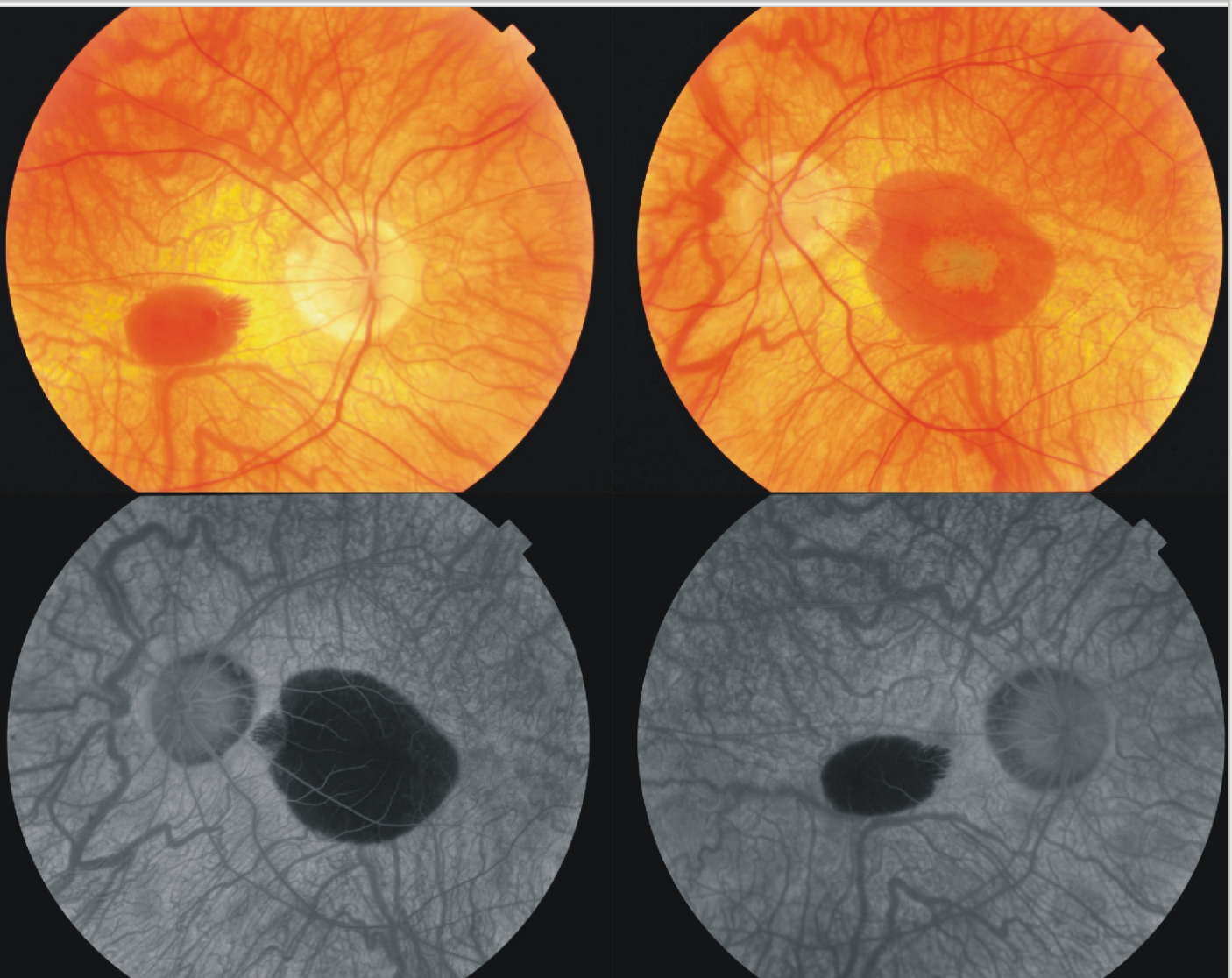


JUNE 2021



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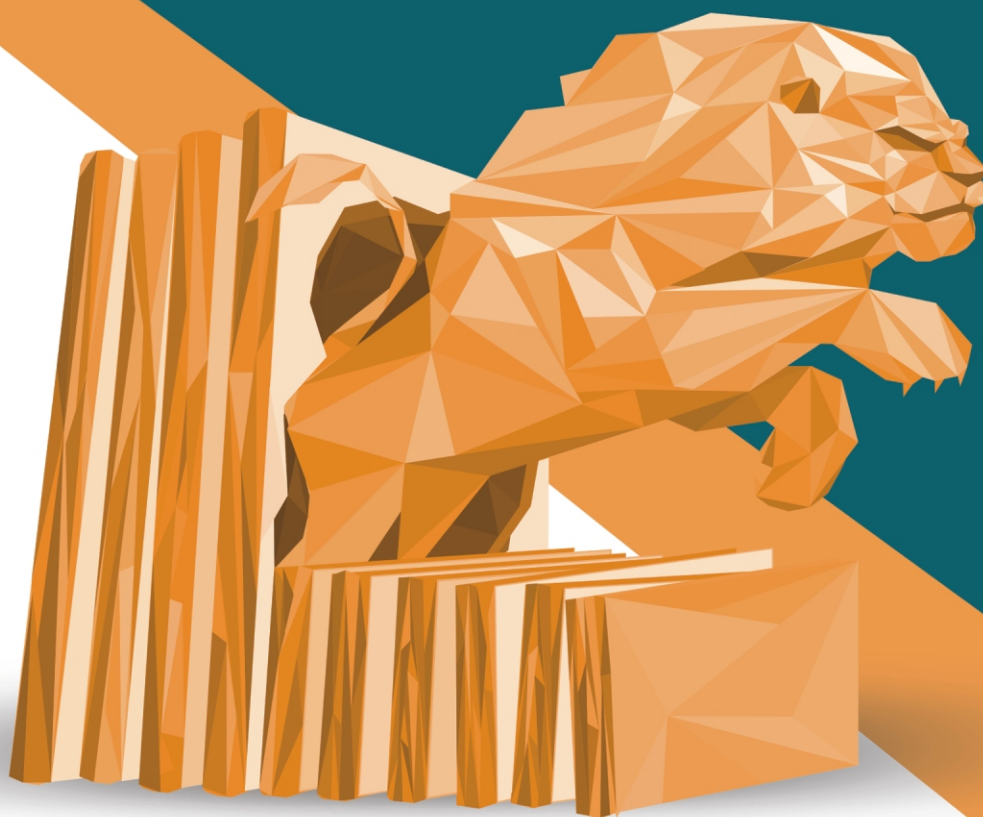
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From the President's Desk

Dr. Shobhit Chawla

Medical Director and Chief - Vitreo Retinal Services
Prakash Netra Kendr
Lucknow
shobhitchawla1412@gmail.com



I'm continually inspired by nature, and the rainbow is one of nature's greatest optical phenomenons. The sighting of a rainbow never fails to bring a smile to people's faces. They signify optimism and positivity: with them comes the sunshine after the rain.

