The human eye is already a work of art, choose the science to perfect it.
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. Yes we can! .......................................................... Rohit Saxena

Major Review

7. Endophthalmitis: A current appraisal
   Anusha Kumar, Vinit Shah, Saurabh Patwardhan, Rajvardhan Azad, Yograj Sharma, Parijat Chandra
14. Duane’s Rectraction Syndrome
   Manish Malhotra, Ramesh Murthy
22. Idiopathic Macular hole
   Varun Gogia, Jaideep Tyagi, Atul Kumar, Rajvardhan Azad, Yograj Sharma
28. Glaucoma Suspect
   Deven Tuli

Preferred Practice Patterns

37. Evaluation of a Visually Inattentive Infant
   Sumit Monga
41. Important Aspects of Paediatric Cataract Surgery
   Abhay R Vasavada, Sajani K Shah, Viraj A Vasavada, Vaishali A Vasavada

Cases

46. Seasonal occurrence of ocular lesions caused by Caterpillar hair-A Case series
   Gajiwala UdayR., Patel Rajesh U., Gangwal Manoj M., Chariwala Rohan A.

Law in Focus

50. Medico-Legal Aspects of Post-Operative Endophthalmitis
   Lalit Verma, Shefali Gupta

History of Ophthalmology

52. Penetrating Keratoplasty: History and Evolution
   Shibal Bhatia, Noopur Gupta, Anita Panda

Instruments Scan

55. Understanding Corneal Topography: Basic
   M. Vanathi

Instructions to Authors
Editorial

Yes we can!

Dear Friends,

It is with great pleasure that I present before you the second issue of the Delhi Journal of Ophthalmology. For this I express my sincere gratitude to Dr Sharad Lakhotia, President DOS, Dr Amit Khosla, Secretary, DOS and the entire DOS executive for their constant support and help. Without their presence and encouragement this would not have been possible.

In the first issue I had wondered if a regular and academically sound DJO is perhaps a bridge too far! That this journal will always remain an after-thought for us! But seeing the response to the first issue and the number of articles that we are now receiving, I think that change is within our reach. I think we can dream of having a good and scientific review journal that we can expect to be eventually accepted as a source of authentic and in-depth information on major scientific topics, nationally and perhaps internationally.

However skeptical we have been of the DJO, but this is what I know. I know that when people say we can’t generate the all the big money, advertisers and articles needed to sustain a journal, I think of all the residents and individual ophthalmologists who forever look forward for good articles and new knowledge to add value to their practice. I know that we have to respond to their need, to answer their questions and to give them the vast information available in the international forum.

So don’t tell us change isn’t possible. It will not be easy, but only with the support and involvement of the readers and the experts I feel we will be able to bring out this journal with the regularity and content that a society of our standing deserves. The first issue I hope was a good start, but there is great scope for improvement. This issue has articles from some of the best in the business. Over the past 4 years this journal has shaped itself to be a review journal providing cutting edge information. Send us your needs and what you want to read in these pages. You as the reader and critique can help us to shape up and give us the much needed direction. Ask for your scientific journal! We, the Editorial Board and the DOS will have to give it to you.

Let us prove wrong the cynics who believe that what began in the first issue was just an illusion.

Yes, we can seize our future! Yes, we can make the change! Yes, we can!!!

Dr Rohit Saxena

Yes we can! was the catch phrase of Senator Barack Obama after he won the Democratic presidential primary in South Carolina. This expression went on to become a national movement for change installing for the first time a black in the Oval Office.
Endophthalmitis: A current appraisal

Anusha Kumar, Vinit Shah, Saurabh Patwardhan, Rajvardhan Azad, Yograj Sharma, Parijat Chandra
Dr. Rajendra Prasad Centre for Ophthalmic Sciences, AIIMS, New Delhi

Endophthalmitis is an intraocular inflammation involving the vitreous cavity and the anterior chamber of the eye. It can be exogenous or endogenous depending on whether the organisms gained entry from the external or the internal environment to the eye. The overall incidence of post traumatic endophthalmitis ranges between 2.4-8%, 0.07%-0.12% for endophthalmitis following cataract surgery, 0.11% for penetrating keratoplasty, 0.05% for pars plana vitrectomy, and 0.2-9.6% for endophthalmitis following filtering surgery.

The different forms of endophthalmitis, their presentation and management strategies are discussed below.

**Exogenous Endophthalmitis**

**Acute Post-operative endophthalmitis:**
It is the most commonly encountered form of endophthalmitis with most patients presenting within 1-2 weeks of ocular surgery. Most commonly seen following cataract surgery, it can also follow glaucoma filtering surgeries, penetrating keratoplasty, kerato refractive surgery, strabismus surgery, scleral buckling and vitreous surgeries and pterygium excision. Reviews of various surgical procedures have shown secondary intraocular lens placement with the highest endophthalmitis rate (0.2-0.3%) and pars plana vitrectomy (PPV) with the lowest rate (0.03-0.046%). The recent increase in the rates of endophthalmitis has been attributed to the increasing use of clear corneal incisions over sclera tunnels for cataract surgery.

**Microbiological isolates:**
Bacteria are the most common infecting agents, although fungal infection may also occur, particularly in association with the use of contaminated ocular irrigation fluids. The eyelids and the patient’s conjunctiva are the the primary source of infection. In the EVS, 94.2% of culture-confirmed cases involved Gram-positive bacteria, mostly coagulase negative (CONS). Others involved Staph.Aureus, Streptococcus, Enterococcus and Gram negative species.

**Signs and symptoms:**
Any eye with inflammation greater than the usual postoperative clinical course should be suspected of having endophthalmitis. Initial symptoms are often rapidly progressive, including pain, red eye, ocular discharge, and blurring. Common signs include decreased visual acuity, lid swelling, conjunctival and corneal edema, anterior chamber cells and fibrin, hypopyon, vitreous inflammation, retinitis, and blunting of red reflex. Wound leak or dehiscence, suture abscess, vitreous incarceration in the wound and eroding scleral sutures predispose to endophthalmitis and should be looked for.

**Differential diagnosis:**
1. Retained nuclear or cortical fragment of the lens.
2. Toxic anterior segment syndrome (TASS) due to non infectious substances entering the eye during or after surgery. Bacterial toxins, preservatives, detergents, intraocular solutions, early postoperative migration of externally applied ointment into the anterior chamber have been implicated in TASS. TASS can be differentiated by its rapid onset (within 12-24 hours of surgery), lack of isolated organisms by Gram stain or culture, and predominance of anterior inflammation.

**Investigations:**
1. Ultrasonography: Media opacities due to vitritis, chorio-retinal thickening in advanced cases, retinal/choroidal detachment, retained lens matter/ intraocular foreign body can be picked up.
2. Specimen to be obtained for slide and culture: A vitreous specimen may be obtained either by vitreous needle tap or by vitreous biopsy with a cutting/aspirating probe. Samples should be obtained for Gram stain, culture (aerobic, anaerobic, and fungal),
as well as antibiotic sensitivities. Anaerobic cultures should be kept for at least 14 days to recover slow-growing species (for example, P. acnes). Fungal cultures should be kept for several weeks.

**Endophthalmitis Vitrectomy Study (EVS) and Treatment of endophthalmitis:**
The purpose of the endophthalmitis vitrectomy study was to determine the roles of immediate pars plana vitrectomy (VIT) and systemic antibiotic treatment in the management of postoperative endophthalmitis. A total of 420 patients who had clinical evidence of endophthalmitis within 6 weeks after cataract surgery or secondary intraocular lens implantation were included in the study. Random assignment to treatment with VIT or vitreous tap or biopsy (TAP) and to treatment with or without systemic antibiotics (cefazidime and amikacin) was made.

**Intra-vitreal antibiotics:**
The main treatment of postoperative endophthalmitis is injection of intravitreal antibiotics as they deliver adequate drug concentration directly to the infected tissues. Therapy should cover Gram-positive organisms, which comprises the majority and Gram-negative organisms as these are associated with high virulence and poor outcomes. Current recommendations for empirical therapy are vancomycin 1.0 mg/0.1 mL and ceftazidime 2.25 mg/0.1 mL. The EVS study employed intravitreal vancomycin and amikacin (400µg/0.1ml). However, concern regarding aminoglycoside related retinal toxicity (macular infarction with gentamicin) has prompted the use of third generation cephalosporins as ceftazidime for Gram negative coverage.

**Pars Plana Vitrectomy (PPV):**
PPV reduces the load of infecting organisms, toxins, inflammatory materials, and opacities and also allows collection of samples for culture and facilitates intravitreal antibiotic distribution.

The EVS addressed the relative efficacy of immediate PPV versus vitreous tap in treatment of postoperative endophthalmitis. In the EVS, patients who presented with light perception only visual acuity had a significant threefold improved chance of obtaining 20/40 vision after immediate vitrectomy (33%) compared to vitreous tap or biopsy (11%). Additionally, there was a 56% chance of obtaining 20/100 or better vision after immediate PPV compared to 30% chance after vitreous tap or biopsy. For subgroups with presenting vision of hand motions or better, there was no significant difference between the two treatment groups in final visual acuity.

The results of the EVS study, however, cannot be applied to endophthalmitis following other intraocular surgeries. The treatment of choice in patient with no light perception cannot be derived from the EVS study. Vision of light perception only and isolation of highly virulent organisms were associated with poor visual outcomes in the EVS.

**Systemic antibiotics:**
The role of systemic antibiotics (amikacin plus ceftazidime or amikacin plus ciprofloxacin) is questionable as the EVS study has demonstrated no statistically significant difference in final visual acuity or media clarity with or without the use of systemic antibiotics. Given the favourable characteristics of broad coverage, good tolerability, and ease of oral

**Table 1: Antibiotics in endophthalmitis**

<table>
<thead>
<tr>
<th>Drugs</th>
<th>Topical</th>
<th>Subconjunctival</th>
<th>Intravitreal (per 0.1 ml)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Amikacin</td>
<td>20mg/ml</td>
<td>40mg</td>
<td>400µg</td>
</tr>
<tr>
<td>Amphotericin B</td>
<td>0.15-0.5%</td>
<td>-</td>
<td>5-10µg</td>
</tr>
<tr>
<td>Cefazolide</td>
<td>50mg/ml</td>
<td>75mg</td>
<td>2mg</td>
</tr>
<tr>
<td>Ceftazidime</td>
<td>50mg/ml</td>
<td>100mg</td>
<td>2.25mg</td>
</tr>
<tr>
<td>Fluconazole</td>
<td>0.2% solution</td>
<td>2%-1ml</td>
<td>10-100µg</td>
</tr>
<tr>
<td>Gatifloxacin</td>
<td>0.3%</td>
<td>-</td>
<td>400µg</td>
</tr>
<tr>
<td>Moxifloxacin</td>
<td>0.5% solution</td>
<td>-</td>
<td>400µg</td>
</tr>
<tr>
<td>Tobramycin</td>
<td>8-15 mg/ml</td>
<td>20mg</td>
<td>200µg</td>
</tr>
<tr>
<td>Vancomycin</td>
<td>50mg/ml</td>
<td>25mg</td>
<td>1 mg</td>
</tr>
<tr>
<td>Voriconazole</td>
<td>1-2% solution</td>
<td>-</td>
<td>100µg</td>
</tr>
</tbody>
</table>
administration of oral gatifloxacin or moxifloxacin, these agents are promising adjunct therapies. However, their role along with intravitreal antibiotics is poorly documented.

Subconjunctival and topical antibiotics:
These are used as adjuvants to intravitreal therapy as they poorly penetration into the vitreous cavity. They achieve significant MIC90 levels in the anterior chamber. Subconjunctival antibiotics have a negligible role in eyes that have received intravitreal antibiotics.

Corticosteroids:
Though oral, subconjunctival and systemic steroids are advocated in the setting of acute post operative endophthalmitis, the role of intravitreal steroids remains controversial.

Retreatment:
The EVS recommends re-treatment if the infection fails to stabilize or improve more than 48 hours after the first injection. Decision to reinject antibiotics should not be taken lightly, since repeated antibiotic injection may increase risk of retinal toxicity. The treatment may be modified based on the reports of culture and sensitivity when an initial response is not obtained.

Chronic post-operative endophthalmitis:
This usually occurs 4 weeks or more after surgery, even months and years later.

Microbiological isolates:
A review of endophthalmitis cases presenting more than 4 weeks after cataract surgery found 63% Propionibacterium species, 16% S. epidermidis, 16% Candida, and 5% Corynebacterium species. S. Epidermidis is usually isolated in cases presenting within 6 weeks, Candida within 3 months and P. acnes anywhere between 2 months-2 years.

Signs and symptoms:
The onset is gradual with minimal loss of vision, minimal pain and mild vitritis. Hypopyon is an unusual finding. P. acnes endophthalmitis is characteristically associated with granulomatous precipitates on the cornea and intra-ocular lens, hypopyon, vitritis and a white intracapsular plaque associated with sequestered organisms and retained lens matter.

A dense and diffuse vitritis usually accompanies endophthalmitis due to S. Epidermidis.

Investigations:
1. Ultrasonography
2. Sample for gram stain, and bacterial, fungal and anaerobic culture by aqueous/vitreous tap. The white plaque, if present, should be sent for histological examination for P. acnes. Average growth time of P. acnes is 10 days and hence cultures should be reported negative only after several weeks of observation.
3. PCR for P. acnes

Treatment:
1. Intravitreal antibiotics: Intravitreal vancomycin and ceftazidime are the main stay of treatment. An intra cameral and intra capsular irrigation with vancomycin is considered appropriate for suspected P. acnes endophthalmitis. Owing to its poor susceptibility to vancomycin newer drugs as etropenem, meropenem and cefepime are being investigated. Intravitreal amphotericin B (5-10mg/0.1ml) is administered in suspected fungal endophthalmitis. Newer drugs as voriconazole (100µg/0.1 ml) can be used in resistant cases of fungal endophthalmitis.
2. Pars plana vitrectomy: It is advocated in cases responding poorly to intravitreal antibiotics. PPV with enbloc capsulectomy along with Intra ocular lens implantation has found to high surgical cure rates, especially in fungal and P. acnes associated endophthalmitis. Recurrences may suggest P. acnes endophthalmitis that may require multiple medical or surgical interventions.
3. Systemic antibiotics: their role in chronic endophthalmitis is poorly documented.
4. Systemic antifungals: Oral fluconazole, itraconazole or voriconazole (200 mg BD) have been found effective when administered along with intravitreal antifungals, with or without PPV.

Note on newer generation azoles:
The azoles (fluconazole and itraconazole) currently used lack broad spectrum coverage. Fluconazole is effective only against Candida species. Itraconazole lacks activity specifically against Fusarium species. The newer generation azoles including ravuconazole, posaconazole and voriconazole, are synthetic derivatives of fluconazole but have a significantly broader spectrum of activity. At present, only voriconazole (Vfend, Pfizer, New York) is
commercially available. It has been approved by the FDA for the treatment of invasive aspergillosis, oesophageal candidiasis and other systemic indications, and is available in oral and intravenous formulations.

Ravuconazole⁵ (Bristol-Myers Squibb, Wallingford, CT) has been shown to have in vitro activity against non-ocular isolates of yeasts, Aspergillus species and other filamentous fungi, black moulds, and some Mucorales. Some Fusarium species were resistant. Posaconazole (Noxafil)⁶ oral suspension (Schering-Plough, Kenilworth, NJ) was recently FDA approved in 2006. In vitro and in vivo studies have shown it to have broad-spectrum activity against non-ocular isolates of most Candida species, Cryptococcus neoformans, Aspergillus species, Fusarium species, Zygomycetes and endemic fungi and organisms resistant to fluconazole and itraconazole, and has been claimed to be the most active of any antifungal agent tested against Aspergillus. Adverse events, most commonly gastrointestinal complaints, are generally mild.

Voriconazole⁷ has been shown to have a broad spectrum of activity against non-ocular isolates of Aspergillus species, Candida species, Paecilomyces lilacinus, Cryptococcus neoformans, Scedosporium species, Curvularia species and other species known to be resistant to amphotericin B, fluconazole and itraconazole. Activity against Fusarium species has been variable. Voriconazole has also become the new standard of care in the treatment of invasive aspergillosis which may occur in immunocompromised patients, including allogeneic BMT, other haematological cancers and solid organ transplants. Visual disturbances, including abnormal vision, colour vision change and/or photophobia, are mild and typically resolve within 1 month, even with continued therapy. Elevations in hepatic enzyme levels can occur.

5. **Systemic corticosteroids:** Inflammation can be initially steroid responsive but recurrent after steroid taper. With some fungal infections, however, inflammation may paradoxically worsen with steroids. Hence oral steroids are generally not preferred in cases of chronic post operative endophthalmitis.

**Outcome:**
Outcomes in chronic endophthalmitis is usually better than acute post-operative endophthalmitis and can be attributed to the low virulence of the organism implicated in the former.

**Filtering bleb associated endophthalmitis:**
The major risk factors for endophthalmitis developing following filtering surgeries include:
1. Use of anti fibrotic agents that lead to a thin cystic and avascular conjunctiva at the bleb site making it vulnerable to infections
2. Inferior bleb
3. Post operative manipulation as suture lysis and needling

**Microbiological isolates:**
Streptococci and coagulase negative staphylococci predominate. Gram negative organisms as H. influenzae are more frequently encountered. The organisms implicated are more virulent. Fungal infections are rarely encountered.

**Signs and symptoms:**
Sudden onset of intra ocular inflammation with pain, redness and decrease in vision are characteristic. Bleb purulence may be noted. The term blebitis is reserved for conditions not extending to the anterior chamber or the vitreous.

