Subhyaloid haemorrhage from positive valsalva manoeuvre
A step ahead in treating bacterial infections

Moxigram
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Dear Colleagues,

Wish you a Great New Year 2016!

This is the second issue TOS Today, the Official Journal of the newly formed Telangana Ophthalmological Society.

The editorial board, specially Dr Padmaja Kumari Rani, has made every effort to keep the Journal relevant to the general ophthalmologist, the sub-specialist, the practitioner, the resident and the researcher alike.

There are several invited Guest Articles from pioneers and stalwarts in their respective fields from across the Country. On behalf of the Telangana Ophthalmological Society the editorial board expresses its deep gratitude to these leaders for their valuable contribution that has improved the quality of the Journal immeasurably.

We hope you enjoy this publication, and look forward to your contributions, clinical or research experiences, for the next issue.

Sincerely,

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From The Secretary’s Desk

Dear Colleagues,

Telangana Ophthalmological Society wishes all its members a happy and prosperous 2016. TOS has completed the formalities of Registration with the Registrar of Societies, affiliation with AIOS, opening bank account etc.

The first Telangana State Ophthalmological Conference was held successfully at Karimnagar between June 19-21, 2015. Delegate and trade participation exceeded expectations, hospitality and scientific content of the conference were excellent. AIOS leaders Dr Debashish Bhattacharya, Dr D Ramamurthy and Dr Partha Biswas have also participated in the conference. The LOC and TOS thanked them for their novel gesture.

Hyderabad Ophthalmologist’s Association has come forward to host the second State Conference at Hyderabad. Dates, venue and other details will be finalised soon.

First issue of TOS Today was released in April and 1500 copies were distributed. The first ARC workshop was conducted at Sarojini Devi Eye Hospital, Hyderabad in September 2015.

Regarding Aarogyasri empanelment of eye hospitals, our honourable members are persuading the concerned authorities and a policy benefitting a larger segment is on the anvil.

A cheque of Rs 5 lakhs was presented by Dr Praneeth to TOS to institute a gold medal in memory of his late father Dr VV Ramana Rao. TOS thanked Dr Praneeth for his generous contribution. TOS membership now stands at 785. New LM numbers, Id cards and certificates will be issued soon.

With regards,

Dr A Ravindra, MS, Hony. General Secretary

From The Desk of the President

Dear Colleagues,

Good wishes and greetings on the occasion of New Year and Pongal to all our members. We are happy to recollect that the First Telangana State Conference was hosted by Karimnagar Ophthalmological Association successfully. It was my privilege to be the first President of our society and also a member of the LOC.

I wish more and more district associations would come forward to host State Conferences in future. This would not only promote fraternity and friendship among members but also strengthen the associations and bring out their organisational skills.

Participation and contributions from members of district units and private practitioners in sharing scientific knowledge and innovative skills has not been very encouraging. Thin attendance of delegates in the scientific halls during sessions has also become a matter of concern. These are some of the things which need to be looked at.

I request all practising ophthalmologists and postgraduates pursuing their PG course in Telangana state to become life members of TOS to improve the strength of our Society.

I am glad to know that the second State Conference will be held under the aegis of Hyderabad Ophthalmologist’s association at Hyderabad.

With regards,

Dr G Hari Kishan, President, TOS
All manuscripts must be sent by email to any of the Editorial Board member.

Manuscripts details

Articles: Randomized controlled trials, intervention studies, studies of screening and diagnostic test, outcome studies, cost effectiveness analysis, case-control series, and surveys with high response rate come in this category. The limit of the text is 3000 words excluding about 30 references and structured abstract of 250 words.

Research methodology: This includes educative articles related to the conduct of research with word count up to 3000 and references up to 30.

Case reports: new/interesting/very rare cases can be reported. Cases with clinical significance or implications will be given priority. However, mere reporting of a rare case may not be considered. The limit is 1000 words excluding references and abstract with a maximum of 10 references.

Announcements of conferences, meetings, courses, and other items likely to be of interest to the readers should be submitted with the name and address of the person from whom additional information can be obtained.

Achievements of Institutions, Hospitals, Centres in Telangana or TOS Members should be submitted with the name and address of the person from whom additional information can be obtained.

The text of original articles should be divided into sections with the headings: Abstract, Key-words, Introduction, Material and Methods, Results, Discussion, References, Tables and Figure legends. For a brief report include Abstract, Key-words Introduction, Case report, Discussion, Reference, Tables and Legends in that order. The text should be in MS Word format. Use double spacing throughout.

Illustrations (Figures), Upload the images in JPEG format.

Figures should be numbered consecutively according to the order in which they have been first cited in the text.

The decision of the Editor and Editorial Board regarding suitability of submitted material for publication will be final.
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Contents

From President’s & Secretary’s Desk 4

APOC 2014 Award Winners 5

APOS-NS Reddy Post-Graduate Best Free Paper Medal 2014 5

Guest Article:
Optical Coherence Tomography assessment of predescemet endothelial keratoplasty (PDEK) graft 6

Guest Article:
ReLEx SMILE – The Future of Refractive Surgery 10

Keratoconus - A practical approach in diagnosis and management 14

Persistent fetal vasculature (PFV) with unusual finding of lenticular cyst 19

Chronic Progressive External Ophthalmoplegia: A Case Report 20

Clinical Profile of Ocular Surface Squamous Neoplasia: A Retrospective Case Series. 22

Intraocular Cysticercosis – A Differential Diagnosis For Leucocoria 24

Perfluorocarbon Liquid as short-term tamponade for posterior giant retinal tear 27

Plateau Iris 28

Retinal Optical Coherence Tomography 30

Minimally invasive eye lid & facial rejuvenation: An evolving world 35

Election for the post of Vice-President TOS (2015-16) 39

First Telangana State Ophthalmological Annual Conference 41
1. Ns Reddy Competitive Free paper session for Post Graduates And Residents
**Winner:**
Dr Pallavi Gupta, Sarojini Devi Hospital, Hyderabad

**Sensitivity of impression cytology is diagnosing OSSN**

**Runner Up:**
Sunisha Gajula, Sarojini Devi Eye Hospital

2. P Ramchander competitive free paper session
**Winners:**
Dr Lumbini V, Hyderabad

**Amniotic membrane graft vs Conjunctival autograft for doing blue Pterygium**

**Runners Up:**
Dr Swathi Kaliki, LV Prasad Eye Institute, Hyderabad

3. Swarup video session
**Winners:**
Dr Vivek Dave, LV Prasad Eye Institute, Hyderabad

**ILM peeling in an unusual case of Terson Syndrome**

**Runners Up:**
Dr Tarjani Dave, LV Prasad Eye Institute, Hyderabad

4. Manoj Mathur Poster session
**Winners:**
Dr Swathi Kaliki, LV Prasad Eye Institute, Hyderabad

**OSSN as the presenting feature of HIV infection**

**Runners Up:**
Dr Jagadesh Reddy, LV Prasad Eye Institute, Hyderabad

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**Ocular Surface Squamous Neoplasia (OSSN) as the Presenting feature of Human Immunodeficiency Virus (HIV) infection**

**Dr Swathi Kaliki, Dr Saurabh Kamal, Dr Jyoti Batra, Dr Dilip K Mishra**

**Purpose:** To study the importance of HIV screening in patients with ocular surface squamous neoplasia (OSSN) and characterize the clinical features, management and histopathological features of OSSN in these cases.

**Methods:** Retrospective chart review of OSSN with HIV infection at tertiary eye center.

**Results:** Of the 200 cases of OSSN screened for HIV, 83 (42%) patients were HIV positive. Of these 83 patients, 57 (69%) of the patients were unaware of their HIV status and OSSN was the sole presenting feature of HIV infection. The mean age at presentation was 41 years. Bilateral involvement was seen in 6 (11%) cases and the male to female ratio was 3:1. The mean duration of symptoms was 16 months. The mean tumor basal dimension was 11 mm and thickness was 3 mm. Limbus was the most common tumor epicenter (77% cases). Orbital involvement was seen in 9% cases. The primary treatment for OSSN included excision biopsy (70%), exenteration (12%), topical chemotherapy with Mitomycin-C (9%), and extended enucleation (5%). Tumor recurrence occurred in 25% cases during a mean follow up period of 10 months. Extended enucleation/exenteration was required in 28% cases. Histopathologically, stromal invasion was evident in 55%, corneal invasion in 19%, scleral invasion in 16%, and orbital invasion in 11% cases.

**Conclusion:** OSSN may often be the presenting sign of underlying HIV infection. We recommend HIV screening in all patients with OSSN.

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Sensitivity of Impression Cytology in diagnosing Ocular Surface Squamous Neoplasia

Dr Pallavi Gupta, Prof Dr Mohd Ather, Dr B Malleshwari, Dr T Kavitha, Gandhi Medical College, Hyderabad

Aim: To evaluate the accuracy of Impression Cytology employing millipore filter paper in diagnosis of OSSN with confirmation by histopathology examination

Introduction: First described by Lee and Hirst in 1995, OSSN encompasses a spectrum of neoplastic lesions, originating from squamous epithelium ranging from simple dysplasia to invasive squamous cell carcinoma (SCC), involving the conjunctiva, limbus and the cornea. Starts in the interpalpebral conjunctiva and then straddles the limbus and may involve cornea(Figure 1). Lesions have a pearly grey appearance with tuft of vessels commonly known as ‘sentinel vessels’ with or without well defined borders (Figure 2).

Egbert et al first introduced ocular surface impression cytology in to ophthalmology in 1977.

Material and Methods: This is a prospective observational & interventional study conducted at a at Sarojini Devi Eye Hospital and Gandhi Hospital. The study included 50 patients presenting with conjunctival mass at the limbus or on the conjunctiva from July 2013 to Feb 2015. Patients with conjunctival or limbal lesions accompanied by feeding blood vessels were included. Impressions were obtained using 0.22micrometre Millipore filter paper (Figure 3) preoperatively from all the patients undergoing excision biopsy for suspected OSSN. The Millipore filter paper was cut in to 3 mm bits by holding the membrane carefully with forceps, to avoid sticking of desquamated cells from the hands.(Figure 4)

Under topical anaesthesia these strips were placed on the ocular surface lesion and pressed gently until the filter paper becomes wet, this takes on an average 10-20 seconds. Then the membrane was carefully peeled off from the lesion and transferred into a container of 95% alcohol immediately
without air-drying. Then the strips were stained with haematoxylin-eosin stain and cytology studied.

In the laboratory the filter paper is stained with Haematoxylin and Eosin stain as follows: the strips are removed from 90% alcohol and dried, stained with Haematoxylin solution for 1 hour and washed under tap water, quickly dipped in acid alcohol followed by a dip in ammonia for 1 second, stained with Eosin solution for 1 second, dehydrated in graded alcohol's 70%, 90% and absolute alcohol with dip in each container for 1 second. Then the paper is thoroughly dried and dipped in Xylene for 10-15 minutes and mounted on glass slides using cover slip. The Millipore filter paper at this stage is fully transparent and allows cytological examination.

Cases were then posted for surgery for wide excision of tumour with 3 mm of normal margin of conjunctiva. Results of impression cytology were correlated with histopathology examination results to know the reliability of impression cytology.

**Results:** A total of 50 excision biopsy of 50 patients suspected for OSSN were performed, there were 40 male patients and 10 female patients. Excision biopsy confirmed the impression cytology in 44 cases and in 6 cases there was poor correlation, among which 4 cases showed mild dysplasia in impression cytology, while histopathology showed invasive squamous cell carcinoma. In 2 cases few dysplastic cells were noted in impression cytology but histopathology showed carcinoma in situ. On analyzing data impression cytology sensitivity was 91.6%, specificity 50% with high correlation rate of 88% with histopathology. It has a positive predictive value of 95.65%, and negative predictive value of 25%.

**Conclusion:** This study shows that impression cytology has a promising role in diagnosing ocular surface neoplasia for its high predictive accuracy compared with tissue histology. However, a fair negative predictive accuracy indicates that impression cytology is a valuable screening technique, but it is not a “gold standard”.

**References:**
1. Lee et al 1994; Newton et al 1996

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Combined intravitreal injection of recombinant tPA (r-tPA) and bevacizumab with SF6 gas for treatment of submacular hemorrhage secondary to AMD

Atul Kumar MD, FAMS; Mayank Bansal MD; Kavitha D

Introduction
Submacular hemorrhage secondary to choroidal neovascularization (CNV) can cause sudden visual loss in patients with age-related macular degeneration (ARMD). Prognosis depends on the size of the bleed; small and thin submacular haemorrhages have better prognosis and treatment is aimed at the underlying choroidal neovascular membrane (CNVM), whereas thick submacular haemorrhage is generally poor.\(^1\)\(^-\)\(^5\) Potential mechanisms of vision loss include toxicity of the released iron on the photoreceptors, shearing of photoreceptors by the contraction of fibrin clots, physical separation of photoreceptors from the retinal pigment epithelium, progression of the underlying CNVM and formation of a macular scar.\(^6\)\(^-\)\(^8\)

In previous attempts to treat massive submacular hemorrhage associated with ARMD, vitrectorbin surgery to remove the subretinal blood clot has been performed. The results of these operations, however, have been disappointing. Surgical removal of submacular bleed and CNVM are no more advised due to poor functional outcomes.\(^1\)\(^,\)\(^8\)\(^,\)\(^10\) Interestingly, several recent studies have indicated that the preoperative or intraoperative use of the fibrinolytic agent recombinant tissue plasminogen activator (r-tPA) may have a beneficial effect, but the true value of this therapeutic approach remains to be investigated. Therefore to avoid surgical manipulation of the macular retina, less invasive procedures have been proposed for displacing submacular hemorrhage like vitrectomy with injection of subretinal tPA and aspiration of liquefied blood,\(^11\)\(^,\)\(^12\)\(^,\)\(^13\) intravitreal tPA with pneumatic displacement,\(^14\) subretinal injection of tPA with pneumatic displacement,\(^17\)\(^,\)\(^18\) intravitreal injection of recombinant tPA with evacuation of subretinal bleed followed by gas tamponade,\(^7\)\(^,\)\(^9\) intravitreal injection of anti-VEGF,\(^15\)\(^,\)\(^16\) subretinal injection of r-tPA during pars plana vitrectomy followed by an intravitreal gas tamponade without evacuation.\(^10\)\(^-\)\(^14\)

One such approach, first described by Heriot (Heriot WJ, American Academy of Ophthalmology Annual Vitreoretinal Update, San Francisco, October 1997), is the injection of intravitreal tissue plasminogen activator and perfluoropropane (C3F8) gas, with the intent to lyse the clot and displace it from the fovea which was further proven by researchers.\(^14\)\(^,\)\(^15\)\(^,\)\(^16\) Haupert et al described the technique of pars plana vitrectomy with subretinal r-tPA injection with pneumatic displacement with gas along with postoperative propped up positioning. Hattenbach L O et al described Intravitreous injection of tissue plasminogen activator along with SF6(6) gas. The efficacy of intravitreous bevacizumab and sulphur hexafluoride (SF6) was described by Hohn F et al.

In our study, we investigated the effectiveness of the modified technique of pars plana vitrectomy with subretinal injection of r-tPA (0.4ml of 12.5mg/0.1ml), bevacizumab (2.5mg/0.1ml) along with 0.3 ml of air. The eye was filled with 20% SF6 followed by postoperative propped up positioning.\(^19\)

Materials and Methods
A retrospective chart review of 10 patients with thick submacular haemorrhage secondary to wet AMD treated with 23G parsplana vitrectomy, subretinal injection of r-tPA, bevacizumab and air with postoperative propped up positioning was done. All were consecutive patients, to avoid any selection bias.

The size of the submacular haemorrhage was 4 to 5 mm in greatest dimension. In all the cases the haemorrhage was involving the fovea extending towards the arcade. The eyes with haemorrhage superior to fovea were not excluded as subsequent to submacular injection of tPA, bevacizumab and air with postoperative propped up position, the haemorrhage gravitated towards the ora serrata at 6 o’clock position.

The bleed was subretinal in 8 cases and only 2 cases had sub RPE bleed along with subretinal bleed. We excluded the cases with major component of subRPE bleed. The thickness of subretinal bleed varied from 500 microns to 750 microns.

Prior to the procedure the solution of r-tPA (Boehringer Ingelheim Pharma GmbH & Co, KG, Germany) was prepared using 20mg r-tPA dissolved in 20 ml of sterile water to give a concentration of 1mg/ml. The solution was further diluted to get a final concentration of 12.5µg/0.1 ml. A 1ml tuberculin syringe was used to load the constituents to be injected. 0.3ml of sterile air was first drawn into the syringe using an air filter. This was followed by loading of 0.1ml (2.5mg) of bevacizumab (Avastin, Genentech, South San Francisco, CA, USA), and finally 0.4ml (12.5µg/0.1ml) of reconstituted r-tPA.

A standard 3 port 23 gauge vitrectomy was done with posterior vitreous detachment under local peribulbar anesthesia. A 41 gauge needle attached to a tuberculin...
syringe (loaded with 0.4 ml r-tPA, 0.1 ml bevacizumab and 0.3 ml air) was used to make a retinotomy superonasal to the submacular haemorrhage. The mixture of r-tPA, bevacizumab and air was injected to create a localised bullous detachment encompassing and extending little beyond the haemorrhage. An air fluid exchange was done and 20% SF₆ gas was injected. The patients were placed propped up for 3-4 days. The patients were followed up at 6 weeks, 3 months and 6 months post-operatively. Pre-operatively, demographic data including age and gender, duration of hemorrhage, preoperative BCVA, anterior segment and fundus examination details were noted. The BCVA was measured with the help of Snellen’s visual acuity chart and then converted into logMAR units. In patients with low vision, counting fingers, hand movements vision was converted in logMAR. Post-operatively, BCVA and displacement of submacular hemorrhage were noted. Investigations included preoperative and postoperative Spectral Domain Optical Coherence Tomography (SD-OCT, Cirrus, Carl Zeiss Meditec), colour fundus photography and fundus fluorescein angiography. A thick submacular haemorrhage was defined as one causing foveal elevation on stereoscopic fundus photography and also extending till the arcades. (Fig 1a & b; 2a & b.)

Results:
Out of 10 patients, 6 were males and 4 were females. The mean age was 66.9 years (range between 56 to 82 years). All patients presented with sudden onset diminution of vision. The average duration from the onset of symptoms to the presentation was 5 days (range between 1 to 11 days). Of the 10 eyes, 4 eyes had history of receiving anti-VEGF therapy prior to the onset of submacular bleed. Mean preoperative BCVA was 1.45 ± 0.3 logMAR units, median 1.47 (range 1.77- 1.07). No intraoperative complications were noted. Table I shows the summary of the patients. All patients were followed up at 6 weeks, 3 months and 6 months. Displacement of the submacular bleed was achieved in all eyes on the 1st postoperative day. All the patients had a complete displacement of the blood from the macula with the inferior displacement of the haemorrhage and subretinal fluid. Mean post-operative BCVA was 0.95 ± 0.4, 0.86 ±0.5 and 0.82±0.6 logMAR units at 6, 12 and 24 weeks respectively. Improvement of vision was defined as more than 2 snellen line improvement. Deterioration was defined as loss of more than 2 lines. There was a significant improvement of BCVA from 1.45±0.3 logMAR units preoperatively to mean 0.95±0.4 logMAR units (median 1) at 6 weeks (p=0.007), 0.86±0.5 logMAR (median 0.97) at 3 months (p=0.01) and 0.82±0.6 logMAR units at 6 months (median 0.62) (p=0.01).

At the end of 6 weeks and at 3 months follow up, best postoperative visual acuity compared with preoperative visual acuity was improved two or more Snellen lines in 9 eyes (90%). Compared to the 6 weeks follow up, 4 (40%)
had improvement in BCVA, 5 patients (50%) did not show any improvement and 1 patient (10%) had deterioration of BCVA at 3 months who subsequently underwent photodynamic therapy with anti VEGF injections. The reason of deterioration of vision in 2 cases was disciform scar. At the end of 6 months follow up, best postoperative visual acuity compared with preoperative visual acuity was improved two or more Snellen lines in 8 eyes (80%) and deteriorate in 2 eyes (20%). Two of the 10 eyes had a rebleed and underwent re-surgery with intravitreal SF6, r-tPA and bevacizumab. In both patients, displacement of subretinal bleed was achieved following resurgery.

Three out of the 4 patients who received prior antiVEGF therapy had discontinued the treatment 3 months prior due to noncompliance. One of the 4 patients had a subretinal bleed one month after the injection. There was no RPE tear in any of the cases. None of the patients were on anticoagulants. Two of the patients had a rebleed. In cases of rebleed, intra-vitreal rather than sub-retinal injection was preferred to minimise the manipulation of the macula thus preventing further damage. Intravitreal tPA (0.2 ml of 12.5mg/0.1ml) with bevacizumab(1.25mg/0.05ml) and with 20% SF6 gas was injected immediately in cases of rebleed.

Discussion

Submacular haemorrhage in AMD is associated with variable visual prognosis depending upon the duration, density of hemorrhage and associated complications. Thick submacular haemorrhages usually carry a poor prognosis. Different treatment approaches have evolved for their treatment. To date, the most commonly used approach has been that of pars plana vitrectomy and aspiration of submacular blood, with or without intraoperative clot dissolution by subretinal tPA. The advent of r-tPA has revolutionised the management of submacular subretinal bleed.

The technique of subretinal injection of r-tPA along with pneumatic displacement was described by Haupert et al. Studies published in the literature describing the role of subretinal r-tPA and gas have shown favourable results in displacing subretinal haemorrhages with minimal complications. However, this technique involved sequential management of the underlying pathology after clearance of the sub-retinal bleed rather than simultaneous management. Joseph N. Martel et al described a modified technique of managing CNVM with subretinal haemorrhage with a 23 gauge pars plana vitrectomy with subretinal injection of 0.4 ml of 12.5 µg/0.1 ml of recombinant t-PA with bevacizumab and air. This technique has the advantage of displacing the subretinal hemorrhage and treating the underlying pathology simultaneously, halting the progression of the neovascular membrane. The rationale is to target both the bleed and the underlying pathology. In the study done by Joseph N. Martel et al they hypothesized the mechanism of each component of the injection. They described the role of subretinal air in decreasing the buoyancy of the subretinal bleed which facilitated the effective displacement of the subretinal bleed.

Based on this hypothesis, we investigated the proposed modified technique in 10 eyes with good results. All patients underwent pars plana vitrectomy, injection of tissue plasminogen activator, bevacizumab and air directly into the subretinal clot, and fluid-gas exchange, followed by prone positioning. The direct injection of subretinal r-tPA gave us a direct access to the bleed for clot liquefaction. A 41 gauge translocation needle was used to create a localised bullous neurosensory detachment covering the subretinal bleed which provided a potential space for the displacement of the liquefied clot. Experimental data has shown that displacement of subretinal blood by intravitreal gas without preliminary clot liquefaction causes irreversible damage to retinal photoreceptors attached to the subretinal clot.

Pneumatic displacement of subretinal hemorrhage damages the retinal photoreceptors. Paper presented at: Macula Society Annual Meeting, 2003; Naples, Florida). This is overcome by directly injecting r-tPA in the subretinal space.

Although we manage the subretinal bleed with the direct injection of r-tPA in the subretinal space but the underlying pathology is not tackled and might cause rebleeds. To prevent that, we also used subretinal bevacizumab (2.5mg/0.1ml) along with r-tPA. F. Treumer et al described the efficacy of pars plana vitrectomy with subretinal co-application of r-tPA and bevacizumab in cases of subretinal bleed associated with CNVM. Although the intravitreal injection of bevacizumab suffices to produce therapeutic effect on the CNVM, the subretinal application ensures the direct delivery of the entire drug to the pathology. The pharmacokinetics of intravitreal bevacizumab in a vitrectomised gas filled eye is not known, hence we used bevacizumab injection in a higher dose directly in the subretinal space. The bevacizumab injection prevents the recurrence of submacular bleed.

The pneumatic displacement of the subretinal bleed was achieved by subretinal injection of 0.3 ml air and intravitreal injection of 20% SF6 gas. The subretinal air complemented the intravitreal SF6 gas to displace the blood from the subretinal space and also prevented the superior migration of the subretinal gas towards the ora serrata.

Complete pneumatic displacement of the subretinal bleed from the macula was seen in all the patients on the first postoperative day. At 6 months follow up, 8 patients had improvement in BCVA of more than 2 lines. Two patients had a rebleed who were further managed by intravitreal recombinant t-PA, bevacizumab and SF6 gas. These patients had partial displacement of the subretinal haemorrhage after the resurgery and slight improvement of BCVA. One
patient presented with an active membrane at 3 months follow up which required a photodynamic therapy with anti-VEGF injections following which the patient had an improvement of BCVA at 6 months.

No other serious complications were seen with the procedure. The 41 gauge translocation needle creates a self-sealing retinotomy which negates the risk of rhegmatogenous retinal detachment and epiretinal membrane.

In conclusion, this is the first study of its kind in the Indian subcontinent in which cases of CNVM with submacular haemorrhage were managed with 23 gauge pars plana vitrectomy with subretinal injection of r-tPA (12.5μg/0.1ml), bevacizumab (2.5mg/0.1ml) and 0.3 ml of air followed by 20 % SF6 gas injection. The 41 gauge needle is used to create a localised bullous neurosensory detachment at the macula which provides a potential space for the pneumatic displacement of the submacular haemorrhage. In addition, use of bevacizumab prevents recurrence of CNVM. The study is limited by its retrospective nature and number of patients. A larger randomized control trial will further elucidate the role of this technique in the future.

References

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Polypoidal Choroidal Vasculopathy: A Review Of Treatment Strategies

Dr Anand Rajendran, FRCS, DNB; Dr Pushpanjali Ramteke, MS

Abstract:
The last few decades have broadened our understanding of polypoidal choroidal vasculopathy (PCV) as a unique form of age-related neovascular maculopathy characterized by subretinal and sub-retinal pigment epithelial bleeding. This was distinct from the classical clinical profile of exudative age-related macular degeneration (ARMD). The natural history of PCV is typically a remitting-relapsing one, with the long term vision being relatively better preserved than in ARMD. Clinically PCV is highlighted by the presence of multiple serosanguineous retinal pigment epithelial and neurosensory retinal detachments. PCV has a higher prevalence in eastern Asiatic populations than in western ones. While optical coherence tomography (OCT) is of high utility, Indocyanine green angiography (ICGA) has emerged as the investigative modality of choice as it delineates the pathognomic choroidal polyps and branching choroidal vascular patterns the best. A variety of treatment options such as laser photocoagulation, photodynamic therapy, anti-vascular endothelial growth factor (VEGF) agents have been used to manage PCV. We review the various therapeutic strategies in an effort to understand the best manner in which we may treat this entity.

Introduction:
Polypoidal choroidal vasculopathy (PCV) as a cause of recurrent haemorrhagic and exudative pigment epithelial detachments (PED) and neurosensory retinal detachments was first described in 1982 [1]. Haemorrhagic detachments of the macula were believed to be part of the spectrum of age-related macular degeneration (ARMD) until the description of this unique disease of choroidal vasculature. It was Yanuzzi et al who first described and coined the term “Polypoidal Choroidal Vasculopathy” about three decades ago [1]. In the past, a variety of terms such as “posterior uveal bleeding syndrome” [2], “multiple recurrent retinal pigment epithelial detachments in black women”[3], and polypoidal choroidal neovascularization (CNV)[4] have been used to designate this disorder.