-Matthew Williamson

After having endured an unusual 16 months we are in to the second half of this year.

Things do look up now .The second wave has left a trail of agony and tragedy but let us move forward with zest and optimism.

The June issue of the VRSI 2021 Newsletter focuses on Macular hole surgery, A Stalwart Speak editorial by William Smiddy and an interesting spotlight on the same anchored by Chetan Rao. The Retina Tech section has all about the current use and status of robotic surgery by Marc De Smet.

I thank Anand Rajendran, our scientific convenor and the contributors for the same.

Once again let's move ahead with prayers and optimism.

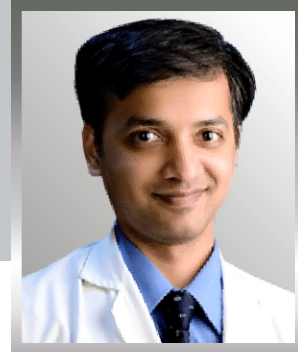
Regards and best wishes

Shobhit Chawla
President, VRSI

From the Honorary Secretary's Desk

Dr. Raja Narayanan

Director-Head, Clinical Research Consultant
Smt. Kanuri Santhamma Centre for Vitreo Retinal Diseases
Kallam Anji Reddy Campus, Hyderabad
narayanan@lvpei.org



Dear Friends:

I hope that you are staying safe during these tough times that our country is facing. We are in a lockdown situation again due to COVID, but there is hope that we will unlock soon. We should be preparing ourselves to see more patients with severe diseases as many of our patients could not come for checkup. Our fraternity has taken up the challenge of providing care during tough times, and VRSI stands in solidarity with our colleagues.

We are continuing to work with Insurance agencies and TPAs for anti-VEGF drugs, and are also working with other Societies such as the Diabetes and Endocrinology Societies to promote inter-disciplinary care for diabetic patients. We are also getting excellent response for VRSI Imaging and video contest, lead by Dr. Madana Gopal. This gives an opportunity for retina specialists to share their interesting images and challenging videos with the VRSI members.

An excellent issue of VRSI Newsletter has been compiled once again by Dr. Anand Rajendran. I am sure that you will find their articles extremely valuable for your daily practice. I take this opportunity to request you all to submit your interesting images, cases, articles and innovations to the VRSI newsletter, which will help improve the scientific knowledge base of our members. Stay safe, and we shall meet online again very soon.

Regards

Raja Narayanan
Hon. General Secretary, VRSI

From the Convenor, Scientific Committee's Desk

Dr. Anand Rajendran

Professor & Head
Vitreo-Retinal Service,
Aravind Eye Hospital, Chennai
anandrjn@gmail.com | convener.scientificcom.vrsi@gmail.com



Dear Friends and Colleagues

I do hope all of you are safe and well in these very trying times. It has been a pleasure bringing out the June edition of the VRSI Newsletter 2021, one that is focussed on Macular Hole Surgery. We have Dr. William Smiddy, an internationally renowned surgeon, giving us a marvellous account on the evolution of Macular Hole Surgery in the 'StalwartSpeak' section. The Spotlight article of the issue, focussed on Challenging Scenarios in Macular hole Surgery and anchored by Dr. Chetan Rao, has an eminent panel of national experts holding forth on an array of complex situations. In the Innovator's Isle section, Dr. Atul Dhawan and Dr. Ashok Natarajan, describe their surgical innovations to tackle tough macular holes –Chromovisco assisted ILM Peels and rotational ILM flaps, respectively. The Retina Tech Section has an enthralling account on a futuristic cutting edge technology – Robotic Surgery and its application in vitreoretinal surgery by one of the pioneers in this area - Dr. Marc deSmet. We have an interesting case report from Dr. Parveen Sen to round off this issue.

We look forward to contributions from all members to future issues of the Newsletter and hope to see the same enthusiastic response and support to VRSI activities.

Dr. Anand Rajendran
Convenor
Scientific Committee
Vitreo-Retinal Society India

Guidelines - Manuscript Submission for VRSI Newsletter



Original articles:

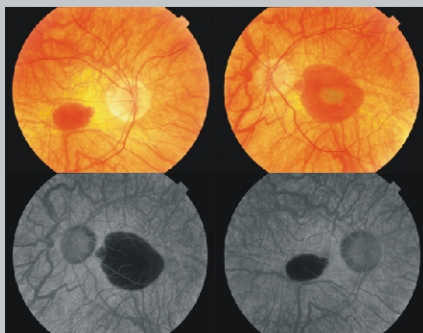
These include randomized controlled trials, intervention studies, studies of screening and diagnostic test, outcome studies, cost effectiveness analyses, case-control series, and surveys with high response rate. The text of original articles amounting to up to 3000 words (excluding Abstract, references and Tables) should be divided into sections with the headings Abstract, Key-words, Introduction, Material and Methods, Results, Discussion, References, Tables and Figure legends.

Case reports / Challenging case /Innovations / Instruments /Techniques :

New, interesting, challenging, rare cases, innovations, instruments and techniques can be reported. They should be unique and providing learning point for the readers. Manuscripts with clinical significance or implications will be given priority. These communications could be of up to 1000 words (excluding Abstract and references) and should have the following headings: Abstract (unstructured), Key-words, Introduction, Case, Discussion, Reference, Tables and Legends in that order.

The manuscript could be of up to 1000 words (excluding references and abstract) and could be supported with up to 10 references. Case Reports could be authored by up to four authors.