**Treatment:**
**Blebitis:** Fortified topical/ subconjunctival antibiotics along with systemic antibiotics may suffice.⁸

**Associated endophthalmitis:** Specimen obtained by aqueous/vitreous tap should be sent for gram stain and culture. Aspiration of the bleb is not advised. Intravitreal antibiotics along with fortified topical fourth generation cephalosporins and systemic antibiotics constitute the main stay of treatment.⁸

**Pars plana vitrectomy:** Twenty-five gauge vitrectomy may provide an advantage over 20-gauge approach when treating endophthalmitis after trabeculectomy as it causes less disruption of the filtering bleb and scarring of the conjunctiva.⁸

**Outcome:**
Visual prognosis is poor.

**Endophthalmitis after intravitreal injection:**
The intravitreal techniques have expanded to include intravitreal anti VEGF agents, antifungals, antibiotics, antivirals, steroids and air as in pneumatic retinopexy.
With the increase in use of these agents, the risk of endophthalmitis is becoming an important concern. Endophthalmitis following injection of anti-VEGF agents have been reported to be in the range of 0.01% - 0.08%. As per a recent study, the incidence of endophthalmitis following intravitreal Bevacizumab and Ranibizumab has been reported to be 1 case per 4500 injections with an overall rate of 0.02%.9,10 The endophthalmitis rates following intravitreal ganciclovir and foscarin; triamcinolone acetate and pneumatic retinopexy have been reported to be 0.0%-0.29%, 0.1-0.87% and 0.1% respectively.11,12

**Microbiological isolates:**
The most commonly identified organism is coagulase negative staphylococcus. Other organisms identified include Streptobacillus parasanguinis, Mycobacterium chelonae, and Streptobacillus species have been reported.13

**Signs and symptoms:**
Same as mentioned for acute endophthalmitis.

**Differential diagnosis:**
In eyes that have received triamcinolone acetonide, the following conditions should be considered.
1. **Sterile endophthalmitis:** inflammation resulting from reaction to the drug, components of the drug vehicle, or sterile microbial toxins in the formulation.
2. **Pseudohypopyon:** triamcinolone acetonide crystals migrating into the anterior chamber can mimic a hypopyon.

**Investigations:**
Same as discussed above.

**Treatment:**
- **Sterile endophthalmitis:** observation with topical steroids.
- **Infectious endophthalmitis:** same as that of acute post operative endophthalmitis

**Prevention:**
This can be prevented by adhering to strict aseptic precautions and cleaning the conjunctival sac with 5% povidone iodine before injection.

**Endophthalmitis following vitrectomy**
Endophthalmitis following pars plana vitrectomy is rare. As per a study performed by Cohen et al, the incidence of endophthalmitis after pars plana vitrectomy is 0.07%. The most commonly identified organism is coagulase negative staphylococci. The dose of intravitreal drugs administered in this case is one tenth of the usual dosage.

As per a recent study, there was no significant difference in the incidence of endophthalmitis following 25 gauge sutureless vitrectomy as compared to 20 gauge vitrectomy.14 Similar results were reported by Barbara et al when comparing the rates of endophthalmitis of 23 gauge vitrectomy with 20 gauge vitrectomy.15

**Post traumatic endophthalmitis:**
The risk of endophthalmitis following open globe injuries is expected to be around 7%.
The risk associated with retained intraocular foreign body is higher, ranging from 11%-30%. Other factors that increase the risk include infected wounds, lens capsule rupture, elderly age group and delayed presentation.16

**Microbiological isolates:**
Bacillus and Staphylococcus are the most frequently encountered organisms in penetrating injuries with retained intraocular foreign body. Other organisms include S. epidermidis, Streptococcus species, Gram-negative organisms and fungi.16,17

**Signs and symptoms:**
The onset may be acute or delayed depending on the virulence of the organism. The signs and symptoms are the same as mentioned above for acute post operative endophthalmitis.

**Investigations:**
The presence of a retained intraocular foreign body (RIOFB) needs to be ruled out. An X-ray of the orbit, an ultrasonography and a non contrast enhanced, thin slice (2/mm) CT scan aid the diagnosis.

According to the National Eye Trauma System (NETS) data, only 3.5% of patients who had primary repair and removal of intraocular foreign bodies within 24 hours after injury developed endophthalmitis as compared to 13.4% who had repair after 24 hours.18

**Management:**
1. Intravitreal vancomycin 1mg/0.1mL and ceftazidime 2.25mg/0.1mL (or amikacin 400µg/0.1mL) in established cases of traumatic endophthalmitis.16,17
2. Early vitrectomy for removal of RIOFB or in cases that fail to respond to initial intravitreal injection within 48 hours.17
3. The role of prophylactic intravitreal antibiotic administration is not fully established.
4. Prophylactic systemic antibiotics are commonly administered. The role, however, has not been fully justified.

The management of RIOFB is beyond the scope of this article.

Outcome:
Post-traumatic endophthalmitis has worse visual outcome than other categories, with only 22% to 42% obtaining a final visual acuity better than or equal to 20/400.

Endogenous endophthalmitis:
As opposed to exogenous endophthalmitis, the source of infection originates within the body. The major risk factors include diabetes mellitus, immunosuppression, intravenous drug abuse, renal failure on dialysis, cardiac disease, malignancy and indwelling catheters. It accounts for just 2-8% of all cases of endophthalmitis.

Microbiological isolates:
The causative organisms may be bacterial, fungal or rarely parasitic.
The organism most commonly isolated in East Asia is Klebsiella species. The spectrum differs in the United States with Gram positive organisms, mostly, Streptococcus being the most commonly isolated organism. The occurrence of fungal endophthalmitis is more frequently seen in West Asia.19-22

Signs and Symptoms:
Similar to exogenous endophthalmitis, findings include decreased vision, eye pain, conjunctival injection, hypopyon, corneal edema, vitritis, and poor fundus view secondary to inflammation. Intra retinal or subretinal nodules, vasculitis and hemorrhages with necrosis and arterial emboli can be seen. Systemic symptoms as fever, chills and rigor may also be seen.

Investigations:
Systemic Cultures: Culture of blood, urine, CSF samples may yield the causative organism. Culture positivity is seen in 75-80% cases.19

Ocular cultures: Positive results are obtained in 36-73% of cases.19

Treatment:
Systemic antibiotics constitute the main stay of treatment. Empirical therapy can be initiated with systemic third generation cephalosporins, vancomycin, ciprofloxacin, and aminoglycosides.19-21 Subsequent therapy can be modified based on the results of culture and sensitivity.
In cases of fungal endophthalmitis, systemic amphotericin-B or oral voriconazole can be administered.
Intra vitreal antibiotics are indicated in cases of progressive vitritis. Vancomycin 1 mg/0.1 mL and ceftazidime 2.25 mg/0.1 mL are recommended for bacterial infections.
In fungal endophthalmitis, intravitreal voriconazole (100µg/0.1 ml) or amphotericin B (5-10µg/0.1ml) are considered effective. The use of intra vitreal steroids has been associated with successful visual outcome in many studies.

Vitrectomy:
A threefold better chance of retaining useful vision and avoiding evisceration or enucleation following vitrectomy has been documented.

Outcome:
The prognosis is poor especially if associated with gram negative or virulent gram positive organisms.19,21

References:


DUANE’S RETRACTION SYNDROME

Manish Malhotra1, Ramesh Murthy2

1. Department of Pediatric Ophthalmology & Strabismus
   L V Prasad Eye Institute, Hyderabad 500034

2. Consultant Pediatric Ophthalmology, Strabismus, Oculoplasty and Ocular Oncology
   Aryan EyeCare Hospital, Hyderabad 500082

Duane’s retraction syndrome (DRS) is a rare, congenital disorder which occurs in approximately 1 in 50 patients with strabismus.1 It was first reported by Hueck in 1879 after which several variations have been described.2 Historically never claimed, but Duane’s retraction syndrome is one clinical entity which is named after Alexander Duane. It has been known to exist for more than a century now and there have been several attempts to explain the cause of the miswiring and the complex eye movements seen in DRS. The purpose of writing this review is to simplify the understanding of this rare disorder and review the best surgical protocols.

Pathogenesis
During 4 to 8 weeks of gestation, injury or maldevelopment of the developing structures of the sixth nerve nucleus leads to redirection of the branches from the third nerve to the lateral rectus. Review of literature shows that specific abnormalities in the anatomy of the extra ocular muscles have also been observed. The lateral rectus has been found to be fibrotic and inelastic.3,4 Co contractions of the medial and the lateral rectus muscle along with the fibrosis of the lateral rectus have been attributed to cause the retraction of the globe.

Various theories have been proposed explaining to explain up shoot or down shoot. An up shoot is an over elevation and down shoot is an over depression of the adducted eye which is seen in 25%-39% of cases of DRS.5,6,7 The up shoots and down shoots can be ascribed to mechanical and innervational causes8. Mechanical theory – the eye remains in the horizontal plane but as it adducts an up shoot or down shoot occurs abruptly due to side slip of the tight lateral rectus muscle. Innervational theory – it says that the eye gradually moves up or down during adduction due to the co innervation of the vertical rectus muscle with the lateral rectus muscle. The upshoots and down shoots seen in DRS are due to the anomalous innervation along with the bridle or the tethering effect of the horizontal muscles, with some amount of slippage of the muscles. This is explained by the fact that with any amount of elevation or depression which occurs outside the primary position there is a change in the horizontal rectus in reference to the rotation of the globe causing the horizontal muscles to become elevators or depressors in addition to horizontal action thus producing up or down shoots.

Surgical, anatomical, cadaveric and electromyographic studies show that it is a complex combination of the mechanical, anatomical, innervational and central nervous system disorders which lead to these clinical findings in DRS.

Associated Anomalies
Literature reveals DRS is frequently associated with various congenital malformations linked to an insult during the embryogenic development. Pfaffenbach et al showed that the patients with sporadic form of DRS had 10-20% times greater risk of congenital malformations,9 which could be divided into:

- Skeletal- involving the palate and vertebral column
- Auricular- involving the external ear, the meatus and the semicircular canals
- Ocular – involving the extra ocular muscles and eyelids and ocular dermoids
- Neural – involving the nuclei of the 3rd, 4th & 6th cranial nerve
- In a study Marshman showed that the patients with DRS had a greater than 50% incidence of having associated skeletal, neural & ocular problems in their first degree relatives.10
Associated anomalies identified in all patients with DRS¹⁰

Skeletal
- High arched palate – 7%
- Webbed toes -6%
- Supernumerary ribs – 3%
- Cleft palate – 2%
- Pigeon chest – 2%
- Club foot – 2%
- Prominent first thoracic vertebra -26%
- Clinodactyly -2%
- Overcrowded dentition – 2%

Auricular
- Deafness – 19%
- Ear tags – 2%
- Malformed auricular cartilage -2%

Ocular
- Ocular dermoids – 3%
- Hypoplastic optic disc – 2%
- Myelinated nerve fibers – 2%
- Latent or manifest latent nystagmus – 2%

Neural
- Fourth nerve palsy – 2%
- Seventh nerve palsy – 2%
- Mobius syndrome – 2%
- Batten disease -2%

Cardiac – 3%

Syndromes
- Crocodile tears – 6%
- Goldenhar syndrome -3%
- Okhiro syndrome 3%
- Holt Oram syndrome- 2%
- Marcus Gunn / jaw winking – 3%

Associated anomalies in first degree relatives of patients with DRS¹⁰

Skeletal (relatives of 10% of patients)
- Webbed toes especially second and third (relatives of 2% of patients)
- Crossover of fifth toe (relatives of 2% of patients)
- Polydactyly (relatives of 2% of patients)
- Absent distal phalanges (relatives of 2% of patients)
- Clinodactyly (relatives of 2% of patients)
- Cleft lip (relatives of 2% of patients)
- Spina bifida (relatives of 2% of patients)
- Flexion deformity of metacarpophalangeal joint (relatives of 2% of patients)

Auricular
- Deafness (relatives of 10% of patients)
- Ear tags (relatives of 7% of patients)
- Malformed auricular cartilage (relatives of 2% of patients)

Ocular
- DRS (relatives of 19% of patients)
- Strabismus (relatives of 10% of patients)
- Dermoids of lid (relatives of 2% of patients)

Cardiac (relatives of 4% of patients)

Classification

The various classifications proposed are as follows-

1. Lyle & Bridgeman classification ¹¹
Type A- Abduction more deficient than adduction, adduction causes globe retraction and narrowing of palpebral fissure.
Type B - Abduction is deficient but adduction is not.
Type C - Abduction is less deficient than adduction, adduction causes globe retraction and palpebral fissure narrowing.

2. Papst classification was based on abnormal co contraction according to EMG studies.¹², ¹³
Abnormal co contraction of
a) medial and lateral rectus
b) superior and lateral rectus
c) inferior and lateral rectus
d) lateral rectus and several other muscles

3. Malbarn classification ¹⁴
Type 1–palsy of abduction
Type 2–palsy of adduction
Type 3–limitation of depression and elevation without impairment of horizontal movements.

4. Huber’s classification¹⁵ (using the above classifications and EMG outcomes), it is the most accepted and widely used classification system.
Type 1-Marked limitation of abduction with minimally defective or normal adduction, retraction of the globe and narrowing of the palpebral fissure in abduction and widening in adduction, EMG showing normal electrical behavior of the medial rectus, with peak impulses on adduction and defective impulses on abduction thus showing paradoxical innervation of lateral rectus. Patient presents with esotropia in primary position and ipsilateral face turn.
Type 2 – Marked limitation of adduction with...
Duane’s Retraction Syndrome

from 40% to as high as 65%.

Laterality:
The predilection for left eye involvement is as high as 67% in unilateral DRS cases as seen in Table 1.6,11,14,15,19,20

Bilateral DRS is seen to range from 10%-24%.6, 11, 14, 15, 19, 20

Types of presentation:
Literature shows Esotropia in primary position was the most common presentation in the patients with DRS. 6, 11, 14, 15, 19, 20

Amblyopia and refractive errors:
It was seen that the average incidence of amblyopia was 3%-25%, Hyperopia was the most evident refractive error seen.6, 11, 14, 15, 19, 20

Approach and Diagnosis
Duane’s retraction syndrome as described by Duane in his landmark article consists of following components:
1) Complete, or less often partial, absence of outward movement of the affected eye
2) Partial or rarely complete deficiency of movement

Epidemiology

Incidence:
The incidence of DRS among strabismus patients varies from 1%-4%.16

Sex distribution:
shows that there is a female preponderance ranging from 40% to as high as 65%.17

Laterality:
The predilection for left eye involvement is as high as 67% in unilateral DRS cases as seen in Table 1.6,11,14,15,19,20

Bilateral DRS is seen to range from 10%-24%.6, 11, 14, 15, 19, 20

Types of presentation:
Literature shows Esotropia in primary position was the most common presentation in the patients with DRS.6, 11, 14, 15, 19, 20

Amblyopia and refractive errors:
It was seen that the average incidence of amblyopia was 3%-25%, Hyperopia was the most evident refractive error seen.6, 11, 14, 15, 19, 20

Approach and Diagnosis
Duane’s retraction syndrome as described by Duane in his landmark article consists of following components:
1) Complete, or less often partial, absence of outward movement of the affected eye
2) Partial or rarely complete deficiency of movement

Table 1. Laterality of eye involvement in DRS

<table>
<thead>
<tr>
<th>Authors/year</th>
<th>No. of patients</th>
<th>Females</th>
<th>Male</th>
<th>Left eye</th>
<th>Right eye</th>
<th>Bilateral</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pfaffenbach et al(1972) 17</td>
<td>186</td>
<td>106(57%)</td>
<td>80(43%)</td>
<td>107(58%)</td>
<td>37(20%)</td>
<td>34(18%)</td>
</tr>
<tr>
<td>Kirkham(1970)18</td>
<td>126</td>
<td>82(65%)</td>
<td>44(35%)</td>
<td>76(60%)</td>
<td>28(22%)</td>
<td>22(18%)</td>
</tr>
<tr>
<td>Isenberg et al (1977)6</td>
<td>101</td>
<td>58(57%)</td>
<td>43(43%)</td>
<td>56(55%)</td>
<td>29(29%)</td>
<td>16(16%)</td>
</tr>
<tr>
<td>O’Malley et al(1982)19</td>
<td>97</td>
<td>60(62%)</td>
<td>37(38%)</td>
<td>54(55%)</td>
<td>26(27%)</td>
<td>17(18%)</td>
</tr>
<tr>
<td>Tredici &amp; Von Noorden(1985)20</td>
<td>70</td>
<td>28(40%)</td>
<td>42(60%)</td>
<td>36(52%)</td>
<td>17(24%)</td>
<td>17(24%)</td>
</tr>
<tr>
<td>Raab et al (1986) 5</td>
<td>70</td>
<td>45(64%)</td>
<td>25(36%)</td>
<td>47(67%)</td>
<td>16(23%)</td>
<td>7(10%)</td>
</tr>
<tr>
<td>Ahluwalia et al (1988)4</td>
<td>20</td>
<td>11(55%)</td>
<td>9(45%)</td>
<td>11(55%)</td>
<td>5(25%)</td>
<td>4(20%)</td>
</tr>
<tr>
<td>Total</td>
<td>650</td>
<td>290(45%)</td>
<td>280(43%)</td>
<td>427(66%)</td>
<td>158(24%)</td>
<td>117(18%)</td>
</tr>
</tbody>
</table>

Table 2. Frequency of types of DRS.

<table>
<thead>
<tr>
<th>Authors/year</th>
<th>No. of patients</th>
<th>Type I</th>
<th>Type II</th>
<th>Type III</th>
</tr>
</thead>
<tbody>
<tr>
<td>O’Malley et al(1982) 19</td>
<td>97</td>
<td>84(87%)</td>
<td>11(11%)</td>
<td>2(2%)</td>
</tr>
<tr>
<td>Tredici &amp; Von noorden(1985)20</td>
<td>70</td>
<td>56(80%)</td>
<td>5(7%)</td>
<td>9(13%)</td>
</tr>
<tr>
<td>Raab et al (1986) 5</td>
<td>70</td>
<td>56(73%)</td>
<td>1(1%)</td>
<td>20(26%)</td>
</tr>
<tr>
<td>Ahluwalia et al (1988)4</td>
<td>20</td>
<td>11(46%)</td>
<td>5(21%)</td>
<td>8(33%)</td>
</tr>
<tr>
<td>Total</td>
<td>257</td>
<td>207(81%)</td>
<td>22(9%)</td>
<td>39(15%)</td>
</tr>
</tbody>
</table>
Duane’s Retraction Syndrome

inward of the affected eye
3) Retraction of the affected eye into the orbit when it is adducted
4) A sharply oblique movement of the affected eye either up and in or down and in when it is adducted
5) Partial closure of eye lid (pseudo ptosis) of the affected eye when it is adducted
6) Paresis or at least marked deficiency of convergence, the affected eye remaining fixed in the primary position while the sound eye is converging

Head posture is also an important sign. A combination of the clinical features should be looked for and not a single sign.

