascularisation of retina, developing after vein occlusions is the most severe complication of lasers. Vitreus hemorrhage, tractional retinal detachment, preretinal membrane formation, cystoid macular oedema or burns in anterior segment can also occur. Recurrent tumour growth either at the margin of tumour or at the centre are reported. Tumour regression is achieved in more than 80% of cases. Shields et al reported an initial response rate of 100% but re-growth was reported in 14% of cases on follow up. COMS has shown that enucleation has no prognostic advantage compared to eye salvaging radiotherapies and other modalities of treatment.

**Transpupillary Thermotherapy**

TTT gives long exposure sub-threshold photocoagulation with a long wavelength. The maximum temperature achieved is 42 to 44 degree C which is used to enhance the cytotoxic effect of radiotherapy. A temperature of 65 degree C with TTT can be considered as directly cytotoxic and no additional radiotherapy is required. Diode laser (810 nm) with large spot size up to 3mm for exposure time of 60 seconds is used. TTT is initiated at the tumour centre and no circumscribing treatment is performed. 3 to 4 sessions are required to destroy the tumour. Tumour regression is seen in 6 to 9 months leaving an atrophic scar with central pigment and visible sclera. In tumours <12mm in diameter and <4mm in height, located behind the equator, regression achieved was more than 90% of the cases. 9% of the tumours showed re-growth in tumours abutting or overhanging the disc or those requiring more than 3 sessions for regression. Complications are comparable to photocoagulation treatment.

**Laser treatment as ancillary therapy**

- Either TTT or photocoagulation is helpful in reducing the risk of continuous or recurrent tumour growth after surgical excision. Routine photocoagulation at the edges of pseudocoloboma is recommended after the trans-scleral local resection.
- The combination of radiotherapy and TTT (Sandwich technique) has shown a tumour control rate of 97% at 5 years. Radiation induced complications are an important indication of supplementary photocoagulation. OCT is useful in the early detection of radiation-induced macular oedema in this situation, before clinical signs of radiation maculopathy develop and before substantial visual loss occurs.
The development of macular oedema is significantly associated with larger tumour size.

- Exudative retinal detachment either primary or secondary to brachytherapy can be treated with scatter photocoagulation combined with vitreoretinal surgery.\(^\text{19}\).

**Recent Advances in Radiotherapy**

**Gamma knife Radiosurgery**

As an alternative for enucleation, gamma knife radiotherapy is proposed using a radiation dose of 30 to 50 cGy. After giving retro-bulbar anaesthesia and fixing the patient’s head in stereotactic frame, tumour is localised with the help of gadolinium enhanced MRI. Maximum dose is delivered at 100% isodose line while periphery of the tumour receives 50% of the isodose. 78 patients with a mean age of 64 years, and tumour more than 3mm thickness showed a 5 year survival rate of 81.9% with local tumour control of 91\(^\text{20}\). These rates are comparable to enucleation. Though the eye retention rate was 89.7%, there was significant absolute reduction in visual acuity. The major ocular complications seen were exudative retinopathy (in 33.3%), neovascular glaucoma (in 18.7%), radiogenic retinopathy (in 13.5%), optic neuropathy (in 15.5%), vitreus hemorrhage (in 10.4%).

**Carbon Ion Therapy**

The vertical (140 MeV/u) and horizontal (170 MeV/u) carbon ion beams from the synchrotron are shaped, using the passive beam delivery system, such as to irradiate the target volume of the tumour. The range modulating ridge filters are designed to produce spread-out Bragg peaks (SOBPs) with a region of uniform cell killing. The apertures and range compensators are designed for individual patients. Dose distributions are calculated with either a broad beam or a pencil beam algorithm using parameters determined by measurements and calculations. In a study by Koyama Ito H\(^\text{27}\), the system was used for 12 patients during one year. For nine patients two-port treatment was assessed to be more effective than mono-port therapy and these patients were treated with two fractions of vertical beams and three fractions of horizontal beams.

**Conclusion**

In future, radiotherapy is likely to become an alternative for enucleation in the management of choroidal melanoma. The COMS gives us the data suggesting that survival following more recently developed brachytherapy is similar to enucleation. In situations when the primary outcomes of differing treatments are essentially the same, any preference of one therapy over another would have to be based on other criteria, such as severity of adverse effects, quality of life issues, or treatment costs. Knowledge about the impact of enucleation for choroidal melanoma on the performance of vision-dependent activities might influence patient selection of equivalent therapies.

**References**

8. Seddon JM, Gragoudas ES, Egan Km et al. Uveal melanomas near the optic disc or fovea: visual results after proton beam irradiation. Ophthalmology 1987;94:354-361
Eyelid trauma still seems to be a taboo among general ophthalmologists. We present a general overview such that these cases may be taken up in most settings.

Work up of a patient with eyelid trauma:

History Taking:
It is of note that periocular trauma can occur as an isolated injury or as a small part of multisystem trauma. It is imperative to obtain adequate details as to how the injury occurred. If it appears that there was deep penetration into the orbit, it is to be determined as to what instrument caused the injury as there could be fragments or foreign bodies lodged within. Other aspects worth mention are history of alcohol intake as this may lead to distortion of facts presented and children may not give adequate history. As almost all cases of trauma are taken up under general anaesthesia the importance of nil per oral status and neurological and cardiopulmonary status is not to be undermined.

Recording of visual acuity:
Preoperative vision is extremely important for medical and medicolegal purposes. Both distance and near vision must be recorded. Importantly the vision should be assessed for both the eyes as you maybe dealing with optic nerve/ chiasmal traction. The pupils as repeatedly quoted are a window to the brain. Direct, consensual and evidence of any RAPD are to be documented.

Evaluation of the globe and orbit:
Examination of the eye is important in periocular trauma. The eye itself should be examined for movements and any sign of perforation. Patients are most likely to be uncooperative so thorough evaluation in General Anesthesia is mandatory, as a globe perforation may be missed in the surgeons zeal to repair the periorbita.

Radilogical studies:
The Periocular trauma needs adequate imaging to assess the underlying anatomical derangements. A CT Scan is the most appropriate study. Be sure the quality is good as many a time the CT may only be assessed when the patient is already under General Anesthesia. An MRI is to be performed when optic nerve injury is of suspect.

Investigations:
A routine Hemogram and a Bleeding Time, Clotting Time may allow to assess the blood loss as the facial area is highly vascular.

Types of Eyelid Trauma and its management:
Injury of the eyelid may be divided into blunt and penetrating trauma.

Blunt Trauma:
Ecchymosis and edema are commonly the presenting signs of blunt trauma. A complete evaluation may be advised as described above.

Penetrating Injury:
A thorough knowledge of eyelid anatomy is essential while repairing a penetrating eyelid injury. The management depends on the depth and location of injury.

We discuss management of four most common types of eyelid trauma:

A Eyelid Margin Repair
B Simple Laceration Repair
C Complex Laceration Repair
D Canalicular Laceration Repair
place give one temporary tie to see the alignment. If you are not happy replace it. Once the alignment is adequate tie the 6 ‘0’ silk and leave the cut end long. Once tied the margins should have a pout upwards as healing and fibrosis of a flat alignment may cause a notch later on.

5. Once this has been achieved two interrupted sutures maybe passed through the anterior lamella and the posterior lamella in a vertical fashion. All these sutures should be left long as there is the risk of corneal abrasion / defect.

6. The remaining defect of the eyelid if deep may be closed in layers, deeper tissues with vicryl and superficial tissues with silk. The lid margin sutures you have left long may be now pulled and included in the knot of one your skin sutures. You can leave these sutures for seven - ten days. A bit longer is better as tissues under traction may tend to gape on removal.

7. Supportive treatment in the form of The Penicillin group and Clavulanic acid may be given for five days, steroids may be included to reduce edema and inflammation.

B. Simple Laceration Repair

Facial lacerations should be repaired within 24 hours for best results or else infection and necrosis may encroach. Before repairing any trauma be sure to thoroughly irrigate the wound with betadine and saline. This will help in washing off any contamination. In our experience it is best to repair complex lacerations in GA. Two reasons may be cited for this. A) Patient cooperation. B) Injecting local anaesthesia makes the cut margins boggy and such margins may not attain adequate apposition. Superficial eyelid lacerations involving just the skin and orbicularis require only skin sutures. Basic principles of plastic repair may be followed which include conservative debridement ( extensive debridement is not necessary as facial wounds heal magically due a generous vasculature), use of small caliber sutures, evasion of wound edges and early suture removal. It is also important to assess that the laceration has not involved deeper structures of the orbit and the brain ( communication through the orbital roof or cribiform plate ). Any orbital fat visible indicates the violation of the orbital septum and requires the need to do a levator exploration. A lacerated levator aponeurosis/ Muscle must be repaired to achieve adequate pre – trauma function. Upper eyelid lagophthalmos and tethering to the superior orbital rim are common if the orbital septum is incorporated into the laceration repair. Orbital septum lacerations need not be sutured as they may heal themselves by local fibrosis.

Figure 1: Eyelid Margin Repair : A. The eyelid margin is aligned with 6 ‘0’ silk grey line sutures, anterior lamella sutures and posterior lamella sutures. The tarsus to tarsus closure is done with 6 ‘0’ vicryl sutures. B. The tarsal sutures are tied and cut. The lid margin sutures are tied and left long. C. The skin is closed with 6 ‘0’ silk and the long lid margin sutures are tied into them to avoid any abrasion.

A. Eyelid Margin Repair (Figure 1)

The eyelid margin repair requires dexterity and in all possibilities should be done under the microscope and if not under adequate light. The repair begins with identifying the adequate landmarks of the eyelid.

The following surgical steps maybe followed:

1. GA or local anaesthesia ( xylocaine + adrenaline + topical anaesthesia)
2. Assess and align the lid margin with two forceps, be careful not to pull as traumatic tissue is lacking strength.
3. If the lid margin is closing with strain a lateral canthotomy may be done.
4. Hold one edge of the cut end with a forceps and your first bite should be taken a bit long on the grey line with a 6 ‘0’ silk suture. Watch the needle as it comes out through the centre of the laceration vertically. Now in the same plane enter the other end of the laceration and come out long through the grey line on the other side (try to keep the distance of entry and exit at the same distance on both sides ). Once this is done you have to go back the same way but with shorter bites. Practice this on two cut ends of thermocol. Once this vertical mattress suture is in
Complex Laceration Repair (Figure 2,3,4):

These lacerations can test the skill and patience of a surgeon. The foremost rule is not to hurry a closure. Assess the laceration, take photographs and if necessary do not hesitate to discuss or ask assistance of your colleague. In most oculoplasty settings a reconstruction in the primary setting is the best, so the dictum is to leave nothing for a second surgery unless absolutely necessary.

Figure 2 A: Complex lid laceration in a 60 yr. old after RTA. The lid is split into multiple strands of tissue.

Figure 2 B: Post – Op Day 1

Figure 3 A: 5yr. old child after falling off a roof.

Figure 3 B: Immediate Post- Op. Pegs are fonix formation sutures

Figure 3 C: 3 months Post – Op

Figure 4A: 18yr. old male after a RTA – Hit and Run case

Figure 4B: Immediate Post – Op

Figure 4 C: After 1 month.

Eyelid trauma
C. Complex Laceration Repair

The following priorities may be cited when dealing with complex reconstructions.6,7

- Development of a stable eyelid margin
- Provision of adequate vertical eyelid height
- Adequate eyelid closure
- Smooth epithelialized internal surface
- Maximum cosmesis and symmetry.

The following steps may be followed in complex lacerations.

A. Clean and inspect the wound
   1. Rule out deep injury
   2. Remove any foreign material
   3. Do not debride any tissue unless it is infected as this may unnecessarily enlarge the defect.