Mail to anandrjn@gmail.com, convener.scientificcom.vrsi@gmail.com



The Cover page Image :

A Winner at the VRSI April 2021 Retina Image Contest - "Naked Retina"

Image Description :

5 year old male with Oculocutaneous Albinism, nystagmus and spontaneous subfoveal hemorrhage due to Bruch's membrane rupture. BCVA OD 6/60; OS 3/60. FFA shows blocking of choroidal fluorescence by submacular haemorrhage with no evidence of CNVM.

Image contributed by :

Dr. Atheeshwar Das, Chennai



STALWARTSPEAK

Macular Hole Surgery: Past, Present, and Future

Dr. William E. Smiddy, MD

Professor of Ophthalmology
M. Brenn Green Chair in Ophthalmology

Bascom Palmer Eye Institute
University of Miami Miller School of Medicine
Miami, Florida, USA



Macular holes (MHs) were first described in 1869, which is a testament to the skills of Dr. Knapp given the rudimentary direct ophthalmoscope (Helmholtz) that was available at the time.¹ While the earliest authors associated MHs with a traumatic etiology, others postulated nontraumatic etiologies², and ultimately a role for vitreous traction³. The concept that an apparent operculum of avulsed foveal tissue pervaded the general dismissive thinking that there could be no possibility of a therapy to correct the anatomic or visual deficit.

It was over 100 years later that, Neil Kelly, the father of macular hole surgery (MHS), demonstrated the potential for substantial visual gain⁴. As I understand it, he discovered this as a consequence of an oversight. The standard treatment at the time for a macular hole-associated retinal detachment (most commonly seen in high myopes, of course) was to perform internal drainage through the MH at the time of vitrectomy, and then to laser the MH edges, sparingly⁵. Evidently, he forgot to laser, but not only did the retina reattached, but the MH closed with a large improvement in vision. To his credit, he understood the significance and purposefully began doing MHS. While the earliest published series yielded only about a 50% success rate, this was likely due, in part, to the fact that it included "all comers". Several others tried various adjuvants to try to enhance the surgical and visual success⁶⁻¹⁰. However, the biggest improvement seemed to be associated with peeling of the internal limiting membrane (ILM), a strategy reported by Brooks

with nearly 100% success¹¹.

Thus MHS became one of the most satisfying and frequent indications for surgery. However, what became apparent is that certain subsets of patients had poorer prognoses¹². In general, the common denominator for poorer anatomic (and visual) prognosis seems to be a larger size. This is inherent in more chronic cases, previously operated but still open cases, and some traumatic cases. However, those with associated retinal detachment, or high myopia also have a lower prognosis, presumably due to other anatomic factors. It is this group that has spurred a more recent search for improved surgical techniques.

When considering strategies to improve the prognosis, especially in such problematic cases, some consideration should be given to the mechanisms of causation and, perhaps more importantly, closure of the MH. These were initially described by Gass (before OCT was available), and his hypotheses remain a fitting framework on which to understand the mechanism of formation of a MH¹³. A parallel development in trying to understand these mechanisms, as well as to have more accurate diagnoses, has been the marked improvement in imaging that has occurred during the past 20 years or so, starting with the development of optical coherence tomography (OCT)¹⁴. With standard OCT, the occurrence of a MH can be definitively detected in virtually all cases. Moreover, the finding of

anomalous vitreoretinal attachment, specifically at the fovea, has been clearly demonstrated. While there are MHs that seem, inexplicably, to occur independent of vitreous traction, clearly the anteroposterior forces involved in such anomalous vitreofoveal separation likely cause the majority of cases. Part in parcel with this observation has been the understanding that the MH is more a perforation than a tractional tear in the sense of peripheral horseshoe tears engender. Better imaging has allowed numerous studies to identify a variety of anatomic parameters that likely influence visual acuity and prognosis. However, it is apparent that probably the most important parameter is the ellipsoid zone (EZ); the smaller the gap in the EZ, the better the VA preoperatively and postoperatively. Indeed, this is likely an important factor in many macular diseases and explains why a smaller MH has a much better prognosis than larger MHs.

Thus, the mechanisms of closure begin with making sure there are no residual vitreofoveal attachments that might impede closure of the edges. In addition, at least as an even subsequent to a breach in the fovea, is inherent traction such as might be generated from glial or other cells migrating around the edge of the MH and onto the ILM immediately surrounding the MH. Finally, it seems critical to provide a surface on which reparative cells may migrate to close the gap of the MH¹⁵. All of this must be accomplished with a minimum of additional trauma to the elements surrounding the MH. The challenges in this, of course, are in cases in which these elements might already have irreparably damaged the remaining border tissues. In particular, the ILM is usually already absent in cases that failed to close with an initial MHS, and the larger size of the MH (especially when chronic) is usually associated with more baseline EZ loss. The importance of the EZ, really a marker for the photoreceptor integrity, was first proposed based on histopathologic studies of closed MHs¹⁶, and confirmed by OCT¹⁷. In the case of myopia, the shape of the posterior pole (as with a staphyloma in the worse cases) might also be an impediment to getting the edges to reapproximate.

Thus, the elements of standard MHS involves removing any posterior vitreomacular attachments (typically with a combination of suction from the cutter or a smaller, end aspirating cannula), peeling ILM (with or without dyes), and then doing a fluid-gas exchange with a medium to long acting gas mixture. These are important steps in the new techniques as well. Postoperative management usually still involves a period of facedown posturing, although some have recommended that

this might be trimmed or avoided, in some instances through early postoperative OCT monitoring¹⁸.

Thus, there are two general strategies involved in newer MHS techniques mobilizing centrally more residual foveal tissue¹⁹ and providing an ILM flap²⁰ as a template for cells to grow, to span the MH, and to facilitate centripetal contraction of the mobilized foveal tissue. It may be that the posterior interface of the gas bubble is an adequate template for primary, standard cases¹⁸, but this needs to be augmented in large MHs. Thus, there are 2 general strategies involved: effecting a central stretching of the retinal tissue (foveoplasty) and various ILM flap techniques. Arguably the retinal patching technique that has also gained much interest is another form of the latter, but we cannot rule out that there might be some cellular transformation to facilitate function also.