Differential Diagnosis

1) Abducens nerve palsy: It can be differentiated from DRS as in abducens palsy there is normal adduction with preservation of convergence, it is an acquired esotropia where the patient complains of diplopia and it does not have any up shoots or down shoots or any co-contraction of the globe.
2) Moebius syndrome: Also called as congenital diplegia it is characterized by unilateral or bilateral limitation in abduction, adduction and convergence. The patient has mask like facies with associated anomalies of the chest, limbs and tongue; it occurs due to paralysis of the sixth and the seventh nerve, the patient with Moebius syndrome will not have up shoots, down shoots or any retraction of the globe. However it poses to be a diagnostic challenge to patients with bilateral Duane’s.
3) Congenital or infantile esotropia: In a patient with congenital esotropia the deviations are larger than in patients of DRS. A doll’s eye maneuver would elicit any abduction limitation and if this maneuver is not possible, patch test would be helpful in eliciting the abduction deficit.
4) Congenital oculomotor apraxia: Children with congenital oculomotor apraxia have characteristic jerky horizontal head thrusts, but the children have normal vertical and full random eye movements.

Clinical Examination

One should always ask the patient to fix in the best possible position so that the presence or absence of fusion can be determined. Office examination should include the cover uncover test in all the gazes as this affects the management strategy, presence of globe retraction, narrowing of palpebral fissure (one of the least dependable signs in DRS), look for up shoots and down shoots (which is due to the anomalously innervated LR slipping over the globe), co contraction of the globe (which is due to the LR muscle slippage above and below the globe) and presence of patterns. Forced duction and forced generation test – this test helps to differentiate whether a muscle is tight or not; in DRS the muscle is tight due to long standing fibrosis whereas in acquired causes like sixth nerve palsy the muscle would not move voluntarily but would move without any restriction on forced duction test.

Surgical Treatment:

Indications for surgical treatment:

Absolute indications:
1) Presence of deviation in primary position.
2) Presence of abnormal head posture.
3) Severe co contraction of the globe.

Relative indications:
1) Marked retraction of the globe on attempted adduction.
2) Severe down shoot or up shoot on attempted adduction.
3) To increase the binocular diplopia free field.
4) To increase the rotational movement of the abduction in the affected eye.

Surgical options:

1) Horizontal muscle surgery- recession or resection: Recession of the tight and over acting muscle is a simple and an effective procedure, though it does not normalize the motility of the eye but if done meticulously it can improve the abnormal head posture, can reduce the co-contraction of the globe and to some extent can improve the up shoots without breaking the fusional ability. The appropriate muscle to be recessed can be judged by the eye which has most marked limitation in adduction or abduction and whether there is any tropia in primary position. If there is esotropia, the medial rectus of the eye with limitation in abduction is recessed. If exotropia is present, then the lateral rectus of the eye with limitation in adduction is recessed. As far as possible a resection of the horizontal rectus muscle should be avoided because a resection increases the risk of enophthalmos and is more traumatic than a recession. Moreover a resection in DRS produces little improvement in the abnormal head posture but markedly reduces the ocular motility and increases
Duane’s Retraction Syndrome

the up shoots, down shoots and co-contraction of the globe. Thus a resection alone or with a recession can be performed but the surgical outcomes are very variable.

**Transposition of muscle:**
Transposition of the superior or the inferior rectus muscle towards the lateral rectus has been described.\(^{21,22}\) This procedure has been known to increase abduction though adduction is sacrificed to some extent. This procedure may produce diplopia in 10-15% of cases.\(^{22}\) The surgery also has the risk of anterior segment ischemia. Literature search shows that the success rate of transposition surgery is 50-75% in eliminating the abnormal head posture. Failures and over corrections of 30% with non correction of esotropia have been seen.\(^{23}\)

**Posterior fixation suture:**
Posterior fixation suture (Faden) along with recession of appropriate rectus muscle surgery has been advocated. However the posterior fixation suture in the lateral rectus muscle produces gratifying results by eliminating the up shoots, but when done on the contralateral medial rectus in order to correct lateral incomitance was not satisfactory.\(^{24}\)

**Managing up shoots, down shoots, A or V patterns:**
The various types of the up shoots or down shoots produced are\(^{25}\)
A) In straight adduction and in adduction and elevation leading to an up shoot and a V pattern
B) In adduction and in adduction and depression causing a down shoot with an A pattern
C) A combination of the two producing a X pattern
One way of managing an upshoot or down shoot is to recess the lateral rectus muscle, the amount of recession to be done is based on the stiffness of the lateral rectus muscle assessed by forced duction test and whether the muscle is found fibrotic during the surgery. Recessing a very stiff muscle can significantly reduce the overshoots. If the lateral rectus muscle is non fibrotic and non stiff on forced duction test a large recession can achieve a similar result. Another approach to correct the shoots can be a posterior fixation suture on the lateral rectus, with or without similar sutures on the medial rectus, along with appropriate recession of the muscle. Jampolsky gave a unique surgical technique in which he showed that splitting of the lateral rectus muscle into Y configuration helped in reducing the globe retraction on adduction.\(^{26}\) The lateral rectus was split into two, 10 mm posterior to the insertion and was reattached to the sclera, forming a Y pattern, all the patients showed an improvement of head turn, esotropia, upshoot or down shoot.\(^{27}\)

**Adjustable suture technique:**
This technique was recommended by Pressman and Scott who performed a lateral rectus recession with an adjustable suture, though the literature shows that the results were not favorable.\(^{28}\)

**Botulinum toxin:**
It was adopted as one of the non surgical modalities in order to correct esotropia in DRS but it was found to be ineffective.\(^{29}\)

**Complications of surgery:**
The possible complications that can occur in surgery are-
a) Anterior segment ischemia – this is seen usually after transposition of one of the horizontal recti to the lateral rectus along with recession or resection of other muscles, one way to prevent is to perform the recession or resection after 4-6 months of transposition surgery.
b) If a large recession of medial rectus is performed then it can compromise adduction and produce exotropia postoperatively.
c) Other outcomes that can be un-desirous for the patient is persisting abnormal head posture, enophthalmos resulting in an attempt to correct the co-contraction of the globe and limitation of the movements.

**Surgical algorithm:**
Based on primary position, presence of face turn, abnormal head posture, upshoots, down shoots with and without A or V pattern.

**Points to remember**
To identify if the up shoot is innervational or mechanical, if innervational it can be taken care by vertical rectus recession.
Transposition of vertical rectus is a beneficial procedure but the risk of anterior segment ischemia is high.
Points to be kept in mind –
A single recession cannot normalize motility of the eye.
It can correct head posture to some extent. Can correct upshoot or down shoot with minimally risking the vertical tropias. A recession should always be preferred over resection as recession causes less enophthalmos. Resection should also be avoided as it reduces motility while increases upshoots or down shoots and co-contraction.
**Synopsis:**
Duane’s retraction syndrome is a complex clinical entity whose diagnosis is based on the group of clinical signs and not just one, these clinical findings are due to abnormalities in the central nervous system and innervational problems which may be mechanical leading to long standing fibrosis of the muscle, or can be innervational. There is a strong co-relation between DRS and associated congenital anomalies in patients and their first degree relatives as shown in the literature. In patients with DRS there is a high predilection among females, they are usually hyperopic with or with out amblyopia and anisometropia, with typical presentation of Duane’s type -1 being more common. There is a chance that it can be mis-diagnosed as congenital lateral rectus palsy but a good clinical examination with proper checking of movements can clinch the right diagnosis. As far as classification is concerned though the Huber classification is the most widely used one but the clinical variants do occur so these variants can be labeled as atypical Duane’s. If there is an indication of surgery like a significant abnormal head posture, a noticeable ocular deviation, or severe contraction of the globe then the appropriate muscle should be operated upon and it yields a good clinical result. Though it has been more than 100 years since Duane’s described this clinical entity, there is a long way to go to as far as surgical outcome is concerned.

**Figure 1:**
1a: An 8 year old girl presented with left type 1 Duane’s retraction syndrome. She was noted to have a right face turn at presentation.
1b: On primary gaze, the child was noted to have a small angle exotropia.
1c: On attempted left gaze, there was abduction limitation and enlargement of the palpebral fissure.
1d: On adduction, there was narrowing of the palpebral fissure and up shoot of the left eye.

**Figure 2:**
2a: In cases with upshoots or down shoots, Y split of the rectus muscle is an effective procedure. The muscle is split with a teaser hook.
The muscle is then secured with 6-0 vicryl sutures and the 2 ends are secured about 2 mm away from each other at the desired position of recession.

References

IDIOPATHIC MACULAR HOLE

Varun Gogia, Jaideep Tyagi, Atul Kumar, Rajvardhan Azad, Yograj Sharma

Dr. Rajendra Prasad Centre for Ophthalmic Sciences, AIIMS, New Delhi

Idiopathic macular hole, a defect in foveal retina, according to current concepts, is believed to be because of vitreofoveal traction. Patients present with blurred vision or metamorphopsia. Biomicroscopic examination of the macula along with OCT help us to detect presence of macular hole and secondary changes in surrounding tissues. There is strong correlation between macular hole diameter and visual acuity and postoperative prognosis. Vitrectomy is recommended for stages 2, 3 and 4 macular holes, wherein the critical step is removal of perimacular traction i.e posterior hyaloid, ILM and ERM for which chromovitrectomy using vital dyes is proposed.

Full thickness stage 4 macular hole

Red free photograph of stage 4 macular hole

EPIDEMIOLOGY AND RISK FACTORS

A macular hole is a full thickness defect in the foveal retina from the internal limiting membrane (ILM) to the outer segment of the photoreceptor layer. A macular hole was first described by Knapp in 1869 in a patient of blunt trauma but since then ophthalmologists have recognized that this condition occurs more commonly in atraumatic settings. Macular holes have been associated with trauma, laser treatment, retinal vascular diseases, retinal detachment treatment, lightning, electrocution, but the vast majority of them are age related and idiopathic macular holes. Women are affected more than men. Most patients are in the 65-74 years age group.

The risk of full thickness macular hole formation in the other eye is 0-5%. Normal fellow eyes with posterior vitreous detachment appear to be at a very low risk of macular hole development. In fellow eyes with persistent vitreoretinal attachments 11% of fellow eyes developed a full thickness macular hole over a 2 year period of observation.

Pathogenesis

Earlier theories on the pathogenesis of macular hole focused on trauma, whereas more than 80% of macular holes are idiopathic.

Trauma related macular holes are suspected to be related to the transmission of concussive force in the countercoup manner. For idiopathic macular hole theories of cystoid degeneration, coalescence of cystic spaces and the development of full thickness macular hole were proposed. Then many theories of vitreomacular traction acting on the fovea and producing macular holes were proposed.

In 1988, Johnson and Gass proposed the current concept of macular hole pathogenesis that tangential vitreofoveal traction is central to the development of and progression to a full thickness macular hole. Gass proposed that contraction of prefoveal cortical vitreous resulted in tangential traction. This traction resulted in the progression of macular hole through multiple stages of development as described by Gass.
Gass classified the macular hole stages as:

- **Stage a**: Foveal detachment. Macular cyst. Tangential vitreous traction results in the elevation of the fovea marked by increased clinical prominence of xanthophyll pigment. This stage is occasionally referred to as the yellow dot stage and can also be seen in cases of central serous chorioretinopathy, cystoid macular edema, and solar retinopathy.
- **Stage b**: As the foveal retina elevates to the level of the perifoveal, the yellow dot of xanthophyll pigment changes to a donut shaped yellow ring. Persistent traction on the fovea leads to dehiscence of deeper retinal layers at the umbo.
- **Stage 2**: This is the first stage when a full-thickness break in the retina exists. It is defined as a full-thickness macular hole less than 400 µm in size. The full-thickness defect may appear eccentric, and there may be a pseudo-operculum at this stage if there has been spontaneous vitreofoveal separation. These opercula have been examined and found to be vitreous condensation and glial proliferation without harboring any retinal tissue.
- **Stage 3**: A full-thickness macular hole in the retina exists. It is greater than 400 µm in size and is still with partial vitreomacular adhesion/traction.
- **Stage 4**: A full-thickness macular hole exists in the presence of a complete separation of the vitreous from the macula and the optic disc. There is recent evidence, however, that, even in the presence of an apparent posterior vitreous detachment, a thin shell of residual cortical vitreous may still remain and contribute to the macular hole.

Gaudric et al (using OCT) and Johnson et al (using B-scan ultrasonography and vitreoretinal surgical observation) demonstrated initial perifoveal vitreous separation extending upto peripheral portions of the macula with the hyaloid remaining focally adherent to the foveola and optic disc, with abnormally strong persistent vitreofoveal adherence. This theory implicates that anteroposterior traction as the causative factor for macular hole and not tangential traction as suggested by Gass.

Smiddy abnd Flynn presented a unifying theory suggesting that a weakened central fovea may be a primary event, followed by attempted repair by foveal proliferation of Muller and glial cells, and persistent anteroposterior traction at the foveola finally resulting in an irreversible development of full thickness hole.

### Clinical presentation

#### History

Patients with idiopathic macular holes present with a variety of symptoms.

- Initial symptoms include blurred central vision or metamorphopsia. Macular holes may only be discovered when patients cover one eye and notice blurred vision and metamorphopsia in the opposite eye.
- A larger macular hole may produce a central defect, or scotoma, in the central vision of the patient.
- Some patients may be asymptomatic.

#### Examination

- Patients with small, eccentric holes may retain excellent visual acuity in the range of 20/25 to 20/40. However, once a macular hole is well developed or full thickness, the usual range of visual acuity is from 20/80 to 20/400, averaging at 20/200.
- A full-thickness macular hole visualized with direct ophthalmoscopy is characterized by a well-defined round or oval lesion in the macula with yellow-white deposits at the base. These yellow dots probably represent lipofuscin-laden macrophages or nodular proliferations of the underlying pigment epithelium with associated eosinophilic material.
- With biomicroscopic examination, a round excavation with well-defined borders interrupting the beam of the slit lamp can be observed. Other associated changes seen may be the presence of a pseudo-operculum, cuff of fluid, cystic changes at the margins of the hole or an epiretinal membrane.
- The Watzke-Allen test is performed at the slit lamp using a 90 D lens and placing a narrow vertical slit beam through the fovea. A positive test is elicited when patients detect a break in the bar of light that
they perceive.
• The laser aiming beam test also is performed similarly, but this time a small 50-µm spot size laser aiming beam is placed within the lesion. A positive test is obtained when the patient fails to detect the aiming beam when it is placed within the lesion but is able to detect it once it is placed onto normal retina.
• In addition, some slit lamps are equipped with a setting to project a small test object, often a star, onto the fovea.

Imaging studies
• Ocular coherence tomography (OCT) allows the physician to detect the presence of a macular hole as well as changes in the surrounding retina, can distinguish lamellar holes and cystic lesions of the macula from macular holes. Also, the status of the vitreomacular interface can be evaluated.
• Fluorescein angiography (FA) may be a useful test in differentiating macular holes from masquerading lesions, such as CME and choroidal neovascularization (CNV). Full-thickness stage 3 holes typically produce a window defect early in the angiogram and do not expand with time. The arteriovenous phase of the angiogram best demonstrates a granular hyper fluorescent window associated with the overlying pigment layer changes. No leakage or accumulation of dye is observed as opposed to other lesions.
• B-scan ultrasonography may be helpful in elucidating the relationship of the macula to the vitreous; but is not sensitive to distinguish a true macular hole from masquerading lesions. Fellow eye PVD status can be assessed and prognosis explained.

Other Tests
• Amsler grid abnormalities, although sensitive for macular lesions, are not specific for macular holes.
• Microperimetry and multifocal electroretinography show loss of retinal function corresponding to the macular hole with subsequent recovery of function following surgical repair of the hole.

Natural history
The vitrectomy for prevention of macular hole study group 21 reported that 40% of the eyes with stage 1 macular holes randomized to observation progressed to full thickness macular hole over 2 years. This progression took an average time of 4.1 months after diagnosis. The spontaneous resolution of stage 1 lesion is associated with a vitrefoveal separation with foveal reattachment and a normal biomicroscopic appearance or may demonstrate an inner lamellar hole. The majority of stage 2 holes continue to progress to stage 3 or stage 4 holes. There is a strong co-relation between macular hole diameter and visual acuity. Similarly the hole diameter and visual acuity are closely related to duration of symptoms.

Differential diagnosis
• Epiretinal Membrane
• Lamellar Hole
• Cystic Macular Edema
• Central Serous Chorioretinopathy
• Solar Retinopathy
• Macular drusen
• Pseudo-operculum.

Treatment
Indications for consideration of the surgical management of macular holes are based on the presence of a full-thickness defect. Once this defect has developed, the potential for spontaneous resolution is low. Thus, surgical management is recommended with documentation of a stage 2 or higher, full-thickness macular hole. Stage 1 holes and lamellar holes are managed conservatively with observation at this time.

In 1991, Kelly and Wende22 demonstrated that vitrectomy, removal of cortical vitreous and epiretinal membranes, and strict face-down gas tamponade could successfully treat full-thickness macular holes. The overall results of their initial report were a 58% anatomic success rate and visual improvement of 2 or more lines in 42% of eyes. A succeeding report by them showed a 73% anatomic success rate and 55% of patients improving 2 or more lines of visual acuity.23 Present anatomic success rates range from 82-100% depending on the series.

Figure 3 Post operative photographs of macular hole patient.