B. Steps of laceration Repair :8,9
   1. To begin the laceration repair identify anatomic landmarks such as the eyebrow, the lid margin and the canthus.
   2. The posterior lamella should be identified and repaired first. In case of large lacerations involving tissue defects advancement flaps may have to be taken which will be discussed later. It is to be noted that during all lid reconstructions it is vital to use the Jaeger type lid guard. The under surface of the guard should be well lubricated with Xylocaine jelly or ointment. Inadverent perforation during lid repair may occur if this precaution is not taken, as you cannot judge your depth in a bloody field.
   3. To reduce the strain you may place temporary ‘tacking’ sutures on the anterior lamella which may ease your suturing of the posterior lamella.
   4. The following structures need to be identified while suturing the posterior lamella, the conjunctiva, the tarsus, the canthal tendons and levator aponeurosis.
   5. The conjunctiva is apposed with 8 ‘0’ vicryl.
   6. The tarsus is closed with 6 ‘0’ vicryl.
   7. Once the tarsus is sutured it is important to identify orbital septum and the levator aponeurosis. Many a time the septum has been breached and the fat prolapsed. This actually can make it easier to identify the levator which lies under the fat. The levator may be found intact, lacerated or disinserted. The levator may be carefully explored and repaired. Levator should be repaired with non absorbable sutures such as 6 ‘0’ Nylon. Again it is important to assess whether the levator is accessible or not, if it is difficult to identify the muscle a secondary repair may be attempted at a later stage.
   8. The lid margin is closed as described above.
   9. The subcutaneous tissue is approximated with 6 ‘0’ vicryl and the overlying skin with 6 ‘0’ silk. Adequate closure of deep structures allows accurate approximation of skin and subcutaneous tissues.
   10. The Canthal tendons are important structures which are commonly damaged in lid trauma. The Lateral canthal tendon is inserted onto the Whitnall’s tubercle. When the tendon can be isolated it can be attached directly to the lateral orbital rim with 4 ‘0’ nylon or ethibond. The effect of the tendon may be recreated by reattaching a portion of the tarsus ( The upper and lower crus of the lateral canthal tendon extend from each tarsal plate to form the stem of the tendon ) to the lateral orbital room. This is done by splitting the lamella towards the lateral edge of the lid. After splitting the lamella a tarsal strip is created. This tarsal strip is hooked with 4 ‘0’ Nylon. Now the lateral orbital rim is exposed and the tarsal strip is attached to periosteum of the orbital rim. It is important to take careful bites near the rim as the globe lies snug out there with the lateral rectus.
   11. The medial canthal tendon is a more complicated structure than the lateral canthal tendon. There are two limbs, the anterior limb is attached to the anterior lacrimal crest and the posterior limb to the posterior lacrimal crest. Avulsion of the anterior limb does not distort the position of the eyelid while the posterior limb does. Suturing the posterior limb of the medial canthal tendon can easily damage the nasolacrimal system. This becomes easier if the Nasolacral Duct system is already damaged. The suturing may be done with 4 ‘0’ Nylon. (Figure 5) 11

Figure 5A : Medial Canthal Avulsion: A punch drunk case of a 40 yr. old male

Figure 5B : Immediate Post – Op
D. Canalicular Laceration Repair

A canalicular laceration should be assumed when there is any injury close to the lid margin and medial to the puncta. These lacerations may occur by 1. Direct trauma 2. An avulsion due to a lateral pull on the eyelid. Direct trauma lacerations are easier to repair as the laceration lies in the centre of the canaliculi while avulsion type injuries tear the canaliculus from the sac itself making visualization of the proximal cut canaliculus difficult.

Methods:
1. Repair using Crawford stents.
2. Repair using the pigtail probe.

1. Identifying the cut ends of the canaliculus:
   - The microscope should be used preferably in all cases.
   - Pouring normal saline to pool in the medial canthal area and then pushing air through the intact canaliculus allows bubbles to be visible in the area of the medial end of the cut canaliculus.
   - Diluted Fluorescein is taken and pushed through the intact canaliculus to become visible through the medial end of the cut canaliculus. Dyes such as methylene blue and gentian violet should be avoided as they may stain the whole operating field.
   - The cut end should be explored under direct microscopic visualization.

2. Passing the stent through the canaliculus:
   - Thread the stent through the punctum and out...
through the distal end of the lacerated canaliculus.

• Next step is to thread the stent through the proximal end of the canaliculus

3. Passing the stent through the Naso Lacrimal Duct.

• This step is similar to all nasolacrimal duct probing. If you are intubating an adult it is helpful to bend the probe of the stent into a curve to maneuver around a prominent brow. Intubation of adults requires more manipulation than children.

• Retrieve the stent and pull it out of the nose, it is simplified if a Crawford hook is available.

• This should only be attempted if you have adequate experience in manipulating the Naso Laerimal Duet system or else you can cause unnecessary damage to the surrounding structures including forming false passages.

• A similar procedure is performed for the opposite canaliculus.

4. Suturing the pericanalicular tissue.

• It is rather difficult to identify and suture the canaliculi together. It is best to suture the pericanalicular tissue around the silicon intubation giving the best anatomical alignment. This maybe done with 6 ‘0’ vicryl.

• Tie the stent in the nose. A high tie may cause blocking of the NLD and a low tie may allow the tube to hang below.

Repair using a pigtail probe (Fig. 8) : 13, 14

• A pigtail probe is a smooth metal probe with an eyelet at one end. It is a curved sickle shaped probe. It is important that the leading end be smooth as a sharp leading end may cause trauma and false passages. There are many sizes available and an appropriate size maybe used.

• The tip of the pigtail probe is passed through the intact punctum and rotated through the normal canaliculus to the medial canthus maintaining a rotatory movement.

• At the medial canthus the probe should go posterior to the medial canthal tendon. You will know that you are in the correct plane if you lift the probe toward the ceiling of the operating room and meet resistance at the canthus.

• Use gentle pressure to guide the pigtail probe through the common canaliculus. The probe should emerge through the cut proximal end of the canaliculus without exerting much pressure.

• The pigtail probe is passed through the distal end of the canaliculus and out through the punctum.

• Now a 6 ‘0’ nylon suture is passed through the eyelet and the pigtail probe is rotated out the same way you entered.

• A hollow silicon tube commonly available for intubation DCR is now carefully threaded over the nylon suture. It is advisable to use a BD stent because their quality and caliber are superior to the stents available locally.

• Now the opposite end of the nylon is gently pulled to allow the silicon tube to thread through the canalicular system, through the distal end of the canaliculus and out through the punctum.

• The two ends of the silicon tube now lie outside the upper and the lower canaliculus.

• The pericanalicular tissue is repaired with 6’0’ vicryl.

• The two ends of the silicon tubing are now sized and sutured end to end with 10 ‘0’ nylon suture forming a ‘ring’. The sutured area is rotated into the canaliculus to give a smooth surface.

In the event of inability to pass the pigtail probe you can try intubating with a Crawford stent.

When a pigtail probe is not available a silicon intubation set maybe used. The metal probe is bent to form a pigtail, care must be taken to keep an adequate internal diameter by assessing before hand the medial canthal area and the distance between the two canaliculi.

Figure 8 : Canalicular laceration repair with the pigtail probe : A. Choose the appropriate curve of the probe. Note the blunt end with the eye. Enter the normal punctum and rotate the probe posterior to the medial canthal tendon. The probe will emerge from the cut end of the canaliculus and then make it come out of the punctum adjacent to the cut end. Thread a 6‘0’ Nylon suture through the eye of the probe. B. Rotate the pigtail probe back the way you came .C. Thread a silicon tube over the nylon suture till it comes out of the opposite end. D. Suture the ends of the silicon tube in the form of a ring and rotate it inwards.
**Post operative care**

Post operative care is of utmost importance, as you may spend hours closing a complex lid laceration but infection can ruin your efforts.

- Don’t be in a hurry to close. Make sure sutured edges are properly cleaned with betadine and ointment applied.
- A good antibiotic preferably Intravenous for the 1st 2 – 3 days (e.g. Monocef + Vancomycin) followed by oral Amoxyciilin + Clavulanic acid.
- Oral Steroids maybe given in clean cases to resolve postoperative inflammation.
- Sutures are to be removed in 7 – 10 days and sutures under traction can be removed after 2 weeks.
- Silicon intubation maybe removed after 4 months.
- Fornix formation sutures are removed in 3 weeks.

**References :**

Corneal grafting in high risk corneas remains a challenge. The management of high risk corneal grafts mandates a thorough knowledge and experience in handling systemic immunosuppressive therapy besides optimal surgical expertise. The immune privilege afforded by the absence of antigen presenting cells in the donor tissue, avascularity of the recipient tissue and anterior chamber associated immunodeviation help to maintain clear corneal allografts without supplemental immunosuppressive regimen in “normal” risk cases of keratoplasty. It is to be observed that primary low-risk corneal transplants have the highest success rate (90%) and lowest rejection rate (11–18%). High risk corneal grafts are those in conditions in which the cornea loses its immune-privileged status, resulting in its susceptibility to immunological rejection induced failure. Eyes under high risk have been defined as having at least two quadrants of stromal vascularisation and/or a history of previous graft rejection. Herpes simplex virus keratitis (HSV), chemical injury, large sized grafts, HLA incompatibility, previous failed corneal graft/s, decreasing age of recipient, glaucoma, and peripheral anterior synechiae may be considered as other risk factors. The risk of graft failure has been found to vary substantially even within a high-risk population. In the Collaborative Corneal Transplantation Study (CCTS) “high risk” was defined as a cornea with two or more quadrants of vascularisation, or one in which a graft had previously been rejected. Two quadrant deep vascularisation is less at risk than in which, all four quadrants are heavily vascularised. It has been observed that the incidence of rejection increases with both the number of quadrants vascularised and with the total number of vessels crossing the proposed graft/donor junction. Gift rejection in a previously grafted eye relates more to the number of blood vessels in the cornea than to the number of previous grafts. Recipient corneas can be divided into low, medium, and high risk depending on the number of quadrants of vascularisation (avascular, 1–2 quadrants, and 3+ quadrants respectively). Reported failure rates vary between 60% and 90% in high-risk corneal grafts. The clonal expansion of graft-specific lymphocytes occurs in lymphoid tissues. With topical steroids not reaching these secondary lymphoid organs, and the effect of systemic steroids not seeming sufficient to interfere with the clonal expansion of activated T cells, it is essential to administer systemic immunosuppressives to achieve clear graft survival in high risk keratopasty cases. Considering that corneal transplantation is not a life-saving procedure, the profile of side-effects is of paramount importance when choosing an immunosuppressive medication. Various immunosuppressive agents (table 1) have been tried with relative safety and efficacy for prolonging graft survival in patients with high-risk keratoplasty especially in those patients who are bilaterally blind. When immunosuppressive therapy is to be considered in high risk corneal grafting cases, appropriate caution should be exercised in its use, with particular attention to side effects, proper patient selection and information before instituting treatment. Obtaining an informed consent is mandatory in all cases. Treatment plan protocols call for exclusion of patients with a history of malignant disorders, serological evidence of HIV or HbsAg positivity, systemic infections that require therapy at the time of entry, active peptic ulcer disease, inadequate contraceptive measures, pregnancy, or patients aged 18 years or below. Corneal surgeons dealing with high risk cases need devise appropriate treatment regimens to enhance graft survival. Intensive topical corticosteroid therapy is beneficial. The addition of systemic corticosteroid has not been shown to add much benefit. With the availability of systemic CsA, and other lesser toxic alternatives, immunosuppression has now become more acceptable. The adoption of appropriate treatment regimens a few days before surgery to optimize the local corneal environment before grafting is advisable. The use of tissue typing in high risk vascularised corneas is still controversial but can be considered. No consensus exists on one single therapy for all these grafts and devising therapeutic regimens, sometimes with multiple medications can help to improve graft survival especially in the bilaterally blind patients. Studies report improvement with both short and long term systemic CsA in high
Some degree of immunological privilege has been considered to be re-established, indicating that CsA can eventually be safely withdrawn. Few authors have found no statistical benefit from the use of systemic CsA, possibly due to variations in criteria for high risk and inclusion of other factors such as chronically inflamed eyes. Intensive topical corticosteroid therapy decreases inflammation, thereby lowering the local population of immunologically active cells and also reducing the expression of class 2 antigens in the recipient cornea. Long-term results of the use of systemic mycophenolate mofetil (MMF) and tacrolimus for the prevention of rejection has been seen to be effective in patients with high-risk corneal grafts.

**Cyclosporine:**
Cyclosporin A is a powerful immunosuppressive agent derived from the fungus tolypocladium inflatum gans. It acts at the early stages of antigenic sensitisation. It is an immunomodulator and works on T cells by binding to an intracellular peptide cyclophilin. It inhibits antigen presentation and thus lymphokine production. By virtue of its anticyclophilin activity, cyclosporin reduces production of interleukin-2 (IL-2), thus limiting the activation of CD4+ and CD8+ T cells. CsA, is a very potent prophylactic agent for preventing corneal allograft rejection and is used in specialised centres after high risk keratoplasty. Although therapy with CsA allows superior graft survival, its use is limited because of a wide range of side effects such as diabetogenicity, arterial hypertension, hyperlipidaemia, nephrotoxicity. Apart from this for CsA to be effective, the daily dose needs to be adjusted to maintain optimal trough levels of CsA which makes its usage expensive and labour intensive with regular laboratory drug monitoring.

**Treatment Schedule:**
After baseline evaluation including clinical history, blood pressure, total cell count, serum urea, creatinine, electrolytes, and liver function tests, oral CsA is given as daily dosage starting immediately after surgery in a loading dose of 10 -15 mg/kg/day. Thereafter, CsA blood levels are to be monitored and measured biweekly. Blood pressure, complete blood cell count, serum creatinine and liver functions are to be evaluated every 2–4 weeks. Patients are to be followed on a regular basis every day for the first postoperative week, weekly in the first month, and then monthly. CsA is given for 12 months unless significant side effects occur. The dose of CsA is to be adjusted to the whole blood trough levels with a target range of 120–150 ng/ml. With stabilisation of the CsA levels the frequency of the tests may be reduced. The advantages of systemic CsA include specific immunomodulation and no ocular surface complications.

Topical CsA is prepared either in olive/ castor oil as 2% solution or in artificial tear as 1% solution. It is prescribed 5 times a day along with the topical corticosteroid drop in high risk patients both pre and post operatively. As blood cyclosporin A level after topical therapy is negligible, monitoring of blood cyclosporine A levels is not required. However, monitoring of renal and liver function is required.

**Side effects:** Elevated serum urea and creatinine, hypertension, gum hyperplasia, increased sweating, backache, nausea, feeling unwell, oral candidiasis, cramps, bone marrow toxicity, hirsutism and parasthesia of the extremities are the commonly occurring side effects. These side effects are rare when the drug blood level is well monitored and kept at low therapeutic levels of approximately 200 ng/ml. The use of CsA in solid organ transplantation has been associated with an increased risk of lymphoproliferative disorders that has been attributed to the long duration of therapy and the use in conjunction with other immunosuppressive drugs. This calls for a stringent reconsideration on our indications for its use in ocular conditions.