PCV is a disorder with choroidal vascular abnormalities [1] a clinical entity distinct from ARMD and has recently been recognized as a type of CNV [5, 6]. The pathognomonic lesion is an inner choroidal vascular network of vessels ending in an aneurysmal bulge or outward projection, visible clinically as reddish-orange, spheroid, polyp-like structures [1]. Clinically multiple, recurrent, serosanguineous detachments of the RPE and neurosensory retina are seen secondary to leakage and bleeding from these peculiar choroidal vascular lesions.

PCV is predominantly seen in middle aged to elderly population, most commonly in patients aged 50-60 years though age group can range from 20-80 years [6, 7]. Initially considered to be a rare condition, reported prevalence of PCV ranges from 22 to 55% of newly diagnosed ARMD and exudative ARMD in eastern Asian population groups [8, 9,10]. In contrast, PCV was identified in 4 to 10% of cases of ARMD in Caucasians [8, 11,12]. Interestingly, in Caucasians, there is a female predominance, while males were more commonly affected in Asian populations [7].

The superiority of indocyanine green angiography (ICGA) over fluorescein angiography (FFA) for the accurate diagnosis of PCV is well established [13,14,15]. ICGA clearly shows the abnormal vascular structure, consisting of a branching vascular network (BVN) and polypoidal lesions characteristic of PCV [1,14,15] as the lesion can be mistaken on FFA for occult CNV or minimally classic CNV secondary to ARMD. Neither are the characteristic polyps highlighted as well on FFA as they are on ICGA. The branching vascular network vessels are often described as a “plaque” on ICGA and regarded as the Type 1 CNV [14,15]. On optical coherence tomography (OCT), the polypoidal lesions appear as a dome-shaped anterior elevation of highly reflective retinal pigment epithelial (RPE) layers with regions of low to moderate reflectivity beneath the RPE line. PCV is characterized by a higher incidence of retinal PEDs, subretinal fluid, and less intraretinal fluid than eyes with exudative ARMD [16].

In PCV, the visual prognosis has been reported to be better than that of exudative ARMD, and a conservative approach is preferred, unless the lesion is associated with persistent or progressive exudative changes threatening central vision [17]. There is an omnipresent risk of subretinal fibrosis and RPE atrophy causing significant and permanent vision loss [18]. Moreover, the incidence of sub-RPE hemorrhage or subretinal hemorrhage is high (30%–64%) in patients with PCV [19,20]. Controversy still remains over the best way to manage the active forms of this disease.

Management Strategies
1. Observation
A conservative approach can be considered in eyes where a benign clinical course is anticipated [21]. Okubo A et al [21] identified, in 4 eyes with PCV, a pattern of reddish-orange nodules alone, or nodules and small subretinal
haemorrhage and followed them for a period of 5 yrs. They found that eyes with this pattern and an absence of hard exudates maintained a benign clinical course or stable vision. Although extensive data on the natural history of PCV is not available but the published data so far suggests unfavourable outcomes with observation alone.

Yuzawa et al [22] reported that only five of 21 eyes (24%) with PCV involving the macula had visual improvement with conservative management over a follow-up period of 3 years and Uyama et al [23] observed that five of 14 eyes (36%) lost more than two lines of vision over a follow-up period of 2 years. Hence, in most instances, where patients have significant symptomatic visual loss, along with subretinal fluid and exudates, treatment is often considered [24].

2. Laser Photocoagulation:
Laser photocoagulation of polypoidal lesions for the treatment of symptomatic serosanguineous maculopathy seems to offer the most benefit for patients with extrafoveal lesions [25]. Yuzawa et al [22] reported visual improvement after conventional laser treatment of the entire PCV complex in nine of 10 eyes. They found photocoagulation of the whole lesion to be more efficacious compared to treating the polyps only.

In a larger study by Lafaut et al [10] fourteen eyes received laser photocoagulation to the polyps. In the five peripapillary lesions they treated, all the polyps regressed with resolution of the fundal lesions. However, among the nine eyes with polyps in the macula or along major vascular arcades, similar success was only achieved in five eyes. Uyama et al [23] performed laser photocoagulation in 17 eyes in which 12 eyes (71%) that had clinical improvement with a decrease of haemorrhage and subretinal fluid. In various studies that analyzed reported vision outcomes, ICGA-guided thermal laser photocoagulation was successful in stabilizing or improving vision in 55% to 100% of eyes [24,25,26,27].

However, no randomized, controlled studies have been performed to prove the efficacy or safety of laser treatment. Moreover, when the lesion extends subfoveally, thermal laser treatment is not indicated due to damage of the overlying neurosensory retina and subsequent reduction in central vision.

Few eyes with extrafoveal polyps may present with foveal exudative maculopathy.

Cheung et al [28] did a prospective study in Asian eyes with extrafoveal polyps including eyes with fluid or blood affecting the fovea at presentation. They found that argon laser therapy with selected use of adjunctive anti-VEGF therapy achieves stable or improved visual outcome in 90.3% of eyes.

3. Photodynamic therapy (PDT)
Photodynamic therapy for subfoveal lesions seems to be the best treatment modality till date [29]. In PDT, the mechanism of selective photothermolysis causes regression of the polyp and allows pathological manifestations of PCV to improve gradually [37]. Quaranta et al [29] first described beneficial functional results in two cases without acute recurrence during a follow-up period of 12 months.

In their retrospective review of 16 patients, Spaide et al [30] reported that 88% of the patients had stable or improved vision after a mean follow-up of 12 months.

Chan et al [31] conducted a prospective study of 22 eyes in 21 patients. At the 12-month follow-up, they observed stable or improved vision in 95% of the eyes with a mean visual improvement of 1.3 lines after a mean of 1.6 PDT sessions. They observed total regression of the polypoidal lesions on ICGA in 95% of the eyes, and noted recurrence in only one eye (5%) during the study period.

Lee et al [32] reported similar clinical and angiographic outcomes in nine patients with no recurrence during a follow-up of 3–18 months. A common limitation of these studies is the relatively short follow-up period, considering the natural history of PCV.

In another prospective study demonstrating the effectiveness of PDT in the treatment of PCV, Silva et al [33] reported recurrence in one (5%) of 21 eyes during the first year of follow-up. However, among the six eyes that completed 2 years of follow-up, there was recurrence in four (67%) during the second year.

In the EVEREST trial [18], both combination therapy and verteporfin PDT monotherapy were found to be superior to ranibizumab monotherapy.

PDT with verteporfin is the treatment most commonly used for PCV, but its efficacy and safety have been questioned. There is recurrence of polyps requiring frequent retreatment [34,35,36]. In the EVEREST trial, in the verteporfin PDT monotherapy arm 14.3% had retinal and 4.8% patients had vitreous hemorrhage [18]. PDT has also been shown to cause choroidal ischemia inducing a secondary angiogenic response with increased expression of VEGF [37].
Figure 1: depicts a case of PCV who underwent PDT treatment

Figure 2 shows the resolution of the PCV with PDT 1 year post-treatment. Newer approaches such as reduced fluence PDT [38] and, more recently, the combined use of VEGF inhibitors, by overcoming these risks, can serve as viable alternatives.

4. Intravitreal anti-VEGF agents

Anti-VEGF therapy has proven effectiveness against CNV related to ARMD [39,40]. The pathogenesis of PCV being controversial, the exact role of anti-VEGF agents in treatment of PCV remains a debatable one [41]. Compared with the CNV seen in ARMD, the vascular changes associated with PCV are more structured and mature and this might explain their limited response to standard anti-VEGF therapy [42]. Matsuoka et al [43] have showed strong expression of VEGF in the PCV specimens, and Tong et al [44] have reported upregulation of VEGF in the aqueous humor in eyes with PCV.

In a comparative, retrospective study by Saito M et al [45], ranibizumab monotherapy was more effective than verteporfin PDT monotherapy in the resolution of retinal fluid at 6 months. In a larger prospective case series by Hikichi et al [46], the response of polypoidal lesions to ranibizumab monotherapy was better, with resolution or improvement in 78% eyes, but the BVN was nevertheless unchanged or worsened in 77.1% eyes.

Inoue et al in their study found that repeated IVR was effective not only for suppressing exudative changes but also for the involution of polyp lesions and had better functional outcome as compared to PDT during a long-term follow-up period of 2 years [47].

Similar results were seen in recent LAPTOP study; a 24-month trial of Ranibizumab (Lucentis) And Photodynamic Therapy On Polypoidal choroidal vasculopathy which compared the vision enhancing effect of ranibizumab and PDT in 97 patients with PCV [48]. The result was similar to that obtained after 12 months of this trial [49] that continuous anti-VEGF therapy can achieve better visual outcome and prevent further visual loss in comparison with PDT monotherapy.

Gomi et al [41] administered intravitreal injections of bevacizumab in 11 eyes with PCV with relatively good vision. Three months after injection, the choroidal vascular abnormalities remained on ICGA in 10 of 11 eyes, although the exudation decreased temporally in most eyes. Ghajarnia et al [50] reported the efficacy of bevacizumab for a patient with PCV refractory to a previous PDT treatment and intravitreal pegaptanib. The weak RPE after previous treatments in this case may have allowed penetration of bevacizumab into the sub-RPE space. Kim et al [51] have found intravitreal bevacizumab to be effective in resolving intraretinal and subretinal fluid originating from polypoidal lesions. However, the favorable effect was maintained for only a limited period of time and required repeated injections.

Cho et al [52] report that intravitreal injections of bevacizumab and ranibizumab have similar effects in stabilization of visual acuity, macular edema, and regression of polypoidal complex with PCV eyes.

Aflibercept, a recombinant fusion protein, has a higher binding affinity for VEGF, and it has been shown to be beneficial for patients with ARMD who were refractory to multiple injections of either bevacizumab or ranibizumab [53, 54].

In addition, recent studies had shown that intravitreal aflibercept may be an effective treatment for serous PEDs in which bevacizumab and ranibizumab were not effective [55, 56].

In a recent study by Inoue et al, intravitreal aflibercept improved the vision of treatment-naive patients with polypoidal choroidal vasculopathy [57]. In this
short term follow-up study intravitreal aflibercept led to involution of polyps and exudation.

Large comparative studies are awaited for proving superiority of one agent over another.

5. Combination Therapy

Combining PDT with an anti-VEGF agent creates a synergistic effect leading to polyp regression and reducing fluid leakage and inflammation [58]. These simultaneous effects enhance the treatment benefits. In a study by Ruamviboonsuk et al [58], combined therapy of PDT with ranibizumab showed was found to be more effective with respect to the improvement of vision when compared to PDT monotherapy. In Lai et al’s study [59], adding PDT to bevacizumab monotherapy was associated with better results than bevacizumab therapy alone.

Combination treatment has been efficacious in lesions with previous unsuccessful therapy and in those with the development of “resistance” to therapy. Significantly improved mean BCVA following combination therapy with verteporfin PDT and ranibizumab at 3 and 12 months was reported by Tomita et al [60] in a recent retrospective chart review, although this was so only in treatment-naïve eyes.

In a study by Romano et al [61], in eyes with recurrence after previous verteporfin PDT therapy combined therapy stabilized vision but VA improvement was not seen. The EVEREST trial [18] found both combination therapy and verteporfin PDT monotherapy to be superior to ranibizumab monotherapy.

In meta-analysis of comparative studies done by Wei wang et al, combination therapy resulted in better visual acuity than PDT alone [62].

Thus, as per the recommendations by Koh et al, a combination of verteporfin PDT and ranibizumab should be considered in the following clinical scenarios –a) when there is leakage from the BVN as well as polyps; b) there is a large amount of subretinal fluid or exudation associated with PED; c) ICGA features are ambiguous between PCV and CNV; or d) lesions are a combination of PCV and typical CNV [63].

In conclusion, this review on the management of PCV recommends ICGA in the diagnosis and monitoring of PCV and considers ICGA-guided verteporfin PDT with or without combination with ranibizumab as the current preferred therapeutic option. However, there still remain unanswered issues such as - the efficacy of PDT combined with ranibizumab versus PDT and bevacizumab; the efficacy of PDT along with intravitreal steroids; the management of polyp and BVN recurrence or persistence post-PDT – all of which necessitate larger randomized clinical trials.

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Legends:
Figure 1a – The fundus picture of a patient with PCV ; 1b – the ICGA highlighting the polypoidal lesions; and 1c – the OCT (with an oblique line scan passing through the fovea and PED) demonstrating the subretinal fluid and exudation. Figure 2a – The clinical picture highlighting the regression of the PCV as further demonstrated by; 1b – the ICGA; and 1c – the OCT.

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### Optical Coherence Tomography (OCT)

Optical coherence tomography (OCT) is a rapid non-contact method that allows in vivo imaging of the retina, choroid, optic nerve head, retinal nerve fiber layer (RNFL), and the anterior segment structures of the eye. It provides high-resolution cross-sectional images of tissue morphology both in situ and in real-time (Figure 1). Owing to the advancement in the OCT technologies from time domain-OCT (TD-OCT) to spectral domain-OCT (SD-OCT), early detection, monitoring and prognosis of the disease are possible.

OCT measures the echo delay and intensity of backscattered light from the internal tissue microstructure. Because the echo time delays of light are too fast to measure directly, an optical correlation technique, known as Michelson low coherence interferometry, is used. Low-coherence light from a super luminescent diode (SLD) is directed through a beam splitter and divided into a sample beam that is focused onto the patient’s retina and a reference beam that travels a calibrated delay path. Light backscattered by retinal structures interferes with light from the reference beam, and the interference of the echoes is detected to measure the backscattering signal versus delay or depth. However, the OCT images are not the direct depiction of the anatomical structures, but they represent the consequence of the optical properties of the tissues being scanned. In SD-OCT, information on depth is transformed from the frequency domain to the time domain, without using a moving reference mirror which allows the image to be acquired rapidly, about 6 & at times faster than with TD-OCT. SD-OCT units allow the improvement of the detection and monitoring of retinal diseases, because these ones have ultrahigh-speed scan rate, superior axial and lateral resolutions, cross-sectional (2D) scan, 3D raster scanning, and a higher imaging sensitivity than the traditional TD-OCT units.

### Ultra High Resolution OCT (UHR OCT)

On a commercial basis, further improvement of B-scan images has been achieved by using UHR SD OCT with an axial resolution of 2-3 \( \mu \)m and speckle noise-reduction technology with SD OCT. This resulted from improvements in light-source technology, replacing the SLD with a broadband femtosecond titanium:sapphire laser and later with multiplexed SLDs. This advance enabled improved delineation of intraretinal layers. However, the expensive cost of the titanium-sapphire laser light source hinders wide commercial use of this technique.

### Enhanced Depth Imaging OCT (EDI OCT)

Recently, a technique termed ‘enhanced depth imaging’ (EDI) has been described by Margolis and Spaide, which uses the commercially available Spectralis HRA and SD OCT device (Heidelberg Engineering) to obtain an enhanced view of the choroid. By placing the OCT machine close enough to the eye to acquire an inverted image within a 5°-30° area centered at the fovea and then performing manual measurements from the outer border of RPE to the inner sclera border the OCT scan (Figure 2).

### Enface OCT

Podoleanu et al. developed a system combining confocal ophthalmoscopy and OCT, producing en-face OCT images that can be directly compared with transversal images provided by the confocal channel. En-face OCT enables the production of transversal C-scans in the X/Y plane at a fixed Z coordinate. By changing the Z coordinate in the OCT channel, OCT C-scans are taken at different depths in the retina.
Combined Depth Imaging (CDI OCT)

In 2013, Barteselli et al.16 described a technique called the Combined Depth Imaging (CDI) in OCT. The CDI gives an average 100 scans where the first 50 scans enhance the vitreoretinal interface while the remaining 50 scans enhance the choroid. It, thereby giving the advantage of both the conventional SD OCT and EDI OCT (Figure 3). It is easy, fast, and sensitive enough to visualize posterior vitreoretinal and choroidal structures together in a single image in case of posterior uveitis.17

CDI OCT: Figure 3: showing (a) Fundus photo of a patient with inferotemporal tubercular choroid granuloma with localized serous detachment. (b),(c),(d) shows the conventional SD OCT, EDI OCT and CDI OCT respectively. Note that while conventional SD OCT shows posterior vitreous detachment and fails to highlight posterior border of the hypo reflective area in the choroid, the EDI OCT gives posterior border but fails to show the PVD whereas CDI OCT enables detection of both the PVD and hypo reflective area of choroid with full visualization of the posterior border.


Applications of OCT in Uvea

Anterior Segment Applications:
The Anterior Segment OCT has been used in few studies to give cross sectional images of the anterior chamber. Using a longer wavelength of 1310 nm, better visualization of the anterior chamber is possible.1 The corneal thickness measurement in corneal edema, anterior chamber exudates and cells, angle of anterior chamber, anterior synechiae can be visualized using the AS OCT. Moreover, its usefulness in monitoring of these signs can also prove to be a useful tool.

Posterior Segment Applications:

1. Uveitis Macular Edema

OCT has become a standard diagnostic technique for the evaluation of uveitic macular edema and measuring the retinal thickness and monitoring response of the treatment (Figure 4). Gupta et al and Roesel et al performed studies comparing the TD OCT with SD OCT in Uveitic Macular Edema and showed the superiority of SD OCT in the diagnosis. Markomichelakis et al.19 using OCT 1, first described three different patterns of distribution of fluid in the macula of patients with uveitis—cystoid macular edema (CME), diffuse macular edema (DME), and serous retinal detachment.OCT has been used for detecting, monitoring and assess the therapeutic response of uveitic Macular Edema20-24. OCT is also highly sensitive in detecting ERM and potential vitreoretinal abnormalities in the macular area, suggesting the hypothesis of a tractional mechanism as a probable origin or cofactor of the onset of macular edema during uveitis.

Uveitic Macular Edema. Figure 4: SD-OCT reveals posterior vitreous cells, posterior vitreous detachment, loss of foveal contour, multiple hyporeflective cystic spaces with intervening high-reflective septae with subfoveal serous retinal detachment along with central retinal thickness of 543 microns in right eye.

2. Vogt-Koyanagi-Harada disease (VKH)

VKH is a granulomatous inflammatory disease affecting the eye, meninges and skin. The main site of inflammation is choroid. Many studies have been done to show the OCT features of VKH using the TD OCT, SD OCT and now EDI OCT. Onal et al extensively described the various OCT features of VKH in acute and recovery phase in all layers. In acute phase, an increase in choroidal thickness leading to RPE undulation and subretinal fluid accumulation is seen.25-28. Also intraretinal cystoid spaces and irregularity in the ellipsoid zone (previously known as IS/OS junction) is seen in the outer retinal layers.29-31 In recovery phase, there is decrease in choroidal thickness seen. Also, peripapillary atrophy showing thinning of RPE/Bruchs membrane with loss of ellipsoid zone, ELM and ONL is seen in VKH. In chronic VKH, loss of stromal choroidal vessels, stromal scarring and choroidal atrophy can be visualized using the EDI OCT (Figure 5).
VKH: Figure 5 showing Spectral Domain OCT image in a case of acute VKH disease with serous retinal detachment. Because of the increased retinal thickness posterior border of the choroid could not be made out.

3. Ocular Tuberculosis
Ocular Tuberculosis encompasses any infection caused by Mycobacterium tuberculosis or any of the three related mycobacterium species. It can be primary or secondary. One of the most common manifestations of Ocular TB is Uveitis in form of either chronic anterior uveitis, panuveitis or choroiditis. EDI has proven as an useful tool for detection of choroidal granulomas in TB (Figure 3). In a study done by Invernizzi et al 32, they showed the EDI OCT appearance of granuloma as homogenous and with increased transmission of OCT signal compared to the surrounding choroid. They also showed that most of the tubercular-related lesions showed lobulated shape and nonhomogeneous internal pattern. Further, Salman et al 33 in their study showed the “contact sign” on OCT in cases of choroidal granuloma which is an attachment between the retinal pigment epithelial- choriocapillaris layer and the neurosensory retina over the granuloma.

4. Ocular Toxoplasmosis
Ocular Toxoplasmosis is characterised by focal retinochoroiditis, an adjacent retinochoroidal scar and moderated to severe vitreous inflammation. Imaging of posterior segment on OCT shows increase intraretinal reflectivity corresponding to area of retinitis with shadowing of underlying choroid, posterior hyaloid thickening and detachment over the lesion and irregular hyperreflective formation (Figure 6). Ore Òrice et al.34 examined the vitreoretinal morphology in patients with active punctate, focal, and satellite ocular toxoplasmosis lesions and observed thinning of retina at the active punctate lesions and thickening in focal and satellite lesions. Also in a study done by Garg et al 35 showing the OCT features in congenital ocular toxoplasmosis showed retinal thinning, RPE hyperreflectivity, excavation and intraretinal cysts. Toxoplasmosis. Figure 6: showing Horizontal line scan on SD-OCT in a patient with Ocular Toxoplasmosis, which reveals presence of posterior vitreous cells with incomplete PVD; hyper-reflective lesion in the retina suggestive of active retinitis with hyporeflective lesion in the outer retina, which is suggestive of fluid-filled spaces with hyperreflective area corresponding to area of chorioretinal scar in outer retina.

5. Sympathetic Ophthalmia
In a study done by Gupta et al studying OCT features of Sympathetic Ophthalmia in 6 patients revealed the involvement of photoreceptors in acute stage along with serous retinal detachments with elongation of photoreceptors. Moreover, disruption of the IS/OS and OS/RPE (COST) junction lines was also seen. There was absence of any intra retinal cystic spaces at presentation. On follow up, there was a resolution of serous retinal detachment as well as of photoreceptors and IS/OS junction line.

The characteristic Dallen Fuchs nodule appears as discrete nodules at level of RPE and associated with mild shadowing and overlying detachment of neurosensory retina on OCT.36 (Figure 7)

6. Sarcoidosis
Ocular Sarcoidosis is primarily a disease of choroid where non caseating inflammatory granulomas are commonly seen. Choroidal granulomas are seen as elevated yellowish-white subretinal masses with fuzzy, ill-defined margins. 37 The key OCT features in this disease are detection of granulomas as homogenous, hyporeflective, well demarcated choroidal lesion.32,38 (Figure 8). With a normal / loss of vascular...
pattern of adjacent areas. Using OCT, we can assess the treatment response of the patients in Sarcoidosis.

Sarcoidosis: Figure 8: (a) The color fundus montage photograph of the left eye shows an orange-yellow lesion superotemporal to the optic disc, suggestive of a choroidal granuloma. (b) Conventional OCT scan shows the presence of posterior vitreous detachment with hyporeflective area in the choroid. The posterior extent of the hyporeflective area could not be made out. (c) The EDI technique clearly demarcates the posterior border of the hyporeflective area. However, the visibility of the posterior vitreous is compromised to a certain extent. (d) The CDI technique demonstrates the entire extent of the hyporeflective area without significantly compromising the posterior vitreous visibility.


7. Behcet’s Disease

Behcet’s disease is a chronic, recurrent, inflammatory systemic occlusive vasculitis affecting both arteries and veins in all organs. It is characterised by triad of recurrent oral and genital ulcers, ocular inflammation and skin lesions. Ocular manifestations in the form of anterior uveitis, posterior uveitis, optic neuropathy and retinal vasculitis is generally seen in BD. Various studies shows the involvement of choroid in terms of diffuse and focal infiltrates with inflammatory cells in choroid leading to increase choroidal thickness which can be determined using EDI OCT (Figure 9). In a study done by Kim et al 39 they found subfoveal choroidal thickening during the active phase of the Behcet’s posterior uveitis. Also the subfoveal choroidal thickness was significantly greater in the quiescent phase compared to normal eyes using EDI OCT.

Behcet’s Disease. Figure 9: shows Fundus photo (a), conventional SD OCT (b), EDI OCT (c) and CDI OCT (d) of a patient with Behcet’s disease. The conventional OCT scan shows posterior vitreous cells but fails to highlight the outer border of choroid while EDI shows outer border of the choroid but fails to show the posterior vitreous. In contrast, CDI shows the presence of posterior vitreous cells as well as good visibility of the outer choroidal border.


8. White dot syndromes

i) Serpiginous Choroiditis.

Serpiginous choroiditis (SC) is an inflammatory disease characterized by a geographic pattern of choroiditis extending from peripapillary area and affects the overlying retinal pigment epithelium (RPE) and the outer retina. Nazari et al in the article Serpiginous choroiditis and Multifocal choroiditis described the OCT features of Serpiginous Choroiditis. The characteristic OCT features of Serpiginous choroiditis in active lesion include normal or slightly increased retinal thickness 40-42 with disruption of photoreceptor layer along with hyperreflectivity in the outer retinal and choriocapillary layer.40,43 Also, the outer retina shows a uniform increased reflectivity sparing the inner retina. The retinal and RPE inflammation in active choroiditis is often associated with limited subretinal fluid overlying the area of choroiditis. In healed choroiditis, the retinal thickness is mildly attenuated owing to the outer retinal atrophy. Also, the outer retina appears as granular and non uniform hyperreflective area. The choroid appears as hyperreflective and it is described as “waterfall effect” due to inflammatory cell infiltration of choroid 40. Also OCT can help to differentiate the Serpiginous choroiditis where the disease limited to the outer retina from multifocal choroiditis where full thickness retinal inflammation is seen.42

ii) Acute Posterior Multifocal Placoid Pigmentary Epitheliopathy
Lofoco et al.44 described the OCT findings in one patient with APMPPE. The characteristic OCT features in APMPPPE includes hyperreflective photoreceptor layer with back scattering, alteration in the COST junction line and subretinal fluid or intraretinal cysts in outer layers of retina. However, the recovery phase shows atrophy of photoreceptors with disruption of RPE and COST junction line.

iii) Acute Zonal Occult Outer Retinopathy
Using SD OCT, Spaide et al45 described the features of AZOOR which included peripapillar ellipsoid zone defects in eyes with blind spot enlargement. Other features seen on OCT are thinning of inner nuclear layers, loss of photoreceptor bodies in outer nuclear layer, indistinctive COST junction line with a normal choroidal thickness in active disease state. In recovery state, OCT shows absence of COST line with decrease in retinal thickness due to shortening of outer nuclear layer and thinning of inner nuclear layer.

iv) Multiple Evanescent White Dot Syndrome
The characteristic features of MEWDS on OCT are widespread loss of ellipsoid zone in acute stage as given by Spaide et al45 in his study. There is also loss of photoreceptors outer segments and subfoveal choroidal thickening with a normal retinal pigment epithelium.

v) Multifocal Choroiditis and Panuveitis
SD OCT in MCP shows absence of ellipsoid zone with thinning of outer nuclear layer over the area surrounding scarring from old choroidal neovascular membrane. Spaide et al showed that patients with MCP who had blind spot enlargement also had corresponding contiguous regions of the IS/OS junction loss around the optic nerve head even in ophthalmoscopically normal areas between visible chorioretinal scars.

vi) Punctate Inner Choroidopathy
Using SD OCT, Stepieen and Carroll46 showed intraretinal fluid accumulation secondary to choroidal neovascular membrane and homogenous outer retinal thickening over chorioretinal lesion in association with recurrences. OCT can also be useful to monitor the effect of treatment on choroidal neovascular membrane secondary to PIC.