The ILM flap technique was first introduced and has been highly developed by Michalewska²⁰, and extensive applications and modifications have been reported by her and other investigators. In its simplest version, the superior ILM is peeled with retention of attachment at the edge of the MH. The remaining ILM is peeled from the inferior margins, and the peeled ILM is hinged inferiorly to cover the MH. In instances where the ILM was previously peeled from the margin of the MH, a free flap from the more peripheral macula is prepared and dragged to the MH as a patch. Initial reports did not seem to contend with trying to make the ILM a monolayer, but later reports have. A difficulty with the free flap in particular is maintaining its position as the fluid air exchange is performed. One solution is to move the flap into place under a bubble of perfluorocarbon liquid; then the fluid-air exchange can be done away from the perfluoron slowly at the end, leaving the flap in proper place. Personally, I will prepare a couple flaps in case the first one is aspirated unintentionally. Also, while I do not use staining for most ILM peeling, I do in this instance so there is no mistaking the ILM distribution and to facilitate seeing that the flap is in proper position.

While other "patch" material has been reported, such as lens capsule and amniotic membrane, the most commonly discussed variation of this technique was reported by Grewal²¹. That technique involves procuring a full thickness piece of retina from the mid periphery and placing it over the MH in a similar manner to an ILM flap²¹. While good anatomic results have been reported, the visual acuity is less impressive, but that is to be expected with such advanced MHs; some evidence as been

reported that the retinal graft might become neurologically functional²², but that requires more study.

An alternative is what I referred to above as a foveoplasty. The principle is to stretch the retina centripetally so as to cover the MH with what was perifoveal retina. Indeed, this principle might be what is achieved with the standard technique when the fluid at the MH is fastidiously dried at the terminus of the fluid-air exchange. With the larger MH defect, however, more of the retina needs to be mobilized. Two general strategies have been reported. In one, a macular detachment is induced by infusing balanced salt through a small gauge infusing needle, typically with automated infusion. I have found it necessary to use a perfluorocarbon bubble to seal the MH during this process to avoid the migration of the infused BSS through the MH before it spread to make a more symmetric serous detachment. An alternative technique has been reported by D'Amico who uses a viscoelastic at the MH to elevate the perifoveal retina directly through the MH²³. When the fluid-air exchange is subsequently performed, the MH is effectively made smaller, allowing a better chance for cellular migration and secondary closure.

Certainly, these are exciting times with great resourcefulness being demonstrated and shared among many investigators. While there will, no doubt, be inherent barriers limiting the final visual acuity in the sorts of difficult MHs being tackled by these innovative techniques, it does provide some important benefit to such patients. Perhaps further advances in neurobiology will allow even more visual benefit in the future to augment (or even replace?) these newer techniques.

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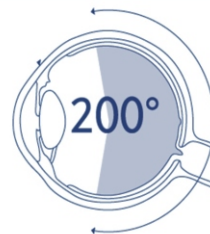


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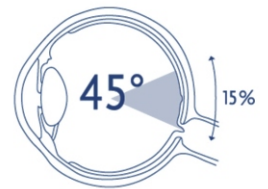
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SPOTLIGHT**Challenging Scenarios in Management of Macular Holes****Author Details:**Dr. Chetan Rao (CR)¹Dr. Lingam Gopal (LG)²Dr. Atul Kumar (AK)³Dr. Shobhit Chawla (SC)⁴Dr. Dhananjay Shukla (DS)⁵Dr. (Air Cmde) Hemant Trehan (HT)⁶Dr. Gopal Pillai (GP)⁷**Author affiliations:**¹Senior Consultant, Shri Bhagwan Mahavir Vitreo-retina department, Sankara Nethralaya, Chennai²Assoc Prof & Senior Consultant, Deptt of Ophthalmology, National University Hospital, Singapore³Professor & Chief, Dr.RP Centre for Ophthalmic Sciences, All India Institute of Medical Sciences, Delhi⁴Director, Prakash Netr Kendra, Lucknow⁵Senior Consultant, Retina-Vitreous Service, Ratan Jyot Netralaya, Gwalior⁶Command Hospital Air force Bangalore⁷Professor & Head, Deptt of Ophthalmology, Amrita Institute of Medical Sciences, Kochi

Idiopathic full thickness macular holes (FTMH) are believed to be caused due to vitreo-macular traction and are managed surgically by vitrectomy that includes inducing posterior vitreous detachment (PVD), peeling of the internal limiting membrane (ILM) around the hole and applying a gas tamponade with face-down positioning. This basic method has been reported to have closure rates of greater than 90% in stage 2 macular holes. For larger holes or recurrent macular holes various techniques of internal limiting membrane flap

management including temporal inverted flap, cabbage leaf peel, folded ILM peel and other modifications have resulted in closure rates up to 98%.¹Use of adjuvants such as amniotic membrane, viscoelastic agent, autologous plasma concentrate, and retinal free flap have been tried primarily for large holes with good success rates. The autologous retinal transplantation for large macular hole is another innovative technique that has been reported to have high functional and anatomical success rate.²

However, the management of macular holes that have developed secondary to trauma, vascular retinopathy, rhegmatogenous retinal detachment, myopic degenerative changes or inherited retinal dystrophies have been challenging as the development of hole is a combination of tractional elements and degenerative changes. We set out to explore and learn about the tactical thinking and management strategy employed by our expert panelist when confronted with unusual macular holes.

Case 1 - Chronic Macular Hole

CR : A 57 year old man presents with decreased vision in the right eye for 10 years with a Best corrected visual acuity (BCVA) of 6/36 Snellen's acuity and near vision of N10, with grade 1 nuclear sclerosis cataract and a full thickness macular hole. The other eye BCVA was 2/60 for distance due to chronic macular hole. The hole dimensions are base diameter-1631 microns, apex diameter- 877 microns, Hole height 317 microns. Hole form factor is 0.53.

There is a zone of retinal pigment epithelium atrophy at the edges of the hole in the optical coherence tomography (OCT) as shown in **Figure 1**, suggesting chronicity.

Would you consider surgery in this case?

LG: The visual prognosis is obviously poor with gross RPE damage around the hole in addition to the large size and chronicity. The rim of the macular hole is also not very edematous. I would not contemplate surgery for the macular hole in this case.