**Mycophenolate mofetil:**
MMF has widely approved safety and efficacy in combination with CsA following renal transplantation. Unlike CsA, MMA does not interfere with IL-2 pathways. It reversibly inhibits the de novo formation of guanosine nucleosides by inhibiting the enzyme inosine monophosphate dehydrogenase. As T and B cells are predominantly dependent on the de novo synthesis of guanosine nucleosides, the purine biosynthesis of these cells is selectively inhibited. MMF is to be given in a dosage of 2 g (1 g twice daily). Routine blood samples need to be taken every 2–4 weeks to check for drug toxicity. Patient follow ups are done at weekly intervals in the first postoperative month, bi-weekly for 2 months and monthly for 3 months and every three months thereafter during which laboratory assessments are performed. The need for blood level adapted dosing for MMF remains controversial. Lesser postoperative visits, reduced cost and logistics of postoperative immunosuppression as a result of the omission of monitoring drug titres are advantages with MMF usage. MMF in
combination with a short postoperative course of oral steroids is just as effective as CsA and oral steroids in preventing acute rejection following high risk corneal transplantation.\textsuperscript{14}

**Tacrolimus:**
Tacrolimus (FK 506) is a macrolide antibiotic isolated from the soil fungus Streptomyces tsukubaensis, with potent immunosuppressive activity, 10 to 100 times more potent than CsA. The mechanism of action of tacrolimus is similar to cyclosporin. While CsA binds to the immunophillin, cyclophilin, tacrolimus binds to a specific cytosol protein: FK – 506 binding protein (FKBP) and inhibits the T cell receptor-mediated signal transduction required for transcription of interleukin 2 and other lymphokines.

**Treatment schedule:**
Baseline evaluation includes clinical history, blood pressure, total blood count, blood urea, creatinine and electrolytes, and liver function tests. Oral tacrolimus is to be initiated at a dose of 2 mg/day (1 mg twice daily) on the day of surgery, and is to be further adjusted (2 – 8 mg/day) aiming for whole blood tacrolimus trough level between 1 and 12 mg/l along with monitoring clinical signs. The recommended levels are the lower end of the range between 1 and 12 mg/l for grafts that do not show any signs of vascularisation or rejection, at the middle of the range for grafts that have vascularisation or rejection features. Trough levels are to be measured 12 h after the last dose of tacrolimus and are not to be allowed to rise above 12 mg/l in order to prevent toxicity.

**Side effects:**
The common side effect of tacrolimus include hypertension or exacerbation of pre-existing hypertension, headaches, malaise and gastrointestinal disturbances, paraesthesia, pancreatitis, folliculitis, reversible increase in serum creatinine, insomnia, diabetes, increased frequency of epileptic episodes and lymphopaenia.

**Recommendations:**
We recommend systemic immunosuppressant therapy for 12 - 18 months postoperatively or until suture removal, whichever is later. Patients with clear grafts in their only seeing eye are usually given the option of continuing therapy indefinitely. For those patients with rejection episodes while on immunosuppressive, the drug is to be continued for a period of one year after the last rejection episode. Those with graft rejection episodes after discontinuing systemic immunosuppression, may be counseled for further treatment for one year. Patients who are unable to tolerate the required dose may be given the option of changing over to an immunosuppressive agent with a different side effect profile or using combination treatment. Patients who do not opt for commencing systemic immunosuppressive therapy again or change in the nature of immunosuppressive agent may be managed with steroids or topical cyclosporine. However, the use of topical cyclosporin is limited by the local discomfort and epithelial toxicity associated with its use.\textsuperscript{15}

---

**Figure 1:** Failing graft secondary to immune rejection

**Figure 2:** Vascularised recipient bed

Repeat grafts usually have extensive vascularised corneal bed preoperatively and a history of previous failure, sometimes immune mediated. (Figure 1 and 2) Repeat corneal transplantation is a subset of high risk condition for immune graft rejection. Rumelt et al 16 followed up 28 re-grafts treated with oral cyclosporine for an average of 26.6 months and reported rejection-related failure in 32% and non-rejection related failure in 36%. They also found that six (21.4%) of 28 grafts on CsA rejected while on treatment and another three (10.7%) failed after treatment was stopped compared
High risk corneal grafting

DJO Vol. 20, No.3, January-March, 2010

have reported use of topical CsA. A randomised controlled study did not demonstrate improved graft survival with topical CsA. Topical cyclosporin A is effective in prevention of immune rejection of corneal graft in humans only when combined with topical corticosteroids. Topical cyclosporin A treatment has been reported to be beneficial in corneal transplanted eyes with no or minimal corneal vascularisation. The treatment of corneal graft rejection with 0.5% topical cyclosporine is effective in eyes with satisfactory preoperative corneal transplantation beds, whereas it has been reported to have only beneficial effects in eyes with poor preoperative corneal transplantation beds. The recurrence of rejection was found to resolve by resumption of the cyclosporine eyedrops. Topical CsA in shields or in fragments have been shown to provide a significant advantage over systemic CsA alone, and CsA fragments also seem to be as effective as shields in preventing corneal allograft rejection. Cyclosporine-treated grafts have been found to contain significantly fewer infiltrating T lymphocytes indicating that the topical application of cyclosporine actively inhibits the entry of T cells into the grafts.

Preliminary results of a randomized multicentre trial estimated the ratio of high risk corneal grafts without immune reactions to be about 89% 1 year postoperatively in patients on MMF, in contrast to only 67% in the control group. Adverse effects of the drug such as gastroenterotoxicity, tachycardia, arthralgia or systemic infections were also found to be reversible. Hence systemic MMF may be an effective and safe immune modulating drug in the prophylaxis of immune reactions after penetrating high-risk keratoplasty. Birnbaum et al report statistically significant, stronger effect of MMF compared with CsA in preventing immune reactions after high-risk keratoplasty with both being comparable in potency regarding clear graft survival. Tacrolimus, systemic and topical, is also evolving as a relatively safe and effective option in reducing rejection and prolonging graft survival in patients with high-risk keratoplasty. Other Immunosuppressives: Rapamycin is more potent than CsA and tacrolimus. It binds to FK-506 binding protein (FKBP) and inhibits the immunophilin activity. It also interferes with the IL-2 induced signals. It suppresses T cell activation at the level of lymphokine production. It is lipophilic in nature and has better corneal penetration. A singly induced signals. It suppresses T cell activation at the level of lymphokine production. It is lipophilic in nature and has better corneal penetration. A singly
Rapamycin and MMF were seen to show similar efficacy in preventing immune reactions after high-risk keratoplasty, until the duration of administration. However rapamycin has been observed to have a broad spectrum of side effects.

In conclusion, immunosuppressive therapy has a definitive role in management of high risk corneal grafts but is to be used with extreme caution and good judgment.

**Summary:** Corneal grafting in high risk corneas remains a challenge. High risk corneal grafts are those conditions in which the cornea loses its immune-privileged status, resulting in its susceptibility to immunological rejection induced failure. High risk corneal graft is defined as a cornea with two or more quadrants of vascularisation, or one in which a graft had previously been rejected. Various immunosuppressive agents have been tried with relative safety and efficacy for prolonging graft survival in patients with high-risk keratoplasty especially in those patients who are bilaterally blind. When immunosuppressive therapy is to be considered in high risk corneal grafting cases, appropriate caution should be exercised in its use, with particular attention to side effects, proper patient selection and information before instituting treatment.

<table>
<thead>
<tr>
<th>Drug</th>
<th>Dose</th>
<th>Side effects</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cyclosporin</td>
<td>10 – 15 mg / kg / day to maintain trough levels of 120-150ng/ml²⁶</td>
<td>Nephrotoxicity, hepatotoxicity anorexia lethargy, viral infection, hypertension, hirsutism, gum hyperplasia, tremor and seizures</td>
</tr>
<tr>
<td>Mycophenolate mofetil</td>
<td>1gm twice daily for 12 months³¹</td>
<td>Vomiting diarrhoea electrolyte disturbances, hepatotoxicity and exacerbation of atopic dermatitis</td>
</tr>
<tr>
<td>Tacrolimus (FK 506)</td>
<td>2gm/day on day of surgery and then adjusted to lower end of range of whole blood tacrolimus level between 1-12mg/l⁴</td>
<td>Hypertension, headache, malaise, GIT disturbances, paraesthesia, pancreatitis, folliculitis, reversible increase in serum creatinine, insomnia, diabetes, increased frequency of epileptic episode and lymphopenia</td>
</tr>
<tr>
<td>Rapamycin (Sirolimus)</td>
<td>Once daily to attain blood trough levels of 4-10ng/m³⁷</td>
<td>Headache, swelling of extremities, gastritis, vomiting, diarrhoea, weight gain, insomnia, fever, rash, tremor</td>
</tr>
<tr>
<td>Azathioprine</td>
<td>1-2mg/kg/day</td>
<td>High toxicity like bone marrow suppression, hepatic necrosis, neoplasia, alopecia</td>
</tr>
</tbody>
</table>

**References:**


Introduction:
As we all know that a post operative Endoophthalmitis is a dreaded complication of cataract surgery. Three most important reasons for developing post operative infection in the order of importance are;
• Inadvertant Touch
• Sterilization failure
• Environment

Failure of sterilization may be due to Faulty technique, inadequate cleaning of the instruments, and tight packing of bin or Contamination after sterilization. Failure to adequately clean instrument results in higher bioburden, protein load, restricted flow in case of cannulated instruments and salt concentration. These will decrease sterilization efficacy preventing contact of sterilant with micro-organisms. So, the instrument cleaning is very important to prevent post operative infection and to increase life of the instruments.

Environment cleaning is the third important reason. So, an attempt should be made to keep it as clean as possible and all activities should be monitored using various methods and check lists.

(A) Preparation of Instruments:
Care of instruments:
(i) Cleaning: Special attention should be given to clean the surgical instruments before sterilization. Surgical instruments vary in configuration from plain surfaces, which respond to most types of cleaning to complicated devices that contain box locks, blind holes and interstices.
• Instruments should be cleaned as soon as possible after their use; this will help avoid drying of debris or blood on the surface of the instruments.
• Sharp and blunt instruments should be separated.
• An ultrasonic cleaner can be used for cleaning the instruments. It is ideal for cleaning instruments. It thoroughly cleans every part of the instrument, including the depths of the cannula, tubes and other unreachable parts, with high frequency sound waves generating bubbles and vacuum zones.
• However, ultrasonic cleaners are not essential. One can use four bowl techniques for the cleaning of the instruments described below.

Technique of using Ultrasonic Cleaner:
• Instrument should be thoroughly cleaned by washing in sterile distilled water or mineral water.
• The tank of the cleaner should be filled above the top of the instruments; suitable detergent as specified by the manufacturer is added.
• The temperature of water should be 80 to 110 degrees Fahrenheit for effectiveness of detergent.
• Enzyme solution or detergent can be added for effective cleaning in the ultrasonic cleaner.
• They should be kept in ultrasonic cleaner for at least 30 minutes.
• After removing the instruments from ultrasonic cleaner, the instruments are first brushed with a soft tooth brush.
• Then washed in four basins containing mineral water or boiled water one after the other, the first one contains mineral water with disinfectant. This should be done even if ultrasonic cleaner is used.
• They are then dried with clean towel; tipped with plastic sleeve and are segregated into separate sets. They are then packed in individual perforated stainless steel trays, which are placed in the bins with indicator and put in the autoclave. (Three indicator tapes should be placed, one in the bottom, one in the middle and one at the top of the bin), of course one strip on the external surface of the bin is required to tell us whether the bin is sterile or not, even without opening the lid.
• Cannulated instruments are first to be flushed with distilled water three times and then with air three times before autoclaving.

N.B.: Chrome plated instruments should not be cleaned in an ultrasonic cleaner.
We can use hot air ovens at 50 degree centigrade for 20 minutes for total drying. However, a hair drier or drying under fan is also good enough. We must make sure that the instruments are totally dry otherwise, droplets of water can contain micro organisms and it is difficult to sterilize the droplets inside the lumen of the cannulated instruments.

(iii) **Instrument sets:** Instruments should be placed in a tray with perforated bottom to allow steam penetration around the instruments during autoclaving and to prevent air trapping in the tray. Each delicate instrument should be physically separated from adjacent one to prevent damage, interlocking and crushing. The size of the instrument pack should not exceed 20” by 20” and weigh not over 5 kgs.

(iv) **Packaging, loading and labeling of instruments for sterilization:**
- Instruments should be arranged in trays to prevent damage. Heavy instruments should be kept in the bottom tray. All detachable parts must be disassembled, syringes separated, caps, plugs removed, etc.
- Lubricated instruments should be thoroughly cleaned as steam or gas can not penetrate. This would lead to improper sterilization.
- The lumens of cannula must be flushed thoroughly with water before being sterilized. Debris inside the lumen prevents steam penetration and will cause permanent blockage.
- Rubber sheets should not be folded or kinked, as steam cannot penetrate or displace the air from the fold or kink. The rubber sheet should be wrapped in linen. Rubber items should not be kept with metal instruments to prevent damage to the rubber items. Rubber items should be powdered before autoclaving, otherwise the heat will make the rubber stiff and disintegrate.
- Perforated metal drums are used for sterilizing large items such as theatre drapes. Smaller items can be wrapped in paper bags, linen or similar material and heat sealed after applying the indicator tape. All the items not in drums should be double wrapped in a cloth or paper bag.
- The articles should not be packed too tightly. Space for the steam penetration and completion of drying cycle should be present.
- All the items are loaded in such a way, that every surface is exposed to the steam. All the instruments should face the same side to avoid air pockets.
- Autoclave tape should be put onto all packs indicating the date of sterilization, who packed them and their contents.