Conclusion:
OCT brings a new light of useful information in helping us understand the disease pathologies. From TD OCT to SD OCT to EDI OCT, this imaging modality has brought revolutionary changes in detection, assessment, monitoring and prognosticating the various diseases of Uvea in anterior as well as posterior segment. Using SD OCT, we can additionally see External Limiting Membrane, Photoreceptor inner and outer segment junction (now called as ellipsoid zone), Photoreceptor outer segment and RPE junction and RPE-choriocapillaris complex. Now with the advent of EDI OCT, the imaging of choroid can be enhanced which was missed on Normal OCT and thus could be a guiding tool especially in Uvea. Thus, invention of OCT should be considered as a historical landmark in Ophthalmology.

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Notification
Dr Ajit babu Majji from Centre for Sight, Hyderabad is contesting as Vice President of All India Ophthalmological Society in the coming AIoC2016 at Kolkata.

TOS Executive & its members wishes him the best in this endeavour!
Development of Geographical Atrophy in Neovascular AMD Treated with Anti-VEGF Therapy: Is it a serious confounder for efficacy of Anti-VEGF?

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Background
Advanced AMD is a progressive, vision-threatening disease that manifests in 2 forms, geographic atrophy (GA) and neovascular AMD (wet AMD). GA generally progresses over several years and may not immediately threaten central vision, while choroidal neovascularization (CNV) in wet AMD can significantly reduce vision within a year. GA and wet AMD may occur concurrently in the same eye.

The pivotal, Phase 3 clinical trials in wet AMD—MARINA and ANCHOR—demonstrated that treatment with ranibizumab, a humanized, monoclonal antigen binding fragment that inhibits all active forms of VEGF-A, can generally improve and maintain vision in patients with wet AMD. Following 2 years of monthly-administered intravitreal ranibizumab 0.3 mg or 0.5 mg, patients achieved mean BCVA gains of +5.4 -10.7 Early Treatment Diabetic Retinopathy Study (ETDRS) letters from baseline, compared with losses of -14.9 letters with sham treatment (MARINA) or -9.8 letters with verteporfin photodynamic therapy (prior standard of care; ANCHOR). Vision improved or stabilized in the majority of patients treated with ranibizumab (90%-92% of patients lost <15 letters at 2 years), and increased areas of GA were not associated with visual acuity loss. After the introduction of anti-VEGF therapy for the treatment of AMD, vision loss or blindness attributable to wet AMD was reduced by ~50% in multiple countries.

The incidence of new GA in patients treated with anti-VEGF agents for wet AMD was retrospectively assessed in the Phase 3 CATT and IVAN studies. These trials evaluated ranibizumab and bevacizumab, administered monthly or as needed (p.r.n.) following a single monthly loading dose (CATT), or in a discontinuous regimen following 3 loading doses (IVAN). In the CATT analysis, GA was defined as areas of hypopigmentation or hypofluorescence ≥ 250 pm on either color fundus photography (CFP) or fluorescein angiography (FA) having ≥ 2 of circular shape, sharp borders, and/or visible choroidal vessels, and nonadjacent to the scar or CNV lesion. IVAN evaluated new GA on CFP or OCT, defined as any area of discrete atrophy that developed since baseline outside the original active lesion with a dropout size >175 µm, well-defined edges, and visible choroidal vessels within the GA area. The 2-year results from CATT suggested a potentially increased rate of new GA with ranibizumab over bevacizumab (ranibizumab, 20.2% vs. bevacizumab, 16.3%), but this finding was not replicated in IVAN (ranibizumab, 28% vs bevacizumab, 31%). Both studies, however, reported higher rates of new GA at 2 years in the monthly compared with p.r.n./discontinuous pooled treatment groups (CATT: monthly, 24.1% vs. p.r.n., 15.5%; IVAN: monthly, 34% vs. p.r.n., 26%). Three potential contributing pathophysiologic mechanisms are considered in the development of atrophy in anti-VEGF-treated eyes. First, the new GA in CATT and IVAN may be the natural progression of underlying dry AMD over time (i.e., true classically defined GA). Alternatively, the pathophysiology of the atrophy observed in wet AMD may differ from true GA and may be better described as macular atrophy. The macular atrophy may be associated with the CNV lesion, caused either by retinal/subretinal fluid and hemorrhage or CNV extension and contraction resulting from ischemia or anti-VEGF-induced regression of abnormal blood vessels. Finally, macular atrophy may be influenced by a class effect of anti-VEGF agents, potentially resulting from the inhibition of constitutive VEGF production in the retina. Macular atrophy in wet AMD patients treated with anti-VEGF agents may be unique compared with classically defined GA.

Methods
This is a retrospective analysis of macular atrophy in the Phase 3 HARBOR trial, which evaluated the efficacy and safety of intravitreal ranibizumab 0.5 mg or 2.0 mg administered monthly or p.r.n. for the treatment of wet AMD. Methods for assessing macular atrophy in the HARBOR data set were developed to address variability in definitions of atrophy, differing grading protocols for atrophy assessment, and the use of different modalities for visualizing atrophy (Figure 1).

1. Method 1, Nonadjacent: FA and CFP images were manually graded for GA following the protocol developed in CATT, described above.
2. Method 2, All-Inclusive: Grading of FA and CFP images was repeated using more inclusive criteria (i.e., to include atrophy immediately adjacent and nonadjacent to CNV lesions).

3. Multimodal grading methods, combining results from FA, CFP, and spectral-domain OCT.

Results

Key findings of the analysis included:

• Mean BCVA improved over time in patients with macular atrophy present at baseline with monthly or p.r.n. treatment (Figure 2).

• Presence of macular atrophy at baseline was not associated with an increased proportion of patients losing ≥15 letters of vision.

• Patients with any concurrent (i.e., new or baseline) macular atrophy at Month 12 and Month 24 had comparable mean BCVA to patients without macular atrophy at Month 12 and Month 24.

Discussion

Atrophy developing in the setting of treated wet AMD may differ from classically defined GA. Therefore, the term “macular atrophy” is used in the HARBOR analysis to describe this entity.

There is currently a lack of “gold-standard” criteria for defining macular atrophy in patients treated with anti-VEGF agents.

Based on existing data, the risk of developing GA or macular atrophy does not appear to outweigh the benefits of ranibizumab therapy for wet AMD.

Vision gains are substantial even in the presence of macular atrophy through 24 months.

The risk of under treatment with ranibizumab therapy remains a potentially greater threat to vision.

There is a need to work toward understanding the pathophysiology of macular atrophy in wet AMD and anti-VEGF-treated wet AMD. This will aid in the development of the best treatment algorithms for patients with AMD.

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Retinal detachment following Giant Retinal Tears: Clinical Presentations and Management

Dr Vivek Pravin Dave, MD, DNB, FRCS; Dr Rajeev R Pappuru, MD

Introduction
Giant retinal tear (GRT) is defined as a peripheral full-thickness retinal break extending circumferentially around the retina for three or more clock hours with a concomitant posterior vitreous detachment.1,2 It is a relatively uncommon form of retinal break which leads to a rapidly progressive retinal detachment with a very guarded visual prognosis unless diagnosed and treated in a timely manner. It needs to be differentiated from a closely mimicking condition known as a giant retinal dialysis which is retinal disinsertion from the ora serrata with 90 degrees or more of circumferential extent.3,4 (Table 1)

Classification
GRTs can be classified on the basis of etiology and on the basis of location of the break. Etiological classification was propounded by Schepens in 1962.5 According to the etiological classification, GRTs are classified as Idiopathic (70%), traumatic (20%) and at the posterior edge of a chorioretinal degeneration (10%). By location, it can be classified as per that proposed by Scott into equatorial, equatorial with posterior extension and oral.3

Occurrence
Around 0.5%-8.3% of all cases of retinal detachment are due to GRTs.2,6,7,8 In pediatric population, the occurrence is higher and varies from 18%-31.7%.9,10 There is a clear male preponderance varying from 65%-91% and most cases occur between the third and the fifth decade.11-14

Etiology
The commonest etiology for GRT formation is idiopathic.5,14 The other common predisposing factors are high myopia, hereditary vitreoretinopathies and trauma.15,16,17 Trauma is by far the commonest predisposing factor. Other uncommon though possible predisposing factors are intraocular surgery, excimer photoablation and necrotic retinitis.

Pathogenesis
Two factors that are central to the pathogenesis of a GRT are abnormal and dynamic vitreous adhesion and traction over an area of white without pressure (WWOP).2 Initially, the central cortical vitreous undergoes liquefaction with condensation of the peripheral vitreous gel. The peripheral vitreous gel contains peripherally attached bands and membranes. With the occurrence of PVD, these strongly adherent bands and membranes cause a large circumferential tear in the neurosensory retina.18 As against small horse shoe tears, GRTs do not have surrounding adequate healthy attached retina to support them due to the large circumferential nature. Consequently, in most cases, the detached retina folds upon itself. Due to the extensively exposed retinal pigment epithelium in such a situation, the stimulus for proliferative vitreoretinopathy (PVR) is very high. This later leads to progressive stiffening of the retina with both longitudinal and circumferential shortening.

Clinical features
Patients with GRT usually present with a macula-off retinal detachment with a visual acuity as low as counting fingers close to face.19,20 30% of cases that present just with a giant retinal tear show progression to retinal detachment over 6 weeks.21 In virtually all patients, the GRT is either partially or completely inverted. (Fig. 1)

When found to be inverted and mobile, proper patient positioning may unfold the flap allowing better clinical examination. PVR is a very high accompanying features with over 50% of cases at presentation having PVR grade B and nearly 15% with PVR grade C.22,23 The large tear releases lot of RPE cells leading to tobacco dusting.19 The formation of the tear may shear a retinal vessel causing concurrent vitreous hemorrhage.24 Most GRTs are less than or upto 6 clock hours in extent. The largest study done to analyse the extent of GRTs showed a mean extent of 166° in a subset of 176 eyes.25,26A major differential diagnosis for a GRT is a giant retinal dialysis (GRD). GRDs occur after severe blunt trauma leading to disinsertion of the peripheral retina from the ora serrata.
Investigations
Ultrasound B-scan is a useful modality in a complete diagnosis of a GRT in cases where there is hazy media either due to cataract or concurrent vitreous hemorrhage which can occur in about 20% of all cases with GRT. A classical “double-linear sign” is seen on B-scan in cases with retinal detachment associated with a GRT. In this sign, two high-amplitude linear echoes extend from the optic disc, one representing the detached retina and one representing the folded retina of the GRT. In general, in the presence of a clear media, GRT and associated retinal detachment is a clinical finding not requiring any investigations.

Management
The management of GRT and the associated detachment is surgical. Though scleral buckling surgery has been attempted in the past with limited success, conventional pars plana vitrectomy (PPV) with endolaser and silicone oil tamponade remains the gold standard surgical procedure for this pathology. A prime requisite for a good outcome in RD with GRT is as complete vitreous clearance as possible. Complete central vitrectomy is quick and simple due to pre-existing posterior vitreous detachment in such cases. A meticulous vitreous base dissection till the extent possible ensures good oil tamponade effect in the post operative period. To ensure this, good pupillary dilatation pharmacologically or with added iris hooks if required should be achieved. Usage of a wide-angle viewing system with occasional usage of a cotton tip applicator for indentation is desirable. This base dissection reduces the chances of formation of new breaks at the edge of the remnant vitreous and reduces occurrence of anterior PVR in the post operative period.

Usage of encirclage buckles in a GRT RD is controversial. Encircling buckle element has been proposed to reduce circumferential traction and reduce the risk of anterior PVR. A contrary view by Hoffman, suggested that use of encirclage buckles without preexistent PVR could lead to possible complications like choroidal hemorrhage, fish mouthing of the break and increased retinal slippage. A routine lensectomy in case of GRT RD is debatable. In pseudophakes and aphakes, a meticulous vitreous base dissection is easily achieved which cannot be achieved in phakics without sacrificing the crystalline lens. Many surgeons suggest that lensectomy should be done routinely to allow base dissection. In a multicentric study by the Vitreon Collaborative Study Group, in the analysis of their data showed there was no difference between lensectomy and no lensectomy in the rate of redetachments or visual acuity.

Role of perfluorocarbon liquid (PFCL)
PFCL is a biologically inert liquid with low viscosity, high specific gravity and a boiling point. It is immiscible with saline and provides excellent hydrokinetic stability to the retina. On completion of vitrectomy, PFCL is invaluable in flattening the mobile retina especially the inverted flap. The flap is teased to revert it back by injecting PFCL over the disc. (Fig. 2)

Figure 2. Reposition of the retinal flap using PFCL

The PFCL is then slowly injected further till PFCL level reaches the ora. Retinopexy is best achieved by applying multiple confluent rows of endolaser. (Fig. 3)

Figure 3. Confluent endolaser done around the edge of the GRT

Some surgeons prefer laser just around the edges of the break while some prefer 360° endolaser. In a study carried out by Al-Khairi et al, the incidence of retinal re-detachments was 25.8% in eyes that did not receive 360 peripheral retinopexy compared with 7.1% in eyes which did. Though statistical significance was not achieved in this study, the results may have clinical application. After endolaser, PFCL can be removed either by a direct PFCL-Silicone oil exchange or a PFCL-air exchange followed by air-silicone oil exchange. (Fig. 4)
During exchange, the flute needle or the vitrectomy cutter on a suction mode is held at the edge of the tear to ensure removal of the residual sub retinal fluid to the extent possible and to minimize the chances of retinal slippage. The silicone oil tamponade is usually maintained for 3-4 months following which it can be removed.

Management Outcomes
Primary and final anatomic outcome in view of retinal reattachment rates vary widely from study to study due to heterogenous inclusion criteria in each study. Most studies suggest an anatomic success rate varying from 70-94% following one or multiple surgeries. Functional outcome too varies from study to study, with BCVA improving in about 50% of patients in most studies. The factors limiting visual improvement are epiretinal membrane formation, cataractogenesis, macular hole formation and posterior capsular opacification. Attempts have been made to ascertain possible pre-operative and intra-operative risk factors associated with final anatomic and functional outcomes. Scott et al reported female gender, age less than 30 years, history of prior vitrectomy, GRT > 180 degrees and presence of PVR at baseline as significant risk factors for a poor outcome. Predictors of poor post operative vision included pseudophakia, aphakia, hypotony, poor preoperative vision, GRT greater than 180 degrees and PVR at baseline.

Management of the fellow eye
Idiopathic GRTs have a high incidence of peripheral retinal pathology in the fellow eye. High risk characteristics in the fellow eye of patients with a GRT include myopia greater than -10 diopters, increasing white-without-pressure areas and increasing condensation of the vitreous base. The vitreous base condensation followed by subsequent traction leads to a GRT. The prophylactic measures propounded for the fellow eye include 360 degrees circumferential encirclage buckling which reduces traction due to the progressive vitreous condensation and 360 degree peripheral retinal laser barrage photocoagulation. Recommendations made by the American Academy of Ophthalmology Preferred Practice Panel on prophylaxis were predominantly based on consensus expert opinion with insufficient evidence to support prophylactic treatment of lesions other than symptomatic flap tears. A Cochrane review also concluded that there was no evidence to support or refute the use of 360 prophylactic treatment in fellow eyes of eyes with a GRT. Some have argued that given the potential morbidity associated with GRTs, prophylaxis may be justified. This could be supported by Freeman's findings of a 51.3% development of retinal breaks or RD in fellow eyes of non-traumatic GRTs after a mean follow-up period of 3.7 years.

Table 1. Differences between a giant retinal tear and a giant retinal dialysis

<table>
<thead>
<tr>
<th>Giant retinal tear</th>
<th>Giant retinal dialysis</th>
</tr>
</thead>
<tbody>
<tr>
<td>PVD present</td>
<td>PVD absent</td>
</tr>
<tr>
<td>Vitreous does not bridge the gap</td>
<td>Vitreous bridges the gap</td>
</tr>
<tr>
<td>Vitreous attached to the anterior margin of the break only</td>
<td>Vitreous attached to both the anterior and posterior margin of the break</td>
</tr>
<tr>
<td>Retina easily inverts upon itself</td>
<td>Retina does not invert upon itself</td>
</tr>
<tr>
<td>Buckling alone often insufficient.</td>
<td>Easily amenable for scleral buckling alone</td>
</tr>
</tbody>
</table>

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X-linked Juvenile retinoschisis (XLRS) is one of the leading causes of macular degeneration in males. It is an X-linked disease caused by mutation in the retinoschisis gene (RS1) located in the Xp22.1, which encodes a 224 amino acid secreting retinal protein, retinoschisin. Retinoschisin octameres is implicated in cell–cell interactions and cell adhesion perhaps by interacting with b2 laminin.[1] This protein helps to maintain the structure of whole retina and structure of photoreceptor–bipolar cell synapse. The mutations cause loss of retinoschisin function that causes splitting of inner retinal layers leading to vision loss. No correlation has been noted between the type of mutation and the disease severity or progression.

Clinical features
The most common presentation is with decreased vision in school going age and it can be presented as early as 3 months of age. The visual impairment can vary from 20/20 to 20/600. Some may present with squint and nystagmus in infancy. Fovealschisis, which is seen, as a cartwheel pattern of folds radiating out from the fovea is the characteristic sign of XLRS and is present in 98–100% of cases. The schisis occurs in the nerve fiber layer (figure 1). This may present as foveal thinning or mild retinal pigment epithelial changes in delayed presentations, esp, after 30 years of age.[2] Peripheral retinoschisis is seen in 50% of XLRS cases. The peripheral retinoschisis may present as a shallow elevation limited to periphery or as bullous elevation of inner leaflet extending to the posterior pole. Retinal breaks can occur in inner layer, outer retinal layer or both (figure 2). The large inner layer breaks and fragmentation of inner layer leads to membranous remnants called vitreous veils that may contain unsupported retinal blood vessels. These blood vessels can cause vitreous hemorrhage and is one of the most common causes of vitreous hemorrhage in children. The other complication is the retinal detachment that can occur when both the inner and outer retinal breaks are present, (figure 1) which can occur in 20% of cases.

Investigations
Optical coherence tomography (OCT) is one of the newer imaging modality in diagnosing XLRS. OCT shows the characteristic macular schisis and helps to differentiate it from retinal detachment.[3] (Figure 2).

OCT studies also showed that outer plexiform layer schisis, micro structure defects in the photoreceptors, abnormalities in cone cell outer segment tips (COST) line and photoreceptor outer segment (PROS) are associated with poor visual acuity in XLRS.[4] OCT is useful in monitoring the therapeutic effect of drugs like acetazolamide that are used in foveal schisis.

Electroretinogram (ERG): The characteristic feature of ERG in XLRS is the negative b wave. This is seen in full field ERG and very rarely cases are reported where full field ERG did not show negative b wave but multifocal ERG showed
negative b wave. So multifocal ERG may be considered in cases with strong clinical suspicion and full field ERG is inconclusive.

Genetic testing can be used to confirm the diagnosis. It is also useful in genetic counselling in females who are at risk of carrying the mutation.

Treatment
Treatment for foveal schisis is generally not indicated. The only treatment option is the low vision aids. Various reports on role of 2% dorzolamide drops and systemic acetazolamide for foveal schisis showed variable response to treatment. No relationship to the kind of mutation and response to treatment was identified. Vitreous hemorrhage and retinal detachment need vitrectomy. Retinal detachment with peripheral retinal breaks may be managed with scleral buckle. The role of barrage laser or scatter laser in peripheral retinoschisis is controversial and is not recommended.

References

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Retinal photocoagulation finds its roots a long way back in the 1940's. Dr. Gerd Meyer - schwickerath used a heliostat (reflective concave mirror with a central viewing ocular) to focus natural sunlight into the eye thus constructing a functional sunlight photocoagulator. This was followed in close succession by introduction of xenon arc laser, monochromatic lasers followed by argon green gas lasers.

Today, the more commonly used Neodymium-doped yttrium aluminum garnet (Nd:YAG) and diode lasers use solid-state platforms that utilize crystals and semiconductors respectively. Modern laser models were introduced in the 1980s and have become popular because of their portability and ability to deliver laser in both continuous and pulse modes. But the need for safe and effective laser treatment for macular disorders without producing overt damage was unmet till the introduction of the micropulse laser technology.

In 1990 Pankratov developed the micropulsed diode laser, producing a train of millisecond laser pulses separated by variable quiet intervals. Micropulsing allowed selective treatment of the retinal pigment epithelium (RPE) and sparing of the neurosensory retina. This article gives an overview of micropulse laser technology i.e. principles of micropulse lasers, its indications and therapeutic stratagem in works so far.

**Principle of micropulse laser**

Subthreshold laser denotes the use of lower energy levels aiming to cause sub-lethal injury to targeted RPE, rather than destroying it and is based on the hypothesis that benefits of photocoagulation are derived from cytokine released by recovering RPE cells.

In the micropulse mode, the laser is delivered in ultra-short pulses (microseconds) that are shorter than the thermal relaxation time of the target tissue. Since, the temperature rise is insufficient to cause ancillary damage to the surrounding retinal tissue it minimizes scarring to the extent that laser spots are generally undetectable on ophthalmic and angiographic examination.

The continuous wave (CW) laser emission is divided into spaced, repetitive micropulses to allow:

- Finer control of photothermal effects
- Lower energy per pulse
- Tissue cooling between pulses (based on Duty Cycle)
- Equivalent or superior clinical outcomes with the benefits of no tissue damage detectable at any time point post-operatively

Heated tissue that remains viable after CW or MicroPulse laser treatment, produces a stress response and induces beneficial intracellular biological factors e.g. PEDF OR Pigment epithelial derived growth factor, SDF1 or Stromal cell derived factor 1, TSP1 or Thrombspondin1 and Beta actin that are primarily anti-angiogenic and restorative.

Table 1 shows distinguishing features between conventional Vs Micropulse laser photocoagulation

**Table 1: Conventional laser Vs micropulse laser**

<table>
<thead>
<tr>
<th>Type of Laser</th>
<th>Conventional / Modified conventional laser</th>
<th>Subthreshold Micropulse Photocoagulation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Laser burn (LB)</td>
<td>Visible, intense/barely visible burns in Modified technique</td>
<td>Invisible</td>
</tr>
<tr>
<td>Direct action (DA)</td>
<td>Destroys tissue results in post treatment scarring which expands with time and modified technique minimal retinal blanching, destroys less tissue damage than conventional laser</td>
<td>Tissue remains viable and produces beneficial intracellular biological factors primarily anti-angiogenic and restorative (i.e. PEDF, TSP1)</td>
</tr>
<tr>
<td>Indirect action (IA)</td>
<td>Tissue remains viable and produces beneficial intracellular biological factors primarily anti-angiogenic and restorative (i.e. PEDF, TSP1)</td>
<td>Negligible</td>
</tr>
<tr>
<td>Tissues affected</td>
<td>Neurosensory retina, RPE, Choroid</td>
<td>RPE</td>
</tr>
<tr>
<td>Temperature rise of surrounding tissues</td>
<td>+20°C to +30°C</td>
<td>Negligible</td>
</tr>
</tbody>
</table>
Micropulse photocoagulation technique divides the laser emission into a “train” of short, repetitive pulses that persist for 0.1 seconds to 0.5 seconds. The “on” time is the duration of each micropulse (typically 100μs to 300μs) and the “off” time (1,700μs to 1,900μs) is the interval between successive micropulses. This “off” time allows for heat dissipation, which decreases collateral damage and confines treatment to the RP e. When a low duty cycle is used, less heat is generated, allowing the RP e to return to baseline temperature before the next pulse is initiated. This eliminates cumulative thermal build-up. This in stark contrast to conventional continuous wave laser, where the same magnitude of energy is delivered throughout the entire exposure cycle of 0.1 seconds to 0.5 seconds. Microscopic, isolated RP e photothermal damage can be achieved with laser powers as low as 10% to 25% of visible threshold powers.

**Duty Cycle:**
The duty cycle is calculated by taking the percentage of the period during which the laser is “on.” For example, with a duty cycle of 15% and a period of 1,000μs, the laser would be on for 150μs and off for 850μs (0.15=150/1,000). If the exposure time was set to 100,000μs, the laser would fire 100 repetitive pulses during that interval. The power and duty cycle are both adjustable, permitting the operator to vary the treatment intensity.

Continuous conventional laser can also be delivered in a subthreshold fashion using lower energy, but this is not the same as dividing the energy into micropulses. Micropulse-divided laser energy will heat the tissue less than a continuous pulse laser even if the total amount of energy is the same.

Frequencies of lasers used in micropulse laser are 532 nm, 577 nm and 810 nm. It can be delivered as a single spot or as a patterned grid in a preselected pattern. Eg: Iridex IQ series, Navilas, Quantel. Figure 1 shows the micropulse settings on Iridex laser system.

### Applications of Micropulse laser
1. Diabetic Macular Edema (DME)
2. Central serous Chorioretinopathy (CSCR)
3. Proliferative Diabetic Retinopathy (PDR)
4. Vascular occlusions
5. Glaucoma
6. Miscellaneous

1. **DME**
The use of micropulse laser in Diabetic Macular Edema (DME) was initiated by Friberg and Karatz in 1997. Luttrell and colleagues in 2005 published, for the first time an account of their experience of using subthreshold MP 810 nm diode laser for DME with complete and contiguous treatment of the entire edematous area without tissue damage. Micropulse laser has been demonstrated to be as efficacious as the conventional laser. A study on Japanese DME patients with Subthreshold Diode Micropulse SDM laser using a 15% duty cycle showed that about 95% of the patients maintained or improved vision over 3 m. The power used was 50 to 90% of the threshold burn. Vujosevic et al, showed that micropulse laser treatment appears to be as effective as the modified-ETDRS laser photocoagulation for treating DME, but it causes far less damage to the retinal pigment epithelium, as judged by microperimetry and fundus autofluorescence. Additionally, it was noted that alterations in macular sensitivity may be demonstrable by microperimetry as early as 1 month post-treatment. This may be prior to detection of changes in retinal thickness by optical coherence tomography (OCT). Though various studies have been conducted to determine the role of micropulse laser in diabetic macular edema with excellent results, till date no large scale study has laid out guidelines with regards to parameters to be used for management of DME as was the case with ETDRS.

2. **CSCR**
The conventional laser treatment debrides the RPe at the site of leakage and this in turns leads to the in-growth of the surrounding RPe and absorption of the sub retinal fluid. Conventional laser in CSCR can lead to central or para central scotoma, contrast sensitivity loss, accidental foveal damage, retinal distortion and choroidal neovascularization (CNV). Difficult to apply conventional laser if the leak is subfoveal or juxtafoveal. Focal laser may close the leak seen on FFA and resolve the sub retinal fluid, but it does not change the amount of choroidal hyperpermeability and leakage, thus the risk of recurrence is not decreased. Photodynamic therapy (PDT) has also been used to treat focal leaks and chronic CSC associated with diffuse compensation of the RPE, but not without side effects.

Various studies have demonstrated the advantages of micropulse laser in CSCR. Lanzetta et al, treated 24 treatment naïve eyes of 22 subjects with the diagnosis of chronic CSC (≥3 m), with non-visible micropulse diode infrared laser irradiation. Treatment was delivered with a micropulse diode infrared laser (Iris Medical Ocvalight SLx). The mean follow-up period was 14 months (range, 3-36 months). One month after laser treatment, nine eyes had complete anatomic resolution of the subretinal fluid in the
central macula and seven had incomplete resolution of the exudative detachment, confirmed by OCT. In another study by Ricci et al, OCT scan revealed complete resolution of the serous neuro-epithelial detachment in five patients and a marked reduction in two patients.