AK: I would not consider macular hole surgery in this case. Macular hole index (MHI) is one of the most important prognosticating factors to predict visual outcome after macular hole surgery (MHI \geq 0.5 is considered as a good prognostic indicator).³ In this case, calculated MHI comes out to be around 0.19 which is quite less than 0.5. The hole is quite old (10 years duration, presence of RPE atrophic changes) and has flat edges. Hence, the probability of macular hole closure and visual gain even with inverted ILM flap technique is low. Right eye cataract surgery (phacoemulsification with implantation of intraocular lens) can be considered in due course to improve his visual field.

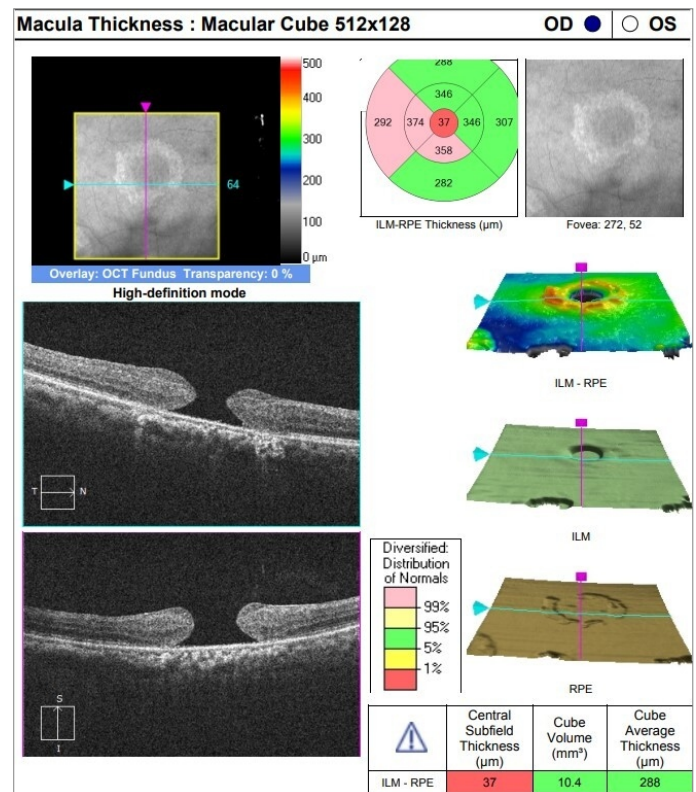


Figure 1 : OCT of the right eye showing full thickness macular hole with retinal pigment epithelium atrophy at the edges of the macular hole.

SC: I would not be enthusiastic about surgery in this patient, especially with history of chronicity and such a lot of atrophy. In a hole of this size ILM flap can only give a type 2 closure. if patient is keen for surgery nonetheless, then I would do a phacoemulsification cataract surgery with multiple ILM Flaps and give the patient a choice of full thickness autologous retinal graft too.

DS: My default answer is NO: the patient is used to this vision for a decade of his working life (or more) and significant visual improvement is unlikely.

HT: 6/36 visual acuity in the presence of a full thickness macular hole implies some visual potential despite presence of RPE changes. The RPE changes appear perifoveal.

GP: Yes, I would like to go ahead with surgery. Prior to surgery I would like to know the status of the optic nerve and do a Visual evoked potential (VEP) to see if the trauma has affected the optic nerve also.

What are the arguments, for and against surgery in a chronic macular hole?

AK: There are various factors which can predict the poor postoperative prognosis in this case.^{4,5} One of the most important factors is macular hole index. In this case, it is less than 0.5, suggesting poor postoperative visual outcome.³ Assessing configuration of the macular hole also helps to prognosticate the case.³ In this case, the edges of the hole are flat suggesting low probability of anatomical closure of macular hole postoperatively.⁶ Preoperative base diameter is one another prognosticating factor, which is also quite large in this case (1631 micron).⁵ Duration of macular hole should also be taken into consideration. Atrophic changes of retinal pigment epithelium at the edge of macular hole are a definitive sign of chronicity which limits the potential of postoperative visual recovery after macular hole surgery.

DS: Let me quickly recap the variables here: Age (youngish), vision (fair), hole size (not too bad) and configuration (not too bad either) favor surgery. However, duration (10 years...could be more) and RPE atrophy all around (OCT) are the obvious contraindications. I am sorely missing autofluorescence imaging here to clearly map out the extent and severity of RPE atrophy. Refraction is not mentioned, but presumably non-myopic status (guesstimating choroid status from low-resolution OCT) is another green signal. So, if the patient is educated and working, would appreciate a 1 or 2-line improvement (again, depends on fellow eye; won't appreciate if 6/6) and the small risk of complications, vis-à-vis the expense of simultaneous cataract surgery, I am game to give surgery a shot, since the hole can be closed.

HT: My reason to operate would be since this is financially a productive age profile and there is a chance of improving quality of life. On the flip side, vision is 6/36, it is possible this may drop if the surgery is unsuccessful, and I will operate only if the patient accepts this risk and is highly motivated for surgery.

GP: FOR: 1) Visual disturbance of 6/36, which makes visual improvement to 6/24 or 6/18 possible. If the vision was less than 3/60, there would be lesser chances of visual improvement. 2) An operable hole morphology. 3) Young age: giving a chance to improve vision in a younger patient is very important.

AGAINST: 1) Long standing for 10 years. 2) less anatomical recovery of RPE and photoreceptors post-surgery due to

preexisting degeneration. 3) Hole forming factor being 0.5 is a bad prognosis. 4) RPE atrophy at the edges of the hole is also a bad prognostic factor.

What would be your preferred technique to manage the macular hole?

AK: I would avoid doing macular hole surgery in this case.

However, in other cases of large macular holes, I do standard pars plana vitrectomy with induction of posterior vitreous detachment to remove antero-posterior vitreous traction followed by internal limiting membrane (ILM) peeling, relieving tangential traction. As the size of macular hole is large, inverted ILM flaps technique is a must. Precautions should be taken to avoid touching the base of the macular hole to prevent photoreceptor damage. Tapping of the edges of the macular hole using Tano diamond dusted membrane scraper (DDMS) can also be tried in some large macular holes.⁷ It facilitates an intraoperative increase in the height of macular hole, improving the chances of closure. 14% perfluoropropane (C₃F₈) gas can be put at the end of surgery as it provides long term tamponade in postoperative period which will improve chances of anatomical and functional success.

SC: If patient is keen for surgery, then I would do a phacoemulsification surgery with multiple ILM Flaps and give the patient a choice of full thickness autologous retinal graft too.