(V) **Cleaning of Phaco-emulsification Instruments:**
- Proper cleaning becomes more relevant in Phaco
emulsification as here we are more dependent on the machine for the surgery.
• The IV set is removed from the bottle. The irrigation tubings are disconnected. The irrigation aspiration tube is disconnected from the hand piece. The hand pieces are unplugged from the console. The tubings are flushed with saline solution before switching off the machine and the saline collected in a bin.
• Cleaning of the components:
• All cleaning procedures must be done immediately after each surgical procedure; otherwise, tissue debris and salts from the saline irrigating solution may collect and cause permanent damage.

Ultrasonic hand piece:
• The hand piece is wiped with soft non abrasive linen and distilled or sterile water to remove residual tissue. Both the irrigation and aspiration ports are flushed thrice with 10 cc syringe filled with warm distilled or sterile water. It is repeated thrice with air.

Irrigation and aspiration hand piece:
• Clean the hand piece, tip and sleeve with gauze piece dipped in isopropyl alcohol or antiseptic. Thoroughly flush all the hand pieces, components and tips with distilled or sterile water.
• Tips and sleeve are usually disposable ones. But they can be re-used if properly sterilized. The tip should be connected to a syringe and flushed with water. Similarly, the sleeve is also flushed with water.
• All are then packed into trays for steam sterilization. Care should be taken to wrap the tubing and hand pieces separately in a cloth i.e. the metal components should not come in contact with the wire.

(VI) Drying and storing:
• When the autoclave cycle is completed and the contents are ready to be removed, place them on wire shelves to allow free flow of air around them so that they cool without developing condensation. If shelves are not available, place the items in a cool place, but do not pile them on top of each other. If drums are used, they should be sealed immediately.
• Once items have cooled they can be wrapped in a polythene bag to prevent dust and external damage. The articles should be used within 48 hours of being sterilized. The sterile packs must be stored above waist level, kept dry, protected from dust, handled only when necessary and used within sterilization date.

(B) Preparation of OT Environment:
(i) Cleaning of OT:
Theatre Cleaning is scheduled as Daily, Weekly & Monthly Cleaning
Daily Cleaning: After Surgery:
• Enough time must be given to the operation theatre staff for the end of the day cleaning activities. This will ensure that the operation theatre is ready for the next day in a desirable manner.
• All the extra items must be removed from the operation theatre.
• The walls are cleaned with 1% sodium hypochlorite solution up to six ft. height daily.
• Equipments such as electrical surgical units should be checked & cleaned first.
• Cabinets & doors are cleaned, especially around handles or push plates where contamination is more likely.
• Walls around the scrub sink need special attention.
• Transportation carts and their wheels are cleaned with special attention after use; equipment is disassembled, cleaned and covered properly.
• Microscope lenses should be cleaned with lens cleaning solutions.
• Microscope head should be cleaned with bacillol spray and covered properly.
• Operating table covers are changed and patient head support also be cleaned.
• Trolleys, Revolving stools, Operating table, Foot stool, I. V. Stand are cleaned with 1% sodium hypochlorite solution.
• The floor is always mopped last. A clean mop is used to clean the floor. One bucket is filled with warm water and another bucket with antiseptic lotion ( for e.g. : Dettol 1:40 dilution in water)
• The mops first dipped in the dettol solution, wrung and mop the floor, then dip in the warm water to wash it. (two bucket technique)
• Again dip in the Dettol solution, wring out and continue the mop in the same manner.
• Slippers should be washed with detergent and
dried completely.
• The washing of slippers should be done away from the instruments washing area or the scrub area.
• At the end of each session, all the waste must be disposed off as per the waste disposal guidelines in colour coded bags. Over the last decade, the disposal of operating room and hospital waste has received much attention. Incineration has been advocated as a viable method of hospital waste disposal. Recently attention has been directed at preventing air pollution from incineration and to find alternative medical waste treatment technologies. These options include gasification, steam sterilization or heat disinfection of certain wastes prior to disposal in landfill.

Weekly Cleaning:
• All the articles removed from shelves and remove the equipments, furniture from the theatre.
• Fans and Air conditioners (A.C. Filter) are cleaned, remove the filter from the A.C. Clean it properly with detergent solution and dry under sunlight before replacing.
• Clean the ceiling and wall mounted fixtures.
• Regular cleaning of all types of sterilizers as recommended by the manufacturers.
• Washing of walls and floor of the OT with detergent and mopping with antiseptic solution (The dilution of the antiseptic according to the manufactures’ advice).
• Bowls and buckets using inside the Theatre cleaned properly.
• When cleaning the theatre operation room first then the scrub area, sterilizing area and preoperative patient waiting area.

Monthly Cleaning:
• Remove all the equipments from the theatre.
• Scrubbing the floor of the OT to make sure that all accumulated deposits are cleaned thoroughly.
• Washing all the furniture and walls of the O.T. Cupboards are removed and areas behind the cupboards are cleaned.
• Trolleys, Stools, Microscopes are cleaned and oiled where necessary, and then routine cleaning procedures are done as described above.

(ii) Disinfection of OT:
Fumigation:
• Aerosol machine/Fogger is used for fumigation (electrically operated) as it can make micro particles. Equal amount of Formalin and distilled water is added depending upon the size of the Theater. After Fumigation, close the theatre for minimum 24 hours. Hydrogen peroxide 1% solution or Glutaraldehyde+ Formaldehyde combination can be used as a change fortnightly to prevent resistance developing against one agent.
• Fumigation needs to be done once in a week.
• Sterile quality of air in the OT is better achieved and maintained by employing air cleaner and ultraviolet lights and through improving the overall cleanliness in and around the OT.
• Air cleaners are turned on while the surgical session is going on. While the ultraviolet lights are turned on over night after the surgical session
when the last person leaves the OT. “Off when the first person enter before the OT next morning”. This will continuously clean the inside air while we are working.

- Humidity of the operation theatre can be maintained by employing De-humidifier.

**Indications:**

- If septic case has been operated, fumigation is mandatory after the procedure.
- If any new construction or reconstruction of any theatre is done, fumigation is mandatory before the functioning of the same.
- When routine surveillance reveals any pathogenic spore former, fumigation is mandatory.

**Details of fumigation of theatre:**

- Formalin 30ml of 40% Formalin dissolved in 90 ml of clean water for 1000 cft by aerosol spray.
- Seal the room air tight. Use adhesive to close gaps
- Pour total amount into the Fogger/Aerosol and switch on. Timings can be set with timer.
- Fogger is the best as the droplet size generated is smaller which can be spread all around effectively.
- Leave it for 24 hours.
- Aldekol-Formaldehyde – 6%, Glutaraldehyde 6% and Benzalkonium chloride 5%
- For 4000 cft 325 aldekol in 350 ml of water sprayed for 30 minutes – close for 2 hrs – Switch on AC – OT ready in 3 hrs
- Open the room wearing a mask. Put small amount of liquid ammonia solution inside the room for half an hour and also turn the exhaust fan of air conditioner on.

<table>
<thead>
<tr>
<th>Procedures and Agents</th>
<th>Routine</th>
<th>Efficacy</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mopping of OT floor, walls, tables, trolleys with 1% Sodium Hypochlorite</td>
<td>Everyday</td>
<td>Reasonably effective against wide range of gram +ve and gram -ve bacteria but little activity against endospores, viruses and hepatitis virus.</td>
</tr>
<tr>
<td>Washing of OT walls floor, tables, trolleys, AC Filter etc with detergent</td>
<td>Once a week</td>
<td>Enhances the effect of daily cleaning and disinfection.</td>
</tr>
<tr>
<td>Fumigation of the operating room with formaldehyde (Occasionally other agents should be used to avoid developing resistance)</td>
<td>Weekly or after surgery on the infected cases</td>
<td>Efficacy uncertain in temperature below 200°C and relative humidity below 70%</td>
</tr>
<tr>
<td>Washing of OT after removing all its contents</td>
<td>Monthly</td>
<td>Enhances and improves the effect of cleaning and disinfection.</td>
</tr>
<tr>
<td>Maintenance, repair of any breaches, cleaning of the ventilation system.</td>
<td>Once in a six month period</td>
<td>Enhances and improves the effect of cleaning and disinfection.</td>
</tr>
</tbody>
</table>
(C) Monitoring of Cleaning Activities:

1. Monitoring of Environment:
   - The environment cleanliness is monitored biologically.
   - Open dish sedimentation method is generally used. A nutrient agar plate is left open in one corner of the operation theatre at the beginning of the activities in the morning. After half an hour, it is taken to the laboratory and incubated. Growth of less than 10 colonies is considered good enough to continue the activities. Otherwise, the cleaning and fumigation need to be repeated and after cross checking biologically only the theatre is used for performing surgeries.
   - Bacterial counters are available but they are very costly and not used commonly.

2. Monitoring sterility:
   - Weekly samples should be taken from sterile items and scrubbed personnel at the end of the surgery and should be sent for biological testing.
   - Weekly samples should be taken from the air conditioner filter, operating table, trolleys, microscopes, and scrub area, area where the drums are stored or kept while using them.
   - Nasal and nail bed swabs also should be taken at regular intervals and sent for testing.

3. Monitoring of hygiene:
   - All the staff members working in the theatre must undergo a regular health check up. Nails should be regularly checked and trimmed.
   - Any skin infection should be treated immediately and the person with skin lesion should not be allowed to work inside the operating room.
   - Nasal carrier state must be checked and all the staff members in carrier state must be treated.

4. Monitoring of activities:
   - All the activities must be monitored carefully using various check lists.

Check lists:
Daily Cleaning Check List Eye O.T.:
Date:
1. Who checked Pre operative check list?
2. Who put 5% Povidone iodine eye drop before giving block?
3. Who checked Autoclave strip register?
4. Who filled drum of gowns - gloves? Who checked it?
5. Who checked clarity of Inj. RL?
6. Who did preparation before arrival of surgeon?
7. Who did Fumigation? With what? (Formalin, 1% Hydrogen Peroxide, Formaldehyde & Glutaraldehyde combination)
8. Who did cleaning before leaving in evening? (Doors should be cleaned every day)
9. Who checked Operation & emergency medicines stock?
10. Who put on the U.V. light at night? Who put it off in the morning?
11. Was the chlorination of water tank done yesterday? Who did it?
12. Who checked anaesthesia trolley?
13. Who replaced Bed sheet of O.T. Table in the evening?
14. Who cleaned Equipments / Instruments (Cautery, Suction machine & O.T. Table) with 1% Sodium hypochlorite??
15. Notes:
16. Signature of O.T. In-charge: Signature of HOD:

Weekly Cleaning Check List Eye O.T.:
Date:
1. List of medicines checked? Who did it? (Daily use + Emergency medicines)
2. Who checked Eye O.T. Check list? (List except medicines)
3. Did in charge prepare the list of O.T. staff posting?
5. In-charge checked the list of Sunday work done?
6. Cleaning done on Saturday by shifting things? (Microscope, O.T. Table)
7. Who did Sodium hypochlorite cleaning of sink?
8. Who cleaned Walls and floor of O.T. with Sodium Hypochlorite?
9. Who fumigated Autoclave room on Saturday after cleaning?
10. Who cleaned A/C Filters?
11. Who cleaned Instruments? (Check blade and change it if necessary) (Check two way cannula)
12. Who checked Staff nail?
13. Who checked Chlorination?
14. Who changed Water in autoclave machine? (Change every fortnight)
15. Who cleaned & autoclaved Bottle of surgical scrub and bottle of liquid soap?
16. Who checked Expiry dates of medicines?
17. Who cleaned the Operating Microscope lenses?

Monthly Cleaning Check List Eye O.T.:
Date:
1. Over book of change of O.T. Boy posting
checked?
2. Swab sample culture done on second Saturday?
3. A/C cleaned by Air Blower on last Saturday?
4. Did in-charge check the washing of O.T. on last Saturday?
5. IOL Report prepared
6. CME Lecture delivered and exam conducted for O.T. staff?
7. Who cleaned the water tank? On which day?
8. Who cleaned Drums? Holes checked?
9. Webs outside windows removed?
10. Note:
11. Signature of O.T. Incharge: Signature of verifying person:

References
3. General Precautions to safeguard against Post Operative Infections following Ophthalmic Surgery – NPCB, India
4. Hinduja Hospital, Mumbai – Infection Control Manual
5. IAPB Guidelines for Eye Care
7. MJ Lights – Operative Operations Theater
10. Textbook of Hospital Infection Control – Shaheen Mehtar
11. Ophthalmic operating theatre practice – A manual for developing Countries, Ingrid Cox and sue Stevens
12. The sterile supply department: Guidelines for planning and quality Management edited by Geetha Mehta.
Case Report

Lenticonus Posterior

Ojaswita Singh, Rashi Shyam, A.K.Gupta
Shroff Eye Centre, New Delhi.