Chen et al treated 26 eyes of 25 patients with persistent CSC and juxtafoveal leakage longer than 4 minutes duration with subthreshold diode photocoagulation (SDM). The eyes were divided into three groups: focal leakage without associated RPE atrophy (group 1), focal leakage with RPE atrophy (group 2), and diffuse RPE decompensation with indeterminate source leakage (group 3). All patients were followed-up for at least 6 months. In group 1, all patients had total SRF resorption after one session of SDM photocoagulation. Eight eyes in group 2 had total SRF resorption after 1 to 3 sessions of SDM laser, whereas 1 patient had persistent SRF. In group 3, only 5 eyes had SRF resorption at the end of the follow-up, and the other 6 eyes needed photodynamic therapy for final SRF resorption. At the end of the follow-up, the average preoperative foveal thickness was reduced by more than half of its original thickness. A gain of visual acuity of 3 lines or more was achieved in 15 eyes (57.7%) and a gain of between 1 and 3 lines was achieved in 6 eyes (23.1%).

3. PDR
The visible burns as the end point (conventional) of pan retinal photocoagulation remain the primary treatment for proliferative diabetic retinopathy. Despite the effectiveness of conventional photocoagulation, there is no evidence that the inherent retinal destruction in visible end point photocoagulation is actually necessary to achieve a therapeutic benefit in the treatment of retinal vascular disease.7, 9

Luttrulllet al performed SDM PRP in 99 eyes of 63 patients with proliferative diabetic retinopathy. All patients were treated with a 500 mm aerial spot size, 0.20 sec pulse envelope duration, with an initial 2.0 W power setting. Within the laser pulse envelope, a 15% duty cycle, each delivering a train of 100 sequential laser pulses of 300 ms ‘on’ time separated by 1700 ms ‘off’ time, was employed for each patient. The median follow-up was 1.0 year (range of 0.3-2.7 years). Treatment sessions per eye ranged from 1 to 6 (with a median of two sessions per eye).

The probability of treatment failure end points at 12 months post treatment was 12.5% for vitreous hemorrhage and 14.6% for vitrectomy. Compared to conventional PRP, the response to SDM PRP developed more gradually and without marked contraction of the neovascular tissue. Thus, SDM PRP may be especially useful in the management of eyes with extensive active neovascularization, which are more prone to retinal detachment following conventional PRP. One-third of eyes required a single treatment session, less than the 45% reported for conventional PRP as noted by Doft et al.27 Moorman et al, in their study found that satisfactory regression of new vessels was achieved using SMD PRP, although the technique required more burns than would be expected using the argon laser. This was a prospective non-comparative case series of 13 eyes with PDR that were treated with 810 nm SMD PRP. Initially, eyes were treated with 1,500 burns. Retreatment was performed as necessary at six week intervals thereafter. The overall number of burns required was approximately 5,250 over three to four treatment sessions, with an average response time of 13 weeks. At six months, 62% of eyes showed complete regression of new vessels, 15% showed some regression and 23% showed no regression.

4. Vascular occlusions
Similar to the diabetic retinopathy experiments, results from studies in vascular occlusions confirm that while SMD treatment may take longer to achieve a similar reduction in edema compared to threshold treatment, long-term visual acuity gain is approximately two times more likely in treated eyes, where photoreceptors are spared.

Parodi et al, in 2006 compared the effect of SMD grid photoagulation to conventional threshold krypton grid photoagulation in 36 eyes with macular edema secondary to BRVO. Both groups demonstrated a reduced mean foveal thickness of half the original value. The result was achieved at six months in the standard laser group compared to one year for the SMD group. After one year, there was no difference in mean foveal thickness or total macular volume between the two groups. At 24-month follow-up, the researchers documented a visual acuity gain of three lines or more in 59% of patients in the SMD group compared to 26% of patients in the threshold group. Visual acuity loss (three lines or more) at 24 months was similar between the two groups (12% in the SMD group and 10% in the threshold group).

Parodi et al, in their study in 2008 showed that treatment with SMD grid photoagulation, in combination with intravitreal triamcinolone injection, resulted in even better visual outcomes—91% of patients gained at least two lines of visual acuity at one-year follow-up.

5. Glaucoma
Micropulse laser trabeculoplasty is being tried as a safe alternative approach to Selective laser trabeculoplasty in the management of open angle glaucomas. Antonio et al, demonstrated that MDLT was effective in reducing IOP in 75% of medically insufficiently controlled OAG eyes without significant complications. This paved the way for further research on this modality in glaucoma. This therapy besides being safe
is also repeatable, thus making it a lucrative proposition. Rantala et al. on the other hand found that MLDLT is a safe but ineffective treatment in patients with open-angle glaucoma. Life-table analysis showed an overall success rate of 2.5% (1/40) and 7.5% (3/40) after up to 19 months of follow-up in their study. Further research is on in this direction and the future looks promising.

Others
A number of small case reports and anecdotal evidence exist on the efficacy of subthreshold micropulse laser in polyoidal choroidal vasculopathy, resistant macular edemas in uveitis and some cases of radiation retinopathy. Recently a report has been published on the use of subthreshold micropulse laser for reactivation of response in anti-VEGF (Vascular endothelial growth factor) resistant choroidal neovascular membranes. These findings are exciting in terms of future potential applications of this therapy.

To conclude one would like to say that, not much is known about micropulse lasers in this time and era but research is on with full gusto to fill in the gaps in our knowledge. Subthreshold micropulse lasers are the lasers of the future, and we keenly await the dawn of time when they shall be the standard of care.

References:

Correspondence
Dr Mahima Jhingan M.S.
LV Prasad Eye Institute
LV Prasad Marg, Banjara Hills, Hyderabad
Unilateral Extensive Myelinated Retinal Nerve Fibers Associated with High Axial Myopia

Dr J Arpitha Rao; Dr C Madhavi

Case Report:
A 35 year old female presented in the Ophthalmology department in a rural area during February, 2014 with complaint of defective vision in right eye for 3-4 years.

Visual acuity best corrected was OD 6/36; OS 6/9 OS.

On slit-lamp examination, anterior segment was normal in OU. Fundus examination showed OD tessellated fundus, extensive peripapillary myelination with white feathery appearance obscuring the optic disc margins and a dull foveal reflex; OS fundus was normal (Figure 1).

Figure 1: Fundus photographs of the 2 eyes

A-scan ultrasound biometry showed axial length of OD 25.75 and OS 21.57 mm.

OCT OD revealed an inner lamellar break with altered foveal contour with inner retinal cystoid spaces; OS was normal (Figure 2).

Figure 2: OCT right eye showing inner lamellar break with cystoids changes.

Discussion:
There is a strong association of myopia with myelinated retinal nerve fibers. Ellis et al found that 83% of patients with myelinated retinal nerve fibers had myopia more than 6 Diopters. However, it remains unknown whether myelination of retinal nerve fiber layer is the reason for or the result of myopia.

Reference:

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Tel: 9550529690
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A 40 year old male patient presented with mild blurring of vision in the left eye of 2 weeks duration. He had a past history of non progressive decrease of vision in the right eye since childhood. He had no history of any systemic diseases. His best-corrected visual acuity was 6/18, N8 in the right eye and 6/24, N36 in the left eye. Both eyes anterior segment was normal and the intraocular pressures were 15 and 10 mm Hg in the right and left eye respectively.

Fundus examination (Figure 1) revealed normal discs with C/D ratio of 0.5:1 in both eyes. The right eye macula revealed an aberrant macrovessel passing under the fovea draining almost the entire macula and emptying into the inferotemporal branch retinal vein. There was no associated foveal avascular zone (FAZ) enlargement, serous detachment, foveolar cystoid spaces or preretinal hemorrhage in the right eye. The left eye revealed a serous detachment about 2 disc diameters and centered at the fovea.

Optical coherence tomography (OCT) of the right eye showed high reflectivity from the macrovessel with posterior shadowing and the left eye showed shallow neurosensory detachment with a small pigment epithelial detachment (PED). Fundus fluorescein angiography (FFA) showed early filling and late emptying of the aberrant macrovessel with no other foveal abnormality in the right eye. Left eye showed window defects superotemporal to the fovea with no focal leak (Figure 2).

A diagnosis of right eye congenital macular macrovessel and left eye acute central serous chorioretinopathy was made. The visual prognosis of both eyes was explained to him and he was counseled about the chances of spontaneous resolution in the left eye.

He was seen again after 2 months. The best-corrected visual acuity remained the same in the right eye while in the left eye it had improved to 6/7.6, N6. A repeat OCT showed similar findings as the previous one in the right eye and no subretinal fluid in the left eye (Figure 1). The patient was reassured, prescribed glasses and asked to follow up as and when required.

Congenital Macular Macrovessel

Dr Annie Mathai

A 40 year old male patient presented with mild blurring of vision in the left eye of 2 weeks duration. He had a past history of non progressive decrease of vision in the right eye since childhood. He had no history of any systemic diseases. His best-corrected visual acuity was 6/18, N8 in the right eye and 6/24, N36 in the left eye. Both eyes anterior segment was normal and the intraocular pressures were 15 and 10 mm Hg in the right and left eye respectively.

Fundus examination (Figure 1) revealed normal discs with C/D ratio of 0.5:1 in both eyes. The right eye macula revealed an aberrant macrovessel passing under the fovea draining almost the entire macula and emptying into the inferotemporal branch retinal vein. There was no associated foveal avascular zone (FAZ) enlargement, serous detachment, foveolar cystoid spaces or preretinal hemorrhage in the right eye. The left eye revealed a serous detachment about 2 disc diameters and centered at the fovea.

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A diagnosis of right eye congenital macular macrovessel and left eye acute central serous chorioretinopathy was made. The visual prognosis of both eyes was explained to him and he was counseled about the chances of spontaneous resolution in the left eye.

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Discussion:

Congenital aberrant or anomalous macular macrovessels or congenital retinal macrovessels are rare. de Crecchio et al reported 13 such cases in a study between 1980 and 2005. Since the first description by Mauthner in 1869, fewer than 50 cases have been reported. They are aberrantly large branches of retinal arteries or veins that typically cross the horizontal raphe to either supply or drain the macular area. These unusual vascular entities must be distinguished from prepapillary vascular loops, racemose hemangiomas, Wyburn-Mason syndrome, or other arteriovenous malformations with potential neuro-ocular sequelae. They may cause visual impairment when they cross the fovea, as in our case with or without associated foveolar cysts and/
or preretinal hemorrhage. Other reported associations of congenital macrovessel include spontaneously resolving serous retinal detachment, a unilateral retinal macrovessel causing bilateral visual impairment due to bilateral retinal pigment epithelial changes, congenital hamartoma of the retinal pigment epithelium, cystoid macular edema and ipsilateral intracranial venous malformation and branch retinal artery occlusion. Sometimes, the visual impairment by the mere presence of a retinal vessel may be subtle causing reduced sensitivity at the macula which can be detected by microperimetry. The visual impairment due to the presence of a retinal macrovessel tends to remain stable, as was shown in a case with a 14 year follow up.

The vessel involved most commonly is a vein, usually arising from the inferotemporal retinal vein, crossing the horizontal raphe and ending up in a vein on the opposite side, or on the same side after double crossing the horizontal raphe. However, the vessel could also arise from the superotemporal retinal vein, or could even be an arterial tree derivative. In the latter, it is reported to be from a 3rd order arteriole. The aberrant vessel could also be an arteriovenous anastomosis from the cilioretinal or retinal artery to the retinal vein. In Brown's series of seven congenital retinal macrovessels, perifoveal arteriovenous communications were seen in four cases and a cilioretinal artery to the retinal vein. In conclusion, the presence of an anomalous vessel crossing the fovea is a rare congenital anomaly. When not associated with complications, the visual impairment is subtle to moderate, usually stable and can be attributed to the loss of the normal architecture of the fovea, as has been observed on OCT.

References:

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Dr. K. Anand Rao
Dr. A. Ravindra
Dr. R. Nagaraj
Dr. P. Praveen
Ocular injuries have been recognized as a major cause for visual morbidity. Until recently, ocular trauma was more of a neglected sub-speciality within the field of Ophthalmology. Among other factors, poor visual outcome even with extensive therapy was probably the most important reason for this neglect. However, with the advances in diagnostic and therapeutic tools available to the clinician in the present day, these limitations have largely been overcome. Hence there is a new found interest in this domain.

**Epidemiology**

Injury to the eye and adnexa is not an uncommon occurrence. Males have been shown to be affected more often than females. This is a reflection of the life-style and choice of professional and recreational activities. Men tend to be employed in heavy manual labour in agricultural and industrial sectors. Hence, the higher likelihood for accidents involving the eye.

The age group often affected is between 18 to 60 years of age. This particular age group is more vulnerable to ocular trauma given their nature of work. Medical care for these patients should also cover rehabilitative aspects and the social and economic limitations due to the visual loss need to be recognized by the health care workers.

**Nomenclature**

Ocular trauma can broadly be grouped under
1. Mechanical injuries
2. Non-mechanical injuries
   a. Chemical injuries
   b. Thermal injuries
   c. Electrical injuries
   d. Radiation injuries

**Mechanical Injuries**

A simplified algorithm for the classification of mechanical injuries is as follows:

**Birmingham Eye Trauma Terminology System (Betts)**

In order to minimize ambiguous terms and to have common glossary when reporting ocular trauma, the BETTS was devised. The definitions are detailed in table 1.

**Table 1: Birmingham Eye Trauma Terminology System (BETTS)**

<table>
<thead>
<tr>
<th>Term</th>
<th>Definition and explanation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Eyewall</td>
<td>Sclera and cornea.</td>
</tr>
<tr>
<td>Closed globe injury</td>
<td>No full-thickness wound of eyewall.</td>
</tr>
<tr>
<td>Open globe injury</td>
<td>Full-thickness wound of the eyewall.</td>
</tr>
<tr>
<td>Contusion</td>
<td>There is no (full-thickness) wound.</td>
</tr>
<tr>
<td>Rupture</td>
<td>Full-thickness wound of the eyewall, caused by a blunt object.</td>
</tr>
<tr>
<td>Laceration</td>
<td>Full-thickness wound of the eyewall, caused by a sharp object.</td>
</tr>
<tr>
<td>Penetrating injury</td>
<td>It is essentially a laceration with an entrance wound into the globe.</td>
</tr>
</tbody>
</table>

In contrast to the above, here, a sharp object opens the globe at its site of impact by an outside-in mechanism.

Retained foreign body

A wound of entry is present and the causative agent is still present within the eye. It is grouped separately as the management differs significantly.
Perforating injury | Both an entrance and exit wound is present and both these wounds are attributable to the same object.

In spite of such clear terminology, practical difficulties occur when encountering a patient in the emergency room. The clinician may use the best suited term or a combination of these to describe the condition of the eye for a particular case.

The mechanism of injury is a very important determinant of the extent and nature of the ocular and adnexal injuries. Penetrating injuries by their very nature lead to open globe injuries with or without retained ocular or orbital foreign bodies. Blunt trauma may cause severe damage to a globe whose walls have not been breached. On occasion, the direct force and the compression-decompression effects may cause defects in the ocular coats leading to an open globe with cases of blunt injury. Hence an open globe indicates either a laceration by a sharp object or rupture due to blunt force.

Ocular Trauma Score
The OTS helps to predict the visual outcome for an individual eye with the assessment of certain parameters at presentation (Table 2 & 3).

Table 2. Ocular trauma Score.

<table>
<thead>
<tr>
<th>Determining The Raw Points</th>
<th>Variable</th>
<th>Raw Point Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Initial Vision</td>
<td>NLP</td>
<td>60</td>
</tr>
<tr>
<td></td>
<td>LP/HM</td>
<td>70</td>
</tr>
<tr>
<td></td>
<td>1/200-19/200</td>
<td>80</td>
</tr>
<tr>
<td></td>
<td>20/200-20/50</td>
<td>90</td>
</tr>
<tr>
<td></td>
<td>&gt;20/40</td>
<td>100</td>
</tr>
<tr>
<td>Associated Findings</td>
<td>Rupture</td>
<td>-23</td>
</tr>
<tr>
<td></td>
<td>Endophthalmitis</td>
<td>-17</td>
</tr>
<tr>
<td></td>
<td>Perforating injury</td>
<td>-14</td>
</tr>
<tr>
<td></td>
<td>Retinal detachment</td>
<td>-11</td>
</tr>
<tr>
<td></td>
<td>Afferent pupillary defect</td>
<td>-10</td>
</tr>
</tbody>
</table>

Table 3. Estimation of final visual outcome

<table>
<thead>
<tr>
<th>SUM</th>
<th>OTS</th>
<th>NPL</th>
<th>PI/ HM</th>
<th>1/200-19/50</th>
<th>20/200-20/50</th>
<th>&gt;20/40</th>
</tr>
</thead>
<tbody>
<tr>
<td>0-44</td>
<td>1</td>
<td>74</td>
<td>15</td>
<td>7</td>
<td>3</td>
<td>1</td>
</tr>
<tr>
<td>45-65</td>
<td>2</td>
<td>27</td>
<td>26</td>
<td>18</td>
<td>15</td>
<td>14</td>
</tr>
<tr>
<td>66-80</td>
<td>3</td>
<td>2</td>
<td>11</td>
<td>15</td>
<td>31</td>
<td>41</td>
</tr>
<tr>
<td>81-91</td>
<td>4</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>22</td>
<td>72</td>
</tr>
<tr>
<td>92-100</td>
<td>5</td>
<td>0</td>
<td>1</td>
<td>1</td>
<td>5</td>
<td>93</td>
</tr>
</tbody>
</table>

Evaluation of a Patient with Ocular Trauma

Triage
The clinician needs to be meticulous in his approach and retain a high index of suspicion with regard to evaluation of any patient presenting with Ocular trauma. Careful documentation of the circumstances during presentation, history given, clinical findings on examination and the reports of ancillary investigations, if any, serve two vital purposes.

Firstly, the medical record serves as a baseline on which treatment, follow-up and further intervention can be planned. An astute clinician can predict the anatomic and visual outcome with a fair degree of accuracy with proper history taking and clinical evaluation alone. Secondly, the primary medical records are often asked for in the court of law if medico-legal issues need to be addressed. Hence it is necessary that findings during examination are noted down clearly. One also needs to have the diagnosis, anticipated action plan and expected outcomes discussed with the patient and caregivers in their native language. The necessary consents for initiation of therapy, investigations and surgical procedures need to be obtained from the patient (if he of legal age to consent) and the caregivers, preferably in their native language.

The first goal of examination and management is “Do no further harm”. An open globe needs to be quickly and carefully handled. Undue pressure on the eye for examination or investigations are best avoided. This may cause prolapsed of ocular contents in a traumatized eye with an open wound. In place of patching the eye, one may use protective eye shields as they are not in contact with the globe but, nevertheless, provide the necessary protection.

History
Detailed history taking is of paramount importance. The age, occupation and location where the injury occurred give an idea about the circumstances leading to the event. Time lapsed since the injury is critical in cases where there is potential for infection with an open globe or with a retained ocular foreign body.

The mode of injury is by far, the most crucial element in history taking. One can anticipate the type of wound, extent of ocular damage, potential for infection, presence of a
retained intra-ocular or intra-orbital foreign body and the urgency for surgical intervention if the mode of injury can be ascertained.

The following table gives an indication of the occupations, modes of injuries and the ocular lesions usually associated with such trauma.

Table 4. Occupation, mode of injury and their relation to ocular effects

<table>
<thead>
<tr>
<th>Occupation</th>
<th>Mode of Injury</th>
<th>Commonly Observed Ocular Lesions</th>
</tr>
</thead>
<tbody>
<tr>
<td>Agricultural worker</td>
<td>Injury with sticks</td>
<td>Corneal or intra-ocular foreign bodies</td>
</tr>
<tr>
<td></td>
<td>Injury with insecticides or toxins</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Corneal ulcers</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Chemical injuries</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Endophthalmitis</td>
</tr>
<tr>
<td>Industrial worker</td>
<td>Injury with mobile machinery</td>
<td>Orbital fractures</td>
</tr>
<tr>
<td></td>
<td>Injury with stationary equipment</td>
<td>Corneo-scleral tears</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Traumatic cataract</td>
</tr>
<tr>
<td>Sports person</td>
<td>Injury with a ball, sports equipment or collision</td>
<td>Blow-out fractures</td>
</tr>
<tr>
<td></td>
<td>during play</td>
<td>Occult globe ruptures</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Angle recession</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Hyphaema</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Retinal detachment</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Retinal dialysis</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Optic nerve avulsion</td>
</tr>
<tr>
<td>Miners</td>
<td>Blast injuries</td>
<td>Lid lacerations</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Corneal tears</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Retained intraocular foreign bodies</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Endophthalmitis</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Vitreous hemorrhage</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Retinal detachment</td>
</tr>
</tbody>
</table>

Drivers and passengers | Road traffic accidents | Lid lacerations                                      |
|                       |                       | Corneo-scleral tears                                  |
|                       |                       | Ruptured lens capsule                                 |
|                       |                       | Retained intraocular foreign bodies                  |
|                       |                       | Multiple retinal breaks                               |
|                       |                       | Retinal detachment                                    |
|                       |                       | Traumatic optic neuritis                              |

Miscellaneous

<table>
<thead>
<tr>
<th>Occupation</th>
<th>Mode of Injury</th>
<th>Commonly Observed Ocular Lesions</th>
</tr>
</thead>
<tbody>
<tr>
<td>Domestic</td>
<td>Domestic accidents</td>
<td>Chemical injury</td>
</tr>
<tr>
<td>accidents</td>
<td></td>
<td>(detergents, glue, fire crackers)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Corneal tears</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Chemosis</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Sub conjunctival hemorrhage</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Lens dislocation</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Vitreous hemorrhage</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Retained intraocular foreign bodies</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Endophthalmitis</td>
</tr>
</tbody>
</table>

**Visual Acuity**

The visual acuity recorded at presentation is a vital parameter in which the Ocular trauma score (OTS) is based. The VA in both eyes should be documented. If there is no perception of light, the same needs documentation as well. Doubtful light perception in an injured eye can be recorded with an indication of the number of correct responses against the total number of times tested. However, in the present, it is no longer believed that absence of light perception is in itself an indication for enucleation or evisceration. Every effort can be made to reconstruct the globe and restore the anatomic integrity of the eye.

In a few instances, the clinician may have more urgent aspects to address at the time of presentation. Road traffic accidents with head injuries in an unresponsive patient are not an uncommon scenario in the emergency room. The surgeon has more pressing issues to attend to and the documentation of VA may be postponed to such a time when there is no imminent threat to life. However, even in such a situation, the reasons for not documenting visual acuity need to be clearly mentioned in the medical records.
OCULAR EXAMINATION

Ocular adnexa and the eye lids
Evaluation of the eye begins with examination of the orbital margins for any defects, irregularities or tenderness. Any of the above points towards fractures of the bony orbit or the bones forming the orbital rim. Blow out fractures of the orbit due to a fracture of the orbital floor will present with enophthalmos due to herniation of orbital contents into the maxillary sinus and loss of orbital volume. In contrast, increase in orbital volume is seen in orbital emphysema arising due to fractures of the medial orbital wall. Traumatic retrobulbar hemorrhage is a rare event that presents with severe chemosis, sub conjunctival hemorrhage, motility limitation and proptosis. It is an ocular emergency and requires immediate attention to relieve pressure on the optic nerve.

Reflex closure of the lids during a traumatic event can lead to eye lid and adnexal injuries (Figure 1). During examination, the extent, depth and number of incisional or lacerated wounds over the eye lid need to be documented. It is preferable to have photographic documentation of the entire anterior segment of the eye (Figure 2). If unavailable, diagrammatic representation can be done. While assessing adnexal injuries, it is also important to note if the lacrimal drainage system and / or lid margins have been involved. Involvement of these structures will need special surgical considerations during reconstruction.

Ocular Motility
Though direct muscle or nerve injury after trauma leading to motility disorders is uncommon, the ocular movements need to be assessed to rule out orbital injury. However, this part of examination needs to be avoided in open globe injuries.

Ocular motility is also restricted in fractures of the bony orbit with associated muscle entrapment.

Conjunctiva, Cornea and Sclera
In accordance with the new terminology for ocular trauma, closed and open globe injuries are usually clinically identified on examination of the cornea and sclera. Obvious defect of the eye wall (cornea and sclera) can be identified on cursory examination (Figure 3). In this scenario, every effort must be taken to ascertain the extent of the tear. The posterior most extent of the eye wall defect is usually not obvious clinically and the definite extent can be detected only after careful surgical exploration. Indeed, some of these tears extend far into the posterior of the globe than can be accessible surgically. Apart from the extent of the wound, signs of infection, prolapsed of ocular contents and the deformation of the globe due to such loss of volume needs to be documented.

The presence of an open globe is often concealed. Self sealed corneal tears and small scleral ruptures beneath the overlying conjunctiva may easily go undetected on cursory exam. Sub-conjunctival hemorrhages, deep anterior chamber or reduced ocular pressure are pointers to occult globe rupture in a patient presenting with ocular trauma (Figure 4). On encountering a victim of trauma who had underwent primary corneal tear repair or who has a self sealed corneal wound, the Seidel’s test with the application of fluorescein to stain the tear film gives an indication of the anatomic integrity of the wound.

If the patient has a history of an ocular surgery, it is rewarding to check the incision sites. Opening of the surgical wound, iris prolapse, extrusion of the IOL or the lens itself through occult limbal tears are known to occur. More subtle clinical findings like corneal abrasions, superficial or stromal foreign bodies have to be carefully looked for. They can be easily overlooked in the presence of more dramatic findings that hold the examiners interest.
Figure 3. Stellate corneo-scleral laceration caused by injury at the workplace. Uveal tissue prolapsed at the wound site is noted.

Figure 4. Extensive temporal chemosis with a deep anterior chamber in a patient who has an occult globe rupture.

Figure 5. Anterior segment photograph of an eye which had an injury with a thorn 3 weeks prior to presentation. One can appreciate chemosis and corneal melting with extensive exudates in the AC suggesting endophthalmitis.

Figure 6. Layered blood in the anterior chamber (hyphaema) in a patient who had an injury with a tennis ball.

Anterior chamber (AC)

The AC may show a post traumatic reaction in the form of cells and flare in the aqueous. In severe cases, they may gravitate down to form a hypopyon. This needs to be differentiated from the hypopyon in post-traumatic endophthalmitis which calls for urgent attention (Figure 5). It is not uncommon to encounter blood in the AC. This traumatic hyphaema can lead to an IOP rise and staining of the cornea with breakdown products of hemoglobin (Figure 6). Other contents not native to the AC like vitreous strands, lens protein or the foreign body may also be seen. A careful gonioscopic examination is essential in this regard to look for the IOFB and to assess damage to angle structures.

The depth of the anterior chamber gives vital clues. As mentioned, an occult globe rupture presents with deep anterior chamber. The same clinical finding may be encountered in cases with dislocated or extruded lens or IOL or in angle recession. The AC may be irregular in depth due to a subluxated lens. In contrast, a flat AC is seen in cases with corneal tears. Often, the iris tissue is found incarcerated at the site of the corneal tear plugging the wound.

Iris tears, dialysis or traumatic mydriasis may be seen. An uncommon finding is total aniridia post trauma. Foreign bodies on the iris or iris defects corresponding to the path taken by the foreign body in its travel into the eye provide vital clues to the examiner.

Lens

Traumatic subluxation or dislocation of the clear lens or an IOL is common following trauma. The integrity of lenticular zonules and the extent of loss in clock hours is an important determinant of the method of surgery necessary for phakic rehabilitation.

Defects in the anterior capsule can be made out on slit lamp Biomicroscopy. Traumatic cataracts take the form of rosettes. A Vossius ring may be noted on the anterior capsule due to compression of the pupillary margin over the lens.