DS: A chronic ERM (even 10 years) can be successfully operated if the OCT parameters are favorable. A similarly chronic macular hole has much worse prognosis.

My preferred technique in this case would be to remove more vitreous anteriorly (for a bigger gas bubble), conventional ILM peeling up to arcades, extending temporally by 1-1.5DD, and using perfluoropropane (C₃F₈, 15%) for tamponade. I reserve inverted flap for bigger, myopic holes, or where large rhexis is difficult.

HT: Although the macular hole parameters are not very favorable, eyeballing the hole configuration reveals the possibility that once the traction is relieved, it is possible that the edges may either just approximate or come to lie close to each other postoperatively. The presence of an ILM flap may further aid in approximation. So, I would operate and do an inverted IL flap with a large area of peel temporally.

GP: Vitrectomy with PVD induction, ILM FLAP and long acting gas

(C3F8). I would carefully look at the periphery for any retina tears or breaks. ILM flap overlay on the hole ensures that there is anatomical closure and with that possible visual improvement. Without ILM flap, the possibility of anatomical closure in this hole is slightly lesser. We may just need a single large ILM flap overlay (temporal ILM overlay). We may not need to consider autologous transplant of retina as I think that with ILM flap itself, this hole will close. However, we may place a drop of viscoelastic, keep air pressure low and suction pressure low during air fluid exchange to ensure that the flap does not dislocate on air fluid exchange.

Take home pearls

A long duration of low vision and a full thickness macular hole that shows flat, thin edges and retinal pigment atrophy at the edges are clues to suggest chronicity of a macular hole. In addition to OCT features, autofluorescence will delineate the area of hypo-autofluorescence at the edges of the macular hole signifying the retinal pigment atrophy. A chronic macular hole morphology has poor functional outcome after surgical intervention. A useful parameter to predict the visual outcome is MHI (macular hole index) albeit more useful for idiopathic macular hole type. A MHI value of less than 0.5 suggests low chance of vision gain after surgery. There may be no change in the size of the central scotoma even if the hole closes due to atrophy of the outer retina and underlying retinal pigment epithelium. If the patient is young and motivated, then an attempt at macular hole closure with ILM flaps can be done after counselling about the very guarded surgical and functional outcome.

Case 2 - Post Traumatic Macular Hole

CR : An 18 year old man with 3 month history of loss of vision in the right eye with a past history of blunt injury with cricket ball 3 years back has a BCVA of 6/60 Snellen's acuity, N24 for near distance. There is a FTMH on OCT as seen in **Figure 2**. The hole measurements are, base diameter 2433 microns, apex diameter 722 microns.

AK: ILM peeling should be as atraumatic as possible. Staining of the ILM becomes an important issue in this case. It can be better stained using newer dyes like ILM blue which are heavy as they contain polyethylene glycol. As a result, dye gravitates to the posterior pole. Use of adjuvants like perfluorocarbon (PFCL) can also be useful in this case as it provides added stability during surgical maneuvers and avoids iatrogenic retinal tissue damage.

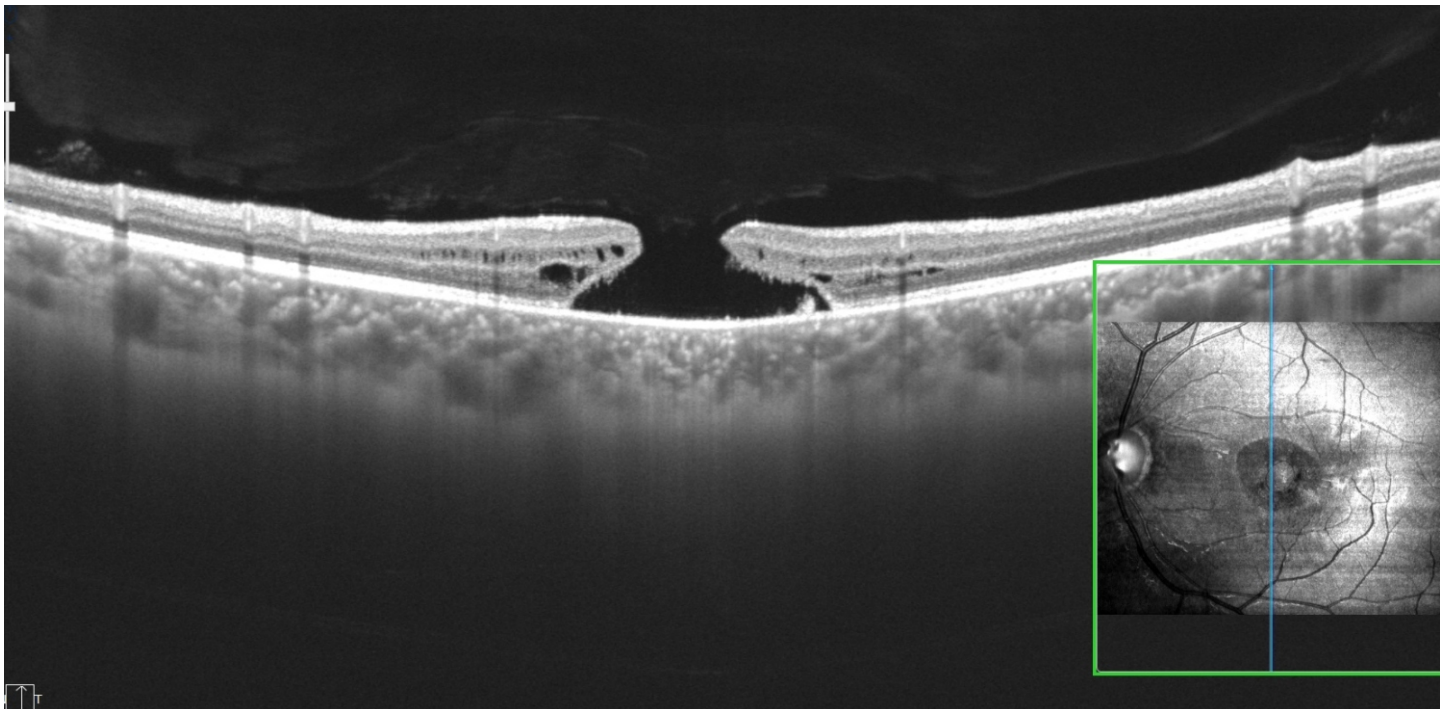


Figure 2 : OCT shows an inverted wine-glass appearance of the macular hole with no posterior hyaloid separation and some debris at the floor of the macular hole. The inset shows the infrared rendering of the macula showing the large ring of the outer hole edge and the narrow ring of the inner edge.