Lenticonus is bulging of the lens capsule and the underlying cortex. It is a relatively uncommon clinical condition. The prevalence is estimated at 1 to 4 of every 100,000 children (1). We are reporting a case of posterior lenticonus because of it rarity and its unusually large size.

Case report

A 16 years old male attended our out patient department with gradual, painless, diminution of vision in the left eye for three years. His best corrected visual acuity was OD 6/6 with -0.50DS, OS 6/18 with -0.25DS. Examination of the anterior segment by slit lamp examination revealed posterior lenticonus in the left eye (Figure 1). Posterior lenticonus showed a cataractous opacity which produced a central dark shadow on retro-illumination (Figure 2). The extent of posterior lenticonus as measured on pentacam was 1920 microns (Figure 3). Intraocular pressure (applanation) was 19mm of Hg OU. Fundus examination was within normal limits in both eyes. The eyes were orthophoric. Ocular movements were normal in both the eyes.

Discussion

Cases of posterior lenticonus can clinically present with reduced visual acuity, amblyopia (1, 4) and strabismus (1). Rarely lenticonus can present with leukocoria (5).

The pathogenesis of lenticonus is not well understood. Collins (2) found a tear in the posterior capsule in a case of posterior lenticonus. Histological investigations by subsequent workers, however, did not confirm this observation (3). Aberrant growth of sub capsular epithelium was found to overlie the lenticonus by Franceschetti and Rickly (6), Makley (3). The case in our study shows a protrusion of 1920 microns which is more than one half the thickness of the lens (3400 microns). Such a large extent of posterior protrusion is more likely due to abnormal development of the lens fibres and not due to protrusions of the cortex through a deficient posterior capsule.

In early cases refractive error should be corrected. Amblyopia should be treated by occlusion therapy, if present. Surgery on lens is indicated if visual acuity is less than 20/100 and can not be corrected with glasses.

References

Cholesterol Granuloma of the Orbit

Mandeep S. Bajaj1, Seema Kashyap2, Neelam Pushker1, Vidushi Sharma1, R. Balasubramanya1, Rachna Meel1, Anoop Kishore Gupta1

1 Oculoplastic and Paediatric Ophthalmology Services, 2 Ocular Pathology Services
Dr. Rajendra Prasad Centre for Ophthalmic Sciences, AIIMS, New Delhi

Abstract
Cholesterol granuloma is a rare, well-defined lesion of the orbit, commencing mostly in the diploe of bone. We report a case of cholesterol granuloma in the orbital part of the frontal bone in a 47-year-old male patient. The patient presented with progressive and painless proptosis and downward displacement of the globe. CT scan showed a superotemporal, homogenous orbital mass involving the frontal bone with expansion and fossa formation with irregular margins. The outer table of the bone was partially absent, with extraperiosteal extension of the lesion. The mass was removed by an antero-lateral orbitotomy. Histopathology confirmed the diagnosis. In conclusion, cholesterol granuloma, though rare, is a readily recognizable lesion with characteristic clinical, radiological and histopathological features.

Introduction
Cholesterol granuloma (CG) is a rare, well-defined lesion of the orbit, commencing mostly in the diploe of the frontal bone.1 It has also been reported in other regions of the head including the zygomatic bone, maxillary and ethmoidal sinuses, and the petrous bone.1-3 In the last decade only three cases have been reported.4-6 We report a case of a cholesterol granuloma in the orbital part of the frontal bone in a 47-year-old male patient.

Case Report
A 47-year-old man presented with right sided painless, progressive proptosis for one year. No history of trauma was elicited. On examination 4mm of proptosis with inferior displacement of the right globe was present (Figure 1). The ocular movements were limited on elevation. A large mass of firm consistency was felt just beneath the irregular bony orbital margins in the superotemporal quadrant. Visual acuity was 20/20 in both the eyes. CT scan showed a superotemporal homogenous, orbital mass involving the frontal bone with its expansion and fossa formation with irregular margins. The outer table of the bone was partially absent with extraperiosteal extension of the lesion (Figure 2).

Figure 1. Preoperative clinical photograph of the patient (Anterior view).

Figure 2. Computed tomographic scan (coronal view showing a superotemporal orbital mass involving the frontal bone. The outer table of the bone was partially absent, with extraperiosteal extension of the lesion.

The mass was approached by an antero-lateral orbitotomy under general anesthesia. The periosteum was reflected from the anterior surface of the frontal bone to the orbital margin, which was irregular in places. The outer table of the bone was thin, eroded and brittle near the orbital rim. On elevating the orbital periosteum from the orbital roof, a yellowish brown mass that was adherent to the bone was visualized subperiosteally. The mass was completely excised along with curettage of granulomatous material from...
the frontal bone. The inner table of the bone was found to be intact.

Gross examination showed two irregularly shaped fragments of yellowish granular tissue each measuring 10x8x6mm and 9x5x4mm respectively. Microscopic examination showed numerous slit like spaces representing cholesterol clefts surrounded by multinucleated giant cells (Figure 3). Deposits of hemosiderin pigment and lipid laden macrophages were also identified. At the periphery of the lesion a few calcified bony trabeculae were also seen. Serial sections failed to reveal any epithelial structures within the tissue. No definite wall was seen. The postoperative period was uneventful. No recurrence was observed at a follow up of 6 months.

**Figure 3. Photomicrograph showing numerous cholesterol clefts along with lipid laden histiocytes (H&E X40).**

**Discussion**

Cholesterol granulomas are usually seen in adult males. The age reported in the largest series of 27 patients ranged from 25 to 68 years (median, 43 years). The superotemporal quadrant of the orbit is involved most often as the predominant site of occurrence of the tumour is the frontal bone.

Computed tomographic (CT) scans usually show a homogenous lesion, isodense with the brain, with expansion of the surrounding bone. The edges of the eroded bone may occasionally be irregular. In a reported series, all the cases had partial or complete erosion of the outer table of the bone allowing the lesion to expand extraperiosteally into the orbit, as was seen in our case. Rarely erosion of the inner table of the skull may be seen with extension into the extradural space or into the sinuses.

The differential diagnosis of this case included orbital lesions occurring at this location, associated with bone remodelling. It includes dermoid cyst, which has an identical clinical presentation but shows a low density lesion on CT scan, in comparison to Cholesterol granuloma which is isodense with the brain. In contrast to the findings in our case, the bone edges in a dermoid are regular and scalloped with adjacent sclerosis and no destructive changes. An organised hematoma with fibrosis was excluded by history. Aneurysmal bone cyst has clinical similarity and evidence of bone destruction, histopathology shows evidence of old and recent hemorrhages and granulomatous inflammation but cholesterol crystals are notably absent. The closest histopathological differential diagnosis of Cholesterol granuloma is the epidermoid cholesteatoma, which is fairly uncommon in the orbit and is encountered more often in the middle ear and mastoid antrum. The only differentiating point is that epidermoid cholesteatomas have a squamous lining which is absent in Cholesterol granulomas.

The exact origin of this lesion is uncertain. Hemorrhage definitely plays a role in its development, which may occur either due to trauma or an anomaly in the dilpoe of the bone. An extraperiosteal approach for its removal allows the surgeon to aspirate its contents, strip or curette the lining, and remain safely extradural, even if a defect in the inner table of the bone is present. Incomplete removals have been attempted with no recurrence. Only one possible recurrence of Cholesterol granuloma is reported in the literature, which was seen in an epidermoid cholesteatoma.

In conclusion, Cholesterol granuloma, though rare, should be considered in the differential diagnosis of lesions involving the superotemporal orbit in adults. It is a readily recognizable lesion with characteristic clinical, radiological and histopathological features. A timely and appropriate surgical intervention carries a relatively favourable prognosis.

**References**

Cancer associated retinopathy
in a patient with non-Hodgkin’s lymphoma and congenital achondroplasia llowed
by drug induced thrombocytopenia and bilateral central retinal vein occlusion

Shahana Mazumdar, Navneet Mehrotra
ICARE eye hospital, NOIDA U.P

Introduction:
Cancer associated retinopathy is a rare paraneoplastic syndrome in which autoantibodies directed at various retinal components cause progressive vision loss. Specific names have been given to the paraneoplastic syndrome depending on the primary malignancy such as cancer associated retinopathy (CAR), melanoma associated retinopathy (MAR), lymphoma associated retinopathy (LAR). Many patients develop the retinopathy before the primary malignancy has been found. Cases, where no malignancy can be found though antibodies can be detected, are referred to as autoimmune retinopathies (AR).

The clinical features are similar in all types and result from rod or cone involvement. Rod involvement results in night blindness, delayed dark adaptation, photopsia, mid-peripheral scotomas. Cone involvement results in reduced visual acuity, photosensitivity, decreased color vision and central scotoma. CAR has been reported with a variety of carcinomas most commonly small cell lung cancer, endometrial, cervical and breast cancers. Several other cancers have been reported with less frequency.

We report a patient with congenital achondroplasia who developed CAR following Non Hodgkins Lymphoma (CNS). He developed it during a quiescent phase but suffered a recurrence of the lymphoma a month later. On being treated for the recurrence the patient developed drug induced thrombocytopenia resulting in bilateral central retinal vein occlusions.

Case report
A 28 years old male with congenital achondroplasia (Figure 1) presented for evaluation for seeing flashes of light since one month in his only seeing eye, the right eye. He was a patient of Non Hodgkins Lymphoma (CNS) B cell type diagnosed three years earlier which had presented as a left orbital mass. The disease had been diagnosed following an orbital biopsy. Subsequently he underwent orbitotomy to remove the tumor but the vision in the left eye was lost due to compressive optic atrophy. There was no evidence of right eye involvement. Bone marrow biopsy and CSF were negative for malignant cells. Patient received CHOP based chemotherapy (Cyclophosphamide, Doxorubicin, Vincristine and Prednisolone) and radiotherapy. A few months later he developed meningeal involvement and received intrathecal methotrexate and high dose methotrexate. He had suffered several recurrences since then receiving many cycles of chemotherapy. On presentation the disease had been quiescent for a few months and he was not on chemotherapy.

On examination his best corrected visual acuity was 6/6 and no PL in the right and left eyes respectively. The anterior segments in both eyes were normal. The left eye showed an afferent pupillary defect. The posterior pole of the right eye revealed a normal optic disc with a dull foveal reflex (Figure 2) whereas the left eye showed a pale optic disc (Figure 3). Both eyes showed generalized arteriolar attenuation and widespread mottling of the retinal pigment epithelium (Figure 4). There was an area of lattice degeneration in the superotemporal retina of the right eye. He revealed an incomplete ring scotoma (Figure 5). His earlier visual fields done 3 years ago were within normal limits (Figure 6). Flash ERG showed mild delay in the implicit time with reduced amplitude in the scotopic response in the right eye. The left eye showed no response.

He did not report for 4 months as he had suffered a recurrence of the malignancy (meningeal) with severe pain in the spine and had been under treatment. By now he had developed debilitating night blindness though his visual acuity was still 6/9. On examination both eyes revealed a marked increase in the retinal mottling (Figure 7). He was scheduled for further sessions of chemotherapy.

Based on the history of malignancy, rapidly progressing positive visual symptoms and night blindness, advancing retinal pigment epithelial mottling, arteriolar attenuation, concentric narrowing of visual fields and a diminished scotopic ERG-a diagnosis of
Cancer associated retinopathy was made. It was not possible to assess antirecoverin antibodies. Though there is no known treatment of the condition we found that various immunotherapies had been used with some success. In consultation with the oncologist the patient was started on oral steroid (1mg/kg body weight to be tapered over six weeks) and the patient felt symptomatically better though he could not come for follow up.

At this point as he had not shown a good response to the earlier chemotherapy regime his treatment was changed to high dose Cytosine-arabinoside. On receiving this he developed Grade IV neutropenia and thrombocytopenia and he reported a sudden drop in vision. On examination the vision in the right eye had dropped to 6/60 and he was found to have bilateral central retinal vein occlusion with the right eye more severely affected than the left (Figure 8 & 9). The right eye showed extensive peripapillary hemorrhages with hemorrhages at the macula. There was some macular edema. The optical coherence tomography showed retinal thickening nasal to fovea (Figure 10). He was given intravitreal avastin in the right eye. Subsequently the macular edema has resolved and on the last examination, 6 months since the venous occlusion, his best corrected visual acuity was 6/18. The retinopathy has progressed further (Figure 11). However there were cells in the anterior chamber bilaterally. Anterior uveitis can be a part of CAR syndrome but on looking up the literature we found one report of anterior uveitis secondary to high dose Cytosine-arabinoside therapy for lymphoma which had resolved readily with topical steroids. The patient has been started on topical steroid drops.

Figure 1 Photograph of the patient with achondroplasia

Figure 2. Posterior pole of right eye showing a healthy disc and macula with arteriolar attenuation and mild RPE mottling.

Figure 3 Posterior pole of the left eye with optic atrophy, arteriolar attenuation and RPE mottling.