Pupil

The shape, size and reaction of the pupils to direct and consensual reflexes need to be looked for. The presence of
relative afferent papillary defect is an important prognostic marker in cases of ocular trauma.

Vitreous
Vitreous hemorrhage is commonly encountered following trauma (Figure 7). If severe, this precludes useful observation of the fundus and the clinician may have to resort to ancillary investigations to judge the extent of posterior segment trauma. The intra-ocular foreign body can also be found in the vitreous. This is often the case when multiple foreign bodies are present after a blast or fire cracker injury.

The traumatic event may induce a detachment of the posterior vitreous with inturn predisposes to retinal breaks and detachment of the retina.

Post-traumatic endophthalmitis presents with extensive vitreous exudation. The response of the patient to therapy is clinically assessed based on the clearance of these exudates with therapy. Hence careful documentation of the severity of the aforementioned vitreous haze at presentation is imperative.

![Figure 7. Vitreous hemorrhage in patients presenting with blunt trauma. The extent of hemorrhage into the vitreous determines the visualization of underlying fundus details.](image)

Retina
The effects of trauma on the retina are myriad. They range from mild retinal edema to total retinal detachment. The spectrum includes commotion retinae (Berlins edema), traumatic macular hole, peripheral retinal breaks and dialysis, partial or total retinal detachment or incarceration of the retina in the open wound.

Reports of retinal vascular occlusions and retinal hemorrhages after ocular trauma are also present. Purbtscher retinopathy is a form of indirect ocular injury due to compressive trauma over the chest or abdomen. It presents with extensive retinal hemorrhages and cotton wool spots. Retinal hemorrhages post ocular trauma may occur at any level – preretinal, intraretinal, sub retinal or below the retinal pigment epithelium (RPE).

The retina is often the final resting place for a foreign body after its eventful journey into the eye (Figure 8). It may be found impacted on the retina with an underlying break or may just be in contact with the retina. Even when a single IOFB is noted, one must be vigilant to look for other areas of impact since a rebound and reflection of the IOFB may have occurred with possible damage at other places on the retina.

![Figure 8. A metallic foreign body seen over the retina in a patient who had an injury at his workplace.](image)

Choroid and Optic nerve
Traumatic choroidal rupture is a common event (Figure 9). Supra choroidal hemorrhages and effusions are also encountered. Sympathetic Ophthalmia needs to be considered when diffuse granulomatous pan uveitis is encountered.

Optic neuritis following trauma occurs in 0.7 to 2.5 % of patients with head injury. Traumatic optic neuritis can present with varied visual loss. Patients may present with no light perception to just a subtle unexplained visual disturbance in the form of colour desaturation. Among others, the former is common in a patient who has been comatose for a period of time following head injury&/or neurosurgery and has regained consciousness to realize that vision is defective in one or, rarely, both eyes. Traumatic optic neuritis may present with mild disc edema but is usually a diagnosis of exclusion when RAPD and visual defects cannot be attributed to other ocular and neurological causes since the optic nerve itself presents no particular clinical sign in the acute setting. In contrast, traumatic optic nerve avulsion is clinically obvious until obscured by overlying hemorrhage (Figure 10).

![Figure 9. Fundus picture showing traumatic choroidal rupture as a whitish streak beneath the retina.](image)
Figure 10. Avulsion of the Optic nerve following trauma in a patient who sustained injury in a road traffic accident.

NON-MECHANICAL INJURIES

Chemical Injuries

Broadly, injuries with chemical agents may be classified as acid or alkali related injuries. Ocular injury due to chemicals may occur in varied settings. Industrial accidents are the most common mode of chemical injury. A considerable proportion of these injuries also occur at an agricultural or domestic setting with insecticides, fertilizers, detergents, solvents or even cosmetics and adhesive glue. Common agents that are handled in daily life are potentially blinding when in contact with the eye.

Alkali burns are caused by chemicals like lime, ammonia and caustic soda. The damaging effects of alkali on the eye are profound. Alkali imbibes water from the cells, causes breakdown cell membranes and hence penetration into ocular tissues is more. On the other hand, acid injuries lead to instant coagulation of proteins in the tissue they come in contact with. This coagulation itself acts as a barrier to further penetration of acid into the eye.

The extent of limbal ischemia and corneal haze has been taken as markers of visual prognosis. Hence during examination, these aspects must be looked into.

Electric and Radiation Injuries

Passage of electric current through the eyes lead to corneal opacities, cataract, retinal hemorrhages and optic neuritis. Intra-ocular inflammation in the form of an AC reaction or vitreous haze is a common finding.

Radiation exposure to the eye may occur in the form of ultraviolet radiation, infrared radiation or exposure to ionizing rays. UV rays cause multiple superficial painful corneal erosions. It is commonly encountered in workers involved with welding. The symptom complex is referred to as photo-ophthalmia. Infra-red rays, in contrast, cause retinal damage with RPE changes and loss of outer segments or a full thickness macular hole.

Prevention of Ocular Injuries

Ocular injuries are a common occurrence. Protective eye wear for sports persons, industrial and agricultural workers can be emphasized. Use of protective films for those exposed to UV rays and helmets for two wheeler riders are a step in this direction. Given its visual morbidity, it is imperative that adequate precautions are taken and safeguards put in place to protect those who are vulnerable to ocular injury. Legislation is a positive step in this regard. Enforcement of ocular protection will achieve intended benefits if practiced along with health education.

Management of Ocular Injuries

After a detailed history and clinical examination, the clinician may need the assistance of investigations to decide the best course of action in an individual case. Commonly utilized investigations are the posterior segment ultrasonography, ultrasound biomicroscopy and computed tomography scans of the bone and orbit. Microbial analysis is also essential when intra-ocular pathogens are suspected.

Management of the injury depends on the presence or absence of an open globe. With an open globe, in the absence of other life threatening injuries, the priority is surgical closure of the ocular wound to maintain the anatomic integrity of the globe. Thereafter, both a previously open globe injury and a closed globe injury may be managed in similar lines. Individual components and traumatic effects of the injury may be taken care of with subsequent surgeries or medication, as appropriate.

References


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Abstract
The “handshake” technique is a modification of the glued-IOL procedure in which the Intraocular lens (IOL) haptic is bimanually transferred from one glued IOL forceps to another under direct visualization in the pupillary plane. This technical modification provides better intraoperative IOL manipulation and extends applicability of the technique to cases that require excess haptic manipulation, such as small pupil, dropped IOL reposition and haptic slippage.

Introduction
The posterior capsule is an essential physical barrier to the normal lens. After cataract surgery, capsular support is crucial for maintaining the normal anatomical position and the ‘barrier’ effect for the IOL. Intraoperative loss of capsules, leads to difficulty in placing the IOL in the bag. Under such circumstances, surgeons opt for either scleral fixated IOLs or anterior chamber lenses. Scleral lens fixation with sutures are prone for UGH (Uveitis-Glaucoma-Hyphema) syndrome and late posterior segment complications. Similarly AC IOL has been known for risk of endothelial loss. Hence in 2007, Dr Agarwal et al has come out with a sutureless fibrin glue assisted scleral IOL fixation technique termed “Glued IOL”. The externalization of the haptic of the intraocular lens (IOL) intraoperatively is the key step in the glued IOL surgery. The important step is to grab the tip of the haptic with the end opening forceps. If one grabs the haptic in place other than the tip with undue pressure to externalize, the haptic may get deformed or sometimes break. However in the learning curve, this precise step of manipulation of IOL haptic should be expertised in order to decrease the surgical time. This is made easier now with the “Handshake” technique. “Handshake” is a modification in the glued IOL procedure in which the IOL haptic is bimanually transferred from one end opening forceps to another under direct visualization in the pupillary plane. This is continued till the tip of the haptic is grasped so that it can be externalized easily.

Handshake Technique
Under peribulbar anesthesia, the partial lamellar scleral flaps are made 180 degrees apart. A 3-piece foldable IOL with a C-loop or a modified C-loop configuration is used. Based on the surgeon’s preference, an infusion cannula or an anterior chamber maintainer is fixed and the flaps and sclerotomies are made 1-1.5mm from limbus. The corneal incision is fashioned with a 2.8 mm keratome, which is slightly enlarged to allow easy insertion of the IOL. The available injectors for the three piece IOLs have either a pushing or a screwing method for unfolding the IOL. In an injector with pushing mechanism, one hand pushes the injector plunger while the other hand holds the end opening forceps ready to grab the tip of the haptic as it comes out of the injector (Fig 1a). This prevents the IOL from falling into the vitreous cavity. In this technique, the end opening forceps is passed through the sclerotomy site while another forceps is in the pupillary plane ready to receive the haptic (Fig 1a, b). Any portion of the leading haptic is initially grasped with the forceps (Fig 1c, d). Then the other forceps starts grabbing the haptic like a “hand shake” and this process are continued till the tip of the haptic is held by the initial forceps. The haptic is then pulled under the scleral flap.

The trailing haptic is grasped with the glued-IOL forceps and flexed into the anterior chamber (Fig 2). A second glued-IOL forceps is introduced through the side port and the haptic is transferred from the first forceps to the second using the handshake technique (Fig 2). Similarly the trailing haptic is externalized. Thus both the haptics are externalized under the scleral flaps. The surgery is completed by tucking the haptics into the scleral tunnel and closing the scleral flap.
Advantage
We have used this “Handshake” technique of bimanual IOL haptic manipulation in injectable glued IOL surgery and the surgical time is shown to be reduced. The retrospective analysis of haptic deformation in injectable glued IOL surgery (n=58) is 6.8%. However after the introduction of the handshake technique (n=45) this has been reduced to 2.2%. There is no haptic break observed. Especially in non dilating pupils and in beginners of glued IOL procedure this step is proven to be useful. One should always remember that in a three piece IOL it is better not to do wound assisted injection. Rather one should pass the cartridge into the anterior chamber and inject. The difficulty occurs if the screwing mechanism injector is used, where both hands are used to inject the IOL. In such a case one should follow the “Handshake” technique. Also in case of difficult situation where the leading haptic tip is not visualized in the pupillary plane or in a non dilating pupil where the iris overlies the haptic, this “Handshake” technique is performed. This technique can be performed in repositioning the decentered and intravitreal dislocated IOLs by Glued IOL procedure.5 Glued IOLs may be more cost effective than alternative means of IOL implantation and fixation as a result of fewer complications and fewer follow-up procedures.6,7 There is no need for special IOL designs, which can be costly for the patient in an unplanned situation, such as intraoperative posterior capsule rupture. The foldable glued IOL has reduced the need for the large incisions initially required for rigid IOLs. The surgical time for suturing the main corneal incision is also less, which decreases the overall operation time.

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Corneal collagen cross-linking with SMILE lenticule: A new modality of treatment for the ultrathin keratoconic cornea

Dr Ritika Sachdev, MS; Dr Gitansha Sachdev, MS, FICO; Dr Deepa Gupta, MS; Dr Mahipal Sachdev, MD

Keratoconus is a non-inflammatory, progressive ectatic disorder of the cornea. Impaired visual acuity results from progressive myopia, irregular astigmatism and central corneal scarring. Treatment modalities described in various stages of the disease process include spectacles, contact lenses, intrastromal corneal ring segments, corneal collagen cross-linking and lamellar and penetrating keratoplasty.

Corneal collagen cross-linking or CXL is a promising treatment modality to increase the mechanical and biochemical strength of the corneal tissue. Using UVA radiation at 370 nm and photosensitizer riboflavin, the photosensitizer is excited into its triplet state generating reactive oxygen species. These reactive oxygen species create additional chemical bonds in the corneal stroma by polymerization, increasing the tensile strength of the cornea.

Based on the current protocol for performing CXL, the cross-linking effect is limited to the anterior 300 microns of the corneal stroma. A corneal thickness of 400 microns is considered as the safe limit to protect the endothelium and intraocular structures from the adverse effects of UVA irradiation and has been established as a clinical standard. Unfortunately, the very patients who are in need of CXL have thin corneas often below the threshold of that considered to be safe for the treatment. In developing countries of the Asian sub-continent such as ours, keratoconus has an earlier onset and is often detected at a later stage making the disease not amenable to traditional CXL.

Currently available treatment modalities for CXL in thin corneas include hypoosmolar riboflavin, contact lens assisted CXL and transepithelial CXL. However these procedures have their own limitations.

We describe a new and innovative procedure to increase the intraoperative corneal thickness using refractive lenticules of patients undergoing small incision femtosecond lenticule extraction for myopic correction.

Technique:
The procedure is carried out under topical anaesthesia. Central 8 mm of the corneal epithelium is debrided using a blunt spatula. The thickness of the remaining stromal bed is measured using intraoperative pachymetry to determine the thickness of the refractive lenticule to be placed for tailored stromal expansion.

The refractive lenticule required for the procedure is obtained from patients undergoing small incision lenticule extraction (SMILE) for myopic correction. The VISUMAX femtosecond laser system is used to perform small incision lenticule extraction. The laser is used to cut a refractive lenticule intrastromally, the thickness of which is determined by the desired refractive correction. The lenticule is then separated manually from the overlying flap and underlying stromal bed. The separated lenticule is extracted from the stromathrough a 3 mm corneal incision. The lenticule procured is thickest at the centre and thinner peripherally. Hence the placement of the lenticule is such that the central portion of the lenticule overlies the thinnest area of the cone (Figure 1).

Figure 1.
The augmented stromal thickness is measured using intraoperative pachymetry and a thickness of more than 400 microns is confirmed, allowing CXL to be performed within the required safety protocol guidelines. 1 drop of isotonic riboflavin (0.1 % solution of 10mg riboflavin 1 phosphate) is instilled every 5 minutes for 30 minutes. Slit lamp examination is performed to confirm the presence of yellow flare in the anterior chamber and ascertain adequate penetration of the dye. UVA radiations of 365nm with desired irradiance of 3mW/cm2 are used at a distance of 5cm. Riboflavin drops are instilled every 5 minutes for the next 30 minutes. Throughout the procedure the lenticule remains firmly attached offering a physiological increase in the intraoperative stromal thickness as seen on anterior segment OCT (Figure 2).
On completion of the procedure the refractive lenticule is peeled off the stromal bed and the surface is irrigated with normal saline. A bandage contact lens is applied which is removed on the fifth postoperative day.

Results:

<table>
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<tr>
<th>S. No.</th>
<th>Patient Details</th>
<th>Kmax (preoperative)</th>
<th>Kmax (6 months postoperative)</th>
<th>Preoperative thinnest pachymetry (in microns)</th>
<th>Epithelium off stromal thickness (in microns)</th>
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</thead>
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<tr>
<td>1</td>
<td>18 V/F</td>
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<td>55.1/57.8</td>
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<td>326</td>
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<tr>
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<td>24 V/F</td>
<td>45.2/50.8</td>
<td>44.7/50.2</td>
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<td>337</td>
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<tr>
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<tr>
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<td>56.6/49.8</td>
<td>55.1/47.2</td>
<td>423</td>
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This procedure was performed in 7 eyes of progressive keratoconus with a stromal corneal thickness of less than 400 microns and a best corrected visual acuity of more than 6/9 with contact lenses. No intraoperative complications were noted in any of the patients. Corneal stability was diagnosed on topography at 6 months follow up (Figure 3). No significant endothelial cell loss was noted in any of the cases. Presence of stromal haze and its resolution was similar to that of conventional CXL. A clear demarcation line at a depth ranging from 280 to 310 microns was noted in all the patients (Figure 4).

Our technique overcomes many of the demerits of the earlier techniques of crosslinking the thin cornea and can be used in both traditional and accelerated crosslinking modalities. Hypoosmalar CXL describes the instillation of hypotonic riboflavin to increase the hydration of the cornea and hence the thickness. Increased intraoperative time, and a relative lower concentration of collagen in the hydrated stroma are some of the limitations of this technique.

Contact lens assisted CXL involves placement of a soft contact lens to increase the corneal thickness intraoperatively. However the biomechanical properties of a contact lens differ from that of the corneal stroma. Moreover the thickness of the contact lens placed is constant and cannot be customised. Additionally buckling of the contact lens intraoperatively can result in an uneven precorneal riboflavin layer with subsequent hot and cold spots.

In our modified technique of CXL using the refractive lenticule, thickness of the cornea is increased in the most physiological manner by adding stromal tissue whose biological and absorptive properties are the same as that of the cornea to be treated. Refractive lenticules of variable thickness (20 to 140 microns) can be obtained following FLE depending on the extent of refractive error to be corrected. Placement of the central lenticule over the apex of the cone enables us to augment the corneal thickness where required while sparing the remaining stroma to be crosslinked normally. Moreover, the relatively rough host stromal surface allows the lenticule to spread easily and buckling is avoided.

In conclusion, our modified technique of tailored stromal expansion for crosslinking the ultrathin ectatic cornea offers a ray of hope for many patients with advanced keratoconus, who have useful vision with contact lenses. Crosslinking these patients offers them the possibility of maintaining their vision and potentially avoiding a future corneal transplantation.

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Introduction
Vernal keratoconjunctivitis (VKC) is a chronic, bilateral, at times asymmetrical, seasonally exacerbated, allergic inflammation of the ocular surface, involving tarsal and / or bulbar conjunctiva. It is more common in children and young adults having an atopic background. The initial seasonal attacks turn into perennial disease after a few years. Although the allergic nature of this entity has been accepted for a long time, its exact aetiology and pathogenesis is still unclear. Pathogenesis of VKC is much more complex than a mere type 1 hypersensitivity reaction.

Geographical distribution and Demography
It is more common in temperate zones of Mediterranean areas, central and west Africa, the Middle East, Japan, the Indian subcontinent and South America. VKC cases are also seen in Western Europe (including the UK and Sweden), Australia and North America. After the recent decline of endemic trachoma, VKC is a leading cause of paediatric outpatient ophthalmic morbidity. The disease starts before 10 years and resolves after puberty, usually around 4–10 years after the onset1,2. Male to female ratio reported in the literature varying from 4:1 to 2:11,3,4. One third of VKC patients have multiple atopic diseases like Atopic allergy, Nasal Allergy and Asthma5.

Clinical features and diagnosis
Symptoms: Itching, redness, photophobia and watering, photophobia and watering are the typical symptoms in VKC. Bilateral in 98%, minor differences in severity between the eyes are common6. Initially there will be several episodes of active inflammation seasonally may become perennial after a few years. The disease will be exacerbated by exposure to wind, dust, bright light, hot weather or physical exertion associated with sweating. Thick mucus hyper-secretion with sticky mucous filaments, called ‘ropy discharge’, is characteristic of VKC.

Signs: Signs of VKC are confined mostly to the conjunctiva and cornea. The skin of the lid and lid margin may be thick and lax. Skin of lid and lid margin are relatively spared as compared to AKC . Amblyopia seen among VKC may be caused by corneal opacity, irregular astigmatism. Sometimes Keratoconus and Dry eye syndrome may coexist. Cataract and glaucoma can be caused by unsupervised use of topical corticosteroids12.. Some time there may be ptosis and blepharospasm.

Conjunctival Involvement
Transient limbal or conjunctival yellow-white points or deposits, known as Horner–Trantas's dots (Figure 1) are degenerating eosinophils and epithelial cell debris . Asians show, perilimbal conjunctival pigmentation the extent of pigmentation did not correlate with the severity of symptoms and signs of VKC. The pigmentation may persist even when the disease was inactive7.

Figure 1: Horner–Trantas's dots

Bonini et al. (2000)14 graded the papillae on the upper tarsal conjunctiva or at the corneoscleral limbus as follows: 1. Grade 0: no papillary reaction. 2. Grade 1+: few papillae, 0.2 mm widespread over the tarsal conjunctiva
3. Grade 2+: papillae of 0.3–1 mm over the tarsal conjunctiva or at the limbus.
4. Grade 3+: papillae of 1–3 mm all over the tarsal conjunctiva or for 360° around the limbus.
5. Grade 4+: papillae of more than 3 mm over the tarsal conjunctiva or gelatinous appearance at the limbus. Papillae with a size of 7–8 mm are known as cobble- stone papillae(Figure 2). Papillae size correlate positively with the persistence or worsening of symptoms over long-term follow-up14. These papillae become quite swollen during the active stage but persist even during the quiescent stage.
Limbal papillae tend to be gelatinous and confluent. Based on predominant involvement of either tarsal or bulbar conjunctiva it can be divided into palpebral or bulbar or mixed forms.

**Corneal Involvement**

Photophobia, pain and foreign body sensation are caused by involvement of the cornea. Corneal changes include punctate epithelial keratitis, epithelial macro-erosions, shield ulcer, plaque formation and late corneal vascularization. Coalescence of punctate epithelial keratitis areas leads to frank corneal epithelial erosion, leaving Bowman's membrane intact. If untreated, a plaque containing fibrin and mucus deposits over the epithelial defect. Epithelial healing is then impaired, and new vessel growth is encouraged. The oval-shaped epithelial defects, known as shield ulcers (Fig. 3), usually have their lower border in the upper half of the visual axis. Healed shield ulcers may leave a subepithelial ring-like scar (Fig. 4). Corneal ulcer is reported to occur in 3–11% of patients. Corneal changes cause permanent reduction in visual acuity in 6% of patients suffering from VKC. Pseudogerontoxon, which resembles arcus senilis, is a waxing and waning grey-white lipid deposition in the superficial stroma of the peripheral cornea.

**Mediators in VKC**

The plethora of mediators and cytokines in VKC compared to controls, seasonal allergic conjunctivitis and giant papillary conjunctivitis provides a new perspective on the complex inflammatory processes occurring on the ocular surface in this chronic disease.

**Cytokines**

Cytokines are small secreted proteins that mediate and regulate immunity and inflammation. Activated helper T cells especially Th2 (CD4), mast cells and eosinophils are the main cytokine-producing cell types infiltrating the conjunctiva during chronic allergic eye diseases.

**Chemokines**

Chemokines, a short term for chemo- tactic cytokine (CC), are potent activators and chemoattractants. Chemokines are produced not only by inflammatory cells but also by stimulated epithelial cells, fibroblasts and vascular endothelial cells in the conjunctiva.

**Histamine**

Histamine an important inflammatory mediator in allergic eye disease, is released by activated mast cells and basophils. Histamine exerts its biological effects by interacting with
four G-protein coupled receptors, classified as H1–H4. Vasodilatation, chemosis and itching of eye are caused by histamine interaction with H1 receptors. Persistent elevation of histamine levels in VKC tears is probably caused by its reduced inactivation by histaminase and increased production by specific or non-specific activation of mast cells and basophils.

**Metalloproteinases (MMPs)**
MMPs are extracellular endopeptidases that selectively degrade components of the extracellular matrix. Inflammatory cells, particularly eosinophils, and structural cells like epithelial cells and conjunctival fibroblasts are the probable cellular source of these enzymes. Increased production and activation of MMPs or imbalance between MMPs and their natural tissue inhibitors (TIMPs) are all probably involved in the pathogenesis of conjunctival inflammation, remodelling and corneal changes in VKC.

**Growth Factors**
Several growth factors, such as epidermal growth factor, fibroblast growth factor and transforming growth factor beta-1 (TGFβ-1), were increased in VKC. These factors induce fibroblast growth and procollagen production.

**Cells in VKC**
Cells that involved in the inflammation of VKC are Mast cells, Eosinophils, T cells, B cells, Epithelial Cells, Fibroblasts and Natural Killer Cells (Fig5).

**Mast Cells**
Mast cells bind IgE on its surface. Cross-linkage of this IgE by specific allergens results in the release of pro-inflammatory mediators, including histamine, proteases, prostaglandin D2 and leukotriene C4, into the local extracellular environment. These mediators are responsible for causing ocular itching, hyperaemia, lacrimation and chemosis in allergic conjunctivitis.

**Eosinophils**
50–90% of cells in the tears during the active phase of VKC are eosinophils. Activated eosinophils release cytokines, chemokines, leukotrienies and epithelio-toxic proteins such as MBP, ECP, eosinophil peroxidase (EPO) and eosinophil protein X/neurotoxin (EPX).

**T cells**
T cells are mainly Th2 type 29. Cytokine flow cytometry has shown that 67% of VKC patients have Th2 cells in tears, while Th1 cells are seen in the tears of only 8% 22. The predominance of Th2-like cells in tears and conjunctival biopsy suggests a local Th2 response in VKC. Th2 lymphocytes, by virtue of their cytokine profile, are responsible for increased production of IgE, recruitment and activation of mast cells and eosinophils. B lymphocytes expressing the ligands CD23, 21 and 40 in conjunctiva specimen from VKC may be a precursor of IgE producing B cells.

**B cells**
B lymphocytes expressing the ligands CD23, 21 and 40 in conjunctiva specimen from VKC may be a precursor of IgE producing B cells.

**Epithelial Cells**
These cells not only act as a mechanical barrier, but also participate in the regulation of allergic inflammation through expressing surface antigens such as adhesion / effector molecules (ICAM-1, vascular cell adhesion molecule-1 and HLA-DR) and releasing many cytokines (eotaxin, IL-8, IL-6, RANTES). Histamine, released from the conjunctival mast cells, might stimulate the synthesis of IL-6 and IL-8 by conjunctival epithelial cells and amplify the allergic response.

**Fibroblasts**
Fibroblasts can modulate the functions of mast cells and eosinophils through the membrane form of stem cell factor and GM-CSF, respectively. Fibroblasts can be affected by inflammatory mediators derived from mast cells and eosinophils, such as TGFβ and nerve growth factor and by the Th2 cytokines, IL-4 and IL-1327.

**Histopathology and Immunohistochemistry**

**Tissue inflammation**
The histopathology of VKC is characterized by infiltration of the conjunctiva by eosinophils, basophils, mast cells, Th2 cells, monocyte/macrophages, dendritic cells, plasma cells and B lymphocytes, frequently organized as small lymphoid follicles without a germinative centre.

**Tissue remodelling**
Conjunctival thickening, subepithelial fibrosis, mucus metaplasia, neovascularization and scarring are typical of chronic VKC. Epithelial changes, connective tissue deposition, oedema, inflammatory cell infiltration and glandular hypertrophy all contribute in the tissue remodelling observed in VKC.

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A Case of Endotheliitis in a Child Following Mumps Infection

Sunita Chaurasia, MD

Abstract:
A 5-year-old boy complained of decreased vision in his left eye. His right eye was completely normal. Visual acuity was perception of hand motions in the left eye. Ocular examination of the left eye revealed diffuse corneal edema more in the central cornea. The patient had been diagnosed as having mumps 3 weeks ago by a pediatrician. He was treated with topical prednisolone acetate eyedrops in tapering doses for a month. Complete recovery of the endotheliitis occurred following treatment. Specular microscopy at 8 months follow up revealed a reduced endothelial cell density and increased mean cell area compared to the unaffected eye. Mumps Endotheliitis resolves well with topical steroids but can lead to endothelial cell loss.

Mumps is a disease caused by Rubulavirus in the family Paramyxovirus. It is characterized by swelling of the parotid glands, salivary glands and other epithelial tissues. With the adoption of immunization schedules, the prevalence of mumps is has reduced as it is a vaccine preventable disease. However, those children who miss their regular vaccination and adults are still at risk of developing mumps. The condition is self limiting but can lead to significant morbidity and at times serious systemic complications such as encephalitis, meningitis and deafness.1

Ocular involvement is relatively rare and most reports suggest a unilateral involvement. Ocular manifestations reported are conjunctival injections, superficial punctuate keratitis, interstitial keratitis, anterior uveitis, keratouveitis, nerve palsies, dacryoadenitis and endotheliitis.2, 3 Reports of endotheliitis are few and described in adults.4, 5

We herein report a 5 year old boy who developed endotheliitis following mumps. Treatment with topical steroid led to complete recovery but specular microscopy revealed a reduced endothelial cell density and increased mean cell area compared to the fellow eye.