A prospective, randomized, and controlled series by the Vitrectomy for Treatment of Macular Hole Study Group24 for stage 2, 3, and 4 holes showed that vision was improved in surgically treated eyes compared with observed eyes. However, more frequent adverse
effects were observed in the surgically treated eyes compared to the control eyes, with the most common adverse effects being macular retinal pigment epithelium changes and cataractogenesis.

**Surgical steps**

Some aspects of the surgery may vary, but the basic technique is the same.

• Core vitrectomy is done via a standard 3-port pars plana vitrectomy. Patients with macular hole frequently undergo vitrectomy using smaller gauge vitrectomy systems (i.e., 25 gauge, 23 gauge).

• The critical step appears to be the removal of the perimacular traction. Factors contributing to this traction, such as the posterior hyaloid, the ILM, and coexisting epimacular membranes, need to be addressed. The traction exerted by the posterior hyaloid on the macula should be relieved by either removing just the perimacular vitreous or combining it with the induction of a complete posterior vitreous detachment.

• The removal of ILM is considered to be a contributing factor in the success of macular hole surgeries. ILM peeling may be accomplished via a “rhexis” not unlike that of a capsulorrhexis in lens surgeries. Very fine forceps may be used to peel the ILM from the underlying retina. Care should be taken not to include the deeper layers in the forceps’ grasp, which may further damage the surrounding retinal tissues.

• Epiretinal membranes, if present, also should be removed. Techniques in completing this procedure vary from surgeon to surgeon.

• After careful indirect ophthalmoscopic examination of the peripheral retina for tears, a total air-fluid exchange is performed to desiccate the vitreous cavity. A nonexpansile concentration of a long-acting gas is exchanged for air. Studies have shown that a longer period of internal tamponade equated to a higher success rate.

• Sterile air and varying concentrations of either perfluoropropane or sulfur hexafluoride have been used based on surgeon preference for internal tamponade. The primary difference achieved using different gases is the duration of the gas bubble and, consequently, the amount of internal tamponade achieved within the first several days after surgery. Silicone oil has also been used as an internal tamponade for patients with difficulty positioning or altitude restrictions. However, the use of silicone oil necessitates a second subsequent surgery to remove the oil. Furthermore, the visual results are not comparable to the use of gas tamponade, possibly as a result of silicone oil toxicity at the level of the photoreceptors and RPE.

**20-gauge versus 23-gauge versus 25-gauge vitrectomy systems**

• Smaller gauge vitrectomy systems, with frequently self-sealing wounds, avoid induced astigmatism from suturing sclerotomies, resulting in a more rapid recovery of vision.

• An initial increase in endophthalmitis appears to have been addressed by changing the means of wound construction but may still be considered a disadvantage to small gauge vitrectomy systems.

**Use of vital dyes**

• Indocyanine green (ICG) dye was the first vital dye used for macular surgery. There is possibility of toxicity of ICG dye to the retina and retinal pigment epithelium (RPE).

• Trypan blue has also been used to stain the ILM without the literature suggesting toxicity. On the other hand; trypan blue does not appear to stain the ILM as effectively as ICG dye.

• Brilliant blue dye can be used to stain and remove ILM.

• Triamcinolone acetonide has also been used to facilitate peeling of the ILM. As of now, it is the only surgical adjunct to peeling of the ILM that is FDA approved for use in the eyes.
Face-down positioning

- Traditionally, it has been believed that the shorter the period of face-down positioning, the lower the rate of successful hole closure.
- Vitrectomy with ILM peeling and SF6 gas tamponade for macular holes without face-down positioning achieves favorable anatomical and functional results.35
- The advent of ILM peeling has encouraged a second look at minimal to no face-down positioning. A critical factor is the size of the gas bubble on postoperative day 1 being greater than 70%. However, that there may be more rapid progression of cataract formation with less face-down positioning. Combined phaco vitrectomy for phakic patients to allow less stringent positioning requirements.
- The use of pharmacologic adjuncts, such as a transforming growth factor-beta (TGF-beta) and autologous platelets, to facilitate hole closure has not been proven to have any added benefit as compared to controls such that their use has not gained much popularity.31, 32

Complications

- Surgical complications include retinal detachments, iatrogenic retinal tears, enlargement of the hole, macular light toxicity, postoperative pressure elevation, and cataractogenesis.
- Failure of hole closure/hole reopening: Histopathologic evaluation of specimens from patients with failed initial macular hole surgery demonstrated massive proliferation of cells and newly formed collagen associated with remaining ILM. The residual ILM and the associated collagen fibrils may become the source of persistent traction that prevents macular hole closure.
- Retinal detachment/iatrogenic tears: The rate of postoperative retinal detachment is reported from 2-14%.

Visual field defects

- Visual field defects have been noted following macular hole surgery.
- They are related to dehydration of the nerve fiber layer.
- The rate is reduced by shorter surgical times, lower air flow, and oblique placement of infusion cannulas caused by beveled incisions of smaller gauge vitrectomies.
- Cataract formation
- There is a small risk of hole reopening in the immediate postoperative period following cataract surgery.
- Consideration of prophylaxis versus cystoid macular edema may reduce the risk of hole reopening after cataract surgery.
- A recent retrospective case series suggest that prior or simultaneous cataract extraction may carry a better long-term visual prognosis than cataract extraction following macular hole repair due to the risk of reopening of the hole following cataract surgery.

Case reports exist that describe the use of autologous plasmin for idiopathic and traumatic macular holes. Ongoing clinical trials are evaluating the role of plasmin as a means of “chemovitrectomy.” In these studies, case illustrations have demonstrated resolution of idiopathic macular holes following intravitreal injection of plasmin and no surgical intervention.

Prognosis

In 1994, Wendel reported a series of 235 consecutive eyes undergoing repair of macular holes. In this series, 93% of patients were successfully managed with only a single operation; 60% patients experienced 4+ lines of visual improvement; and 84% patients experienced 2+ lines of improvement. Within this group, 58% of patients achieved 20/40 or better final visual acuity. Multiple other studies cite similar success rates, though vision recovery may be protracted and also further delayed by onset of cataract formation. Use of ILM peeling may further increase the rate of single operation success, though it may potentially slow or affect final vision recovery.

References

Glaucoma suspect describes a person with one or more risk factors that may lead to glaucoma, but this individual does not have definite glaucomatous optic nerve damage or visual field defect. The number of individuals with eye findings that raise a suspicion of glaucoma, usually elevated Intraocular pressure (IOP) or asymmetric optic disc morphology, far exceeds the number of people who have been diagnosed with glaucoma. Increased IOP, family history of glaucoma and thin cornea are the main three contemporary risk factors. Optic nerve, retinal nerve fiber examination, visual fields and pachymetry are crucial in setting target pressures for glaucoma suspects and deciding the appropriate follow up/therapy.

Introduction
A glaucoma suspect is an individual with clinical findings and/or a constellation of risk factors that indicate an increased likelihood of developing POAG. The clinical findings that define a glaucoma suspect are characterized by one or more of the following in an individual with open angles by gonioscopy. Appearance of the optic disc or retinal nerve fiber layer that is suspicious for glaucomatous damage A visual field suspicious for glaucomatous damage Consistently elevated intraocular pressure (IOP) associated with normal appearance of the optic disc and retinal nerve fiber layer and with normal visual field test results A great overlap can exist between findings in patients with early glaucoma and those who are glaucoma suspect without the disease. These patients with suspicious findings are at increased risk of developing glaucoma. The Ocular Hypertension Treatment Study (OHTS) recently demonstrated the rate of participants developing glaucomatous optic neuropathy to be 9.5% in five years or close to 2% per year. The rate was previously estimated to be close to 10 per 1000 (1%) per year. More than 100 million people have elevated IOP. More than 3 million people worldwide are blind secondary to POAG; about 2.4 million people develop POAG each year. Five to 10 million Indians have elevated intraocular pressure (IOP) above 21 mm Hg without evidence of damage (ocular hypertension). Many of these patients are being treated, but the indications for treatment are not clear cut. Many others are glaucoma suspect based on the suspicious appearance of the optic nerve head or other risk factors.

With an early diagnosis and timely therapy, the goal is to prevent glaucoma-related blindness. The goal of identifying and treating patients who are glaucoma suspect is to preserve visual function by monitoring them for the earliest signs of glaucomatous damage. In individuals who are at a high risk of developing glaucomatous damage, preventive measures, including lowering IOP, may be indicated.

Pathophysiology
In general, the cause of glaucomatous optic neuropathy is unknown. The disease affects the individual axons of the optic nerve, which may die by apoptosis, also known as programmed cell death. Multiple theories exist concerning how IOP can be one of the factors that initiates glaucomatous damage in a patient. Two of the fundamental theories include the mechanical and vascular theory.

Mechanical Theory
The mechanical compression theory (Quigley et al., 1981) suggests that elevated IOP causes a backward bowing of the lamina cribrosa, resulting in kinking of the axons as they exit through the lamina pores. This may deprive the axons of neurotrophins or interfere with axoplasmic flow, thereby triggering cell death. Neurotrophins increase RGC survival and may be produced by the RGCs themselves (Perez and Caminos, 1995). Damaged RGC axons are affected by neurotrophin deprivation. With the loss of neurotrophic support of the RGCs, slow death is inevitable. Brain derived neurotrophic factor (BDNF) is one such neurotrophin that can temporarily increase the survival of the RGCs by inhibiting the...
excitotoxicity related cell death. Deficiency of these protective factors may contribute to glaucomatous optic neuropathy.

**Vascular Theory**

The vascular theory (Hayreh et al., 1970) proposes that cell death is triggered by ischemia, whether induced by elevated IOP or as a primary insult. Studies done to evaluate the circulation of the optic nerve using the laser Doppler flowmetry have shown diminished blood flow in the optic nerves of eyes with POAG. (Grunwald et al., 1998).

**Contributory Mechanisms**

Complementary to vascular compromise and mechanically impaired axoplasmic flow, additional pathogenic mechanisms that underlie glaucomatous optic neuropathy include excitotoxic damage from excessive retinal glutamate, peroxynitrite toxicity from increased nitric oxide synthase activity, immunemediated nerve damage, and oxidative stress (Naskar and Dreyer, 2000).

Beyond the neuronal degeneration that results from the primary insult or risk factors in glaucoma, there is an expanding cascade of events called “secondary degeneration”, during which RGCs are gradually damaged from the unfavorable “milieu” of the neighboring degenerating neurons (Schwartz, 2005). Many pathophysiological mediators common to all neurodegenerative diseases, including glaucoma, have been identified, such as increase in glutamate, conformationally altered self proteins, increase in inflammation-associated factors (cyclooxygenase-2 [COX-2], tumor necrosis factor [TNF alpha], nitric oxide [NO]), increase in extracellular matrix proteins and growth-associated inhibitors (myelin-related proteins), oxidative stress, and malfunctioning of local immune cells (microglia). These mediators evoke a response for which the tolerance of the neural tissue is minimal.

**Risk Factors**

The overall likelihood of developing glaucomatous optic neuropathy increases with the number and strength of risk factors, which include the following: Elevated IOP, increasing age, family history of glaucoma, thin central corneal thickness, black race and increased cup to disc ratio. In addition, migraine headache and peripheral vasospasm have been identified in some studies as risk factors for progressing to glaucomatous optic nerve damage. The association between factors such as concurrent cardiovascular disease, systemic hypertension, and myopia and the development of glaucomatous optic nerve damage has not been demonstrated consistently. The relationship between diabetes mellitus and progression to glaucomatous optic neuropathy is unclear.

**IOP**

IOP is a definite and important risk factor for developing glaucomatous damage but is not sufficient for a diagnosis. The prevalence of POAG is higher with increasing IOP. One fifth of patients with ocular hypertension develop field loss within 10 years. Each year, about 2% of all individuals with increased IOP progress to glaucomatous damage. As many as 50% of patients with glaucomatous optic neuropathy or visual field changes have IOP of less than 21 mm Hg on initial evaluation. Some eyes undergo damage at IOP of less than 18 mm Hg; others tolerate IOP of more than 30 mm Hg. A pressure range of 10-21 mm Hg is considered normal; a nongaussian distribution occurs with a skew toward higher pressures. Diurnal variation- 2-6 mm Hg is considered normal, greater than 8 mm Hg variation is suggestive of glaucoma with peak usually occurring in the morning hours. However, the probability of injury increases exponentially with higher IOP (Heijl et al., 2002). Large diurnal fluctuation in IOP is another independent risk factor for glaucoma (Nouri-Mahdavi et al., 2004).

**Age**

Age older than 40 years is a risk factor for the development of POAG. Up to 15% of African-American men are affected by the ninth decade of life (Friedman et al., 2004). Consequently, glaucoma is found to be more prevalent in the aging population, even after compensating for the mean rise in IOP with increasing age. However, the disease itself is not limited to only middle-aged and elderly individuals. The prevalence of POAG is 3-10 times higher among individuals older than 80 years (than people in their 40s).

**Family history**

Family history is a definite risk factor. Heritable susceptibility has been shown. Between 10-20% of patients with glaucoma have a positive family history. Ask about family history of glaucoma, especially in first-degree relatives. Family history of glaucoma in a
sibling is the greatest risk factor, followed by glaucoma in a parent. Also, ask if glaucoma in other family members resulted in vision loss (the individual may have only had ocular hypertension). The Baltimore Eye Survey found that the relative risk of having glaucoma is increased 3.7-fold for individuals who have siblings with POAG. The Rotterdam Eye Study (Wolfs et al., 2000) concluded that the prevalence of glaucoma was 10.4% in siblings of patients with glaucoma and the relative risk of having POAG increased 9.2-fold for individuals with a relative with POAG. High IOP may be the inherited feature of glaucoma, or inherited risk factors independent of IOP may be involved.

**Thin Cornea**

OHTS showed central corneal thickness as a significant predictor of the development of POAG. Patients with a central corneal thickness of less than 555 µm had a 3 times greater risk of developing POAG than patients with a central corneal thickness of greater than 588 µm. CCT is an important risk factor for the development of glaucoma (Gordon et al., 2002). CCT likely influences the measurement of IOP with many tonometers, including applanation techniques (Brandt et al., 2001). Increased CCT beyond the mean of 545 µm causes overestimation of IOP; lower CCT translates into underestimation of the IOP. A thin cornea (for example, 480 µm) may occur with glaucomatous visual field loss despite normal applanation IOP because the measurements are fallaciously low. Conversely, a thick cornea (for example, 620 µm) might occur in an eye with high IOP, normal visual fields and a normal optic nerve because it results in false overestimation of true IOP. Ehlers et al. (1975) extrapolated that applanation tonometry is overestimated or underestimated by approximately 5 mm Hg for every 70 µm difference in measured CCT from mean thickness. It is likely that central corneal thickness may itself constitute an intrinsic risk (or protective) factor for glaucomatous optic nerve damage independent of its ability to affect the IOP measurement.

**Race**

As evident from Figure 1 black races are particularly at risk followed by Hispanics. Indian races are more at risk as compared to Whites. Nearly 50% of all primary glaucomas in adult Indians are angle closure glaucoma types.
**Work Up**

**History**

Glaucoma in one eye is associated with increased risk of future damage in the other eye. Development of visual field defects in an average of 5 years in about 29% of untreated undamaged fellow eyes.

**Retinal vascular occlusion** - In individuals who are susceptible, increased IOP is associated with a risk of developing central retinal vein occlusion (CRVO).

**Current or past use of steroids** - Topical steroids may elevate pressure in certain individuals. Optic nerve damage may be residual from previous increased IOP associated with steroid use. The elevation of IOP is usually seen within a few weeks of starting topical steroids. Homeopathic and Ayurvedic medication arguably have been shown to accelerate glaucoma process.

Systemic history includes the following conditions that have been associated as risk factors for developing glaucoma:

- **Low blood pressure** - Also includes overmedication of systemic hypertension. A previous episode of hypotensive shock, trauma, vascular surgery, or hemorrhage can be significant; it may indicate that optic nerve damage is not progressive but may have been a onetime insult.

- **History of vasospastic disorders** - A higher prevalence of migraine headaches and Raynaud syndrome exists with normal-tension glaucoma.

- **Medications** - In individuals who are susceptible, steroids may cause a rise in IOP. Anticholinergics (antihistamine and antipsychotics) may precipitate angle-closure glaucoma.

- **Cardiovascular disease** may be a factor in low-tension glaucoma.

- **Hypertension**

- **Diabetes mellitus** - Small association, some studies have reported a higher prevalence of increased mean IOP and POAG with diabetes mellitus. Diabetes is a questionable risk factor for glaucoma. The association may be a result of self-selection into the health care system.

**Family history**

Review of old records - Note previous IOP, cup-to-disc ratios, ocular surgery, and past visual fields.

**Examination**

**IOP measurement** - Goldmann type applanation tonometry is the criterion standard. Often in a busy general ophthalmic practice, the tendency is to diagnose glaucoma based solely on a single Schiotz or Non Contact Tonometry. With well understood fallacies associated with both of these tonometers it is a must to perform applanation tonometry or short of that Pneumatonometry, at least more than once before calling the patient as glaucoma suspect. Wherever possible, preferred practice is to check IOP numerous times before initiating treatment to assess diurnal variation. Abnormally thick corneas may result in artificially high IOP measurements by applanation tonometry, while abnormally thin corneas may result in artificially low IOP measurements.

**Corneal pachymetry** - Must be done in all glaucoma suspects. Important for two reasons - Influences IOP measurement by all tonometers including Goldmann Applanation as discussed above. Also following OHTS study it is an independent risk factor for developing glaucoma (Figure 3). It can be performed optically with an attachment to slit lamp but now popular is the ultrasonic pachymeter. Taking an average of three central corneal readings is good practice.

**Slit lamp examination** - Look for signs of secondary causes/risk factors of glaucoma:

- **Corneal endothelium** - Krukenberg spindle, Keratic precipitates, Pigmentary changes on endothelial cells

- **Anterior chamber angle depth** - Identification of narrow occludable depth

- **Iris** - Mid iris spoke like transillumination defects seen in pseudoexfoliation and pigment dispersion. Dandruff like material on pupillary margin and on lens capsule (pseudoexfoliation) and neovascularization.