Figure 4 Superior retina of the right eye showing mottling of the RPE and arteriolar attenuation.
Cancer associated retinopathy

Figure 7 Fundus photograph after 4 months with marked increase in the RPE mottling

Figure 8 Acute central retinal vein occlusion in the right eye following thrombocytopenia secondary to chemotherapy

Figure 9 Other eye had multiple retinal hemorrhages with milder central retinal vein occlusion

Figure 10 OCT of the macula with intraretinal thickening

Figure 11 Progression of retinopathy with pigment clumping and degranulation

Figure 5 Visual fields (24-2) showed incomplete concentric visual fields

Figure 6 Normal visual fields of the patient in 2005
Discussion

We have discussed this case with the following in mind: to report the occurrence of CAR syndrome in a patient with achondroplasia and to report the occurrence of CAR in association with Non-Hodgkins lymphoma. We also wanted to report the ocular complications of cytosine-arabinoside therapy in our patient namely bilateral central retinal vein occlusion and possibly anterior uveitis.

Cancer Associated Retinopathy (CAR) is a remote manifestation of cancer and is a paraneoplastic disease. The retinopathy can precede detection of the cancer and a high index of suspicion must be maintained in patients presenting with unexplained visual loss and symptoms. Cancers most commonly associated with CAR are small cell lung cancers, breast cancer1-7, endometrial, cervical and ovarian cancers. Less commonly it has been found with pancreatic cancers, other lung cancers, lymphomas, prostatic, bladder and laryngeal cancers, colon cancers and metastases of unknown origin. Similar paraneoplastic retinopathy has been described in patients with melanoma and is specifically referred to as melanoma associated retinopathy (MAR). Lymphoma associated retinopathy (LAR) is also used to describe the retinopathy when associated with lymphomas8.

Symptoms are usually bilateral and can be due to affection of rods, cones or both. Cone related symptoms include photosensitivity, reduced vision, decreased color sensitivity and central scotoma. Rod related symptoms include prolonged dark adaptation, night blindness, positive visual phenomena such as flashes of light and mid-peripheral field defects. Both ways the retinopathy is progressive and ultimately the patient loses vision.

Signs commonly seen are mottling of the pigment epithelium, arteriolar attenuation and mild optic disc pallor. Rarer signs include periphlebitis, arteriolar sheathing and cells in the vitreous and anterior chamber. Visual fields may show a generalized depression, ring scotoma or central scotoma. ERG shows a diminished rod or cone response9. CSF reveals a non-specific mild lymphocytic pleocytosis. Histopathologically there is diffuse photoreceptor degeneration of both rods and cones with or without inflammation. Inner retinal layers are spared. The photoreceptor degeneration is believed to be immune mediated. Antibodies against retinal proteins have been found and there appears to be cross reactivity between cancer cells and retinal proteins. Several antibodies have been identified but the most extensively studied is antirecoverin antibody10 directed against retinal recoverin – a calcium binding protein located within retinal photoreceptors. Detection of this antibody has been used for diagnosis though it can also be found in some non-paraneoplastic autoimmune retinopathies. Treatment with systemic steroids (oral and high dose intravenous)11 and other immunomodulators have met with mixed results. Treatment of the primary malignancy does not alter the visual prognosis.

References
11. Nippon Ganka Gakkai Zasshi: Case of cancer-associated retinopathy in which immediate treatment succeeded 2008 Sep; 112(9): 806-11
MESECTODERMAL LEIOMYOMA OF CILIARY BODY

Sabia Rashid, M. Q. Keng, M. Farooq, A. R. Khan
Department of Ophthalmology, Government Medical College, Srinagar

SUMMARY
A 20 year old female presented with a mass over the right iris with decreased visual acuity. Clinical examination revealed a ciliary body mass. Iridocyclectomy was performed. Histopathology and immunohistochemistry proved the mass to be a Mesectodermal Leiomyoma of the Ciliary Body. Ciliary body mesectodermal leiomyomas are rare, benign smooth muscle tumors which need to be differentiated from the malignant lesions. These tumors display a mixed myogenic and neurogenic morphology on microscopy. This case report presents a clinical, microscopical and immunohistochemical features of mesectodermal leiomyoma.

INTRODUCTION
Smooth muscle tumors of the ciliary body are extremely rare. A “Hybrid” smooth muscle and neuroectodermal tumor of ciliary body was first reported in 1977. It was proposed that the unusual neural appearance of this smooth muscle tumor reflected its origin from neural crest cells. These neural crest cells also contribute to the formation of bone, cartilage, connective tissue and smooth muscles in the head and neck region. These neural crest derived tissues have been termed “Mesectoderm”, hence the ciliary body tumor is called ‘Mesectodermal Lieomyoma’.

The published reports of mesectodermal leiomyoma reflect the rarity of this entity and highlight the clinical presentation and outcomes. It is a benign tumor and is treated by complete excision. Incomplete excision may result in recurrence.

CASE REPORT
A 20 year old female presented with history of a progressive swelling over the iris of her right eye of 3 months duration, compromising her vision. Visual acuity in the right eye was 6/18 and 6/6 in the left eye. Slit lamp examination revealed no signs of inflammation. A large smooth, pinkish, vascular tumor was seen over the iris originating from the angle on temporal side between 2 and 3 O’clock position (Figure 1). Iris was seen moving freely over the mass once the pupil was dilated. The adjacent lens was indented and displaced with a sectoral cataract on the side of the mass. The tumor appeared confined to the ciliary body on gonioscopy with open angle. B-scan of the right eye was also suggestive of a ciliary body tumor. Left eye was normal, Fine needle aspiration cytology (FNAC) of the mass was done but proved inconclusive.

In view of the clinical features, the diagnosis of a benign tumor of ciliary body was made. Iridocyclectomy of the right eye was performed under peribulbar anesthesia. Excision of the clear margins of the normal ciliary body and iris surrounding the tumor was performed via a T-shaped limbal scleral incision of ‘stallard.’ (Figure 2,3).
Macroscopically it was an ovoid tumor 8mm in diameter with smooth, dark external surface. Cut surface was yellowish grey, firm and solid. Microscopic examination revealed well circumscribed tumour enclosed by ciliary epithelium. The tumor cells were loosely packed and having round to ovoid vesicular nuclei with 1-2 small nucleoli and abundant fine eosinophillic fibrillary cytoplasm. Nuclear atypia, mitosis and tumor necrosis were absent. The stroma contained widely spaced thin ectatic capillaries lined by single layer of cells. The diagnosis of mesectodermal leiomyoma was made (Figure 4). Immunohisto-chemistry profile revealed strong immunoreactivity for smooth muscle actin and negative reaction for S-100, epithelial membrane antigen (EMA), glial fibrillary acidic protein (GFAP), Desmin and Mib-1 labeling index of < 1%.

DISCUSSION
The ciliary body tumors include Naevi, Melanomas and benign smooth muscle neoplasms. Smooth muscle neoplasms are either leiomyomas or mesectodermal leiomyoma. Mesectodermal leiomyomas are extremely rare and only 15 cases have been reported in literature so far. The differential diagnosis of mesectodermal leiomyoma includes melanomas, gliomas, peripheral nerve tumors or paraganglioma because of its resemblance to ganglionic, astrocytic and peripheral nerve tumors.

Published literature reported on leiomyomas has shown that most of the cases are treated by resection but some may need enucleation. Recurrences have also been reported in a few cases and are attributed to incomplete removal of the tumor. Maximum follow up and reports of recurrence has been six years. In our case the follow up is of 2 years and there is no evidence of recurrence.

REFERENCES
One of the greatest tributes a physician can have is to have a medical condition named after him. Eponymy is “the most enduring and perhaps most prestigious kind of recognition institutionalized in science”. Scientists are not known to be modest about themselves. They would not appreciate Thomas Jefferson’s famous remark after being named America’s second ambassador to France after Benjamin Franklin. Arriving in Paris in 1785, Jefferson was asked, ‘So you are to replace Dr. Franklin?’ he replied, ‘I succeed Dr Franklin. No one can replace him.’ Scientists love eponyms and they have the maximum eponyms after them and painters and musicians have the least number.

The vocabulary of medicine is replete with eponymous descriptions. “There are, for instance more than 15 eponymous designations credited to Fanconi, and there are at least 13 Smiths who independently described different and completely unrelated syndromes, thus rendering the eponyms Fanconi’s Syndrome and Smith’s Syndrome practically useless”. Ophthalmology is hardly less culpable: at least 15 eponyms hang around Ernst Fuchs’ neck, (Fuchs’ adenoma, angle, coloboma, dellen, corneal dystrophy, gyrate atrophy, heterochromic iridocyclitis, lines of clearing, nodules, phenomenon, spot, spur, stomas, superficial keratitis and syndrome) Anton Elschnig is almost strangulated by the weight of 8 eponyms, (Elschnig’s pearls, conjunctivitis, spots, operation, syndrome, theory, intracapsular forceps, scleral ring) David Cogan limps along with 5, (Cogan’s syndrome, Cogan Reese Syndrome, lid twitch sign, epithelial dystrophy and congenital oculomotor apraxia) Goldmann has the unbearable lightness of being with 7, (Goldmann perimetry, applanation tonometry, Goldmann-Favre syndrome, fundus contact lens, 3 mirror, gonioscope, adaptometer) and so on. The Eponyms Dictionaries Index edited by James A. Ruffner, lists 20,000 eponyms overall!

In 1974, US National Institutes of Health held a conference on the naming of diseases and in 1975, JAMA published an article and decided to drop the possessive “s” in all its publications (Down Syndrome not Down’s Syndrome) but has not been able to convince journal editors. Eponyms can be of various kinds:

- Named after male physicians (2,880 on whonamedit.com)
- Named after female physicians (101)
- First and last name
  - Austin Flint
  - Foster Kennedy Syndrome
- Named after more than one person
  - Charcot-Marie-Tooth-Hoffman disease
- Named after famous people with the disease
  - Lou Gehrig Syndrome (ALS – Amyotrophic Lateral Sclerosis)
- Named after fictional characters
- Ulysses (side effects of extensive diagnostic tests
because of false positive results in course of routine investigation, like Ulysses, patient ends up weary after much fruitless exploration)

- Lazarus (symptoms that occur due to prolonged cerebral anoxia mostly of temporal lobes, seen in post resuscitation states after cardiac arrest, “moving through a tunnel, seeing a light, feeling outside one’s body”, named after the Biblical Lazarus whom Jesus raised from the dead after 4 days)
  - Named after the first patient with the disease
  - Christmas Disease (after Stephen Christmas)
  - Hartnup Disease (family name of first patient diagnosed in England in 1956 of this metabolic disorder)

Not everybody agrees. Stephen Stigler is definitely disgruntled about it. He believes that eponyms do not reward the achievement of an original discoverer, because they are usually wrongly attributed. In fact Stigler’s law of eponymy states that eponyms are never named after the original discoverer (exception; Pott’s fracture has to be a legitimate eponym because it describes - not a patient’s fracture - but his own. Pott suffered this fracture of the lower leg bones when he was thrown from his horse). Mark M. Ravitch goes so far as to say that one of the following is usually true of eponyms:

“The eponymized person was not the first describer; the eponymized person didn’t understand the discovery; the eponym’s current meaning is different from that which was described; the eponym has no historical basis”

**Story of the two Hans**

And now we will talk about two scientists, one a German bacteriologist-Hans Reiter, the other, a Swiss ophthalmologist-Hans Goldmann.

**The Double Life of Hans Reiter**

In 1916, Dr. Hans Reiter examined a German military officer who complained of burning micturition, joint pains, high fever and red eyes following a sexual liaison. The term Reiter Syndrome comprising of features of conjunctivitis, urethritis and arthritis, particularly when associated with HLA B-27, was adopted in English language journals in 1940. Reiter’s name was first attached to the syndrome, at least in the English language, in an arthritis textbook in 1941 and a journal article in 1942 by Dr. Bauer and Engleman. But behind the eponym lay the dark side of the moon. Dr. Hans Conrad Julius Reiter was one of the first physicians to swear an oath of allegiance to Adolf Hitler in 1932, and was one of the most prominent members of the eugenics movement that advocated killing all people who had any physical or mental disability. He declared that it is medicine’s responsibility to stop further transmission of inferior genes. He sanctioned medical experiments that killed thousands of people at Buchenwald Concentration Camp. One documented experiment was to infect 250 prisoners at Buchenwald with rickettsia and then massacre them. He wrote a book on racial hygiene and was a strong supporter of Hitler’s anti smoking campaign, medically progressive at that time, and was a talented teacher who was popular with his students and was even included in a book of the top 5000 Nazis. After the Nazis were defeated, he was arrested by the Red Army in Soviet Union-occupied Germany, tried at Nuremberg and was interned in an American prisoner of war camp. He was not convicted of any crimes in the Nuremberg Trials but that may have been partly because of the difficulty in obtaining proof; the two top-ranking Nazi doctors with whom Dr. Reiter had planned experiments at Buchenwald committed suicide in 1945. After the war, Reiter continued publishing research papers and practiced medicine in Kassel. He died in 1969 at the age of 88. In 2000, Dr. Daniel J. Wallace and Dr. Michael Weisman, writing in the Journal of Clinical Rheumatology, unveiled this stunning revelation. Acting on this information, the Spondylitis Association of America, a patient advocacy group, that represents people with Reiter’s Syndrome, voted to call Reiter’s Syndrome, Reactive Arthritis.

Another eponym deriving from this period is that of Wegener’s Granulomatosis. Friedrich Wegener was a member of the Nazi brownshirts and his mentor Martin Staemmler an enthusiastic supporter of the Nazi Racial Hygiene Programme. At present there is no direct evidence that Wegener participated in any way in war crimes although he appears to be on a wanted list by the Polish Ministry for the Interior. Interestingly, Wegener himself did not refer to the disease as Wegener’s Granulomatosis, but at most referred to it as the “so called Wegener’s Granulomatosis”.