Case Description
A 5 year-old boy presented with complaint of blurred vision of 1 week duration. He was on treatment with oral acyclovir 200 mg 5 times/day prescribed by a local ophthalmologist. He had been diagnosed to have mumps 3 weeks ago by a pediatrician and was recovering. At the time of this visit, the child was afebrile, and there was no swelling and tenderness over the parotid region. On examination, his visual acuity was 20/20 in the right eye and hand motions in the left eye. The left eye showed an edematous cornea localized more in the central part. (Figure 1).

The conjunctiva was quiet and anterior chamber was deep, pupil was round, regular and reacting to light. The B scan ultrasonography showed a normal posterior segment. Intraocular pressure was 12 mm Hg in the left eye. The right eye was unremarkable. An anterior segment OCT (Visante, Zeiss) was performed which showed a central corneal thickness of 1.13 mm. The patient was diagnosed with endotheliitis and started on prednisolone acetate eye drops every 4 hourly for a week, followed by tapering every one weekly. At 1 month follow up, the vision improved to 20/20. The cornea was clear and compact. At the last follow up 8 months later, the specular microscopy of the left eye showed an endothelial cell density of 2404 cells/ mm², mean cell area of 416 mm², CV 38%. The right eye had a cell density of 3267 cells/mm², mean cell area 306 mm² and CV 34% (Figures 2a and b).

Discussion
Corneal endotheliitis is more commonly implicated with herpetic/cytomegalic infection and relatively rarer with mumps virus. Endotheliitis due to herpetic causes are usually localized and often disciform. Corneal endotheliitis due to cytomegalus infection presents with coin shaped lesions on the lesions which can worsen with steroid use.

Mumps endotheliitis is typically seen in the resolving phase of mumps when active viremia is subsiding. The corneal endotheliitis that occurs in mumps is likely to be due to immune response as has been reported earlier and was seen in this patient. The rapid and complete response to topical corticosteroids also favors an immune pathology as the underlying cause. Although the condition responds well with steroids, it can lead to endothelial cell loss.

Most of the cases of mumps endotheliitis have been reported in young and middle aged adults. As mumps vaccine is a part of regular immunization programs, those children who...
Figure 2: Specular microscopy of the right and the left eye at 8 months follow up. The left eye shows a reduced cell count and increase in the mean cell area.

miss their vaccination are inherently at risk of developing mumps and its ocular complications. This case serves to raise an awareness of the Public Health issues and potential ocular manifestations associated with mumps vaccination non compliance.

References:

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Definition:
Persistent fetal vasculature (PFV) is a most common developmental ocular anomaly, characterised by a complex clinical spectrum, which develops due to failure in the complete involution of the hyaloid vasculature system (HVS) and primary vitreous.

Clinical Spectrum:
- Leukokoria
- Cataract (Posterior polar cataract)
- Persistent pupillary membranes
- Microphthalmia
- Retrolenticular fibrovascular tissue
- Elongated ciliary processes
- Usually unilateral (90% of cases)

Associated Features:
- Late-onset angle-closure glaucoma
- Intraocular hemorrhage
- Optic nerve hypoplasia
- Retinal detachment
- Amblyopia

Introduction:
The hyaloid vascular system (HVS) in the primary vitreous is an example of developmentally regulated vascular involution. In the HVS, the hyaloid artery arises from the ophthalmic/central retinal artery posteriorly and branches in the vitreous to form the vasa hyaloidea propria (VHP) and the tunica vasculosa lentis (TVL), which envelops the lens.(1),(2) The pupillary membrane makes up the anterior component of this system(1, 2). These fetal vessels start to grow during the first month of gestation, reach their maximum proliferative activity by the second to third month, and begin to involute at 4 months of gestation. Normally these fetal vessels disappear by birth.(1, 3) These vascular beds normally regress to create the avascular cornea, lens, and secondary vitreous composed of collagen, fibronectin, and other extracellular matrix macromolecules(4).

Reese(3) in his 1955, Jackson Memorial Lecture, described this entity as a Persistent hyperplastic primary vitreous (PHPV), process associated with persistence of the hyaloid vascular structures in the vitreous(1),(5). Recently, Goldberg(1) revisited the nomenclature in his 1997, Jackson Memorial Lecture, and proposed the term Persistent fetal vasculature (PFV), to reflect more accurately the broader manifestations of failed regression of other vascular beds in the eye. Depending on whether the major abnormalities are in the region of the TVL/VHP or the hyaloid artery, PFV is subdivided into anterior or posterior forms, respectively. (6),(7) Most reported cases show overlap, though, with abnormalities in both anatomic compartments. (7) Fibroblast-like cells, occasionally intermixed with pigmented cells, form a fibrovascular mass surrounding the remnants of the HVS(6),(8). In anterior or combined PFV, the retrolental tissue lies adjacent to a rent or posterior bulge in the posterior lens capsule, typically resulting in cataract formation.(3, 6),(8) In some cases, it can lead to intralenticular hemorrhage, lens swelling with secondary glaucoma; and total lens absorption, calcification, or even replacement by adipose tissue.(6, 8) In posterior or combined PFV, the retina may be detached by congenital nonattachment or by traction from the retrolental tissue adjacent to the innerneuroretina.(6),(8) This may be associated with retinal folding, dysplasia, and reactive retinal pigment epithelial (RPE) cell accumulation.(8) In PFV some, or all, components of the fetal intraocular vasculature remain after birth. This malformation may affect the anterior, retrolental, and/or posterior parts of the infant eye. Although, historically the visual outcomes have been poor, recent advances in clinical evaluation and surgical technique have lead to more frequent anatomical and functional restoration of the afflicted eye.

Etiopathogenesis:
The etiology of PFV is not well established. PFV is sporadic and frequently unilateral, but bilateral disease has been described in 2% to 30% of children. (6),(8),(9) One report exists of possible vertical transmission with both a mother and son affected, which suggests autosomal dominant inheritance.(10) In two other reports, autosomal recessive inheritance was suggested; these included one case of fraternal twins, both of whom had PFV.(11),(12) Although no definite risk factors or confirmed systemic associations occur, maternal cocaine use during pregnancy of affected newborns has been reported.(13), (9) Rarely, PFV occurs in conjunction with other ocular or systemic abnormalities, for example, microcephaly, mental retardation, spasticity, cleft palate, and short stature in the oculopalatocerebral dwarfism syndrome.(14) One case of PFV accompanying with bilateral megalocornea,(15) Reigers anomaly(16) and another with optic nerve coloboma(17) have been noted. There is no reported gender predilection.
Clinical Manifestations:
Understanding of the embryological milestones responsible for the development of fetal vasculature is crucial for the understanding of the diversity of symptoms and signs appearing in PFV. Persistence of some or all fetal vasculature may have profound morphological consequences. If this regression does not occur completely, subsequent contraction and opacification of the primary vitreous along the hyaloid vascular system lead to the clinical presentation of PFV. The broad clinical spectrum of PFV depends on the anatomic region involved (i.e., Anterior, Posterior, or both) and the extent of residual hyaloid vessels. Typically, PFV presents in children as unilateral microphthalmia, leukocoria, or cataract, and is associated with retinal folding and detachment.(6) Visual acuity can be nearly normal, but is 20/200 or less at diagnosis in most cases of posterior PFV.(7) Depending on its severity, surgical treatment focuses on vision preservation by lensctomy, vitrectomy, or membranectomy to prevent the sequelae of glaucoma and phthisis. Occasionally, enucleation of a blind eye is required for pain control or cosmesis.(1),(6)

Persistent pupillary membrane(18). A medusa head like appearance pupillary membrane with pigmented iris strands are usually tenacious, with fine fibrils attached to the iris collarette on one side, with the other end either free floating or attached to the anterior lens capsule or iris focally on the opposite side. This condition is thought to represent ectopic iris tissue on the lens with abnormal iris stromal caused by aberrant involutional changes in the primitive embryological tunica vasculosa lentis (TVL) of anterior segment. These vessels may deform the pupil. A thin fibrous sheet may appear and congenital iris ectropion (entropion uveae) may also occur. Vision may be unaffected or reduced, depending on the extent of pupillary occlusion. The presence of a pupillary membrane may aid in the diagnosis of PFV in an eye with total cataract or whitish retrolental mass.

Iridohyaloid blood vessels. These fetal vessels appear as radial, short, and parallel vessels by the equator of the lens. They constitute a vascular connection between the posterior and the anterior tunica vasculosa lentis. When these vessels do not regress by the second trimester of gestation, they contribute to the appearance of radial superficial vessels in iris stroma. Often, white limbal connective tissue malformation may be seen in the same meridian. When the vessels reach the pupil they make hairpin loops, inducing a small pupillary notch.

Posterior fibrovascular sheath of the lens(19) Persistence of the posterior tunica vasculosa lentis may cause the appearance of a fibrovascular mass behind the lens. Reese(3) described this as the hallmark of PHPV syndrome. The retrolental membrane may be small or may cover the entire posterior capsule of the lens. It may be associated with a clear lens or cause variable degrees of lenticular opacification. Typically the retrolental membrane is white or pink in color, differentiating it from the yellow tissue seen in Coats disease or the snow-white tissue typical of calcified retinoblastoma. Formation of a retrolental membrane in PFV is often accompanied by traction on and elongation of the ciliary processes, which may become visible as the pupil is dilated. Although prominent and centrally displaced ciliary processes were once considered pathognomonic for PHPV, they are also seen in Retinopathy of prematurity stage V (ROP), Norrie disease(9), Trisomy 13, and congenital subluxated lenses.(1)

Posterior capsular plaque: The association of posterior capsular plaque and PFV has only recently been described. Mullner-Eidenboch(20) and colleagues first reported that a high proportion of unilateral cataracts had associated findings such as posterior capsular plaque that could represent a subtle form of PFV. Wilson et al in their Infant Aphakia Treatment Study (IATS) later confirmed the association of posterior capsular plaque and unilateral cataract.(21) In this multicenter study of primary intraocular lens (IOL) placement versus contact lens use at the time of surgical removal of unilateral congenital cataract, 88% of all children with cataract, and 100% of infants with unilateral nuclear cataract had an associated posterior capsular plaque. In the IATS study, plaque was hypothesized to be formed by fetal vessels perforating the lens capsule during lens development. Mullner-Eidenboch(20) and colleagues theorized that these perforating fetal vessels create an abnormally strong adherence of the lens to the posterior capsule. The vessels subsequently resolve but the lens opacity and posterior capsular plaque will remain.

Unilateral posterior lenticonus or lentiglobus in association with PFV or minimal fetal vascular remnants (MFVR) has been previously described(20, 22-25). Kilty et al(22) hypothesized that the traction exerted by the remnants of the hyaloid artery on the posterior lens capsule may have a role in causing posterior lentiglobus. Abdel-Hafez et al(25) reported progressive lentiglobus in an eye with PFV. According to them, PFV and the development of lentiglobus may have been concurrent events due to random developmental arrests, rather than lentiglobus being a consequence of traction caused by PFV. Though conventionally believed to be a common cause of unilateral infantile cataract, posterior lenticonus has been demonstrated to be responsible for a significant proportion of unilateral as well as bilateral childhood cataracts.(26) In a large series reporting surgical outcomes in childhood posterior polar and posterior lentiglobus cataracts, all cases with posterior lenticonus were monococular, and visual outcomes were generally good.(27) In contrast to this, bilateral PFV generally presents with both anterior and posterior components, and visual outcomes are disappointing with or without intervention.(28)
Mittendorf dot: This small white dot on the posterior surface of the lens is typically found 0.5 mm to the nasal side of the center of the posterior pole and designates the point of incomplete regression of the hyaloid artery, where it attaches to the posterior surface of the lens. It is normally found in 0.7 to 2 % of the population and rarely causes any visual disturbance(29).

Persistent hyaloid artery: The fetal hyaloid artery lies within the Cloquet canal and normally loses perfusion around the seventh month of gestation. When this vessel persists, it extends from the optic nerve to the lens. It may be filled with blood but is usually bloodless(30).

Bergmeister papilla: (31) This is term used to describe a benign remnant of the posterior part of the hyaloid artery and can be seen as an epipapillary vascular tissue. Its effect on vision depends on the presence of other associated optic nerve abnormalities.

Congenital tent-shaped retinal detachment: Congenitally detached retina can result from PFV traction on the retina. It typically has the shape of a tractional retinal detachment and it adheres to the posterior surface of the lens, ciliary body, or both. The detachment may progress, and it has grave visual consequences.

Macular abnormalities: Various dysplastic and hypoplastic abnormalities of the macula may occur in PFV and these will inevitably affect vision.

Optic nerve abnormalities: Both primary and secondary abnormalities of the optic nerve, including optic disc hypoplasia, may be seen in PFV.

Microphthalmos: Microphthalmia was defined based on previously published normal axial length measurements stratified by age.(32) Anaxial length difference (catactous eye vs. the noncataractous fellow eye) of 1.5 mm was classified as mild microphthalmos, and an axial length difference of 1.6 mm was classified as severe microphthalmos. Retention of fetal vasculature may be accompanied by an arrest in the growth of the eye globe. Typically, eyes with severe forms of PFV have some degree of microphthalmos. Additional changes include a decreased corneal diameter and distortion of the configuration of globe wall, with colobomatous microphthalmos as a result.

Minimal fetal vascular remnants of the posterior lens capsule/anterior hyaloid face such as radiating perfused vessels and/or nonperfused spidery ghost vessels, an abnormally thickened anterior hyaloid face or a membranelike structure contiguous with the posterior capsular lens opacity, and the type of posterior lens clouding (axial/paraxial translucent opacity, axial/paraxial lenticytic area, axial/paraxial calcified plaques) were documented. The degree of lens opacification (20) i.e., clear lens, nuclear clouding, anterior or posterior pole opacification, intralenticular bleeding, and total cataract.

Clinical Diagnosis:
PFV is typically a clinical diagnosis. The young age at which affected children are usually diagnosed means that an examination under anesthesia to confirm the diagnosis and to examine the fellow eye is often indicated. Despite the continuum of clinical manifestations, diagnosing PFV may sometimes be challenging. When the full constellation of clinical findings is present unilaterally, the diagnosis is usually straightforward. True bilaterality rarely observed. It is in these cases of bilaterality, as well as in those that have atypical clinical presentations, that additional history and ancillary testing may be useful. Positive family history, complicated birth history, and bilaterality, as seen in retinoblastoma, Norrie’s disease, and retinopathy of prematurity, are frequently absent in PFV. Any child with a cataract, unilateral or bilateral, especially when associated with a microphthalmic globe, should be suspected of having PFV.

Role of Imaging:
When clinical signs are indecisive, adjunctive imaging may help in making the accurate diagnosis. The most useful and noninvasive tool is ultrasound biomicroscopy. Both posterior segment and anterior segment ultrasound biomicroscopy is invaluable. Posterior segment ultrasound biomicroscopy typically shows a small globe with a retrolental membrane and a vitreous band extending from the posterior lens capsule to the disc area. (33) It can also reveal whether a retinal detachment is present, which may influence the choice of surgical technique used to remove the cataract. High-frequency ultrasonography (34) may demonstrate an anteriorly placed and swollen lens with a resultant shallow anterior chamber, centrally dragged ciliary processes, and thickened anterior vitreous face appearing as a double linear echo near the pars plana or pars plicata. In addition, color Doppler imaging of the persistent hyaloid artery may detect blood flow within the stalk. Computerized tomography and magnetic resonance imaging have also been reported as excellent adjunctive devices in the evaluation of PFV. (35), (36) Computed tomographic scanning can be used to evaluate the presence of intraocular calcification (37), which is seen frequently in retinoblastoma but not in PFV. Conversely, microphthalmia is a consistent feature of PFV but not most other causes of leukokoria. Bilateral axial length measurements with ultrasonography, therefore, are helpful. The rare occurrence of retinoblastoma in an eye with PFV has been reported. (38)

PFV and Associated Anomalies:
Although PFV mostly appears as a single anomaly, sometimes it may be associated with other ocular abnormalities such as Peters anomaly, (39) Rieger anomaly, (16) and morning
glory syndrome.(40),(41) Only 5 to 10% of children with PFV have binocular involvement. Bilaterality represents a more widespread degree of abnormal embryological development. Associated systemic anomalies may occur, especially neurological abnormalities.(42) Haddad et al(8) have reported systemic abnormalities including cleft palate and lip, polydactyly, and microcephaly in association with bilateral PFV. Goldberg reported on the association of PFV with trizomy 13.(1) A few pedigrees with familial PFV have been described,(43) suggesting the possibility of an autosomal recessive(44) or autosomal dominant(10) inheritance pattern in selected cases. In animal models, the presence of PFV has been associated with Arf tumor suppressor gene deficiency,(45),(46) angiopoietin-2 deficiency,(46) abnormalities of macrophage-induced programmed cell death,(47) and abnormalities of astrocyte cell migration.(48)

Management:
Historically, eyes with PFV, particularly when accompanied by dense cataracts obscuring the visual axis during the critical period of visual development, were considered to be blind.(49), (50) Surgery was indicated only to avoid or treat complications such as angle-closure glaucoma, vitreous hemorrhage, progressive retinal detachment, and phthisis(3). Many eyes eventually required enucleation, with a resultant poor cosmetic outcome.(6) Since the advent of closed-system vitreoretinal instrumentation, removal of the cataract, retrolental mass, and persistent hyaloid stalk has been made possible. By surgical release of the traction on the ciliary body in eyes with PFV, the eye is allowed to grow and acceptable cosmetic improvement achieved. (51) Even though initial surgical goals in eyes with PFV were mainly to avoid the complications of the disease and improve cosmesis, reports of useful postoperative vision following microsurgical vitrectomy techniques began to appear in the 1980s.(52-55) Successful visual rehabilitation has been reported for the most part in PFV with anterior presentation, i.e., without disc or macular involvement. (56) Recent introduction of sophisticated microsurgical techniques, in combination with aggressive amblyopia therapy, has resulted in more favorable visual outcomes for eyes with PFV.(9, 57-59) Therapeutic goals should therefore be expanded to include saving useful vision. Several surgical approaches removing the cataract and the retrolenticular fibrovascular membrane in PFV have been described. During the past two decades, the modified surgical strategy in the management of PFV, similar to that previously described for uncomplicated cataracts.(60)

Surgical techniques: includes either the pars plana or the limbal approach. In both, the cataract is removed by a lensectomy in combination with an anterior vitrectomy and removal of any retrolenticular membranes or fibrovascular tissue associated with persistence of the hyaloid system.

Posterior Approach: A micro vitreoretinal (MVR) blade (20 gauge) is used to perform a sclerotomy at the 10 o'clock position 1.5 to 2.0 mm from the limbus. The MVR blade is then pierced through the lens by the equator, leaving an opening in the anterior capsule. A cannula with irrigation fluid is introduced through a similar sclerotomy site at the 2 o'clock position. A vitrectomy handpiece is then introduced through the sclerotomy site at 10 o'clock and inserted in the opened bag, and the lens material is aspirated within the bag. After removal of all lens material, the lens capsule and adjacent retrolental membranes are removed by the vitrector. When the retrolental membrane is too thick to be cut with a vitrector alone, intraocular scissors may help to segmentally cut the membrane into fragments small enough for the vitrector to remove.(61) An anterior vitrectomy with removal of the anterior part of the hyaloid stalk is then performed. Possible bleeding from the patent hyaloid artery can usually be controlled by raising the infusion set or by applying diathermy to the bleeding stump. Since all lens capsule material is removed when using this posterior approach, the eye remains aphakic. When posterior abnormalities are present, a complete posterior vitrectomy is suggested, with peeling of membranes to release retinal traction and folds. In some cases air–fluid exchange may be indicated.

Anterior Approach: The main advantage of this technique is that, it allows implantation of IOL, aid in better visual rehabilitation and final cosmetic outcome.(57) In addition, the anterior approach facilitates surgery by avoiding the peripheral retina, which might be attached to the ciliary body with an absent pars plana.(8) Briefly, in the anterior approach an MVR blade or 15 degree super sharp blade is used to create two paracentesis incisions at the limbus or in the peripheral cornea near the limbus: one for the vitrectomy hand piece and a second for the anterior chamber maintainer. These incisions can be made in the superior nasal and superior temporal locations or placed superotemporally for the vitreor for inferotemporally for the infusion. Anterior capsulotomy is performed with vitrectomy cutter. The lens is aspirated within the bag. A posterior capsulotomy and removal of retrolental membranes are performed using the vitrector. In cases where the membranes are thick and stiffened, scissors may be used to fragment the membrane before final removal with the vitrector. Through the opening in the posterior capsule, the anterior vitreous and anterior portions of the persistent hyaloid artery are amputated by using the vitrectomy-cutting instrument. An IOL can then be inserted either in the bag or in the sulcus, similarly to uncomplicated childhood cataract.

Visual Outcome:
To achieve satisfactory vision in children with PFV, regardless of surgical approach, timely intervention followed by aggressive antimyopic patching therapy is indicated.
Anteby et al.(57) reported the visual outcome in 89 eyes with unilateral PFV, comparing 60 operated eyes to 29 nonoperated eyes. In this large series a final visual acuity of 20/200 or better in 25% of 60 operated eyes was achieved. Mittra et al.(59) described even more promising results of 14 eyes managed by surgery and prompt amblyopic treatment, with 66% achieving 20/100 or better vision. However, in this series no long-term follow-up was available and possible late complications affecting vision were not taken into account. Alexandrakis et al. (58) in a study of 30 eyes managed by surgery, reported 47% of the operated eyes achieving 20/400 or better, compared to 12% in nonoperated eyes. Sisk et al. (62) reported that 49/70 eyes had the presence of form vision after surgery defined as counting fingers or better. In all of these studies, more severe, posterior forms of PFV were associated with a poorer final visual and structural outcome. (9), (62) However; successful visual results have been reported after surgery with PFV of the anterior approach. (6), (56)

Conventionally, operated PFV eyes remained aphakic. With an anterior surgical approach, the insertion of an IOL during the initial surgery is practicable. IOLS assist postoperative care by avoiding the need for contact lenses. Due to the microphthalmos often encountered in PFV, contact lenses need a high refractive power and can be difficult to fit on a small cornea. Recent reports have included attempts to rehabilitate vision in PFV eyes by IOL implantation. Anteby et al.(57) inserted IOLS in 30 eyes with unilateral PFV over the past 15 years. A good visual acuity, 20/50 or better, was seen in 20% of these eyes and a fair visual acuity, 20/200 or better, was obtained in 33.3%. Mittra et al.(59) stated on the use of IOL in two eyes with PFV with a satisfactory visual outcome. Although the IATS was not powered to detect differences in IOL versus contact lens use in children with PFV, results for children with PFV from this study have been reported separately. (63) The IATS had specific inclusion criteria for enrollment regarding PFV. Infants had to have unilateral lens opacity present before 7 months of age with a corneal diameter of 9 mm or greater. (64) An absence of posterior findings such as ciliary process stretching and retinal traction was also mandated. In the IATS, 18 infants were determined to have evidence of persistent hyaloid remnant or retrolental vascular membrane. Eleven kids were randomized to receive aphakic treatment with contact lenses and seven received a primary IOL. Median logMAR visual acuity at 1-year was 0.88 for patients with PFV and 0.80 for patients without PFV. This difference was not stastically significant. One or more adverse event during the first year after surgery occurred in 67% of infants with PFV and 46% of infants without PFV.

**Postoperative Complications:**
The principal post-operative complications in PFV eyes include glaucoma, secondary membrane formation, vitreous hemorrhage, retinal detachment, and strabismus. Dass and Tresie(9) described a general reoperation rate of 32% in 27 eyes with PFV. The rate of glaucoma in eyes with PFV varies from series to series. Anteby et al.(57) reported a 15% overall rate of glaucoma in 89 PFV eyes. Glaucoma developed twice as often in eyes with aphakia (22%) as in nonoperated eyes (11%). The rate of glaucoma in PFV eyes with pseudophakia was only 8%. (57) This suggests that IOL implantation in these eyes does not increase the risk for glaucoma, although eyes selected for IOL implantation may have less severe forms of PFV. Others reported glaucoma to occur in up to 30% of eyes operated for PFV. (65) Glaucoma associated with PFV is often diagnosed within the first year after lensectomy but may also develop several years after surgery. (65) Despite the performance of a relatively large posterior capsulotomy in eyes with PFV, the rate of secondary cataract and membrane formation necessitating further surgery is high up to 30%. (57), (65) Possibly this high rate of secondary cataract can be attributed to the microphthalmos itself, as typical postoperative complications have been found to be less common in PFV eyes that are myopic. (65) It is thought that a more complete relief of circumferential traction on the ciliary processes will reduce reoperation rates. Cutting the retrolental membrane between each stretched ciliary process until they all fall back into a more normal position does this. The reoperation rate seems unaffected by insertion of an IOL during the initial lensectomy. (57) In the IATS, adverse events in the first year after surgery were significantly higher in patients with PFV compared to patients without PFV in the contact lens group (55% vs. 20%), but not in the IOL group (86% vs. 71%), possibly because all children receiving IOLS had higher rates of adverse events when compared to aphakic children (73% vs. 29%) in the study. (63, 64)

**Cosmetic Outcome:**
The primary objective of surgical treatment of eyes with PFV should be focused in maximizing the cosmetic outcome. Complications i.e., secondary glaucoma, vitreous hemorrhage, pseudo-Peter anomaly and phthisis have been attributed mainly to eyes with PFV that are not receiving surgical intervention. These complications often result in disfigured globe. In addition, poor vision consequences in development of sensory strabismus further blemishing the child. The quality of life for children with PFV is severely affected by the development of phthisis and deformed eye or the need for a cosmetic shell or prosthesis. Anteby et al. reported 30% of PFV eyes developing a visible cosmetic blemish due to advanced microphthalmos, buphthalmos, extensive corneal leukemia, or total phthisis. Interestingly also operated eyes may become cosmetically blemished. Anteby et al(57) showed that 25% of aphakic eyes and 7.1% of nonoperated eyes needed a prosthesis or cosmetic shell during the years of follow-up, whereas none of the eyes with pseudophakia needed this type of rehabilitation. (57)
the good cosmos is in eyes with PFV. Must include maximizing the visual potency and achieving complications such as secondary angle closure glaucoma. Amblyopic care after surgery. These children should be Visual success depends not only on the severity of the PFV, but also on the timely intervention combined with prompt amblyopic care after surgery. These children should be scrutinized for the potential development of postoperative complications such as secondary angle closure glaucoma and cataract formation. The objectives of rehabilitation must include maximizing the visual potency and achieving the good cosmos in eyes with PFV.

Conclusion:
The prompt diagnosis and timely intervention is the key augmenting the surgical success in a child with PFV. In eyes where visual axis obscuration owing to cataract and retrolental membranes, early intervention should be attempted to achieve good visual outcome and cosmetic result. Meticulous surgical steps, whether using an anterior or a posterior approach, lenectomy, anterior vitrectomy, release of ciliary body traction, and removal of the anterior portion of the hyaloid stalk. When technically possible, these eyes can be optically rehabilitated by suitable IOLs. Visual success depends not only on the severity of the PFV, but also on the timely intervention combined with prompt amblyopic care after surgery. These children should be scrutinized for the potential development of postoperative complications such as secondary angle closure glaucoma and cataract formation. The objectives of rehabilitation must include maximizing the visual potency and achieving the good cosmos in eyes with PFV.