- **Pseudoexfoliation** - Increases with advanced age Pigment dispersion -25-50% risk of developing glaucoma

- **Gonioscopy** - Perform on all patients who are glaucoma suspect, and repeat it periodically. It is especially important in the following cases:
  - The chamber is shallow, and IOP raised.
  - Angle problems (e.g., hyperopia, symptoms of sub acute/acute angle-closure glaucoma, narrow angle) are evident.
  - The patient is diabetic.
  - Vein occlusion is present.
  - A history of ocular trauma exists.

Evaluation of angle depth, signs, and risk factors for secondary glaucoma

- Narrow-angle depth
- Angle recession
Evaluation of the optic nerve head

The best examination method is a slit lamp combined with a 78-D, or 90-D lens through a dilated pupil. Advantages are high magnification, stereoscopic view, and excellent illumination. Special attention should be paid to contour and color of optic nerve head (ONH).

Normal vertical cup-to-disc ratio is 0.3. In a normal rim, the inferior portion is thickest, followed by the superior rim, then nasal and last temporal (ISNT rule). Patients with myopia have larger eyes and larger discs and cups. Assessing optic nerve damage in small optic discs with minimal cupping may be difficult. Large optic discs may appear pathologic when they actually show only physiologic cupping.

- Heavy pigmentation of TM (pigment dispersion)
- Patchy pigmentation of TM (pseudoexfoliation)
- Hemorrhage
- Inflammatory changes

- Angle closure
- Peripheral anterior synechiae
- Neovascularization of the angle

Signs of early glaucomatous damage can be subtle (Figure 4).
- Generalized enlargement of cup, progressive enlargement of cup, vertical elongation of cup, cupping to rim margin.
- Focal thinning or notching of neuroretinal rim, Thinning of inferior-temporal rim, acquired change in disc rim appearance
- Superficial splinter hemorrhage
- Asymmetry of cupping or rim between 2 eyes
- Nerve fiber layer dropout or deficit, acquired change in retinal nerve fiber layer
- Exposure of lamina cribrosa
- Peripapillary atrophy
- Baring of circumlinear vessels

Figure 3: Risk for developing primary open glaucoma varies with central corneal thickness. The numbers and percentage of participants in the observation group who developed primary open-angle glaucoma (median follow-up, 72 months) are indicated below each bar. Participants are grouped by baseline intraocular pressure of ≤23.75 mm Hg, >23.75 mm Hg to ≤25.75 mm Hg, and > 25.75 mm Hg and by central corneal thickness measurements of ≤555 μm, >555 μm to ≤588 μm, and >588 μm. These percentages are not adjusted for length of follow-up. From: Gordon MO, Beiser JA, Brandt JD, Heuer DK, Higginbotham EJ, Johnson CA, Keltner JL, Miller JP, Parrish RK 2nd, Wilson MR, Kass MA (2002) The Ocular Hypertension Treatment Study: baseline factors that predict the onset of primary open-angle glaucoma. Arch Ophthalmol 120: 714–20.
Search for other abnormalities that may account for the visual field defect.
- Tilted disc
- Disc drusen
- Optic pits
- Retinal disease
- Optic atrophy

**Document the appearance of the optic nerve head.**
- The preferred technique - Baseline stereo disc photographs for future comparison
- Detailed description and drawings
- Automated techniques
- Scanning lasers ophthalmoscopy (HRT)
- Polarimetry (GDx)
- Optical Coherence Tomography (OCT)

Figure 4: Glaucomatous Optic Neuropathy.  
- a) Vertical oval cup. ISNT rule not met.  
- b) Wedge shape RNFL defects on red free photography
- c) Advanced glaucoma changes with asymmetry

Figure 5: OCT is a preperimetry tool for diagnosis and follow up of glaucoma suspects. In this scan Left eye is definitely glaucomatous and in right eye, OCT helped diagnose glaucoma and treatment was instituted. Note the superior RNFL defects and corresponding inferior notch in right eye.

**Evaluation of the retinal nerve fiber layer** - Look for nerve fiber layer defects/dropout.

**Techniques**
- Ophthalmoscopic examination with red-free (green) filter
- Stereo color photography and Red-free photography
- Instruments for retinal nerve fiber layer analysis: GDx, HRT and OCT
**Visual field testing**
- Absence of visual field defects does not always suggest absence of glaucoma.
- As many as 50% of optic nerve fibers in a single optic nerve may be damaged before visual field defects are found by perimetry.

**Interpretation of visual field testing**
- Use comparable tests when comparing fields. For example, one cannot directly compare Swedish interactive threshold algorithm (SITA) with Fastpac or HVF 30-2 threshold testing.
- If a field defect is detected, ensure that it is reproducible by doing repeat field analysis.
- The abnormal points should be contiguous, paralleling the pattern of the nerve fiber layer in an arcuate pattern respecting the horizontal midline.

![Figure 6: Showing a classical reproducible nasal sign of early open angle glaucoma](image)

The greater the abnormal points and the deeper the defects, the more likely it represents a true scotoma. The standard testing strategy used by many ophthalmologists in past evaluations has been HVF 30-2 or 24-2 traditional threshold testing with statistical analysis. Newer strategies are:

- **Humphrey Fastpac** - Requires less testing time; decrease in precision of threshold algorithm estimate.
- **Swedish Interactive Test Algorithm (SITA)** - Reduces testing time by nearly half without sacrificing accuracy. Less inter individual variability occurs, and gray scale printouts easier to interpret.
- **Short wavelength automated perimetry (SWAP)** - Uses blue target on a yellow background to isolate those visual pathways that are believed to be damaged selectively in early glaucoma. Many studies suggest that it is capable of earlier detection of glaucomatous defects, which may be useful in those patients who are glaucoma suspect and are at a high risk. More difficult to perform for the patient and requires longer testing time.
- **Frequency-doubling technology (FDT/FDP) perimetry** - Uses a coarse striped grating of rapidly alternating dark and light bands. Takes 4-5 minutes for each eye; screening test takes less than 1 minute. Potential role exists in screening for early glaucoma.

**Differential Diagnoses**
- Glaucoma, Angle Closure, Chronic, Plateau Iris
- Glaucoma, Angle Recession
- Glaucoma, Aphakic And Pseudophakic
- Glaucoma, Drug-Induced
- Glaucoma, Pseudoexfoliation
- Glaucoma, Juvenile
- Glaucoma, Uveitic
- Glaucoma, Low Tension
- Optic Neuropathy-Anterior Ischemic, Toxic/Nutritional
- Glaucoma, Neovascular
- Optic Neuropathy, Compressive

**Other Problems to Be Considered**
- Physiologic cupping
- Intermittent angle-closure glaucoma
- Optic atrophy (e.g., chiasmal tumors, syphilis, ischemic optic neuropathy, drugs, retinal vascular or degenerative disease)
- Previous ischemic damage to the optic nerve (e.g., previous hypotensive event, vascular surgery, hemorrhage, shock)
- Hematologic disease (e.g., anemia, polycythemia vera)
- Congenital optic nerve abnormalities (e.g., myopic discs, coloboma, optic nerve pits)

**Management**

**Target Intraocular Pressure**
In managing the glaucoma suspect, the ophthalmologist strives to achieve a stable range of measured IOPs deemed likely to protect against optic nerve damage. The estimated upper limit of that range is considered the “target pressure.” The target pressure will vary among patients, and in the same patient it may vary during the clinical course. For glaucoma suspects...
not being treated, the target pressure can be viewed as that pressure over which treatment would be recommended (i.e., the threshold for the initiation of treatment).

If therapy is initiated, the ophthalmologist assumes that the measured pretreatment pressure range is that which places the optic nerve at risk for damage. The OHTS (which limited enrollment to patients with an IOP of 32 mmHg or below) used a target pressure 20% lower than the mean of several baseline IOP measurements and 24 mmHg or below. This seems an appropriate initial goal. At present, there is not a prior way to determine the pressure below which optic nerve damage will be prevented in any particular patient. The initial target pressure is an estimate based on experience and judgement of the glaucoma specialist. Current IOP and its relationship to the target IOP should be evaluated at each visit. The status of the optic nerve in relation to glaucoma changes is good start point to set target pressure. With each additional risk factor for glaucoma that the individual patient may have the target should be reduced appropriately. Failure to achieve and maintain a target pressure should trigger a reassessment of the treatment regimen in light of the potential risks and benefits of additional or alternative treatment. In a glaucoma suspect, a definite deterioration in optic nerve structure or visual field (i.e., conversion from glaucoma suspect to glaucoma patient) indicates that the target pressure should be reduced and the patient managed as glaucoma patient.

Therapeutic Choices
If the decision to begin treatment is made, the choice of initial therapy depends on numerous considerations, and discussion of treatment with the patient should include appropriate options. In most instances, topical medications constitute effective initial therapy. The prostaglandin analogs are the first choice monotherapy and the beta adrenergic antagonists are often used second choice for lowering IOP in patients with glaucoma. Other supplemental agents used include alpha2 adrenergic agonists, topical and oral carbonic anhydrase inhibitors, and parasympathomimetics. To determine the effectiveness of topical therapy, it may be useful to begin by treating only one eye and comparing the relative change of the IOP in the two eyes at follow-up visits. If a drug fails to reduce IOP, it should be replaced with an alternate agent. If a single medication is ineffective in lowering IOP to target levels, combination therapy should be tried. Generally, beyond three topical medications it is good practice to consider Laser Trabeuloplasty as supplement before considering filtering surgery or drainage devices.

Follow-up
The frequency and the composition of follow-up evaluation depend on the age of the patient, the level of elevation of IOP, the appearance of optic nerve head cupping, a family history of glaucoma, the presence of additional risk factors, and the stability of the patient’s clinical course.

In general and depending on the patient’s risk factors, check IOP every 3-12 months. If the patient is a low-tension glaucoma suspect with normal IOP but suspicious optic nerve head cupping, conduct a diurnal assessment of IOP.

Perform visual field examinations every 6-12 months. If a new visual field defect is suspected, the test should be repeated to ensure that the defect is reproducible. Gonioscopy and optic nerve head evaluation are generally performed annually.

Baseline documentation, such as stereo disc photographs, should be obtained for future comparison to objectively evaluate any possible subtle progression. In selected patients, some ophthalmologists prefer to obtain this documentation yearly for detailed comparison.

If the target IOP has been achieved and there is no progression of damage, it is recommended (American Academy of Ophthalmology: Preferred Practice Patterns, 2005b) to follow up with ONH exam and visual field testing at 6 to 18-month intervals. On the contrary, if there is progression of damage and the target IOP has not been achieved, more frequent testing is required until stability is achieved.

Conclusion
Most patients who are glaucoma suspect do not develop glaucomatous optic nerve damage and/or visual field loss. Overall, about 2% of individuals with ocular hypertension develop glaucoma per year. The risk is higher for patients with additional risk factors. Glaucoma causes silent damage; follow-up care is essential to exclude any progressive change over time that may warrant treatment. Left untreated, patients with optic nerve damage may progress, resulting in progressive loss of nerve fibers and eventually total optic nerve atrophy and irreversible blindness.
References
Evaluation of the visually inattentive infant

Sumit Monga

Associate Consultant, Pediatric Ophthalmology Service,
Shroff Eye Centre, Kailash Colony, New Delhi. Email: drsumitmonga@gmail.com

It is not unusual in ophthalmic practice that one encounters a visually inattentive infant who is unable to fix at parents’ faces and is not able to respond appropriately to the immediate surroundings. The treating ophthalmologist is faced with an arduous task of not only having to unravel the mystery of the child’s inappropriate responses but also to predict a reasonable visual prognosis of the child. This requires a sequential multi-disciplinary assessment.

The visually inattentive infant is unresponsive to visual stimuli without auditory accompaniments. The child does not respond to parents’ smiles and faces (Figure ).

Blindness or visual impairment in infants could be due to severe congenital malformations of ocular structures or of parts of the brain that sub serve vision. Infants with variants of albinism, congenital stationary night blindness (CSNB), monochromatism and Lebers’ congenital amaurosis (LCA) may present with apparent blindness in infancy. Delays in visual maturation may occur in otherwise healthy and normal infants.

The diagnosis and severity of involvement determines the future prospects for the infants’ future vision range from nearly normal to complete blindness. Hence, there is a need to secure a reliable diagnosis as soon as possible. This requires close coordination between the ophthalmologist, genetici st and the pediatrician.

Evaluation of a visually inattentive child

The important historical points include the family history, with special reference to consanguinity and history of blindness in other family members. The birth history, including occurrence of perinatal hypoxia, gestational age, birth weight, and possible presence of retinopathy of prematurity should be determined. The presence of other associated medical problems may provide important clues, e.g. tachypneic infant with poor vision may be a case of Joubert’s syndrome, which is also characterized by presence of hypotonia, ataxia, oculomotor apraxia, seizures and severe developmental delay.

Some of the eye conditions, causing visual inattentiveness in infants, like high refractive errors, bilateral microphthalmia, congenital cataracts, optic nerve hypoplasia, foveal hypoplasia associated with albinism, choroidal colobomas involving the maculae and optic nerve heads, are conspicuous ophthalmoscopically. When present, such anomalies account for the abnormal visual behavior of the infant and predict the guarded visual potential in future. Other anomalies of retinal function like CSNB and monochromatism are invisible ophthalmoscopically but can be diagnosed in infancy when appropriate tests are done (Table ). Still other infants have diseases of the retina, visual pathway or the brain that severely or permanently impair the vision.
Some of the eye conditions, causing visual inattentiveness in infants, like high refractive errors, bilateral microphthalmia, congenital cataracts, optic nerve hypoplasia, foveal hypoplasia associated with albinism, choroidal colobomas involving the maculae and optic nerve heads, are conspicuous ophthalmoscopically. When present, such anomalies account for the abnormal visual inattentiveness of the infant. Rule out media opacities, high refractive errors, and gross retinal pathologies.

Foveal Hypoplasia
Endocrine evaluation
ERG evaluation
Neuroimaging, VEP assessment

Optic nerve hypoplasia
Optic atrophy

Normal Fundus or pigmentary retinopathy
Optic atrophy

Examine for albinism, aniridia, rod monochromatism
Endocrine evaluation
ERG evaluation
Neuroimaging, VEP assessment

ERG abnormal without neurological problems
Rule out LCA, Rod monochromatism, CSNB

ERG abnormal with neurological problems
Rule out peroxisomal disorders, neuronal ceroid lipofuscinosis

ERG normal
VEP normal
Observe for delayed visual maturation

VEP abnormal
Neuroimaging, lactic acid, pyruvate estimation

Algorithm for working up a visually inattentive infant

Table 1: Important Diagnostic Parameters in Ophthalmoscopically inconspicuous causes of visual impairment in infants

<table>
<thead>
<tr>
<th>Cause of visual Impairment</th>
<th>Important Diagnostic features</th>
</tr>
</thead>
<tbody>
<tr>
<td>Leber’s Congenital Amaurosis</td>
<td>Poor pupillary response to light, hyperopia, Decreased rod- and cone- mediated ERG responses</td>
</tr>
<tr>
<td>CSNB</td>
<td>Paradoxical pupillary response to dark, myopia, Small b- wave amplitudes on ERG</td>
</tr>
<tr>
<td>Monochromatism</td>
<td>Paradoxical pupillary response to dark, Hyperopia, Decreased cone- mediated ERG responses</td>
</tr>
<tr>
<td>Cortical Visual Impairment</td>
<td>Normal pupillary response and refraction, normal ERG, subnormal VEP, CT/MRI to document cranial pathology</td>
</tr>
<tr>
<td>Delayed Visual Maturation</td>
<td>Normal pupillary response and refraction, normal ERG and VEP, normal neuro-imaging</td>
</tr>
</tbody>
</table>
Leber’s Congenital Amaurosis (LCA) and Early Retinal Degenerations

LCA actually represents several different diseases that include clinical features of visual impairment before 6 months of age, poor pupillary reactions, moderate to high degrees of hyperopia and markedly attenuated or absent electroretino-graphic (ERG) responses. Some infants with LCA have associated brain anomalies and mental retardation. The fundus picture is very variable from a normal appearing fundus to subtle abnormalities like mild attenuation of the retinal vasculature or tiny clumps of bony spicules. (Figure 2). The ERG amplitudes are markedly attenuated (<10% of the normal amplitude for age) in infants with increasing age. Conditions other than LCA can cause retinal degeneration, visual impairment, and attenuation of an infant’s ERG responses. These include neuronal ceroid fuscinosis, mitochondrial and peroxismal disorders and rare metabolic conditions such as methylmalonic academia that become apparent as the child’s general medical problems are pursued.

Monochromatism

Rod monochromatism (achromatopsia) is a stationary, inherited retinal disorder of cone function. Monochromats have deficits in color vision, reduced visual acuity, nystagmus and aversion to light. In infant monochromats, invariably, there is aversion to light but no tearing, photophobia or corneal changes that accompany congenital glaucoma. The diagnosis is aided by the presence of paradoxical pupillary response to darkness; immediately after change of light to dark, the pupil diameter becomes smaller rather than larger. Normally, ERG demonstrates absent or attenuated photopic (cone mediated) responses. (Figure 3). Infants with monochromatism have normal fundus, although irregular distribution of macular pigment and slight optic disc pallor may occur (Figure 3).

Congenital stationary night blindness (CSNB)

It is a retinal disorder with a presumed defect in neural transmission between the photoreceptors and second order retinal neurons. It is characterized by stable, non-progressive defects in scotopic visual sensitivity, visual acuity and often nystagmus. The fundi appear normal but some patients show pale or tilted optic discs. In infancy, the diagnosis of CSNB depends largely on finding of relative attenuation of the b-wave amplitude with normal or nearly normal a-wave amplitudes. Other features that may be present are paradoxical pupillary response to darkness, nystagmus and myopia.

Albinism

Infants with albinism are typically slow to see, often appearing blind in early postnatal weeks. In the early weeks, wandering eye movements may be present, which progress to nystagmus and oculomotor instabilities. Diaphanous irides and foveal hypoplasia are other features of an albino’s eyes. The misrouting of the visual pathways can also be documented by using the VEP techniques.