This macular hole has a large basal diameter and a narrow inner diameter (inverted wine glass appearance).

What will be the ILM peeling strategy since the inner edge is thinner due to possible loss of retinal elements secondary to degenerative changes?

LG: Considering the large basal diameter, I would prefer an inverted flap technique. My preferred method is to make a large temporal flap and fold across the hole (not tucking into it). I may also remove the nasal ILM carefully without disrupting the residual attachment of the temporal flap.

Dye can also be injected under the PFCL bubble to stain the ILM. Pinch and peel technique is the preferred one to initiate ILM flap. Multiple large flaps can be peeled and placed on the macular hole. ILM peeling should be circumferential and avoid peeling across the fovea.⁸

I have found NGenuity (Alcon Inc.), 3D visualization system to be highly useful in such cases where ILM is thin and retina is degenerated. It provides enhanced visualization of the membranes using various filters. It also provides increased depth of focus and magnified view of the field (more than the conventional microscopes). These features help in better

surgical maneuvering.⁹ Also, we have to keep in mind that post traumatic macular holes can spontaneously close with time.¹⁰

SC: This case requires surgery with ILM flaps, I would start away from the hole and do multiple flaps like an envelope which would stop short of the thin edge and invert the flap over the hole. I would surely do a more extensive temporal horizontal raphe area extra peel away from the hole and as it is a large hole, multiple flaps are necessary. As it is a traumatic hole there is tissue loss and not tissue dehiscence, so visual prognosis is also guarded.

DS: RAPD and traumatic retinal dialysis have not been mentioned: they must be looked for and ruled out in blunt trauma. Presuming there is no traumatic optic neuropathy (TON), this is a reasonably good prognosis case where the hole size matches the visual loss.

The basal diameter has been mentioned twice by now; it represents junk data like height, form factor, index etc.: One way of closing a large, refractory macular hole is to detach the underlying retina (hydro-dissection), i.e., enlarge the base diameter. Since International VMT study group (Ophthalmology 2013) has set a clear, reproducible, evidence-based method of measuring the hole aperture, we should stop using redundant parameters and communicate in a uniform language. In this case, narrow aperture is the key prognostic factor in closure. I agree that lack of edema at edges indicates tissue loss, but it is not a big deal. Tissue loss happens in most large macular holes, even when not traumatic. Many can still be closed... and vision improved to some extent. ILM peeling strategy will be same as in the previous case.

HT: This patient has a poor prognosis with retinal tissue loss. The only way this tissue could be filled would be with a retinal autograft, even if we use scaffold material like ILM or AMG, the loss of neural elements is very apparent and would preclude visual improvement. I would be prepared to do a retinal tissue transplant.

GP: Here we have a large hole with inverted wine glass configuration without PVD. After a good triamcinolone assisted PVD induction, we will do a large ILM peeling and stop peeling just short of the edges of the hole 360 degrees. I would like to overlay the ILM flap on the hole from multiple sides (Petal ILM flaps). Care during air fluid exchange like low pressure and low suction will help retain the position of the flap. Make sure that we don't peel the flap across the hole, it may further cause damage to the edges.

Take home pearls

The development of macular hole in trauma is a combination of neuro-degenerative changes due to the contusion injury and traction due to the vitreous. A macular hole that develops acutely at the time of contusion injury has a 10-30% chance of spontaneous closure in the ensuing months. Surgical intervention is attempted if the hole does not close after a reasonable period of observation which could be a few weeks to 6 months from presentation. Visual prognosis can be complicated by presence of concomitant traumatic optic nerve injury and must be ruled out before surgery.

Since there is evidence of neural tissue loss, special attention should be paid to peel ILM atraumatically so as to preserve the retinal tissue at the edge of the hole. A preferred technique is to develop a temporal ILM flap hinged at the edge of the macular hole, that is then inverted over it to cover it completely followed by a low pressure fluid gas exchange to avoid turbulence and displacement of the flap.

To visualise the ILM better, dye can be applied under a PFCL bubble or use a digital visualization system like Ngenuity with high magnification and colour filters.

If the tissue loss is large then a retinal autograft could be used to close the hole in exceptional cases.

Case 3 - Acute Traumatic Macular hole with submacular hemorrhage

CR : A 24 year old man came with a history of blunt injury with stone in the left eye 12 days back and was diagnosed as macular hole with high intraocular pressure (IOP) then. He was treated with IOP lowering measures. The Left eye BCVA was 1/60 Snellen's acuity.

The posterior segment (**Figure 3**) shows vitreous hemorrhage inferiorly with a FTMH and submacular hemorrhage that is confirmed on the OCT (**Figure 4**).

Considering the submacular haemorrhage that appears to be mobile and the FTMH, would you consider surgical treatment?

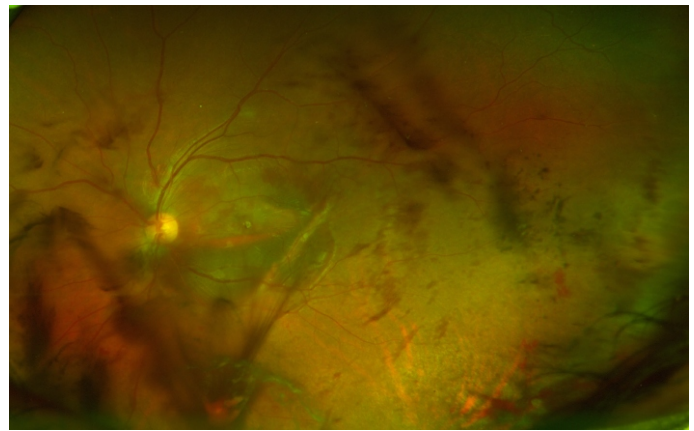


Figure 3 : Optos image of the left eye shows inferior vitreous hemorrhage with linear choroidal rupture temporal to the fovea with a large area of submacular hemorrhage collection that shows layering and a FTMH.