References:
Amblyopia

Orbit, Oculoplasty
Amblyopia also known as “lazy eye” is one of the most common visual disorders in children and persistent visual impairment in adult hood. It is caused by abnormal visual experiences early in life.

**Definition**

Amblyopia has been defined as a unilateral or bilateral decrease of visual acuity caused by deprivation of pattern vision or abnormal binocular interaction, for which no cause can be detected by physical examination of the eye and which in some cases can be reversed by therapeutic measures. (1)

Amblyopia is a disorder of development of the visual system that can present with varying levels of severity and usually affects one eye. This visual disorder is potentially reversible if detected and treated at young age. Timely diagnosis and appropriate treatment is extremely important and usually gives rewarding results.

**Indian scenario**

The prevalence of amblyopia in India is not exactly known, but studies in school children have shown it to be in the range of 1.1% to 5% of all children (2,3). This results in a huge public health burden although this is a preventable cause of low vision.

**Clinical definition**

Clinically, a reduction in best-corrected visual acuity to less than 6/9 (20/30) monocularly in Snellen optotype or a two-line difference or more in best-corrected visual acuity, is considered to be amblyopia.

However, it must be remembered that amblyopia cannot always be defined in terms of visual acuity in children. Fixation preference is also a very important tool in detection of amblyopia in young children and infants. Clinical tests such as resistance to monocular occlusion, fixation testing with prisms may be useful in the younger age group.

**Pathophysiology**

Amblyopia is considered to derive from the degradation of the retinal image associated with abnormal visual experience during the developmental period of the visual system in infancy and early childhood.

The concept of ‘Period of critical development’ is the key to understanding of the pathophysiology of amblyopia. The normal development and right function of the cortical neuronal circuits fundamentally depend on the clarity of the visual image and the equal perception of the visual stimuli with respect to both eyes. In the stages of early visual development, there exists visual plasticity due to adaptive nature of cortical neurons. Period of visual plasticity begins at 4 months of age, peaks at 2 years after which gradually declines and ceases by 12 years of age.

Children with anisometropia, strabismus or any other condition causing a reduction in the clarity of the image in one or both eyes, thereby disrupting equal binocular vision, are at risk of developing amblyopia. (3,4)

Hubel and Weisel's pioneering work explained the mechanism of deprivation induced competitive inhibition in cortical cells. They established, that by alternating ocular dominance columns of visual cortical cells reacted selectively to the stimulus of the opened or the occluded eye. (5,6) Multiple animal models have been used to study the effects of deprivation on the cortex and now functional MRI (fMRI) studies are of further use in elucidating the possible mechanisms in this disease. (7)

**Etiological types**

- **Strabismic**
- **Refractive - Anisometropic**
  - Isometropic (Ametropic)
  - Meridional
- **Stimulus Deprivalional** (8)

**Strabismic Amblyopia**

It is the most common form of amblyopia and is usually unilateral. Constant, non alternating or unequally alternating strabismus, most commonly esodeviations, cause strabismic amblyopia. Other characteristics include reduced contrast sensitivity, unsteady and eccentric fixation and pursuit abnormalities. One can expect to find amblyopia far more often in esotropes than in exotropes, because exotropia is often intermittent at its onset.

**Anisometropic Amblyopia**

It is the second most common type of Amblyopia. It develops when unequal refractive error in two eyes causes image in one retina to be more defocused than the fellow eye. Visual acuity is lower binocularly than when tested monocularly. A difference in hyperopic error of more than +1.50 D, astigmatism of +1.50 D and myopia of more than -3D is usually considered significant enough to cause amblyopia.
Ametropic Amblyopia

Ametropic amblyopia results from large uncorrected, bilateral refractive errors, commonly due to high hyperopia or astigmatism. Hyperopia of 4.5D or 2.50 D of astigmatism (meridional amblyopia) and high myopia (usually > -5 D) usually cause amblyopia.

Visual deprivation Amblyopia

Visual deprivation amblyopia results from partial or complete obstruction of visual axis due to various causes which result in blurred retinal image. It is caused by a total lack of pattern stimulation, e.g.: Congenital cataracts, vitreous hemorrhage, corneal opacities, congenital ptosis.

Occlusion Amblyopia is a specific form of deprivation amblyopia that results from therapeutic patching or image blur due to prolonged cyclopia. It is also called reverse amblyopia.

Detection of Amblyopia

Visual acuity testing:
Linear acuity with single optotype is better than letters present in a row. This is called crowding phenomenon. By age 3 years, most children are able to perform recognition visual acuity tests, such as Allen or Lea pictures, Tumbling E, HOTV and Snellen letters.

Neutral density filter effect
With reduced illumination, acuity of an amblyopic eye declines less sharply than that of an organically diseased eye, this can be demonstrated with neutral density filters which improves visual acuity.

Assessment of fixation pattern
Fixation preference in one eye suggests poorer vision in the other eye. Presence of alternate fixation generally rules out amblyopia. The degree of fixation preference is estimated by briefly covering preferred eye to force the fixation in nonpreferred eye, if the fixation immediately goes back to the preferred eye after cover is removed then it indicated strong fixation preference. If the patient maintains fixation through a blink with smooth pursuit movement then milder degree of amblyopia is suspected.

Management of Amblyopia

Amblyopia treatment starts with elimination of the cause of visual deprivation or abnormal binocular interaction. For example, treatment of cataracts by surgery, management of corneal opacity, ptosis surgery, correction of refractive errors by glasses or contact lenses.

The next step is to provide additional stimulus to the affected eye by penalising the ‘good’ eye. This is usually accomplished by occlusion therapy which is considered the gold standard of amblyopia treatment. The other methods include pharmacological penalisation and optical penalisation.

Occlusion therapy

Occlusion therapy involves complete closure of the unaffected eye. This can be done by adhesive patches which are specially made for the purpose, other modalities include Doyne’s occluders, graded transparent filters and occluder contact lenses. Frosted spectacle glasses are usually not very effective since the child can easily peek over them.

While the traditional thought was to patch the good eye full time, most pediatric ophthalmologists now prefer part-time regimens which result in better parental compliance. The PEDIG – Amblyopia Treatment Studies (ATS) have shed light on multiple aspects of occlusion therapy. The ATS – 2A showed that 6 hours of patching is equivalent to full time patching for severe amblyopia (20/100 to 20/400) in children younger than 7 years.(9) The ATS – 2B showed that in children younger than 7 years with moderate amblyopia (20/40 to 20/80), 2 hours of patching was equivalent to 6 hours. (10)

Pharmacological penalisation

Pharmacological penalisation with atropine has also been used with or without spectacle correction as a modality of treating amblyopia. The PEDIG group showed with their ATS – 1 study that atropine penalisation and patching are equally effective in the treatment of moderate amblyopia in the age group of 3 to 7 years. (11) Also, there was an additional study which proved that weekend instillation of atropine was as effective as daily atropine used, and had a better compliance rate. (12) However, it must be kept in mind that atropine is a powerful drug and can lead to irreversible ‘reverse amblyopia’, specially in children with hyperopia. (13)

Pleoptics

Pleoptics is a method of treating eccentric fixation that may be associated with amblyopia. A bright ring of light is flashed around the fovea and leads to temporary saturation of the photoreceptors surrounding the fovea and eliminates vision from the eccentric fixation point and forces fixation to the fovea. Pleoptic treatments usually require several sittings, several times a week.

Newer advances

Multiple new avenues are being explored with regards to amblyopia management. One such avenue is the use of Levodopa – Carbidopa in addition to patching. Levodopa is a catecholamine precursor used to treat adults with Parkinson’s disease. Carbidopa – a peripheral decarboxylase inhibitor, prevents peripheral conversion of levodopa to catecholamine metabolites, thus allowing more levodopa to cross the blood–brain barrier. Depletion of catecholamines by a neurotoxin (6-hydroxydopamine) hals the plasticity
of the visual cortex. Plasticity can be re-established by infusion of exogenous catecholamines into the brain or by direct electrical stimulation of a locus, which projects catecholamine fibers to the visual cortex. Levodopa – Carbidopa can increase plasticity – thus help in amblyopia treatment. However, a corresponding increase in the chances of reverse amblyopia must be kept in mind. (14) Latest studies in adults are exploring possibilities of plasticity in adults that may change our management protocols in the near future. (15)

**Conclusion**

Amblyopia is a very common cause of visual impairment in both adults and children. If diagnosed early, amblyopia treatment can be rewarding. A great amount if patience in needed on the part of the treating physician and good counseling of the parents and care-givers is essential for a successful outcome.

**References**


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Hands on training for PG Students, TOC 2015
Blepharoplasty
Shubhra Goel, MD, Cat N Burkat, MD FACS

Introduction
With aging, the periorbital tissues undergo histopathological changes that result in decreased tissue elasticity, tone, and volume. In the eyelid region, this may manifest as droopy eyelids that can lead to a tired appearance, and eventually obscure vision. Blepharoplasty is one of the most popular cosmetic procedures for eyelid and facial rejuvenation.

Blepharoplasty, whether for aesthetic or functional purposes, is performed by removing redundant eyelid skin, excess herniated orbital fat, and hypertrophic orbicularis oculi muscle along the upper or lower eyelids. The primary goal of aesthetic blepharoplasty is the restoration of a youthful and natural eye. Enhanced superior and peripheral vision are the primary goals for those undergoing medically necessary blepharoplasty.

Indications (Figure-1)
1. Dermatochalasis (skin and/or fat) leading to obscured vision and heaviness over the upper eyelid margin
2. Dermatochalasis (skin and/or fat), due to age-related changes, for cosmetic improvement of the eyelid appearance
3. Improvement of eyelid symmetry
4. Improvement of eyelid contour and crease definition, such as in Asian eyelids
5. Lower eyelid dermatochalasis (skin and/or fat) leading to eyelid bags or fullness
6. Thyroid eye disease for sculpting of prolapsed orbital fat

Contraindications
1. Upper or lower eyelid skin anterior lamellar shortening (secondary to previous surgery, burns, lasers, or trauma)
2. Severe sulcus deformity (post trauma, aging, previous surgery)
3. Cicatricial entropion of the eyelids

Relative Contraindications
1. Severe dry eyes
2. Lower eyelid retraction
3. Anticoagulation that cannot be discontinued for surgery
4. Unrealistic expectations

Surgical technique:
A. Upper lid blepharoplasty

1. Preoperative evaluation (Figure-2)
   - In addition to the routine general and ophthalmic examination, a detailed periorcular and facial assessment is necessary. Schirmer’s test or measurement of the tear lake meniscus should be performed.
   - It is often helpful to assess old photographs together with the patient to better understand the patient’s desired outcome and whether a realistic goal is achievable.
   - Brow position- Normal aging initially manifests as noticeable lateral brow ptosis and lateral skin hooding, due to the lack of frontalis muscle fibers attached to the lateral brow soft tissues. Significant brow ptosis leading to pseudo-dermatochalasis should be addressed via a brow elevation procedure, rather than blepharoplasty. If true dermatochalasis is present in conjunction with brow ptosis, a combined brow lifting procedure and blepharoplasty can be performed through the same or different incisions(Figure-3).

   - Assessment of the upper eyelid skin is of utmost importance, and is measured in millimeters from the lash line to the thicker brow skin centrally, medially, and laterally. Approximately 18-20mm of the upper eyelid should be retained for normal function and closure of the eyelids, and should be individualized for each patient.
• The margin to reflex distance (MRD-1) should be documented for both sides. In some patients, the actual eyelid margin is in good position under the redundant overhanging skin, thus requiring only blepharoplasty surgery. In other patients, the eyelid margin approaches or covers the pupil, as opposed to the overhanging skin approaching the pupil and causing functional obstruction. These patients will require levator surgery for optimal results.

• Lid contour and shape - The eyelids should ideally have a positive canthal tilt, such that the lateral canthus is approximately 2mm superior to the medial canthus.

• Lid crease - In most females, the upper lid crease should be 8-12mm above the lash line. In males, it is typically lower at 7-9mm above the lash line. Patients of Asian descent have anatomically lower creases at 4-6mm, or absent creases.

• Orbital fat - The degree of prolapsed orbital fat within the upper eyelids should be noted, and can be rated on the scale of 1+ to 4+. Excessive fullness of the lateral eyelid may be secondary to lacrimal gland prolapse that should be resuspended under the supraorbital rim during surgery (Figure- 4).

2. Surgical technique (Figure 5-a-d)

• A surgical marking pen and calipers are used to demarcate crescentic upper eyelid blepharoplasty incisions with the lid crease placed at the desired height based on natural location, symmetry, gender, and ethnicity. Approximately 18-20 mm of skin should be retained in both upper eyelids.

• Avoid incising medial to the upper punctum and lateral into the thicker temporal skin to avoid webbing of the skin.

• The pinch technique is used to confirm the adequacy of retained skin. Eversion of the lashes or lid margin, or excessive vertical tightening of the remaining skin suggests overly aggressive skin excision.

• Brow ptosis may be measured by lifting the lateral brow up to the desired height above the superior orbital rim and measuring its excursion with a ruler. Brow ptosis should be corrected simultaneously in order to avoid the appearance postoperatively of residual skin redundancy laterally.

• Local infiltrative anesthesia using 2% lidocaine with 1:100,000 units epinephrine mixed in equal parts with 0.5% bupivicaine is given to both upper lids.

• A 15 blade, CO2 laser, or monopolar cautery tip, is used to incise the eyelid skin along the previously placed markings.

• The skin and orbicularis muscle are removed as single flaps generally, although a skin flap alone could be removed if there is preoperative concern for poor eyelid closure function. Such patients may include those with a history of Bell's palsy, dry eyes, decreased tear film or exposure signs, or elderly patients with no frank lagophthalmos but slow spontaneous blinking. Another benefit of preserving the orbicularis muscle is that volume is maintained in the sulcus to optimize the youthful appearance.

• The orbital septum is incised if there is significant fat proplape preoperatively to isolate the underlying preaponeurotic fat pads. The fat should be conservatively removed over a hemostat clamp or forceps, and the fat stump cauterized before release, in order to avoid orbital hemorrhage.

• Avoid aggressively pulling or cauterization of the medial fat, as permanent injury to the trochlea and superior oblique tendon may occur.

• Fullness and prolapse laterally suggests lacrimal gland dystopia as there is no upper eyelid lateral fat pad anatomically. Failure to recognize this during surgery may lead to inadvertent lacrimal lobectomy and possible postoperative hemorrhage or chronic dry eye.

• The upper eyelid incisions are closed with 6-0 or 7-0
vicryl buried interrupted suture to close the orbicularis muscle layer, followed by 6-0 nylon suture to close the skin edges in a running or interrupted fashion. Closure is performed from medial to lateral, as any dog-ear deformity, if present, is better addressed and camouflaged laterally (Figure-6).

**Pearls**

- Make the skin markings with a fine tip marker prior to local anesthetic infiltration as this may distort the eyelid tissues.
- Coexisting findings, such as levator dehiscence, lacrimal gland prolapse, and brow ptosis should be addressed concurrently to achieve the best results.
- Avoid excessive removal of the orbital fat pads to prevent a sunken superior sulcus appearance.
- Upper eyelid skin should measure approximately 18-20mm to avoid postoperative lagophthalmos.
- Conservative skin excision should be performed medially to avoid webbing and medial ectropion.
- If the skin pinch leads to vertical striae, a tight upper lid, and/or eversion of the eyelid margin and lashes, the skin excision is too aggressive.
- The medial marking should not extend more than 1-2mm past the upper puncta to avoid webbing.
- The lateral markings should not extend into the thicker temporal skin or below the level of the lateral raphe.
- Aggressive skin and fat removal in men may lead to a hollow, feminized appearance. Retaining slightly more skin will create a more natural skin fold and fullness that is typical for men.
- Avoid aggressively pulling or cautery of the medial fat, as permanent injury to the trochlea and superior oblique tendon may occur.
- Closure of the orbital septum is contraindicated.

**B. Lower lid blepharoplasty**

**Preoperative evaluation (Figure-2)**

In general, the goal of lower lid blepharoplasty is to eliminate redundant skin, smooth the underlying musculature, tighten supporting structures, and sculpt or redrape excess orbital to blend the transition over the orbital rim between the lower eyelid and the cheek. In addition to the examination discussed in the upper lid blepharoplasty section, other important findings to evaluate include:

- Skin- Skin turgor, texture, and tone, as well as dyschromias should also be considered. The presence of festoons or chronic eyelid edema may indicate chronic inflammation, and may persist postoperatively (Figure-7). The amount of redundant skin can be evaluated by asking the patient to look upward, or to open his or her mouth. Dyschromias can often be addressed with laser procedures or topical bleaching agents.

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• Fat prolapse or lack of fat- The location and amount of prominence of each of the three lower lid fat pads is important to note in the upright position. Herniated orbital fat can also be noted by looking for areas of fullness in the lower eyelid that become more prominent as the patient looks up (Figure-9).

2. Surgical technique
Lower lid blepharoplasty includes several approaches:
• Transcutaneous technique
• Transconjunctival technique
• Combined technique
• Adjunctive midface lifting

Transcutaneous technique
Markings are made preoperatively in the upright position. A subciliary incision is made 1-2mm below the lash line and gently curved laterally for 1cm into a crow's feet crease. Skin with or without the orbicularis oculi muscle are dissected off the orbital septum. The orbital septum is button-holed over the prominent fat compartments and the fat isolated and completely freed circumferentially from the multilamellar orbital septum, cauterized at the base, and then excised with scissors or monopolar cautery. Fat pedicles can also be redraped, particularly medially and centrally, to add volume to significant tear trough sulci.If any degree of lower lid laxity was present preoperatively, a lateral canthopexy or tarsal strip procedure should be performed. The lateral skin excision often angles downwards and should blend into a natural relaxed skin tension crease. The subciliary incision is closed with 6-0 nylon, or prolene running or subcuticular suture.

Transconjunctival technique (Figure10 a-d)

• This technique is ideal for patients with fat prominence but no significant skin redundancy. Alternatively, the transcutaneous incision and skin removal can be performed as above, and the fat debulking performed via a posterior approach.

• A Desmarres retractor is used to evert the lower eyelid and a Jaeger plate is used to protect the globe and provide gentle posterior pressure in order to prolapse the fat pads anteriorly toward the conjunctiva.

• Each of the three lower lid fat pads, the medial, central, and lateral, are accessed through a minimal 3-4 mm button-hole conjunctival incision over each fat pad. Through each button-hole, the fat pad can be drawn forward with minimal tension.

• The fat pads can be clamped, if preferred, prior to excision using monopolar or radiofrequency cautery. The stump of the resected fat should be controlled and cauterized prior to release in order to avoid orbital hemorrhage.

• Fat should never be debulked posterior to the anterior orbital rim.

• The lateral fat is often undercorrected and therefore should be carefully addressed.

• When debulking the central fat pad, care is taken to neither instrument nor cauterize the inferior oblique muscle.

Combined technique
As mentioned previously, orbital fat is addressed through the transconjunctival approach while the skin-orbicularis complex is addressed transcutaneously. This theoretically avoids dissection in the plane of the orbital septum and thus may lead to less postoperative eyelid malposition (Figure -11)

3. Pearls
• It is important to be conservative when removing orbital fat. Draping or repositioning fat over the inferior orbital rim is often preferable to significant fat excision.

• Meticulous hemostasis is critical. It is a good practice to follow the clamp, cut, and cautery technique.

• Excessive skin removal laterally can lead to distortion, rounding, and ejection of the lateral canthus, known
as “lateral canthal syndrome”.

- Always recreate a sharp lateral canthal angle to avoid rounding or web formation laterally.
- In the transconjunctival technique, the incisions should be placed 4-5 mm below the inferior tarsus, which corresponds to the inferior point of fusion between the lower lid retractors and the orbital septum.
- When possible, the transconjunctival approach is preferred as it leads to less lower lid retraction, scleral show, and postoperative ectropion. If excess skin is present, this can be removed through the subciliary approach or via a more conservative skin pinch that removes skin without muscle.
- The inferior oblique muscle between the medial and central fat pads should always be left undisturbed.

**Complications**

1. Retrobulbar hemorrhage with or without visual loss
2. Infection
3. Asymmetry
4. Incomplete removal of redundant tissue, need for additional procedure
5. Hypertrophic scarring or prolonged erythema of scar
6. Aggressive skin resection leading to lagophthalmos (transient or permanent), cicatricial ectropion of the upper lid, web formation (incision placed too far medial)
7. Blepharoptosis (levator muscle injury)
8. Diplopia (superior or inferior oblique muscle damage)
9. Hollowed out appearance (overly aggressive fat removal)
10. Milia, suture granulomas

**Postoperative Care**

- Avoid strenuous activity, bending, or lifting heavy weights for 1 week
- Sleep with two pillows (approximately 45 degree elevation) for 1 week
- Start cold compresses immediately after surgery. Cold compresses should be applied continuously as much as possible while awake for the first 48 hours
- Clean the operated area gently with warm water and a clean cotton tip applicator if needed
- Avoid rubbing the eyelid area, forehead, or face to prevent dehiscence of the incisions
- Allow the shower water to hit the back of the head, rather than the face for 1 week
- Use topical lubricating eye drops frequently to prevent dry eye issues
- Avoid aspirin or ibuprofen for pain relief in the first several days after surgery
- Bruising typically resolves by 2 weeks
- May resume normal activities after one week
- Avoid makeup around the eyelids for at least one week; after that, light makeup can be applied, but should be carefully removed with a gentle cleanser to avoid incision dehiscence
- Contact lenses may be worn at 1 to 2 weeks after surgery, taking care to avoid pulling on the surgical eyelids during placement or removal of the lenses

**Conclusion**

Upper and lower eyelid blepharoplasty procedures can be very gratifying for both the surgeon and patient. It is important to identify the expectations of the patient preoperatively, and determine preexisting conditions that may lead to postoperative complications. A meticulous and individualized surgical approach, combined with appropriate postoperative care can yield excellent results.

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Introduction
Anophthalmia is derived from the Greek word anophthalmos1, which means the absence of an eye. Congenital anophthalmia is a relatively rare congenital eye anomaly in which the eye is absent as a result of a defective development of the primary optic vesicle.2 True absence of ocular and orbital contents is rare and more often a phenotypic range between microphthalmos and anophthalmos is seen. Microphthalmia is commonly associated with coloboma or an orbitopalpebral cyst (figure 1).

Figure 1: 1a. Anophthalmia with orbitopalpebral cyst
1b. Appearance of the cyst on opening eyelid

Coloboma is defined as a defect in any tissue(s) consistent with failure of closure of the fetal fissure. These 3 ocular anomalies are collectively termed as Microphthalmia anophthalmia coloboma (MAC) complex.3 Both anophthalmia and microphthalmia may be unilateral or bilateral (figure 2), and may be associated with systemic abnormalities such as the CHARGE syndrome.4

Figure 2: 2a Unilateral and 2b bilateral congenital clinical anophthalmos, 2c unilateral and 2d bilateral microphthalmos

In the case of unilateral anophthalmia or microphthalmia, there may be developmental anomalies of the other eye, including coloboma, lens, and optic nerve abnormalities.5 This cosmetic disfigurement and disability is less well understood even by specialists and its management needs timely intervention in an orderly manner.6-8 Missed window period leads to permanent unmanageable socket contracture and mid-facial hypoplasia (figure 3).

Figure 3: 3a. CT showing orbital and midfacial hypoplasia; 3b soft tissue hypoplasia; 3c small prosthetic eye on right side

This review addresses the basis for classification of MAC, the known etiologies, clinical assessment and management options in congenital anophthalmia.

Basis for Classification
The eye derives from three embryological germ layers: the neuroectoderm which gives rise to the optic vesicle, the neural crest cells which migrate to the anterior chamber of the developing eye, and the ectoderm from which the lens placode is derived. Neuroectodermal and mesodermal cells participate in the closure of the optic fissure.9 This multiple embryological derivation explains the variability of the phenotype and the aetiological heterogeneity of congenital ocular defects.

The term anophthalmia is used when there is no visible ocular remnant. However, clinically it might be difficult to discern if ocular structures are present. Ultrasound B Scan and Computed tomography scan will often show rudimentary eye tissue, extraocular muscles and the optic nerve. Hence the term congenital clinical anophthalmos is a more appropriate (figure 4).

Figure 4: 4a Right congenital clinical anophthalmos with orbitopalpebral cyst and left microcornea; 4b axial Ct showing optic nerve and extraocular muscles, 4c sagittal CT showing the orbitopalpebral cyst and 4d coronal CT showing the lacrimal gland. All 3 scans show a microphthalmic eye on right side.
Microphthalmos stands for “an abnormally small eye” which is less than 2 standard deviation for that age group. This would equate to axial length less than 16 mms at birth and 19 mms at 12 months of age and a corneal diameter at birth of 10 mm or less.3 Severe microphthalmia refers to a globe that is severely reduced in size, with a corneal diameter <4 mm and an axial length of less than 10 mm at birth or less than 12 mm after one year of age. The size of the globe can be measured ultrasonographically, by computerised tomographic scans (CT scans), or by magnetic resonance imaging (MRI). Fetal ultrasonograms of the eye can be obtained both transabdominally10 and by a transvaginal procedure in the transverse section.11 Normal values for fetal axial length12 and for the growth of the fetal orbit10 are available. In hereditary cases, microphthalmos may be diagnosed prenatally by expert ultrasonography.11

The growth of the eye is fast during the first three years; the adult size is reached at around 13 years, when the outer sagittal diameter is 24mm, the inner sagittal 22mm, the anteroposterior 25mm and the transverse 24 mm.9 Without orbital imaging studies, severe microphthalmia can be mistaken for anophthalmia; thus, the term “congenital clinical anophthalmia” is often interchangeably used for severe microphthalmia. Thus in essence congenital clinical anophthalmos and microphthalmos are diseases along the same line with varying phenotypic features. Congenital cystic eye is a malformation resulting from failure of invagination of the optic vesicle and should therefore be classified as a case of microphthalmos.

Classification of MICROPHTHALMOS
Considering the clinical heterogeneity microphthalmos can be classified based on phenotype and on etiology.

The Phenotypic classification5 allows the clinician to describe the anomalies in a systematic way. The phenotype does not predict the etiology but a systematic description of ocular and systemic anomalies improves syndrome identification. There are two major classes, total and partial microphthalmos, and a sub classification which follows the embryology of the anomalies (Table 1).