**The Astonishing Universality of Hans Goldmann**

“Hans Goldmann took the torch and illuminated vast areas of ophthalmology. Every patient and every ophthalmologist benefited from his contributions.” Born in 1899 near Prague, this Renaissance man of ophthalmology wanted to become an astronomer but was persuaded by his parents “to do something practical” so he became an ophthalmologist. His universality was overwhelming. A list of his
achievements reveals the vast scope and range of his interests.
1. Pathogenesis of cataract in general, but glassblower’s cataract in particular, he clashed with Alfred Vogt in a 300-page long academic battle which was dangerous for him because he was a foreigner and Jewish in Switzerland. Vogt thought that the cataract was a result of infrared energy being absorbed directly by the lens, while Goldmann held that it was caused by heat transferred to the lens from the energy absorbed by the iris. Later experiments proved they were both right.
2. First to determine in 1950 aqueous flow to be 2.5 μL per minute.
3. Designed the Goldmann cupola perimeter in 1945 which remained the gold standard for perimetry for almost 50 years before his own student and chief guinea pig Franz Fankhauser ushered in the era of automated static perimetry with the invention of the Octopus.
4. Researches on colour vision - his first research activity was to reassemble a Hering colorimeter that had been left in a highly disordered state by the former chairman of the Institute. He had observed earlier that his own color sense was somewhat defective as he was unable to see the tuberculosis bacilli present in a stained microscopic slide. He proved to be an anomalous trichromat, his name was mentioned in a handbook of physiology as the “protoanomalous Goldmann”.
5. Experiments related to the Stiles-Crawford effect. The Stiles-Crawford effect is one of those discoveries that came by chance. In 1933, Stiles and Crawford were working in the National Physical Laboratory in Middlesex, England. They were building an instrument to measure the area of the pupil based on the photometric principle, but it did not work and after eliminating all possible sources of error they decided to study the cause of their failure and discovered the effect!
6. Worked on the dark adaptometer which is the standard for dark adaptation (Goldmann - Weekers adaptometer)
7. Researches on the fluorophotometer.
8. The construction of a new slit-lamp, the Haag Streit 360, in 1933 and later, the Haag Streit 900 which is the world standard. The Gullstrand slit lamp was manufactured by Carl Zeiss. (Typically a classic Haag Streit has converging optics with a flip lever to change from 10x to 6x, where as a Zeiss has parallel optics and has a drum wheel to change magnification)
9. In 1954, he invented the applanation tonometer, the most accurate measurement of intraocular pressure till date.
10. Stereochronoscopy of the optic nerve head, which involves rapid detection of changes in the optic disc in glaucoma.
11. Clinical research on Retinopathy of Prematurity and Diabetic Retinopathy
12. Philosophical writings relating to the philosophy, spirit, power, limits and reasoning in science.
Such was Hans Goldmann, the greatest of scientists in the branch of ophthalmology and such was the despicable double life of Hans Reiter. Both were honored with eponyms but what a world of difference!
Sometimes eponyms can be frankly nationalistic. A case in point is Takayasu’s arteritis. Makito Takayasu presented the case of a young woman with retinal artery changes to the Japanese Society of Ophthalmology in 1908. It seems that Takayasu did not even recognize the peripheral artery changes of the disease. Nevertheless, by 1941 Yasuo Niimi was proposing to honor Takayasu by calling the disease after him, something which must be seen in context of the political situation of that time, at the dawn of World War Two and the height of Japanese imperialist designs.
So what’s in a name, you ask? Plenty of heartburn, jealousy, intrigue and infighting. It reminds us of the friction between two great English Prime Ministers William Gladstone and Benjamin Disraeli. In one such encounter Gladstone said derisively, “You will die either on the gallows or of a vile disease”, Disraeli came back with one of history’s greatest retort; “That will depend upon whether I embrace your principles or your mistress”. So it’s a refreshing change for us when a perfectly delightful gem of a syndrome emerges from the dreary, desert sand of the world of eponyms. It is a new syndrome, which has never been previously reported in the medical literature. It is called Tashima’s syndrome (C. K. Tashima, JAMA 194:208; 1965) and in its full-blown state (forme fruste manifestations may be described in the future) is defined as a condition in which a physician earnestly searches for a new sign, disease, or syndrome to which his name can be attached!
With rapid development in imaging techniques their role in diagnosis, management and follow up of cases in Ophthalmology has increased multifold. Need for imaging can be diverse more so when there is a diagnostic dilemma. Neuroimaging however can only supplement a good clinical examination and never replace it. In all cases of suspected intracranial lesion, it is important to clinically localise it so that the appropriate investigation with correct site is imaged. A wrong investigation or one that does not image the site of likely lesion is more dangerous than no investigation as it leads us into a sense of complacency and may delay diagnosis with fatal outcomes. The current article gives a set of guidelines for imaging in cases where good clinical examination may not provide the answer.

**Common Indications for Neuroimaging in Ophthalmology**

1. Unilateral or bilateral visual loss (transient visual loss, unilateral or bilateral optic neuropathy, junctional scotoma, bitemporal hemianopia, homonymous hemianopia or cortical blindness)
2. Efferent pupillary defects (anisocoria due to Horner syndrome or third nerve palsy) or afferent pupillary defects (e.g. RAPD or light-near dissociation of the pupils)
3. Proptosis (thyroid eye disease, orbital tumours, idiopathic orbital inflammation, orbital cellulitis or CCF)
4. Diplopia or external ophthalmoplegia
5. Lid abnormalities (lid retraction, lid lag, ptosis or orbital-lid lesion)
6. Oscillopsia (nystagmus)
7. Fundus abnormalities (papilloedema, optic atrophy, optic nerve hypoplasia, optic disc head drusen or choroidal folds)
8. Ocular or orbital trauma (e.g. intraocular/intraorbital foreign body or suspected fracture)
9. Isolated headache or facial pain

**Pre requisites for scanning**

1. Decide whether CT or MRI is needed – MRI is used in most neuro-ophthalmic conditions except in cases of – calcification, acute haemorrhage, emergent situations and if MRI is contraindicated.
2. Contrast is ordered for most cases in CT & MRI except in cases of renal failure and allergy to contrast
3. The contrast is not required in cases of acute intracranial or intraorbital haemorrhage as a non-contrast study is superior for assessing the hyperdensity of acute blood. It is also not required in cases of trauma (e.g. orbital, facial or skull base fracture, orbital or intracranial FB or traumatic optic neuropathy), birth hypoxia, hydrocephalus, sinus disease (sinusitis) and thyroid eye disease (iodinated contrast may even worsen systemic thyroid disease.)
4. When ordering a CT scan it is important for the ophthalmologist to specify whether the study should include the orbit, the head or both after topographically localizing the lesion.
5. Although the most commonly obtained orbital CT planes are axial and coronal, sagittal views can also be obtained with computerized reconstruction - the only option in a patient who is unable to extend or flex their neck for direct imaging (e.g. trauma, cervical collar or cervical disc disease)
6. Order specific imaging sequences – e.g. Fat suppression for orbital post contrast study especially for optic nerve sheath meningiomas,optic neuritis and thyroid ophthalmopathy, FLAIR sequence for white matter lesions and GRE for stroke.
7. If the clinical picture suggests a particular lesion but the imaging is normal – repeat the imaging with thinner slices and higher magnification of the area of interest.
8. An ophthalmologist should communicate to the neuroradiologist - the clinical findings, suspected lesion location, differential diagnosis and urgency of the imaging request to optimize the interpretation of imaging studies. When in doubt get another radiologist to give an independent report
9. Better neuroimaging techniques should be used if the clinical signs definitely suggest a lesion.
Indications of CT scan

1. Acute orbital trauma (e.g. suspected orbital fracture, orbital hematoma, metallic or wooden foreign bodies, or traumatic optic neuropathy)
2. Acute proptosis (e.g. orbital cellulitis, idiopathic orbital inflammation, thyroid eye disease with compressive optic neuropathy, post-surgical or spontaneous retrobulbar hemorrhage)
3. Acute bitemporal hemianopia (e.g. pituitary apoplexy)
4. Acute homonymous hemianopia or cortical blindness (e.g. acute stroke, acute hemorrhage from tumor or arteriovenous lesion)
5. Acute, severe headache (‘worst headache of my life’) with or without acute third nerve palsy (subarachnoid hemorrhage due to ruptured intracranial aneurysm)
6. Acute papilledema (e.g. to rule out intracranial tumor or bleed in the emergent setting)
7. Acute visual loss
8. Acute diplopia
9. Demonstrating calcification in ophthalmic conditions (retinoblastoma, optic nerve head drusen, meningioma, craniopharyngioma)

Right homonymous hemianopia (Figure 1) with left PCA infarct and occipital tuberculoma

Figure 1: Right homonymous hemianopia

Figure 2: Non contrast Axial CT scan Spontaneous intracranial haemorrhage involving thalamus and extending to optic radiation

Figure 3: Acute spontaneous haemorrhage Left occipital lobe

Figure 4A
Role of Radio Imaging in Ophthalmology

Figure 4: A (left) B (above) HIE with Right hemiparesis and Left cerebral hemiatrophy seen on Axial NCCT scan.

Figure 5: Sagittal CT scan (right) showing right superior orbital haematoma along SR/LPS complex.

Figure 6: Traumatic blowout fracture Right.

Figure 7: Traumatic blowout fracture Left.
Indications of MRI
For almost all neuro-ophthalmic indications, MRI provides superior soft-tissue discrimination (e.g. meninges, cavernous sinus, posterior fossa, dural venous sinuses and optic nerve).
Normal anatomy is best demonstrated on T1WI. In distinction, T2WI are typically better for demonstrating intracranial or other pathology.

CT is often used in conjunction with MRI
CT & MRI are often complimentary & can offer a complete picture of nature of lesion. It is used in conjunction in disorders of bone or sinuses which includes the following -

1. Orbital, maxillofacial, calvarial or skull base fracture,
2. Hyperostosis of bone associated with meningiomas,
3. Fibrous dysplasia,
4. Sphenoid wing agenesis in neurofibromatosis-1,
5. Craniosynostosis syndromes,
6. Sinusitis or sinus tumours,
7. Clivus or other skull base pathology,
8. Primary bone tumors, bone destruction or erosion
9. Demonstrating calcification
10. Craniopharyngioma, meningioma or retinoblastoma
11. Hyperdensity of acute haemorrhage
12. Orbital, subdural, subarachnoid intraventricular or intraparenchymal haemorrhage associated with tumor, stroke or ruptured aneurysm.

MRI is contraindicated in case of severe claustrophobia, marked obesity exceeding table weight limit, uncooperative patient showing excessive mobility, cochlear implant, ferromagnetic aneurysm clip, pacemaker and in cases of metallic foreign body.

Gadolinium contrast material in MRI
As with CT, MR scanning with intravenous contrast material improves detection of pathology by demonstrating areas of blood–brain barrier breakdown. Unlike the iodinated contrast used for CT, the contrast material used for MR is a paramagnetic material called gadolinium. The gadolinium paramagnetic metal ion enhances the local magnetic field and increases signal intensity. Gadolinium contrast agents are chelated to form a larger, more stable complex around the more toxic, free gadolinium. This stable complex is excreted via the kidneys. Fortunately, unlike iodinated contrast material, serious side-effects from gadolinium are uncommon. The most common allergic reactions are typically mild (e.g. skin rash, sweating, itching, hives and facial swelling), but there are reports of rare reactions that can be severe or even fatal. Until now MR with gadolinium was considered the ‘procedure of choice’ in patients with renal insufficiency that required a contrast study. Unfortunately, a relatively new disorder related to gadolinium contrast has emerged called nephrogenic systemic fibrosis (NSF). NSF is a disorder seen weeks to months following gadolinium contrast administration in patients with kidney failure. Clinically, NSF is characterized by thickening and hardening of the skin, typically over the extremities and trunk.

Specific imaging sequences
1. Fat suppression for orbital post-contrast study
2. Fluid attenuation inversion recovery (FLAIR) for white matter lesions,
3. Gradient recall echo imaging (GRE) for haemorrhage and blood products
4. Diffusion-weighted imaging (DWI) for stroke or PRES
5. Perfusion weighted imaging (PWI)
6. Dynamic perfusion CT (PCT)

Fat suppression (Figure 8) is used for all post-contrast orbital T1WI – in cases of optic neuritis, optic nerve sheath meningiomas.

FLAIR sequences in cases of suspected demyelinating disease as it may not be routinely performed as part of a standard MR protocol. Axial and Sagittal T1 weighted Gadolinium enhanced MRI brain – asymmetric periventricular plaques suggestive of demyelination in a young female patient with optic neuritis.
Role of Radio Imaging in Ophthalmology

**Gradient recall echo imaging (GRE)** (Figure 9) can show blood products effectively, the approximate age of a haematoma may be determined, haemorrhage associated with AVM or cavernomas, intra- or extra-axial intracranial haemorrhage

**Diffusion Weighted Imaging** (Figure 10) is based on the microscopic random brownian motion of water. These changes in water molecular diffusion can be measured as signal intensity in vivo with DWI. In certain pathological states, diffusion becomes restricted (i.e. hyper intense or bright on DWI). DWI can differentiate the various phases of cerebral infarction namely hyperacute, acute, subacute and chronic and therefore can detect hyperacute ischemic stroke even before abnormalities are detected on conventional T1W, T2W and FLAIR sequences (Figure 8). It can also be used in cases of acute ischemic homonymous hemianopia, cortical visual impairment, acute cerebral emboli in patients with embolic retinal arterial occlusion and in the basilar syndrome with acute brainstem ischemia producing a progressive ophthalmoplegia.