Delayed visual maturation (DVM)

This entity encompasses infant who have normal eyes, visual pathways and brain. Typically, these infants behave as blind up to the age of 5 to 6 months, despite good pupillary responses and normal fundi. The parents often report that vision mediated behavior characteristically turns on suddenly in these infants. There is a frequent association of delayed visual maturation and premature birth.
Cortical Visual Impairment (CVI)

It is characterized by bilateral loss of vision with a normal ocular examination and an intact pupillary light reflex. Decreased vision is due to injury or dysfunction of either optic radiation or visual cortex. Most children with so-called cortical blindness have some degree of residual vision. In infants, hypoxia is the most common cause of CVI in infants. At preterm ages, the hypoxic or ischemic injury typically involves the periventricular white matter (periventricular leukomalacia). By term or later in infancy, the “watershed” zones (border zones at the end of arterial vascular distributions) are affected. The other common causes of CVI in infants are seizures, perinatal stroke, brain malformations, hydrocephalus, brain tumors, meningitis, metabolic disorders, drugs like anticonvulsants, cyclosporine, poisons like lead, CO, Nitrous oxide, head trauma etc.

The absence of nystagmus helps in differentiating CVI from the anterior visual pathway disorders. Pediatric patients with CVI have short visual attention span; do not look directly at faces and objects and exhibit light gazing. At times, they may show aversion to light. The ERG is usually normal in patients of CVI and may be used to rule out concomitant retinal diseases. The use of VEP may or may not correlate with the severity of visual loss or with the outcome. Neuroimaging including the CT and MRI scans can show the extent of damage to the CNS and may help in determining the prognosis of the visual potential in these children. The ophthalmologist should work in tandem with the pediatric neurologist to ensure that the visual rehabilitation process of the ill-fated baby is carried in a meticulous manner.

References

IMPORTANT ASPECTS OF PAEDIATRIC CATARACT SURGERY

Abhay R Vasavada, Sajani K Shah, Viraj A Vasavada, Vaishali A Vasavada
Iladevi Cataract & IOL Research Centre, Raghudeep Eye Clinic, Ahmedabad, India

Introduction
Pediatric cataract surgery is a complex issue best left to surgeons who are familiar with its long-term complications and lengthy follow-up. Treatment is often difficult, tedious and requires a dedicated team effort, the most important members being parents. At one point lenscetomy was the standard treatment for such cases. However, in recent times, small incisions, anterior capsulorhexis, bimanual irrigation/aspiration and primary posterior capsulectomy with or without anterior vitrectomy have become acceptable treatment options for pediatric cataract. Intraocular lens implantation has become the standard of care for the optical rehabilitation of children with cataracts from the toddler age group and up.

Anterior Capsule Management
The anterior capsule in children is very elastic, and therefore it may be difficult to perform a controlled manual continuous curvilinear capsulorhexis (CCC). However, it provides a very strong margin that is resistant to tearing. Manual ACCC, therefore, remains the gold standard and should be accomplished whenever possible. Difficulties of performing manual CCC in infantile eye led researchers and surgeons to search for alternative methods to open the anterior capsule in children. Alternatives to manual CCC currently available include vitrectorhexis, radiofrequency diathermy with a Fugo plasma blade, the two incision push pull technique, and the four incision technique. Wilson et al. analyzed pediatric anterior capsulotomy techniques using porcine model and found that manual capsulorhexis produced the most extensible capsulotomy with most regular and stable edge. In eyes with poor anterior capsule visibility, trypan blue (0.0125%) should be used to stain the anterior capsule. The shape, size and edge integrity of anterior capsulotomy are very important for long-term centration of the IOL. (Figure 1)

Management of the Posterior Capsule and Anterior Vitreous Face
The most frequent and significant problem following pediatric cataract surgery is Visual axis opacification (VAO) (Figure 2). Maintenance of a clear visual axis remains a high priority when planning management of the posterior capsule in the amblyogenic age range. An important question that remains is when should the posterior capsule be left intact? Primary posterior capsulectomy (with or without anterior vitrectomy) are considered “routine surgical steps”, especially in young children. A manual PCCC offers the advantage of a controlled size and strong edges but is more difficult to perform. Many investigators have observed that performing manual PCCC is technically difficult. A potential complication associated with this procedure is the disruption of the anterior vitreous face (AVF). The signs of AVF disruption vary from subtle to obvious- the presence of vitreous strands in the anterior chamber, the attachment of the vitreous to the capsular flap and distortion of the capsulorhexis margin. Recently we described a technique to render the vitreous visible to the anterior segment surgeon using intracameral injection of 0.1 ml of a suspension of preservative free triamcinolone acetonide (Aurocort, Aurolab India). Preservative-free Triamcinolone can

Preferred Practice Patterns
While dramatic advances have occurred in this field over the past 10 years, some technical aspects of surgery, changing refraction and functional outcome continue to pose significant problems. Primary management of the posterior capsule is mandatory depending on the age of the child at surgery. With refinements in surgical techniques, improvisation of IOLs and better understanding of growth of the pediatric eye, in the coming years IOL implantation is likely to become an established mode of treatment of children even in the youngest age group.
be injected at three time points during the surgery. First injection, on completion of PCCC, for visualizing the anterior vitreous face as well as the presence and extent of vitreous disturbance. Second injection, after performing a vitrectomy, to ensure that there are no superficial vitreous strands remaining, which may have otherwise gone unnoticed. Third injection, after IOL implantation and removal of the residual OVD. Additional anterior vitrectomy can be performed if vitreous strands were identified in the anterior chamber.

**Intraocular Lens (IOL) Implantation**

One of the most important preoperative considerations is whether to implant an IOL or not. Surgeons should be prepared for the common question “Would you implant an IOL if this was your child?” The capability of the IOL to offer constant visual input is an important advantage for a better visual outcome after pediatric cataract surgery. Use of IOL provides at least a partial optical correction at all times. Because of the advantage it offers, primary IOL implantation has slowly gained acceptance for the management of childhood cataracts. However, as of now, use of IOL remains controversial for the management of infantile cataract. The important concerns about primary IOL implantation during infancy are the technical difficulties of implanting an IOL, selecting an IOL power, and the higher rate of visual axis opacification (VAO) and complications like glaucoma. At present, only adult sized IOLs are available, which are often difficult to implant in these small eyes and may cause complications over the long run.

The size of the posterior capsulorhexis should be large enough to provide a clear central visual axis, but smaller than the IOL optic, so as to allow stable in-the-bag IOL fixation. (Figure 3) Even if the surgeon is not planning to implant an IOL, it is important to leave behind sufficient anterior and posterior capsular support at the time of cataract surgery to facilitate subsequent IOL implantation. The common practice is to perform posterior capsulotomy and anterior vitrectomy before IOL implantation if the limbal approach has been used, whereas, if a pars plana vitrectorhexis is performed, it is done after the IOL is implanted. Further there is no agreement on whether the IOL should be implanted before or after the primary posterior capsulotomy. Some surgeons perform a pars plicata capsulotomy and vitrectomy with the vitrectome after implanting the IOL in the bag. Both PMMA and hydrophobic acrylic foldable IOLs have been widely used in pediatric eyes. However, several studies have now shown that hydrophobic acrylic IOLs are preferable as they offer better uveal biocompatibility and decreased incidence of VAO, with hydrophobic acrylic IOLs causing a delayed onset of PCO. For bilateral cataract during first year, aphakic glasses and/or contact lens use may be a reasonable option; however, for unilateral cataract, we are truly equipoised between whether or not to offer primary IOL implantation at the time of infantile cataract surgery. A large randomized clinical trial - the Infant Aphakia Treatment Study (IATS) is currently underway to compare primary IOL implantation to contact lens correction in children undergoing unilateral cataract surgery in the first six months of life.

**Glaucoma**

Glaucoma is a recognized complication of pediatric cataract surgery. Despite improved surgical techniques, the incidence of glaucoma following successful cataract removal remains high. A significant number of surgeons regard aphakia as a cause of glaucoma. However, the glaucoma occurring postoperatively may be better described as “glaucoma in aphakia” and “glaucoma in pseudophakia”. The most common type of glaucoma that develops following congenital cataract surgery is open-angle glaucoma. The risk factors include age at the time of surgery, pre-existing ocular abnormalities, type of cataract and the effect of lens particles, lens proteins, inflammatory cells and retained lens material. In addition microcornea, secondary surgery, chronic postoperative inflammation, the type of lensectomy procedure or instrumentation used, pupillary block and the duration of postoperative follow-up have been found to influence the likelihood of glaucoma after pediatric cataract surgery. Undergoing lensectomy at a young age, especially in the first year of life, may be a risk factor for the development of glaucoma. It has been suggested that the immaturity of the developing infant’s angle leads to increased susceptibility to secondary surgical trauma. Hence some surgeons feel that it is prudent to consider delaying surgery until the infant is 4 weeks old in bilateral cases. Glaucoma can occur at any time after congenital cataract surgery. Therefore, patients who have undergone congenital cataract surgery should be monitored for glaucoma throughout their lives.
Visual Rehabilitation and Amblyopia Management

The visual rehabilitation of a child following cataract surgery is of utmost importance. If the child is left aphakic bilaterally, aphakic glasses or contact lenses may be prescribed. Contact lenses have the advantage of providing vision free of prismatic aberrations, a less magnified image and a wider field of vision. On the other hand, they are often difficult to fit in very young children, are expensive, have a potential risk of infection and high rate of loss. However, with unilateral aphakia, contact lens fitting becomes almost mandatory.

Even when primary IOL implantation is performed, optical correction of residual refractive error should be given with changing refraction. Bifocal glasses should be advised once the child approaches school going age.

Inspite of a successful cataract surgery and appropriate optical correction, visual development may be limited by amblyopia. Untreated amblyopia in childhood may be a cause of permanent visual loss. Progression of amblyopia in early childhood is inversely proportional to the child’s age: the younger the child, the faster the progression. Most cataractous patients are predisposed to all three causes of amblyopia – pattern deprivation, strabismus, and optical defocus. Even when optically corrected, these eyes are focused at a fixed distance and continue to optically defocus promoting amblyopia, especially if the other eye is normal.

Although refractive correction alone can result in improved vision in anisometropic amblyopia, it is generally believed that most post-cataract cases will need amblyopia treatment. Amblyopia therapy requires a high-level of effort. First and foremost, any residual refractive error should be corrected in the form of spectacles or contact lenses. Non-compliance is a major hurdle during the treatment process. Occlusion should be prescribed over the better eye. The parents should be advised to make the child spend at least part of the patching period each day performing near-vision eye–hand coordination activities, such as crafts, coloring, tracing, cutting out shapes with scissors, completing workbook games (connect the dots, hidden pictures, and word finders), computer-generated or video games, computer or internet-based activities, reading, or writing.

NEWER APPROACHES

Sealed Capsule Irrigation

Maloof and associates have designed a sealed capsule irrigation device (Perfect Capsule TM) that can help to selectively irrigate the capsular bag. This may help pediatric cataract surgeons to eliminate or delay VAO by using chemicals to selectively target lens epithelial cells through this device.

Manual PCCC via Pars plana approach

Vasavada and coauthors recently introduced a technique of performing manual PCCC via pars plicata. After implantation of the IOL in the capsular bag all the incisions are sutured with 10-0 nylon and residual provisc is left in the anterior chamber. The pars plicata entry is made 1 - 1.5 mm behind the limbus and an initial puncture is made in the center of the posterior capsule and later a coaxial capsulorhexis forceps was introduced and a flap is generated .The edge of the flap is grasped and then re-grasped every 2 clock hours fashioning the PCCC in a clockwise manner.

Bag-in-the-lens implantation

Tassignon and colleagues reported the outcome of a surgical procedure they called ‘bag-in-the-lens’ in pediatric cataractous eyes. In this technique, the anterior and posterior capsules are placed in the groove of a specially designed IOL after a capsulorhexis of the same size is created in both capsules. The principle behind this IOL design is to ensure a clear visual axis by mechanically tucking the two capsules into the IOL, thereby preventing any migration of proliferating lens epithelial cells.

Heparin in irrigating solution

Heparin has been used in intraocular irrigating solutions to reduce inflammatory reactions after pediatric cataract surgery. It has been documented that infusion of enoxaparin, a low molecular weight heparin, during pediatric cataract surgery may minimize the postoperative inflammatory response.

Posterior capsulorhexis combined with optic buttonholing

Recently R. Menapace introduced Posterior optic buttonholing (POBH) a safe and effective technique which not only excludes retro-optical opacification,
but also withholds capsular fibrosis by obviating direct contact between the anterior capsular leaf and the optic surface.

Legend for Figures:

Figure -1: Anterior continuous curvilinear capsulorhexis (ACCC) under trypan blue dye.

Figure -2: Visual Axis Opacification at 2.5 years follows up of IOL implantation with primary posterior continuous curvilinear capsulorhexis (PCCC) and vitrectomy.

Figure -3: Retro illumination photograph showing Anterior and Posterior capsulorhexis and well centered IOL.

References:
16. Mandal AK, Netland PA. Glaucoma in aphakia and
Seasonal occurrence of ocular lesions caused by Caterpillar hair-A Case series

Gajiwala Uday R., Patel Rajesh U., Gangwal Manoj M., Chariwala Rohan A.
SEWA Rural, Jhagadia-393110, Dist.-Bharuch, Gujarat.

The ocular reactions to caterpillar hair are diverse in nature and location, ranging from a toxic reaction to the external foreign bodies, keratoconjunctivitis or the formation of conjunctival nodules, to iritis, vitritis, papillitis or endophthalmitis.

Twelve patients of ocular caterpillar hair injuries were observed at our hospital from 1st November 2008 to 31st March 2009. Ophthalmic nodosa is common in the winter months in India when the caterpillars are in plenty. The incidence of caterpillar hair injuries is essentially dependent upon the life cycle of the caterpillar in the locality. Shorter quiescent period was due to hair being sharp and smooth and without barbs. Pattern of corneal abrasions was very useful in indicating the presence and location of setae.

Mechanical removal of the setae and patching the eyes with antibiotic and atropine ointment found useful in alleviating the symptoms. All cases resolved without sequelae.

Introduction:
Ocular caterpillar hair injuries are quite common but often missed during routine examination especially deeply buried caterpillar hair. Clinical features of caterpillar injuries differ slightly in India from those seen in the western world. Not many cases have been reported from India. We found seasonal trend of ocular caterpillar hair injuries from October to March. Purpose of this case report is to highlight its seasonal trend, spectrum of involvement, diagnostic method, and prognosis and to create awareness about these injuries for proper control and management.

Case History:
Twelve patients of ocular caterpillar hair injuries were observed at our hospital from 1st November 2008 to 31st March 2009. As shown in table, the patients of various age groups attended ophthalmic OPD with complaint of foreign body/ pricking sensation in the eye, watering, redness, photophobia and inability to open the lids following fall of insects while working outdoor/indoor. Few patients had taken treatment from elsewhere but with no relief of symptoms. Presenting visual acuity with pinhole using snellen’s chart and time since injury was noted.

After putting 1 % Proparacaine to relieve photophobia, patients were examined on the slit lamp. Cornea was stained with 1 % fluorescein to detect corneal abrasions, its location and pattern. Cornea showed punctate stippling, vertical/oblique superficial linear scratches in the part coinciding with the presence of caterpillar hair. Cornea was carefully searched for embedded caterpillar hair.

Figure 1 Caterpillar hair in Upper tarsal conjunctiva
<table>
<thead>
<tr>
<th>No.</th>
<th>Age</th>
<th>Date of Presentation</th>
<th>Quiescent period</th>
<th>Presenting Complaint</th>
<th>Indoor/Outdoor/Work</th>
<th>Ocular Involvement</th>
<th>Staining pattern</th>
<th>Remarks</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>M/32</td>
<td>1/11/08</td>
<td>4 days</td>
<td>H/O fall of Insect</td>
<td>Outdoor</td>
<td>Cornea</td>
<td>Linear Abrasions</td>
<td>4 hair removed</td>
</tr>
<tr>
<td>2</td>
<td>M/6</td>
<td>18/11/08</td>
<td>7 days</td>
<td>H/O inability to open the eye</td>
<td>Not Aware</td>
<td>Eyelids Conjunctiva Cornea</td>
<td>Big Epi. Defect</td>
<td>24 Hair removed Under GA</td>
</tr>
<tr>
<td>3</td>
<td>M/40</td>
<td>22/11/08</td>
<td>2 days</td>
<td>H/O fall of Insect</td>
<td>Outdoor</td>
<td>Eyelids Cornea</td>
<td>Linear Abrasions</td>
<td>3 Hair removed</td>
</tr>
<tr>
<td>4</td>
<td>M/48</td>
<td>1/12/08</td>
<td>5 days</td>
<td>H/O fall of Sugarcane leaf</td>
<td>Outdoor</td>
<td>Only Cornea</td>
<td>Linear Abrasions</td>
<td>3 Hair removed</td>
</tr>
<tr>
<td>5</td>
<td>M/40</td>
<td>3/12/08</td>
<td>3 days</td>
<td>H/O fall of Insect while travelling on a bike</td>
<td>Outdoor</td>
<td>Lower Lid Margin, Palpebral Conjunctiva, Cornea</td>
<td>Lower Corneal</td>
<td>2 hair removed</td>
</tr>
<tr>
<td>6</td>
<td>F/42</td>
<td>25/12/08</td>
<td>1 day</td>
<td>H/O FB sensation</td>
<td>Indoor</td>
<td>Upper palpebral Conjunctiva, Papillary Reaction, Cornea</td>
<td>Abrasion Upper Corneal</td>
<td>3 hair removed</td>
</tr>
<tr>
<td>7</td>
<td>M/40</td>
<td>26/12/08</td>
<td>1 day</td>
<td>H/O fall of Insect while travelling on a bike</td>
<td>Outdoor</td>
<td>Upper palpebral Conjunctiva, Cornea</td>
<td>Abrasion Upper Corneal</td>
<td>4 hair removed</td>
</tr>
<tr>
<td>8</td>
<td>M/30</td>
<td>29/12/08</td>
<td>26 days</td>
<td>H/O fall of Insect</td>
<td>Outdoor</td>
<td>Lower Lid Margin, Papillary Reaction, Cornea</td>
<td>Abrasion Lower Corneal</td>
<td>3 Hair Removed</td>
</tr>
<tr>
<td>9</td>
<td>M/20</td>
<td>10/01/09</td>
<td>3 days</td>
<td>H/O fall of Insect</td>
<td>Outdoor</td>
<td>Upper &amp; Lower Palpebral Conjunctiva, Lids, Cornea,</td>
<td>Abrasion Large Epithelial Defect</td>
<td>17 hair removed</td>
</tr>
<tr>
<td>10</td>
<td>F/72</td>
<td>24/01/09</td>
<td>3 days</td>
<td>H/O FB sensation</td>
<td>Indoor</td>
<td>Lower Lid, Lower Cornea</td>
<td>Lower Corneal Abrasion Lower</td>
<td>2 hair removed</td>
</tr>
<tr>
<td>11</td>
<td>M/60</td>
<td>03/02/09</td>
<td>1 day</td>
<td>H/O fall of something</td>
<td>Outdoor</td>
<td>Lower Lid, Lower cornea</td>
<td>Corneal Abrasion Upper</td>
<td>3 hair removed</td>
</tr>
<tr>
<td>12</td>
<td>M/36</td>
<td>20/3/09</td>
<td>2 hours</td>
<td>H/O fall of Insect</td>
<td>Outdoor</td>
<td>Upper Palpebral Conjunctiva, Cornea,</td>
<td>Corneal Abrasion</td>
<td>2 hair removed</td>
</tr>
</tbody>
</table>
The eyelids were everted to look for follicles and embedded caterpillar hair in the upper tarsal conjunctiva. The follicles were also stained with fluorescein to detect conjunctival epithelial defect in the centre of the follicles. Dark brown/black caterpillar hairs were also searched for in upper and lower eyelid margin. We found caterpillar setae more commonly in tarsal conjunctiva, eyelid margin, and superficial cornea.