Table 1: Phenotypic classification of microphthalmos

<table>
<thead>
<tr>
<th>I. Total microphthalmos</th>
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<tbody>
<tr>
<td>A. Congenital cystic eye</td>
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<tr>
<td>B. Apparent anophthalmos</td>
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<tr>
<td>C. Simple microphthalmos</td>
</tr>
<tr>
<td>D. Microphthalmos with intraocular malformations</td>
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<tr>
<td>i) congenital cataract</td>
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<tr>
<td>ii) anterior segment malformations</td>
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<tr>
<td>iii) coloboma of the uvea and posterior colobomatous cyst</td>
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</table>

| E. Microphthalmos with multiple ocular malformations |

<table>
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<th>II. Partial microphthalmos</th>
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</thead>
<tbody>
<tr>
<td>A. Anterior segment microphthalmos</td>
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<tr>
<td>B. Posterior segment microphthalmos</td>
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(I) Total microphthalmos is where both anterior and posterior segments are foreshortened. The most severe type of microphthalmos is (A) Congenital cystic eye 14 which occurs due to failure of invagination of the optic vesicle. Clinical examination shows complete absence of eye tissue with rapid expansion of the cyst. Treatment consists of excision of the cyst and placement of a conformer followed by subsequent orbital expansion. (B) Anophthalmos is the clinical term for extreme microphthalmos where a rudimentary eye may be found by serial histopathological sections. (C) Simple microphthalmos without other malformations is called nanophthalmos. The eye is deeply set in the orbit, refraction is high hypermetropic, and the cornea is small. Both the anterior and posterior segments of the eye are foreshortened, the relative lens volume is increased, and there is sclerochoroidal thickening.15,16 (D) Microphthalmos with intraocular malformations is common. Microphthalmos is associated with congenital cataract as in Hallerman-Streiff syndrome and Nance-Horan syndrome.17 Microphthalmos is also associated with anterior segment dysgenesis and sclerocornea.18 Iris and retinochoroidal coloboma are common with microphthalmos. Sometimes the posterior coloboma becomes staphylomatous and forms a cystic lesion called microphthalmos with posterior colobomatous cyst.19

Figure 5: 5a Right congenital clinical anophthalmos with orbitopalpebral cyst and left microphthalmos, 5b right b-scan showing a posterior colobomatous cyst

(E) Microphthalmos with other malformations, that is, malformations deriving from different embryological germ layers are also found; examples are Delleman syndrome (the oculo cerebro cutaneous syndrome)20 and the osteoporosis-pseudoglioma syndrome.21

(II) In partial microphthalmos, the anterior chamber and the cornea may be of normal size even if the posterior segment is small.22 Posterior segment microphthalmos is seen, for instance, in patients with high hypermetropia.23 In some patients the anterior chamber is small while the posterior compartment is of normal size24 or larger than normal.25
The etiological classification consists of three classes: (1) Genetic (monogenic and chromosomal). Chromosomal duplications, deletions and translocations are implicated in context of genetic derangements. SOX2 has been studied extensively in literature with reported mutation prevalence of 4.6 to 17.6%. Most of genetic disturbances are de novo and loss of SOX2 gene is seen in bilateral cases. Polygenic inheritance with low penetrance is postulated to be involved in unilateral cases. (2) Prenatally acquired (teratological agents and intrauterine deformations) The term prenatally acquired indicates that the embryo or fetus had a normal genetic background but that the intrauterine environment resulted in disruptions or deformation. Microphthalmos has been described as a consequence of maternal ingestion of teratogens, among them alcohol, thalidomide and isoretinoic acid are known. Maternal diabetes and maternal rubella are the known diseases which may delay the normal growth of the eye. (3) Associations. If an underlying genetic cause is suspected but not known, a condition may be referred to as an “association”. By definition, an association shows that the collection of signs and symptoms occurs in combination more frequently than would be likely by chance alone. Microphthalmos and coloboma presenting as associations make up for a small number of cases. Microphthalmos is associated with CHARGE association and the VATER association.

Epidemiology
Incidence of anophthalmia is reported to be 0.2-0.4 per 10000 births and microphthalmia around 1.5-19/10,000 births. Various studies carried out in India have shown prevalence in the range of 30 to 57% among blind school population. True incidence depicting epidemiological studies are still lacking from the Indian subcontinent.

Clinical Evaluation
Congenital clinical anophthalmia can be unilateral or bilateral. Microphthalmia has microorbitism and microblepharon. Every child should be examined under anesthesia, both for diagnostic as well as therapeutic purposes. It involves measurement of horizontal palpebral fissure, superior and inferior fornical depth, socket inspection, assessment for orbitopalpebral cyst and detailed ocular examination for any visual potential as well as fellow eye evaluation in unilateral cases. Associated anomalies like coloboma, sclerocornea, glaucoma have been found in fellow eye. Ultrasonography of eye and orbit is an easy to do and readily available imaging tool for assessment of globe. Orbital imaging like computed tomography is essential to look for associated orbitopalpebral cyst and also if planning orbital expander for volumetric assessment. Visual assessment with the help of electrodiagnostic studies should be done wherever potential vision is expected. A flash visual evoked potential (VEP) is performed in cases of apparent anophthalmia or severe microphthalmia; a pattern VEP for level of acuity and any optic nerve function, and an electroretinogram for retinal dysfunction.

Systemic evaluation by pediatrician for associated craniofacial anomalies like cleft lip, cleft palate, cardiac and genitourinary malformations should be carried out. Craniofacial anomalies have been reported to be the most common in MAC complex. Rehabilitation services to educate parents and children to cope up with the disability associated issues should be included in initial treatment.

Investigations
Pediatric evaluation includes MRI brain, renal ultrasonography to look for midline abnormalities and renal anomalies. MRI is preferable to CT scanning since there is higher resolution of the structures of interest and no radiation exposure. Screening for TORCH infections should be done whenever suspected, as congenital rubella has been found to have an association with microphthalmia with cataract. Examination of other family members and siblings, pedigree charting is required to look for hereditary pattern. Consanguinity was seen in 50% of bilateral anophthalmia cases in our centre (unpublished data). Genetics assessment will include chromosome analysis and genomic sequencing.

Treatment
Goals of treatment in MAC complex remain – management of soft tissue hypoplasia and management of orbital hypoplasia. Graduated socket expanders followed by orbital expansion is the way to manage. Normal orbital and facial development is affected by ocular volume. The normal infant eye is approximately 70% of its adult size and grows most rapidly in the first 12 months of age. At 3 months of age, the face is only approximately 40% the size of the adult face, and by 2 years of age, the face reaches 70% of the adult size. By 5.5 years of age, the pediatric face is approximately 90% of the adult dimensions. Reduction in orbital volume in turn leads to maxillary hypoplasia and expansion of ethmoidal and maxillary sinuses. Timely intervention is a must for good outcome to prevent permanent bony contracture. Various modalities for treatment are discussed below.

Conformers
These are made up of clear polymethmethacrylate (PMMA) material which are fitted under general anesthesia or topical anesthesia for a cooperative child. Conformers help in expansion of the lid dimensions, both horizontally and vertically, expansion of the conjunctival cul-de-sac and formation of normal adequate fornices. Every 3 weeks the conformer is exchanged for a larger one. (figure 6)
This expansion is carried out till the horizontal palpebral fissure equalizes with the contralateral normal eye or is within 2 millimeters of it. For bilateral cases the expansion is aimed towards the age matched normal values. Hemispherical socket expanders can also be used for achieving surface expansion. Hemispherical socket expanders are made up of copolymers of polymethyl methacrylate and polyvinyl pyrrolidone which can be moulded in any shape. They expand after imbibing tears and stretches the fornices. However PMMA conformers are cheaper and can be customized too. Following fornical expansion one should proceed for orbital expansion.

Expanders

1) Spherical implants
Solid implants like silicon spheres or PMMA are static implants of increasing size can be implanted for orbital expansion. It is increase in orbital volume, which maintains a constant pressure and results in bony expansion. Multiple surgeries and general anesthesia are required to replace a larger implant every 6 months.

2) Dermis fat graft
Study of 7 patients with primary anophthalmia with dermis fat graft43,44 has shown promising results but it lacks longer follow up and numbers to infer its usage. It can be used as second modality for socket expansion in these cases. In pediatric patients the fat component of the dermis fat graft has been known to expand with the growth of the orbit and this may be advantageous like a self-expanding implant.45

3) Inflatable expanders
In cases of anophthalmia and severe microphthalmia when aggressive orbital expansion with spherical implants cannot achieve the desired orbital bony growth, tissue expanders have been used to stimulate orbital growth. Inflatable tissue expanders are placed intracorally in the orbit via bicoronal approach or lateral orbitotomy. Gradually, over time, saline is injected into a port in the parietal region that is connected to the tissue expander. Grossman et al.46 reported good orbital expansion using a gradual saline expander and recommended placement of the expander during the first year of life. Furthermore, they recommended waiting 3 months after expander placement before inflation, and a target inflation period over 20 to 36 weeks to decrease the risk of expander extrusion. However it is difficult to control the direction of expansion and distribute equal amount of pressure in every direction. These expanders might displace the conformer if they balloon out anteriorly and may compromise surface expansion.

4) Self expanding implants
Hydrogel implants47-48 are spheres, made from either poly(2-hydroxyethyl methacrylate) or copolymers of polymethyl methacrylate and polyvinyl pyrrolidone, can increase up to 30 times their original volume. (figure 7)

Disadvantage with them is the loss of orbital expansile forces once it reaches its water equilibrium which further requires resurgery. Long term results of use of hydrogel implants are not available in literature. Complications like brittleness of hydrogel while removing makes resurgery difficult. Amount of orbital pressure required to achieve an adequate expansion is not known for human socket. However study by Reedy et al49 determined the optimal orbital pressure of 20 mm Hg in Yorkshire piglets resulted in near normal orbital volumes, that less than normal pressures resulted in reduced orbital growth, and that greater than normal pressures resulted in larger orbital volumes. This can’t be extrapolated to human socket.

An ideal expander would be biocompatible, easy to place and would gradually enlarge over a relatively short time, minimal complications in terms of extrusion, exposure; and would require minimal manipulation or revision. No implant so far matches all above stated requirements.

5) Orbital tissue expander
The Orbital tissue expander (OTE) designed by David Tse has been introduced in 2007.50 It consists of Inflatable silicone balloon attached to cylindrical titanium injection chamber held in place by fixation plate anchored to lateral orbital wall. (Figure 8).
The device is implanted into the orbit via a lateral orbitotomy. (Figure 9)

Bone plate prevents extrusion and facilitates self-centering of the device in the orbit. A 30 gauge disposable hypodermic needle connected to a 1cc disposable syringe filled with sterile saline is inserted into the OTE through an injection port at 3-6 monthly intervals. The injection track seals upon removal of the needle. Five cc volume is equivalent to an anterior-posterior diameter of 24mm and a 22mm equatorial diameter. Simple inflation and deflation eliminates the need for successive surgeries and the implantation of larger devices, hence requires minimal follow-up. Preliminary results in 9 patients showed average increase of 5cc in orbital volume after mean 18.89 months of follow up. Complications encountered were inadvertent device puncture in two and one implant failure. This device looks promising but issues like bending of T plate, end point of orbital expansion, puncture of silicone sphere during inflation need further evaluation.

Table 2: salient features of the orbital expanders

<table>
<thead>
<tr>
<th>Hard Spherical Implants</th>
<th>Dermis fat graft</th>
<th>Inflatable balloon tissue expanders</th>
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<tbody>
<tr>
<td>• Repeated conjunctival trauma and general anaesthesia</td>
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Summary

Children with MAC complex require a multidisciplinary team approach by ophthalmologist, paediatrician, geneticist and expert oculist for socket expansion and acceptable cosmetic results. Treatment should begin immediately after birth and is aimed at socket expansion with graduated conformers. Achieving and maintaining orbital expansion seems promising with the introduction of a novel orbital tissue expander than with orbital osmotic hydrogel expanders. Management is time consuming and challenging but early and timely intervention achieves good results.

References:

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Unilateral Proptosis Due to Nasal Polyposis With Allergic Aspergillus Sinusitis

Dr Sree Kumar Vaggu; Dr Preethi B

Introduction
• Allergic aspergillus sinusitis or Allergic Fungal Sinusitis (AFS) with nasal polyposis is a rare cause of proptosis.
• Nasal polyps are benign lesions arising from the mucosa of the nasal sinuses or the nasal cavity.
• Nasal polyps are common, affecting up to 4% of the population. Known to have associations with allergy, asthma, infection, cystic fibrosis and aspirin sensitivity.
• They present with nasal obstruction, anosmia, rhinorrhea, post nasal drip, and less commonly facial pain.
• Here is a rare presentation with non axial proptosis, nasal polyposis with allergic fungal sinusitis associated with orbital bony erosion and optic nerve compression.

Case Report
• A 56 year old female referred from ENT department for ophthalmic evaluation presented with complaint of prominence of the left eye with epiphora
• History of recurrent sinusitis in the past. Known diabetes for 5 years
• There was painless left sided proptosis, epiphora and nasal obstruction

Ophthalmic examination:
• BCVA: RE 6/6, LE 6/6
• IOP OU 20 mm Hg
• OS 15 degrees exotropia with non axial proptosis
• Exophthalmometry: OD - 20mm; OS - 24mm
• OU anterior segment within normal limits
• Pupils – OD brisk, OS RAPD grade-1
• OU Extraocular mobility full range
• Fundus examination : OD within normal limits. OS pallor of the inferior Neuroretinal rim +. Peripheral retina normal.

Perimetry HVF 30-2
• OD unreliable due to high fixation loses
• OS superior hemifield defect

Nasal examination, anterior rhinoscopy:
• Single nasal polyp in the left nasal cavity

Investigations:
• Blood counts normal
• Bleeding Time 2min 2 sec
• Clotting Time 5min 9sec
• Absolute eosinophil count – 510
• ESR – 55mm/1hr, 99 mm/2hr
• RBS-280mg/dl
• Renal parameters normal
• Liver Function Tests Normal
• Urine exam normal
• HIV 1 & 2 Non reactive
• HbsAg Non reactive

CT Orbits
• Large lobulated iso to hyperdense lesion in the left ethmoid and sphenoid sinuses, bilateral maxillary and frontal sinuses
• Erosion of the middle & inferior turbinates, widening of maxillary ostium, erosion of medial & inferior walls of left orbit, erosion of cribiform plate.
• Erosion of left lateral wall of sphenoid sinus
• Extension of lesion into the left orbit in the extraconal compartment causing lateral displacement of medial rectus and optic nerve compression
CT PNS

- Polypoidal mucosal thickening with areas of hyperdensity noted in bilateral frontal & maxillary sinuses, left ethmoid & sphenoid sinuses, left nasal cavity & post nasal space.
- Erosion of bilateral cribriform plates, left lamina papyracea with extension of soft tissue into left orbit.
- Features suggestive of sinunasal polyposis with superadded fungal infection, bony erosions and left intraorbital extension.

Discussion

- Allergic fungal sinusitis (AFS) is a non-invasive pansinusitis that occurs in young immunocompetent individuals, with a strong history of atopy and elevated levels of total immunoglobulin (Ig)E and peripheral eosinophilia.
- The diagnosis is based, by analogy with the findings in broncho-pulmonary aspergillosis, on the presence of allergic mucin within the sinus. Macroscopically, the secretions are thick, viscous, and green and microscopically, the allergic mucin contains eosinophilic polynuclear cells, charcot-leyden crystals, and scattered mycelial filaments without tissue invasion.
- CT scan shows heterogeneous opacities or calcifications. The presence of bony erosion of the skull base and orbit in AFS has been well documented in the literature. Patients with AFS have intractable sinusitis that fails to respond to repeated courses of antibiotics. Ophthalmologically the close proximity of the orbit to the paranasal sinuses may lead to the spread of the diseases of the sinuses into the orbital space.
- Therefore proptosis can be a presenting sign of the sinus disease which requires thorough evaluation by the ophthalmologist.
- Treatment of AFS consists of surgical debulking with endoscopic sinus surgery and use of systemic steroids or immunotherapy. Surgery is necessary to remove the allergic mucin, polyps and other debris and to re-establish sinus ventilation.
- To prevent vision loss early surgical debulking is indicated to avoid compression of optic nerve. After surgical and medical intervention improvement in optic nerve function has been reported.
- Proposed theories of vision loss include direct or indirect compression of optic nerve; orbital inflammatory changes causing optic neuritis; venous congestion of optic nerve due to thrombophlebitis; and retinal artery occlusion due to increase in orbital pressure.

Summary and Conclusion

- A 56 year old female patient referred from ENT department with fungal nasal sinusitis and polyposis causing proptosis for ophthalmological evaluation.
- EXAMINATION revealed OS non axial proptosis with XT 15 degrees, RAPD, pallor of inferior NRR with corresponding superior hemifield defect and radiological evidence of optic nerve compression.
- Allergic fungal rhinosinusitis is an uncommon cause of optic neuropathy.
- Early cases of optic neuropathy are difficult to diagnose because the typical signs may not be present. The symptoms develop gradually as further damage to the...
nerve occurs. High level of clinical suspicion is required in all cases of proptosis with normal vision. Early treatment can prevent permanent damage to the vision.

- This case highlights the need for ophthalmologists to be aware of nasal polyp with secondary aspergillus infection as a cause of proptosis with compression of the optic nerve which needs appropriate measures to safeguard the vision.

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Hands on course on Intravitreal Injections, TOC 2015
A Rare Case Report of Unilateral Optic Nerve Sheath Meningioma

Dr Manogna A, Dr K Vijay Kumar

Introduction
Optic nerve sheath meningiomas (ONSM) are rare, slow-growing benign tumors, which approximately constitute 2% of all orbital tumors of the anterior visual pathway and 1-2% of all meningiomas. Middle-aged females are primarily affected. Primary orbital meningiomas originate from the optic nerve sheath or from the extension of an intracranial meningioma into the orbit. ONSM left untreated always leads to progressive vision loss and color blindness which is often associated with optic atrophy. Most of the time management is conservative in long-standing ONSM, as there is not much of visual improvement after the surgery. Here we report a case of 55 year female patient with unilateral optic nerve sheath meningioma.

Case Report
55 yr old female patient presented to ophthalmology OPD with complaints of protrusion of right eye and gradual painless loss of vision in right eye for 5 years with mild intermittent headache. She did not give any history of eye pain, eye trauma or projectile vomiting. She was not a known diabetic or hypertensive.

Patient underwent detailed examination, ocular, medical and neurological from which the following observations were made.

Right eye had exotropia. Best corrected visual acuity was perception of light. Axial proptosis of about 26mm was present. All extra-ocular movements were restricted. Pupil reaction was sluggish to direct light. Anterior segment was normal. Fundus showed optic atrophy with retino-choroidal collaterals.

Left eye anterior segment and fundus were normal and visual acuity was 6/6.

General investigations including CBP, RBS, CHEST X-RAY, T3, T4, TSH were normal

Contrast enhanced CT (CECT) orbits: revealed a hyperdense mass lesion measuring 45 X 32mm arising from right optic nerve sheath with areas of calcification. It was seen extending intracranially along the optic nerve sheath through optic foramen into the right frontal horn, suprasellar & parasellar regions and had a mass effect on frontal lobe. Post contrast scan showed intense, almost homogenous pattern of enhancement. Findings were suggestive of right optic nerve sheath meningioma with intracranial spread.

Discussion
ONSM are rare benign tumors of the optic nerve. 60-70% of cases occur in middle age females, and is more common in older adults. It is also seen in children, but this is rare. The tumors grow from cells that surround the optic nerve, and as the tumor grows, it compresses the optic nerve. This causes loss of vision in the affected eye. It is mostly unilateral. Rarely, it may affect both eyes at the same time.

It is typically a slow growing tumor and has never been reported to cause death. However, there is concern that the tumor can grow into the brain and cause other types of neurological damage. In some patients, the tumor grows so slowly that the treatment is not necessary.
Conclusion

Often meningiomas show homogenous and well-defined mass on CECT with areas of calcification. CECT is the procedure of choice for diagnosis of ONSM.6

The findings in optic nerve sheath meningioma in this case are quite typical. The benign non-invading, well-defined growth pattern, the clinical manifestation strongly suggests the possibility of meningioma

References


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Orbital metastasis - A Case Report

Dr J Arpitha Rao; Dr C Madhavi

Purpose
To report a rare case of orbital metastases from a nasopharyngeal carcinoma.

Case Description
A 50 year old heavy smoker male presented to our Ophthalmology Department in February, 2015 with the complaints of bulging of right eyeball and blurring of vision in right eye, which he had noticed one month prior to the presentation. There was a history of nasopharyngeal carcinoma with metastases to 3rd and 4th cervical spine one year ago. He had received chemotherapy and radiotherapy at the time of diagnosis. The last radiotherapy session was about 5 months prior to presentation.

Examination revealed OD severe proptosis, dystopia and limitation of eye movement in all directions. Visual acuity was OD hand movements close to face with no improvement and OS 20/20. Hertel exophthalmometer readings were OD 25 and OS 18 mm with an inferior displacement of OD. Intraocular pressure and fundus was normal in OU.

An orbital CT scan and MRI showed large soft mass 6(CC)*4.5(TR)*5(AP) cm in the right anterior temporal region causing proptosis (Figures 1& 2 respectively).

Conclusions:
Orbital metastases are a rare manifestation of systemic malignancies; such orbital metastasis account for only 1% - 13% of all orbital tumors. Although nasopharyngeal carcinoma rarely metastases to the orbit, it should be considered in every rapidly progressing proptosis, especially in elderly.

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Dear Friends and Colleagues

As the Secretary of WOS Telangana state, I would like to brief you about what is WOS and its goals.

BSE recently published this data. In India, in top 100 companies there are only 5% women on board and in top 240 companies only 11% have female CEOs.

Existence of Women’s Organization, availability of support systems for women - External & Internal, Networking, feminist leadership and favourable policy climate will be important facilitating factors for empowerment of women.

With this mission the Women Ophthalmologists Society (WOS) was formed in February 2015 at AIOC 15, Delhi.

**WOS - Aims and objectives**

- To improve the professional working environment as well as professional knowledge and skills for women
- To create a forum where women can put forward and discuss their problems and their solutions, which they had been facing till now in isolation
- To develop training surgical programs for women to encourage more women into the world of surgical ophthalmology
- To encourage more women to come in lead positions in practices, leadership positions and chairs in academic events
- To develop a mentor-mentee program for encouraging junior women by making senior women as their mentors
- To give recognition to women who have been doing good work by giving awards and felicitations, this in turn would encourage other women to follow these role models
- To act as an intermediary forum for helping in placements of women ophthalmologists as per the requirements of recruiting institutions by creating a forum where they can access these opportunities

**WOS Telangana state chapter**

*State Secretary:* Dr Alpa Atul Poorabia

*State representatives:* Dr Somasheila Murthy, Dr Sirisha Senthil, Dr N Sreelakshmi, Dr Anita Kamarthy

*Male Advisers of WOS Telangana:* Dr G Chandra Shekhar Rao; Dr Kasu Prasad Reddy

On 8th November 2015, WOS Telangana state chapter has successfully organised first state meeting.

The time is long overdue to encourage more women to dream the possible dream and encourage more men to support women in the workforce and in the home.

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Indications: Improvement and maintenance of visual acuity and function and for reduction of vascular leakage and retinal edema, in patients with neovascular age-related macular degeneration (AMD), the treatment of visual impairment due to diabetic macular edema (DME), the treatment of macular edema following retinal vein occlusion (RVO) in the treatment of visual impairment due to choroidal neovascularization (CNV) secondary to pathologic myopia (PM).

Dosage: The recommended dose is 0.5 mg (0.05 mL) given as a single intravitreal injection. The interval between two doses injected into the same eye should not be shorter than 1 month. Wors AMD, DME, RVO, PM. Treatment is initiated with one injection per month until maximum visual acuity is achieved and/or there are no signs of disease activity. Thereafter, monitoring and treatment intervals should be determined by the physician and should be based on disease activity as assessed by visual acuity and/or anatomic parameters.

Monitor for disease activity may include clinical examination, functional testing or imaging techniques (e.g., optical coherence tomography or fluorescein angiography). While applying the treat-and-extend regimen, the treatment interval should be extended by two weeks at a time for well AMD and central RVO, or by one month at a time for DME and branch RVO. Accentrix and laser photocoagulation in DME or branch RVO. Accentrix has been used concomitantly with laser photocoagulation in clinical studies. When given on the same day, Accentrix should be administered at least 30 minutes after laser photocoagulation. Accentrix can be administered in patients who have received previous laser photocoagulation. Accentrix must be administered by a qualified ophthalmologist using aseptic techniques. Broad-spectrum topical microbiolide and anesthetic should be administered prior to the injection. Not recommended in children and adolescents.

Contraindications: Hypersensitivity to ranibizumab or to any of the excipients, patients with active or suspected ocular or pericellular infections, patients with active intraocular inflammation.

Warnings/Precautions: Intravitreal injections have been associated with endophthalmitis, intraocular inflammation, rhegmatogenous retinal detachment, retinal tear and idiopathic traumatic cataract. Therefore proper aseptic injection techniques must be used. Patients should be monitored during the week following the injection to permit early treatment if an infection occurs. Transient increases in intraocular pressure (IOP) have been seen within 60 minutes of injection of Accentrix. Sustained IOP increases have also been reported. Intravenous pressure and the perfusion of the optic nerve head must be monitored and managed appropriately. There is a potential risk of arterial thromboembolic events following intravitreal use of VEGF inhibitors. A numerically higher stroke rate was observed in patients treated with ranibizumab 0.5 mg compared to ranibizumab 0.25 mg or control, however, the differences were not statistically significant. Patients with known risk factors for stroke, including history of prior stroke or transient ischemic attack should be carefully evaluated by their physicians as to whether Accentrix treatment is appropriate and the benefit outweighs the potential risk. Available data do not suggest an increased risk of systemic adverse events with bilateral treatment. As with all therapeutic proteins, there is a potential for immunogenicity with Accentrix. Accentrix has not been studied in patients with active systemic infections or in patients with concurrent eye conditions such as retinal detachment or macular hole. There is limited experience with treatment of patients with prior episodes of RVO and of patients with ischemic branch RVO (BRVO) and central RVO (CRVO). In patients with RVO presenting with clinical signs of irreversible ischemic visual function loss, treatment is not recommended. Should not be used during pregnancy unless the expected benefit outweighs the potential risk to the fetus. For women who wish to become pregnant and have been treated with ranibizumab, it is recommended to wait at least 3 months after the last dose of ranibizumab before conceiving a child; use of effective contraception recommended for women of child-bearing potential. Breast-feeding not recommended. Following treatment patients may develop transient visual disturbances that may interfere with their ability to drive or use machines. Patients should not drive or use machines as long as these symptoms persist.

Interactions: No formal interaction studies have been performed.

Adverse reactions: Very common adverse reactions are: intraocular inflammation, vitreous detachment, retinal hemorrhage, visual disturbance, eye pain, vitreous floaters, conjunctival hemorrhage, eye irritation, foreign body sensation in eyes, lacrimation increased, blepharitis, dry eye, ocular hyperemia, eye pruritus, intraocular pressure increased, xerophthalmia, headache, anopia. Common adverse reactions are: retinal degeneration, retinal disorder, retinal detachment, retinal tear, detachment of the retinal pigment epithelium, retinal pigment epithelium tear, visual acuity reduced, vitreous hemorrhage, vitreous disorder, visual, irita, iridocyclitis, cataract, cataract subcapsular, posterior capsule opacification, punctate keratopathy, corneal abrasion, anterior chamber flare, vision blurred, injection site hemorrhage, eye hemorhage, conjunctivitis, conjunctival alergic, eye discharge, photophobia, photophobia, ocular discomfort, eyelid edema, eyelid pain, conjunctival hyperemia, strike, inflammation, urinary tract infection, anemia, anorexia, cough, nausea, allergic reactions (rash, pruritus, urticaria, erythema). Uncommon adverse reactions are: blindness, endophthalmitis, hypopyon, hypopyon, keratopathy, iris adhesions, corneal deposits, corneal edema, corneal striae, injection site pain, injection site irritation, abnormal sensation in eye, eyelid irritaiton. Serious adverse events related to Intravitreal Injections included rhematogenous retinal detachment, retinal tear and idiopathic traumatic cataract.

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