**Diffusion Perfusion Mismatch** (Figure 11) The volume difference of DWI and PWI gives an approximate measure of hypoperfused, but not yet infarcted and potentially salvageable tissue (the ischemic penumbra). It represents an indication for thrombolysis based on potential reversibility of DWI lesions.

**MR or CT Angiography**

**Indications** -
1. Hemispheric transient ischemic attacks
2. Homonymous hemianopia
3. Transient monocular visual loss
4. Completed cortical strokes
5. Isolated third nerve palsy
6. Carotid cavernous fistula
7. AV malformations

**MRA over CTA:** Advantages are lack of ionizing radiation exposure, less nephrotoxic contrast material (gadolinium vs. iodinated contrast), increased signal-to-noise ratio and easier post-processing techniques.

**CTA over MRA:** Advantages are increased spatial resolution, technically easier and faster study to acquire, less motion artefacts

The sensitivity and specificity of both MRA and CTA are probably equivalent.
Role of Radio Imaging in Ophthalmology

MR Venogram (MRV)(Figure 12)
MRV preferred over CTV due to iodinated contrast material, ionizing radiation exposure, problems subtracting bone adjacent to the venous sinuses in CTV
Thus with the advent of newer sequences and techniques in a widely available array of existing imaging modalities, it is important to not to over-investigate the patients. For that an apt knowledge about the utility of each sequence and technique is a must for those dealing especially in neuro-ophthalmology to get to the root cause of symptoms and make correct diagnosis and management. This article attempts a brief overview which may be useful in routine clinical practice to decide investigation of choice for a particular disease and to get correct imaging done.

Figure 12: Sagittal and oblique MRV: Dural venous sinus thrombosis in a patient presenting with papilledema from increased intracranial pressure
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:
1. The views of experts on current advances in the field, in a clear and readable format.
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.
7. Book review and letters to Editor.

All correspondence shall be acknowledged within six weeks of receipt by the editorial board. Authors shall be intimated about the acceptance of their articles for publication within six weeks of the acknowledgment. The journal expects each contributor to have made a significant contribution in writing the article.

The authors must take full responsibility to ensure that the manuscript contains no matter that is, to the best of contributors’ knowledge, libelous or unlawful, or infringes upon any Indian copyright laws. All contributors are requested to sign a ‘letter of transmittal’ at the time of submitting their manuscript for consideration for publication.

Submission of the Manuscript
Authors are requested to read these instructions carefully before submitting manuscripts.
All Manuscripts/contribution should be sent by post to Dr. Rohit Saxena, Editor, Delhi Journal for Ophthalmology, Room No. 479, Dr. RP Centre for Ophthalmic Sciences, AIIMS, New Delhi-110029. All manuscript and illustrations must be submitted in triplicate along with a CD. The author(s) should retain a copy for their future reference. To hasten the process of review, Manuscripts may be submitted electronically by email to Dr. Rohit Saxena at editor_djo@gmail.com. These should be in MS-Word format & the image should be in jpeg format (email) and Tiff format (Disc). Graphs and line drawing/diagrams must be sent in graphic format that is EPS, LOTUS/EXCEL Spreadsheet files, PICT/CHART files, or Harvard graphic. Do not send graphs and diagrams in freehand. The disk should be labeled with the title of the article, author’s name, the file name, and software used including version. The disk should be sent in proper packaging to avoid damage and corruption of the information during transit. Unreadable disks will be returned to the author for substitution.

Manuscript Preparation
1. Type using font size of 11 or 12 with black ink.
2. Use double spacing, throughout the manuscript including references, tables, and legends.
3. Do not use vertical lines or underlining, anywhere in the text or the table.
4. In the upper right-hand corner, identify each page with a number and a running title.
5. Number pages consecutively in Arabic numerals beginning with title page.
6. Other than the title page, do not identify authors else-sm-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: Cambridge 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.

Manuscript Layout
The manuscript (including reference, legend, and tables) must be typed in double spacing on a 21.6 x 27.6 cm (8.5”x11”) paper with at least 2.5 cm
(1") margins. The pages should be numbered in the following order, title page, abstract, text, reference, legends for illustrations and tables.

**Title Page**

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.

**Abstract**

The abstract should be structured for Original Articles and unstructured for others. It should not exceed 250 words. The Structured abstract should have the following sections: Purpose (or background), Methods, Result and Conclusion. Key Words: should be submitted to assist indexing. These should not exceed five.

**Text**

The body of the text should include Introduction, Materials and Methods, Results and Discussion.

**References**

All references must be numbered consecutively by their order of appearance in the text. In the text, please indicate the reference number as superscripts. Use “INDEX MEDICUS” type of abbreviations. Please follow precisely the format and punctuation shown in the following examples.


**Tables**

Each table must be numbered consecutively in the order mentioned in the text and titled. Please do not type more than one table on a page.

**Illustrations**

Line drawing or graphs must be printed on glossy paper. Each illustration should be numbered in Arabic numerals and cited consecutively in the text. Attach a label on the back of each print giving the illustration number, an arrow indicating the orientation “top”, and the article’s running title (without author’s name). Do not write on the print. Do not damage illustrations with paper clips or by bending them. The legend or illustration number should not be incorporated into the illustration. Published illustration and photographs will not be returned to the author(s). Patients should have their identity concealed (including names and hospital numbers) or their photographs should be accompanied by the patient’s written permission to publish. Any figure that has been published elsewhere should have an acknowledgment to the original source and proof of permission to use from the holder of copyright. Graphs, original illustrations, and line drawings may be drawn with India ink, photographed and submitted as photographic prints, or may be drawn on a computer in graphic format and submitted as laser printouts. All photographs, graphs and line drawing should be included in the electronic file (see Electronic Manuscript).

**Photographs**

All photographs (black & white and colour) must be top quality prints and should be 5x7 or 4x6 inch in size. A smaller size will result in a poor quality. All photographs are printed free of charge. However, the number of photographs to be selected for printing will be decided by the Editorial board. Very sharp contrast is essential for colour representations.

**Colour Photographs**

Authors wishing to include colour photographs in the text should send either on colour transparencies or on a CD in TIFF format. Colour photographs may be printed at the discretion of the editor if it is felt that they would contribute substantially to the understanding of the text.

**Legends**

Figure legends must be numbered consecutively in Arabic numerals as they appear in the text. For histological figures, stain and magnification should be noted. Legends must identify all symbols or letters appearing in the figure.

**Reprints**

One reprint shall be sent free of cost to each contributor of the columns: review articles, current practice e, recent advances, original article, surgical.
Acknowledgments
Acknowledgment are accepted for sponsoring organizations and grants, and for those who referred patients, provided statistical assistance, supplied essential tissue, equipment, or other material without which the study could not have been accomplished. Acknowledgment will not be published for those who reviewed, discussed, edited or typed the manuscript; clinic coordinators, ophthalmic photographers, or technicians.

Letter of Transmittal
The letter of transmittal, the text of which is given below must accompany all manuscripts. This must be typed separately on a fresh sheet of paper and signed by each of the contributor/s.

In consideration of my submission entitled..... being reviewed and edited by the Editorial board of Delhi Journal of Ophthalmology (DJO) the contributor(s) undersigned hereby transfer(s) and otherwise conveys all copyright ownership to DJO in case the work is published in DJO. The contributor(s) declares that the manuscript contains no matter that is, to the best of contributor(s) knowledge, unlawful or that infringes the Copyright Acts of India.

Disclosure and copyright transfer statement
All manuscripts should be accompanied by the disclosure and copyright transfer statement which must be signed and dated by all the authors without which the manuscript will not be accepted for review and possible publication. The statement should read as follows: “The enclosed manuscript is hereby submitted to the Delhi Journal Ophthalmology. The undersigned confirm that the typescript and illustrations have not be been published in any other Journal, and on acceptance will not be offered to any other Journal, and on acceptance will not be offered to any other publisher without the consent of the Editorial Board. The undersigned transfers, assigns or otherwise conveys all copyright ownership of this manuscript to the Delhi Ophthalmology Society in the event of its publication in the Delhi Journal of Ophthalmology. Such conveyance includes any product that may derive from the published journal, whether print or electronic.” If the data in the manuscript were presented at a scientific meeting, the place of data of presentation, and name of the meeting should be stated on the title page. Any proprietary or financial interest in any product mentioned in the manuscript should be stated on the title page.

Methodological guidelines
The journal recommends that authors ensure statistical expertise for a study that has statistical content. Authors are requested to follow the following guidelines.

Analysis and Presentation
1. There must be a satisfactory statement about the source of subjects.
2. Criteria for the selection of subjects must be stated.
3. Treatments must be well defined.
4. Duration and post-treatment follow up must be stated.
5. A statement adequately describing or referencing all statistical procedures is mandatory.
6. Statistical analyses used must be appropriate.
7. An appropriate sample size should be used. If the sample is small, the statistical power needs to be mentioned.
8. Confidence intervals along with exact probability values must be stated for the results.
9. Conclusions drawn from statistical analysis must be valid and justified.

Conduct of Trials
1. Concurrent or historical controls must be used.
2. Treatment and control groups must be comparable in relevant parameters.
3. Randomization, if used, must be specified.
4. The followup period and proportion of dropouts must be comparable for both the groups, with the reasons for dropouts specified.
5. Side effects of the treatments must be reported.

Manuscript Processing
All manuscripts are acknowledged and a number is assigned to the manuscripts. In future correspondence the same number must be quoted.

Peer Review
All manuscripts are subject to editorial review. Manuscripts may be processed by section editor. Manuscripts involving statistics are, in addition, subjected to statistical review. Accepted manuscripts become the permanent property of the journal and may not be published elsewhere without permission from the Editor.
Revision of Manuscripts
Manuscripts sent for revision must be returned within the time stipulated in the Editor’s letter. Failing this, the manuscript must be resubmitted.

Rejected Manuscript
The manuscripts of rejected articles are not returned due to high postal expenses.

Miscellaneous
Patients should have their identity concealed (including names and hospital numbers) or their photographs should be accompanied by the patient’s written permission to publish. Any figure that has been published elsewhere should have an acknowledgment to the original source and proof of permission to use from the holder of copyright.

Categories of Manuscripts

Review Articles
Reviewers write short articles in which they present developments in their topic, emphasizing the aspects that, in their opinion are the most important. In addition, they provide short annotations to the papers published in their topic during the period reviewed. This selected bibliography is printed at the end of each review. Your review should be 10-14 typed pages. The article should highlight and discuss all interesting developments in your subject, as reflected in the recent literature. In addition to describing recent trends, you are encouraged to give your own opinions of the topics discussed. However, be careful of expressing conclusions in a way that might be construed as biased against a particular researcher, product or manufacturer.

Original Articles
Original article should generally not exceed 3,000 words or 12 double-spaced pages.

Brief Reports
These should not exceed 1000 words with a maximum of 4 illustrations. They should follow the following format: introduction, case(s), and discussion. No more than 8 references should be cited. Each brief report must begin with a 75-100 words summary that highlights the significance of the article. Besides these requirements, the general instructions for author should be followed.

Letter to the Editor
Comments about an article published in the journal, or topics of ophthalmic interest are considered. Comments regarding articles in the journal should be submitted within 3 months of publication, and the author(s) of that article will be given an opportunity to reply. The general instructions for authors should be followed. The letters should be accompanied by the disclosure and copyright transfer statement. Authorship is limited to three, and signatures of all authors are required.

Book Reviews
Book reviews should be accompanied by photocopies (one set) of the title page (citing page numbers, indexing, illustrations, year of publication, publishing company), and contents page(s).

Journal Abstracts
Abstracts of interesting articles published in other journals may be submitted. Those contribution journal abstracts should include a photocopy of the published article.

Electronic Manuscript
This should accompany the paper text at the time of submission. Upon acceptance for publication or at the time of revision when a manuscript is likely to be accepted for publication, the corresponding author will have to submit an electronic file on disk, in addition to the original manuscript. Disks that are IBM PC compatible (non Macintosh) will be accepted. Floppy disks should be MS-DOS based in word perfect 5.1 of MS Word for windows. Files in formats other than these should be converted to MS-Word DOS text format (ASCII) before submission. The disk should be labeled with the title of the article, author’s name, the file name, and software used including version. The disk must contain exactly the same material as the revised manuscript including the tables, legends, and graphs. Graphs and line drawing/diagrams must be sent in graphic format, that is, EPS, LOTUS/EXCEL Spreadsheet files, PICT/CHART files, or Harvard graphic. Do not send graphs and diagrams in freehand. The disk should be sent in proper packaging to avoid damage and corruption of the information during transit. Unreadable disks will be returned to the author substitution. Disk with their packaging will be returned to the author after use by the journal on request.

Complimentary Copies
Complimentary copies are sent to authors of published articles even if they are neither DOS members nor subscribers. This request should be made in the reprint order form.

Errat
Substantial errors in published material are corrected at the earliest possible after being brought to the notice of the Editorial Board.