Caterpillar hair on the surface (conjunctival, corneal, eyelid margin) were picked up with fine forceps under topical anaesthesia. The hair embedded in the follicles were removed after rupturing the follicles and gently stroking its base with a 26G needle. In few cases, caterpillar hair was removed on subsequent follow up. These were mounted on a glass slide and examined under the microscope. All the patients were prescribed topical antibiotics, atropine ointment and the eyes were patched. All cases resolved without sequelae.

**Discussion:**
The term “Ophthalmic Nodosa” is derived from the nodular conjunctival reaction produced as an inflammatory response to certain insect or vegetable hair or fragile spines. It is one of the manifestations of caterpillar hair injuries. These hair or the fragile spine protects the caterpillar. Urticating hair can be of two types.

1) **Envenomating Hair**
Tubular or porous spine capable of holding a venom or irritant produced by a gland at the base

2) **Non- Envenomating Hair**
These produce a mechanical irritation on contact. These hair are fragile and easily dislodged from the caterpillar. They adhere to the surface of skin, or become air borne and can drift and settle on nearby washing or other surfaces, which human will contact.

The black and the brown hairy caterpillars belong to the order Lepidoptera. Gesick et al observed a seasonal incidence between July and November and the autumn as the usual season. Sethi et al reported its occurrence between September and December. We observed all cases from November 2008 to March 2009, but in February and March, we found only two cases. Thus, in India ophthalmic nodosa is common in the winter months when the caterpillars are in plenty. The incidence of caterpillar hair injuries essentially depends on the life cycle of the caterpillar.
Caterpillar Hair

in the locality.
Sethi et al observed caterpillar hair injuries in the house itself, mostly while asleep. The caterpillar tend to seek dark and secluded spaces for population and for that, they wander into the houses. Western literature reported caterpillar hair injuries while outdoor work. In our series, we found injuries in nine (out of 12) patients while doing outdoor work and so male preponderances.

Duke Elder described that after the initial symptoms of trauma there was a quiescent interval followed by a recurrence of symptoms. He mentioned this interval to be usually several weeks and occasionally 3-5 days.

Sethi et al described shorter quiescent interval as short as three hours and as long as few days. Patel and Shanbhag et al also observed short quiescent interval. We observed this period as short as 2 hours to as long as 7 days in 11 patients. Only one patient presented to us after 26 days, mostly because all setae buried deep in the follicles. This shorter quiescent interval from India may be due to the quicker and easier passage of the smooth and sharp hair through the tissues of the eyelid, by the muscular movement of the eyelid and by rubbing over the eyelid because of irritation.

Most of the western literature has described barbed hair in their cases, which must be dependent on the type of the caterpillar existing there. Sethi et al described sharp, smooth caterpillar hair without barbs. In our cases, we observed brown caterpillar hair without barb with one sharp end and other blunt.

The tissue reaction to the caterpillar hair in palpebral conjunctiva is due to mechanical and toxic effects. Caterpillar setae-incited inflammatory reaction is classified into five types:

Type 1: An acute reaction - chemosis and inflammation.

Type 2: Chronic mechanical keratoconjunctivitis

Type 3: Formation of grey-yellow nodules in the conjunctiva (granulomas).

Type 4: Iritis

Type 5: Vitreoretinal involvement

We found type 2 involvements in almost all cases (10 Cases); with type 3 involvement in only two cases. There are four types of presentations of caterpillar hair in palpebral conjunctiva:

1) Hair projects out of the yellow follicles
2) Hair remains embedded and seen as a black point
3) Initially no hair seen, subsequently hair projects out from the follicle.
4) No hair visible but on mechanical rupture of follicle and gently stroking its base, the caterpillar hair could be coaxed out.

Usually prognosis is good in patients with caterpillar hair injuries.

**Key Messages:**

History of foreign body sensation after fall of insect should arouse suspicion of caterpillar hair injury and these should be actively searched for. The pattern of corneal abrasion is very useful in searching the presence and location of the setae.

**References:**

5. Fraser SG, Dowd TC, Bosanquet RC. Intraocular caterpillar hair (Setae): Clinical course and management. Eye. 1994; 8 (pt 5) : 596-8
9. Choi JTL, Rauf A, Ophthalmic nodosa secondary to tarantula hair. Eye 2003;17,433-434
Medico-Legal Aspects of Post-Operative Endophthalmitis

Lalit Verma, Shefali Gupta,
Centre for Sight & Indraprastha Apollo Hospital, New Delhi

---
Endophthalmitis occurs in best of hands in best of set ups.
Only people who do not get endophthalmitis are those who do not operate

For endophthalmitis to occur what is required is a breach or a cut in the integrity of ocular coats and introduction of microbial inoculum. We all know that during intraocular surgery both of these happen. Inoculum means microbiological load resulting in endophthalmitis. Inoculum can be of various sizes and types. To measure size of the inoculum one can use the concept of colony forming unit (CFU). It is a measure of viable cells in which a colony represents an aggregate of cells derived from a single progenitor cell. CFU is used to determine the number of viable bacterial cells in a sample per mL. Hence, it tells the degree of contamination in samples of water, vegetables, soil or fruits, or the magnitude of the infection in humans and animals. It is different from the direct microscopic counts that include both dead and living cells. Types of inoculum mean different types of microorganisms. Like for Staph. aureus if 19 colony forming units enter intravitreally or 50 colony forming units enter anterior chamber during surgery, endophthalmitis will occur. For Pseudomonas if only 5 colony forming units reach vitreous cavity or 197 colony forming units reach anterior chamber fulminant infection results.

Corneal incision is at least three times more potent than tunnel incision for causing endophthalmitis.

Intra-ocular infection has always brought disrepute to the ophthalmologist and this problem is not only rampant at eye-camps but also in hospitals, which include the five star ones. Only surgeon who does not have endophthalmitis is the one who does not operate. The problem is general and it is not the surgeon who is to be blamed although he is responsible for the surgery. Despite the best possible care, mishaps cannot always be avoided because the error in one link of the entire chain may sometimes result in a disaster. But in the court of law if you have a misfortune of infection then how to save yourself?

- Record all findings including vision including projection of rays, intraocular pressure, status of cornea, anterior chamber reaction, pupillary reaction, details of iris, IOL (if present) and fundus. Get B-Scan ultrasound done if fundus cannot be seen at all.
- Record them daily and keep a copy with you.
- Do not give telephonic treatment. e.g if patient calls up in the night and complains of pain, redness, watering and if you tell him to continue or add steroid drops, then this is asking for disaster. That means that instead of giving telephonic treatment tell the patients to go to the nearby available ophthalmologist available and show him.
- Patient who is on treatment for endophthalmitis, examine him daily and always write on the prescription to report SOS. If patient is from far flung area write on his card or prescription slip that in case of any pain or redness or decreased vision or unusual symptoms report to nearest ophthalmologist and mention “do not ignore.”
- Even at the cost of …………. please document, document and document.
- When in doubt seek peer review, refer to a retinal surgeon or a higher level hospital.
- Involve multiple people or hospitals to safeguard your self.

Greatest malpractice risk associated with...
endophthalmitis- Analysis of claims show that liability arises from a delay in diagnosis or treatment, including a delay in referring the patient to a vitreo-retinal specialist.

To reduce the risk of delay in diagnosis-
• If the surgery was complicated and took a long time or required extensive instrumentation, you should have a higher index of suspicion for the development of endophthalmitis.
• Give all patients written discharge instructions stating the symptoms that warrant contacting you (blurred vision, red eye, pain, photophobia).
• Educate your staff members who handle telephone calls about the risk of endophthalmitis. Instruct them to schedule emergent appointments for such patients. Err on the side of patient safety when deciding to treat over the phone versus examining the patient.

To reduce the risk of delay in treatments-
• Document your decision making process in the medical record, especially when the patient calls with symptoms of a possible infection.
• Obtain a thorough interval history and perform and document the clinical examination. Note the presence and absence of signs of endophthalmitis (the cardinal sign is intraocular inflammation greater than expected for that point in the recovery process.)
• If in doubt, consult with and/or refer patients to a vitreo-retinal specialist for management.

Measures to take to reduce liability:
• During the informed consent discussion (a must for all surgeries), warn patients about the risk of infection and possibility of vision loss. Emphasize the risk specially if the patient has diabetes or is immunosuppressed. You have to tell the patient and relatives that you are going to do the best and leave no stone unturned in this regard – but still complications including infection happen in the best of hands and in best of set ups including in all developed countries. Explain in lay language- where ever there is a cut (however small it may be ), bacteria or other organisms can enter.
• Have a prudent follow up plan, especially in the symptomatic patient, and ensure that the patients make the appointment before leaving your office.
• Diligently follow up on all the patients who miss or cancel appointments, again ensuring that they understand that not receiving appropriate treatment could result in blindness.
• Carefully instruct patients to call you immediately if vision loss, pain or other ocular symptoms develop before their next scheduled visit.
• Make sure to DOCUMENT, DOCUMENT, DOCUMENT.
• Take anterior segment and fundus photographs, if possible.

After the catastrophe in Khujra, practically a National Alarm was created and Supreme Court intervened & passed certain guidelines for eye camps:
1) Qualified, experienced ophthalmic surgeons registered with Medical Council of India or any State Medical Council should only perform the operations. Camps should not be used as training ground for post-graduate students, and operative work should not be entrusted to post-graduate students.

View point: Students or fellows or inexperienced doctors should operate under guidance and avoid doing surgery in one eyed and other high risk patients.

2) There should be a pathologist to examine urine, blood, sugar etc.

3) It is preferable to have a dentist to check the teeth for sepsis and a physician for general medical check-up.

View point: Physical presence of pathologist is not essential. What is required is a proper work up of patients, proper preoperative evaluation, and clearance from physician or cardiologist if needed.

4) All medicines to be used should be of standard quality duly verified by the doctor in-charge of the camp.

View point: This is of utmost importance especially so, for irrigating fluids, viscoelastics, sutures, intraocular lenses etc.

5) The necessity of maintenance of the highest standards of aseptic and sterile conditions at places where ophthalmic surgery - or any surgery - will be conducted was emphasized. The Supreme Court said: “The necessity of maintenance of the highest standards of aseptic and sterile conditions at places where ophthalmic surgery - or any surgery - is conducted cannot be over-emphasized. View point: It is not merely on the formulation of the theoretical standards but really on the professional commitment, with which these are implemented, followed and periodically reviewed and appropriate action taken, that the ultimate result rests.

Remember, a surgeon is best known or assessed by the way he handles complications or unusual situations. The way he talks to the patient and their relatives is of paramount importance. All the problems arise when patient’s expectations are sky high and he is not explained the reality by the treating surgeon and someone else tells, makes the patient aware or even instigates (not a uncommon situation ).
Penetrating Keratoplasty: History and Evolution

Shibal Bhartiya, Noopur Gupta, Anita Panda
Cornea and OSD Services
Dr R P Centre For Ophthalmic Sciences, AIIMS, New Delhi, India

“Two kinds of blindness are easily combined so that those who do not see really appear to see what is not.”
Tertullian.

Introduction
The history of the evolution of keratoplasty into a distinct clinical entity reads as would a fairy tale: the glorious triumph of the human spirit against all odds, and ingenious methods to combat hurdles coming ones way. The almost impossible dreams and fights of fancy that have morphed into the contemporary technique, making it the most successful of transplant surgeries, worldwide. This article is an acknowledgement of the contribution of the pioneers in this field who conceived the notion of keratoplasty, stumbled along with countless failures, and laid the groundwork for what has become a twentieth century success story.1,2

Review
The notion of completely replacing dysfunctional, opaque cornea was first suggested by a Frenchman, Guillaume Pellier de Quengsy in 1789 who proposed the use of a thin glass disc, the size of the cornea, set in a silver ring which could be stitched to the sclera with cotton threads, after removal of scarred tissue. This actually was the first proposed keratoprosthesis: although this innovative operation was not performed, the circumstances under which the surgery should be performed, the instruments to be used, postoperative care and even possible complications were described by him.
Franz Reisinger, coined the term "keratoplasty” and suggested the use of animal tissue to replace the scarred human cornea. His work was confined to rabbits and chickens, with most of his experimental works failing because of either clouding of the graft or panophthalmitis.
In nineteenth century Germany, K Himly suggested replacing an opaque cornea of one animal with a clear cornea of another animal (1813). In 1837, came the report by Dr. S. L. Bigger, of his successful attempts at keratoplasty, a story as captivating in its inception as its results. While traveling, Bigger was taken captive by Sahara Bedouins near Cairo. During his captivity he successfully performed a corneal homograft on a blinded pet gazelle using the tissue of another gazelle.
Dr. Richard Sharp Kissam in New York was the first to attempt this procedure in the only functional eye of an Irishman in 1838. He removed the opaque cornea with a Beer cataract knife and then with only two sutures at 3 and 9 o’clock positions, secured the donated tissue from a 6-month-old pig, into position. These sutures were removed after 36 hours, at which time the porcine graft appeared to have been united with the host tissue. Like all other surgical procedures at that time, this operation was also performed without any anaesthesia. After the “severe chemoisis” had subsided, the patient’s vision was “improved” (i.e., the patient received more light), but the cornea
The early nineteenth century saw a multitude of innovations which were to revolutionize surgical technique and prognosis globally. Ether was introduced in 1846, chloroform in 1847. Cocaine came a decade later in 1858, and infiltration anaesthesia was introduced in 1889. Lister’s principles of antiseptic surgery were postulated in 1867, increasing awareness of the importance of asepsis, careful handling and placement of graft, and the use of homograft.

In 1886, Arthur von Hippel transplanted a full thickness rabbit cornea on to the lamellar bed of a young girl’s cornea, reporting an improvement in thickness rabbit cornea on to the lamellar bed of a young girl’s cornea, resulting in an improvement in vision from finger counting to 6/60. This was the first successful corneal transplant on a human being. Von Hippel used cocaine anaesthesia and iodoform as an antiseptic. He focused on lamellar keratoplasty reiterating that the main determinants of corneal transparency were the integrity of corneal endothelium and Descemet’s membrane. His most significant contribution was the circular trephine which is still the mainstay of surgical instrumentation keratoplasty.

In 1906, Eduard Konrad Zirm performed the first full thickness graft that remained clear in a 45 year old laborer named Alois Gloat. His patient had suffered lime burn in his both eyes while cleaning a chicken coop and had a vision of counting fingers in both eyes.

The donor cornea was from the blind traumatized eye of an eleven-year-old boy, Brauer, who had been enucleated just prior to transplantation. The bilateral penetrating grafts were carried out under general anaesthesia (chloroform). The left eye graft remained clear with a vision of 6/36.

Zirm elaborated the basic principles for successful keratoplasty, which with minor technical modifications, hold true even today. He recommended the use of cornea from young, healthy human donors, use of trephine and eserine, adequate anaesthesia, strict asepsis, protection of the graft in gauze moistened with physiologic saline, use of overlay sutures and careful selection of cases.

In 1908, Plange replaced the scarred cornea of a blind eye with a lamellar graft from the patient’s other eye which, although blind, had a normal cornea. This was the first auto-keratoplasty to be performed.

The list of significant twentieth century contributors to the evolution of keratoplasty is long and illustrious including luminaries like Arthur Elschnig, Vladimir Filatov, R.T. Paton, Arruga, Ramon Castroviejo, Barraquer, Paufigue, Sourdille and Franceschett, Forstot and Kaufman. They have all made significant technical and biological advances in the evolution of the art and science of keratoplasty.

VP Filatov rightfully earned the epithet: the father of modern eye banking. His work involved the usage of cadaver cornea as the donor material and highlighted the importance of protecting the intraocular tissues while trephining the host tissue and advocated direct suturing.

In 1940s, corneal transplant surgery evolved dramatically with the availability of antibiotics and introduction of the use of steroids in corneal surgery. In late 1950s small fine needles were used for the first time for suturing.

At the same time, Paufigue and Charleux advocated lamellar corneal grafting techniques, and introduced limbal and eccentric grafts.

Corneal transplant surgery evolved in the first half of the 20th century in leaps and bounds, however, the greatest advances in corneal grafting have taken place in the last 30 years.

The understanding of corneal anatomy and physiology especially with regard to the corneal endothelium, introduction of microsurgical techniques, advances in corneal preservation, the elucidation of corneal immunology and the development of usage of anti-inflammatory and immunosuppressive agents have resulted in a high success rate of corneal grafting.

Edward Maumenee was the first to recognize graft rejection as the greatest limiting factor in graft survival. The classic scientific description and experimental models were elegantly designed by Khodadoust. Ramon Castroviejo performed the world’s first successful human corneal transplant and devised numerous instruments including the Castroviejo Calipers, Forceps, Corneal Scissors, Corneoscleral Punch, Cyclodialysis Spatula, Needle Holder, Tying Forceps, Suturing Forceps. His original suturing technique used a continuous silk suture coursing across the external surface of a square graft, with many of his square grafts providing good visual acuity for many years.

Richard Troutman was the pioneer in the field of astigmatism control. He is credited with the design of a microscope and numerous microsurgical instruments including the surgical keratometer and technique of wedge resection.

The first Eye bank was set up by Townly Paton in New York in 1959, and the Eye Bank Association of America was founded in 1961. This organization laid down the standards for obtaining, preservation, storage and usage of donor tissue.

The Specular microscope developed by Maurice provided the means of studying donor and transplanted endothelium, proving that a healthy functioning endothelium is the key to success of a corneal graft.
First successful transplantation using a cryopreserved human donor tissue was reported by Eastcott in 1954. Capella and Kaufman developed the basic method of cryopreservation in 1965. Introduction of the MK medium by McCarey and Kaufman in 1974 was a significant development heralding a revolution in corneal preservation. This medium proved to be reliable for storage of donor cornea for at least 3-4 days, allowing the elective planning for surgery.

**Conclusion**

Anterior segment and corneal surgery have undergone significant change and greater emphasis is now placed on quality of visual rehabilitation. Increasing interest has developed in alterations of the cornea to effect refractive change. Also, with the recent focus on component surgery of the cornea and stem cell transplantations, a whole new vista awaits the corneal transplant surgeon.

**References**

Instruments Scan

Understanding Corneal Topography: Basics

M. Vanathi
Asst Prof of Ophthalmology – Cornea Services
Dr R P Centre For Ophthalmic Sciences, AIIMS, New Delhi, India

Corneal topography provides an in depth understanding of the shape of the cornea. Corneal profile on topography might be categorized into three profiles viz prolate, oblate or mixed. The advent of reflection based topography systems and projection based systems have made expanded the horizons of corneal topography mapping. Reflection-based systems measure the slope of the corneal surface from which the curvature and power is derived with additional measurements and certain assumptions being made. Projection-based systems measure the true shape of the cornea in terms of the height, or elevation, above a reference plane, from which the surface slope, curvature and power. Radius of curvature of the cornea, power of the cornea, elevation and pachymetry are among the various details that may be elicited for corneal topography maps. More recent developments in terms of wave-front analysis provide information about the refractive power of the eye as a whole, rather than just the effect of the anterior corneal surface. Selecting the appropriate display format remains the key in maximizing the information obtained from a topography examination. Interpretation of maps involves as systematic approach to study the topographic pattern. This article aims to provide a basic understanding of the various corneal topography and their interpretation.

Corneal topography by way of which measurements of the corneal shape is recorded may be done by several ways\(^1\). These include the conventional, reflection based topography systems (keratometry, photokeratoscopy, videokeratoscopy) and the recent, projection based systems (rasterstereography, laser interferometry, moiré interference etc\(^2\)). Data measurement and presentation by various corneal topography systems depends on the proper image acquisition. Projection-based systems measure the true shape of the cornea in terms of the height, or elevation, above a reference plane. This data is used to calculate surface slope, curvature and power. Reflection-based systems measure the slope of the corneal surface from which the curvature and power is derived with additional measurements and certain assumptions being made. Computer assisted videokeratography, the most commonly used, combines concentric ring keratoscopy with analysis by computer programs to produce a high resolution imaging of the corneal topography. The computer calculates the dioptric power and the radius of the curvature at hundreds of points on the anterior corneal surface on the basis of the captured video image of the reflected keratoscopic rings from the corneal surface. A graphic depiction of this information is presented as colour coded corneal topographic map.

Radius of curvature of the cornea can be calculated either globally (axial radii of curvature) or locally (tangential radii of curvature), and then converted to dioptric power using the standard keratometric index. Global (axial/sagittal) radius of curvature measures the distance of points from the optical axis, and therefore has a spherical bias. Local (instantaneous/tangential) radius of curvature calculates the curvature at each point with respect to its neighbouring points, and is therefore more accurate for local irregularities and in the peripheral cornea\(^3\). Axial curvature (sagittal curvature) measures the curvature at a certain point on the corneal surface in axial direction relative to the center. Meridional curvature (tangential/instantaneous curvature) measures the curvature at a certain point on the corneal surface in meridional direction relative to the other points on the particular ring. Meridional curvature maps are more sensitive measures of local curvature change. Axial curvature maps can be derived from meridional maps. Axial value at a certain point equals the average meridional curvature along the radius from the map center to the point of interest, thereby approximating the average refractive power. Axial and meridional maps should theoretically be displayed in the units of radii of curvature (i.e., mm) at each corneal surface point. When the display curvature is in units of keratometric diopters they are called axial or meridional power maps. The power of the cornea (in dioptres) is a measure of the anterior corneal refractive effect. Radius of curvature is converted to power using the standard keratometric index (SKI = 1.3375). This is an approximate figure derived from assumptions about the thickness and
refractive index of the cornea, and the shape of its posterior surface. Height or elevation maps define the distance of each point on the surface from a reference surface flat reference plane, which is used to interpret the overall shape of the cornea\(^3\). Elevation is not measured directly by Placido-based topographers, but certain assumptions allow the construction of elevation maps. Elevation of a point on the corneal surface displays the height of the point on the corneal surface relative to a spherical reference surface (with the reference surface chosen in most instruments being a sphere). Best mathematical approximation of the actual corneal surface called best-fit sphere is calculated by instrument software for every elevation map separately. More recent developments in terms of wave-front analysis provide information about the refractive power of the eye as a whole, rather than just the effect of the anterior corneal surface.

Most videokeratoscopes use similar mechanisms but vary in some of their features, such as the size of the cone of placido rings, and whether focusing is manual or automated. Alignment and focusing of the reflected placido rings should be optimized while using videokeratoscopes in order to obtain good measurement data. Accurate alignment, centration and focusing is important to avoid the introduction of artificial abnormalities. Tear film irregularities also contribute to artifacts. The reflected image of the placido rings is captured on video camera and digitized. The computer analyses the position of each of the 15-38 circular mires along 256-360 semi-meridians, theoretically providing about 6,000 to 11,000 data points\(^1\). Algorithms then compute the curvature at each point. The accuracy of measurements is about 0.15D in the central zone of a normal cornea, but is commonly less in other situations due to the assumptions and approximations made by the algorithms.

Selecting the appropriate display format remains the key in maximizing the information obtained from a topography examination\(^4\). The raw image of the reflected placido rings depicts the corneal pathology or tear film abnormalities. Videokeratoscopy mires appear closer together in steep areas and are further apart in flat areas. Every topographic map has a color scale assigning a particular color to a certain keratometric dioptic range. Interpretation is not to be based an on color alone, but also on the keratometric value. The most common display presentation format of topography in the colour-coded scale is representation of the steep areas by the warm colours (red and orange) and the flat areas by the cool colours (green and blue). When using the absolute (standardised) scale, the same colours represent the same power on every map\(^4\). The normalized (relative) scale, the step interval fills the range of each map. In systems permitting an adjustable scale, the step interval can be selected by the operator\(^5\). As absolute maps have a preset color scale with the same dioptic steps, dioptric minimum and maximum assigned to the same colors for a particular instrument, they allow direct comparison of two different maps. However, because the steps are in large increments (generally 0.5 D), their disadvantage is that they do not show subtle changes of curvature and can miss subtle local changes (e.g., early keratoconus). Normalized maps have different color scales assigned to each map based on the instrument software that identifies the actual minimal and maximal keratometric dioptic value of a particular cornea. Hence the dioptic range assigned to each color generally is smaller compared to the absolute map depicting more detailed description of the corneal surface. Two different maps can then be compared based on the interpretation of the keratometric values of their different colour scales.

Statistical indices depict a particular feature of the cornea, such as its symmetry, regularity or asphericity. Interpretation of maps involves systematic approach to study the topographic pattern. This includes the following:

- Patient details, laterality of eye, date of examination
- Scale
- Type of measurement (e.g. elevation, curvature, power)
- Step interval
- Map
- Statistical information (indices)
- Comparison with prior maps of the same eye (check to ensure that the scale is the same)
- Comparison with the topography of the fellow eye (check to ensure that the scale is the same)

Corneal profile on topography might be categorized into three profiles viz prolate, oblate or mixed. The normal cornea is prolate, with the centre being the steepest and gradually flattening towards the periphery.
Topographic patterns of the normal corneas classify astigmatic patterns of the cornea in accordance to its pattern characteristics as follows (figure 1) 6, 7, 8:

(i) Round pattern: the ratio of the shortest to the longest diameter at the colour zone is 2/3 or more
(ii) Oval pattern: the ratio of the shortest to the longest diameter at the colour zone is less than 2/3.
(iii) Regular astigmatism pattern: this is seen when the two principal meridians are oriented at approximately right angles to each other. The angle between the axis of the two halves of the bowtie of less than 200 is defined as regular astigmatism
  • Symmetrical bow tie (Figure 1, 2):
    i. a central constriction is identified in the colour zone,
    ii. the ratio X0 / X1 or X0 / X 2 is 1/3 or less
    iii. the ratio of X1 / X2 or Y1 / Y 2 is 2/3 or more

• Asymmetrical bow tie (figure 1, 3)
  i. a central constriction is identified in the colour zone,
  ii. the ratio X0 / X1 or X0 / X 2 is 1/3 or less
  iii. the ratio of X1 / X2 or Y1 / Y 2 is less than 2/3
(iv) Irregular astigmatism pattern: This pattern is defined when the angle between the two steepest semimeridia is greater than 200 and would represent a bi-oblique bowtie pattern or when no pattern is discernable.
Astromtic corneas show bow tie appearances with the red bows lying along the steep meridian. In oblate corneas (as those undergone flattening by surgery), the bows are blue and lie along the flat meridian. Based on the various pattern descriptions 8, combination patterns such of regular astigmatism such as prolate symmetric bow tie, prolate asymmetric bow tie, oblate symmetric bow tie, oblate asymmetric bow tie and irregular astigmatism such as prolate irregular, oblate irregular, mixed patterns and others such as steep/flat pattern, localized steepness pattern, triple pattern, horseshoe pattern have also been elaborated.

Common indications for corneal topography in practice:
(i) Refractive surgery patients
  • Preoperative assessment
(ii) Diagnostic
  • Screening for ocular disease
  • Keratoconus
(iii) Planning surgical incision (cataract, astigmatic keratotomy)
  • Incision location, length, depth
(iv) Contact lens fitting in irregular corneas
(v) Intraocular lens power calculation in special situations
(vi) Management of astigmatism
  • Adjustment of incisions or sutures
(vii) Keratoplasty follow up
(viii) Others:
  • Suture manipulation/removal
  • Patient education
  • Communication with colleagues
  • Documentation for medico legal purposes.

Corneal topography in common clinical situations in cornea practice:
CL-induced corneal warpage: Corneal topographic changes following contact lens wear are thought to occur directly as a result of the mechanical pressure exerted by the lens. Patients with corneal warpage may be asymptomatic, have reduced spectacle corrected acuity or contact lens intolerance. The changes are most severe and persistent in wearers of RGP lenses. Many topographic patterns can be induced, but they tend to comprise flattening in the areas of lens bearing, with possible adjacent steepening. The topographic changes produced by CL induced warpage are highly variable with the most common one being a presentation similar to early keratoconus. Others include central irregular astigmatism, changed axis of astigmatism, loss of normal progressive flattening form the centre to the periphery and a correlation between the resting position of the CL and topographic pattern. Contact lens wear should cease six weeks prior to pre-operative assessment for hard or rigid lenses, and two weeks prior to soft contact lens fitting. Surgery is not advisable till stabilization of topography pattern.

Post-keratoplasty: In such highly irregular corneas, topography assessment using computer assisted videokeratography is more accurate than refraction or keratometry for determining axis of greatest astigmatism, and the axis of tight sutures. Prolate patterns of topography are commonly seen after single continuous suturing. Suture adjustments are effective in bowtie patterns. Suture removals may effect decrease in astigmatism in bowtie patterns and not in oval/steep flat patterns.
Corneal Ectasias: Keratoconus and pellucid marginal degeneration (PMD) is characterized by presence of irregular astigmatism and inferior (commonly) corneal steepening on topography (figure 4).

Corneal topography serves as one of the most sensitive methods for detection of early keratoconus, as it may provide the clinician with characteristic clues before clinical signs become evident. It is also imperative to be able to differentiate true early keratoconus from other similar conditions such as a normal cornea with asymmetric bowtie or contact lens induced warpage. Corneal topography of mild inferior steepening with normal corneal thickness and no evident clinical signs of keratoconus is termed “keratoconus suspect” and need apt attention of the clinician in decision making to proceed with refractive surgery.

Terrien’s marginal degeneration of the cornea is characterized on topography by noticeable flattening of the cornea with high against the rule astigmatism. Refractive surgery:

Refractive corneal procedures alter the central corneal curvature and hence the asphericity of the cornea. Myopic refractive ablation treatments flatten the central optical zone resulting in a cornea that is less prolate, or even oblate, while hyperopic treatment steepens the optical zone, causing the cornea to become increasingly prolate. Changes in corneal topography can be depicted in difference or subtraction maps in which a later map is subtracted from an earlier one. When topography is used to guide ablation, height maps are used so that the treatment can be applied to the peaks, rather than the steep sides, of any elevation.

Myopic treatment zone is delineated by a central zone flattened zone while hyperopic correction shows central steepening surrounded by a ring of relative flattening at the edge of the treatment zone, where most corneal tissue has been removed. In astigmatic treatments, the treatment zone is oval. Decentration is identified by comparing the first week post-operative map with a pre-operative map. Similar post-operative appearance may also be seen in pre-existing asymmetric astigmatism, or an asymmetrical healing response. Decentraions of large diameter (6mm) optical zones tend to be clinically significant if greater than 1mm, or in patients with relatively large pupils.

Eight topographic patterns after PRK have been identified. Patients with a homogeneous pattern have least astigmatism. Those with regular patterns (homogeneous or toric) have a better refractive predictability, visual acuity and level of satisfaction than those with irregular patterns. The irregular patterns include semi-circular, keyhole, central islands, focal irregularities and irregularly irregular. A central island is present when any part of the treatment zone surrounded by areas of lesser curvature on more half of its circumference. They are classified according to the power and diameter of the central steep area. The refractive and topographic changes after LASIK are similar to PRK, but the overcorrection is not as large, and usually early stability is achieved. Decentration is more common and tends to be more significant. Epithelial in-growth at the periphery of the flap-stromal interface is characterized on topography by an area of steepening at the edge of the treatment zone, which can progress centrally.

Quantitative descriptors of corneal topography:

Quantitative descriptors of corneal topography provide useful topographic information that enhance the utility of these machines in clinical practice and research. Simulated keratometry provide an estimate of measurement obtained with a keratometer. Simulated keratometry measurements characterize corneal curvatures in the central 3-mm area. The steep simulated K-reading is the steepest meridian of the cornea, using only the points along the central pupil area with 3-mm diameter. The flat simulated K-reading is the flattest meridian of the cornea and is, by definition, 90° apart. These readings give an idea about the central corneal curvature that is frequently visually most significant. The 3-mm diameter is chosen primarily from historical reasons for the purpose of comparison with standard keratometry that is used for analysis of 4 central points, 3.2 mm apart.

Central corneal regularity correlates BCVA in normal eyes, such as the SRI (surface irregularity index) on the Tomey topography, the CIM (corneal irregularity measurement) on the Humphrey topography and the diagnostic summary on the Eyesys topography. Keratoconus detection programs are automated algorithms and those such as the Klyce/Maeda and the Rabinowitz algorithms are available on the Tomey autotopgrapher. A few orbscan maps of patients under followup in my cornea practice have been included (figure 5 – 9).
Summary:
Corneal imaging techniques rapidly evolving into higher standards and understanding their significance is imperative in management of common corneal refractive clinical situations. Corneal topography instruments used in clinical practice most often are based on Placido reflective image analysis. In this method, the anterior corneal surface is imaged using the analysis of reflected images of multiple concentric rings projected on the cornea. The basics of the most common clinical method of Placido-based corneal topography has been briefly reviewed. Limitations of corneal topography include errors of corneal topography under optimal conditions of the range of ±0.25 D or 2-3 µm, and could be higher in abnormal corneas. Corneal topography imaging based on the placido based systems requires an intact epithelial surface and tear film. Different technologies use different measurement methods and algorithms; thus, the output data are not directly comparable. As technologies undergo advancements, interpretation of the results of studies comparing the instruments, become rapidly redundant and difficult for practical clinical purposes.

References:
Rabinowitz YS, Rasheed K. KISA% index: a quantitative videokeratography algorithm embodying minimal topographic criteria for diagnosing...
Figure 6: Orbscan quad map – OS post Lasik ectasia

Figure 7: Orbscan quad map of Keratoconus suspect

Figure 8: Orbscan quad map of Keratoconus suspect

Figure 9: Orbscan quad map of patient undergone triple procedure: 11 months post op with sutures off
Instruction to Authors

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 editordjo@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-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 27.6cm (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 crossindexing. 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 them either on colour transparencies or on a CD in TIFF format. Colour photographs sent by email should be in a JPEG 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
and diagnostic techniques, history of ophthalmology and case report. All other contributors shall be entitled to one reprint of their article free of cost.

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 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.