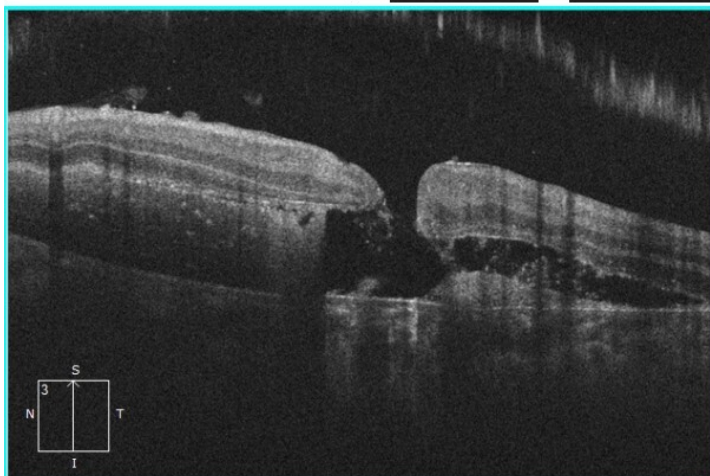
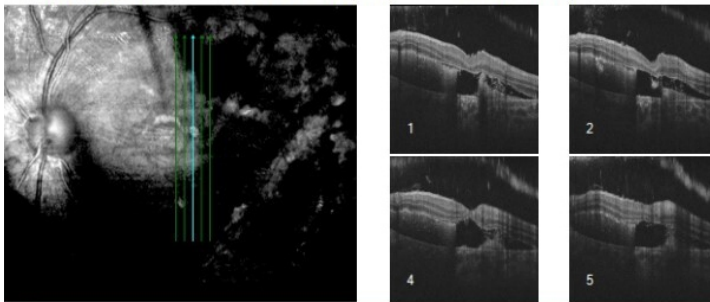
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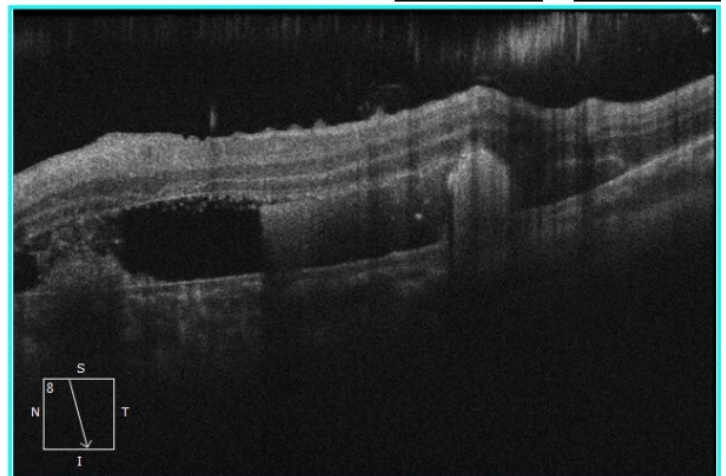
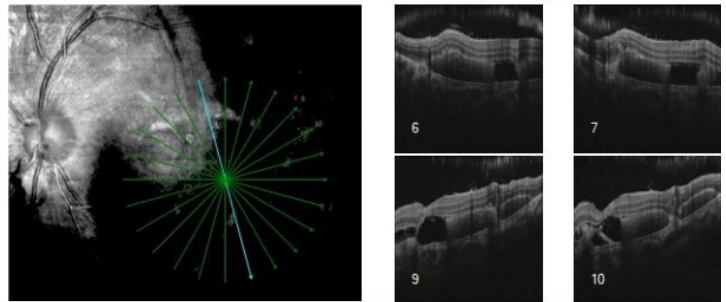


Figure 4 : OCT shows 2 radial scans through the fovea showing full thickness macular hole with subretinal low to medium reflective homogenous mass with hyper reflective dot like echoes with areas of clear space in between the masses and between the mass and the overlying retinal detachment.

LG: I would perform early surgery to evacuate the sub retinal hemorrhage same time as repairing the macular hole. Care is needed to avoid damaging the RPE while trying to remove the hemorrhage. One probably should avoid directly sucking on the blood through the macular hole. Usage of a small bubble of PFCL to squeeze the blood out through the macular hole would be better. The aim is to remove as much of the blood as possible with least trauma to the RPE underneath. Subsequent to that step, ILM peel can be done as usual followed by gas tamponade. The choroidal rupture should not be an immediate issue since it is significantly temporal to macula. Critical post op follow up is needed for detecting CNV from rupture site (patient may not be very sensitive in picking up early CNV in view of the relatively compromised visual status).

AK: As the submacular hemorrhage is still mobile, we can consider initial trial of intravitreal injection of 50 microgram of recombinant tissue plasminogen activator. This technique will help in liquefaction of blood and displacement of blood from the fovea. We can serially follow up the patient for 48-72 hours monitoring displacement of the bleed and size of the macular hole. If there is no displacement of bleed and macular hole closure by 48-72 hours, then other surgical options can be tried.

SC: Observation for a week or two weeks would be ideal as there is edema I would give a trial of steroids for three days in any acute blunt trauma eye. Once a complete IOP control is achieved, gonioscopy, angle imaging and status of lens has been ascertained I would repeat OCT one month from injury date, re-study the OCT picture, as in the initial stage the haemorrhage is mobile and edges are edematous. Another approach would be to just do an early vitrectomy with gas tamponade and see how this situation responds to displacement and hole closure. So the merits have to be weighed here on an individual case basis. Using an adjuvant like tPA is not indicated as small amount of subfoveal bleed and presence of macular hole.

DS: Let us first separate wheat from chaff: macular hole and vitreous hemorrhage are small and insignificant at this time; both are likely to resolve spontaneously. I'd fuss about the missing wheat: TON and additional choroidal ruptures (latter commonly associated with submacular bleed). The key element is submacular bleed, which is THIN (underlying RPE visible) and patchy. BCVA does not appear to correspond to media clarity, macular hole or submacular hemorrhage. Since the patient is young (healthy retina & RPE) and coexisting TON is a possibility, I would observe him for at least 1-2 months for spontaneous

resolution of subretinal hemorrhage and simultaneous closure of the small macular hole: both events are likely.

HT: I would operate. The immediate danger here is the submacular hemorrhage and it is already 12 days old. The macular hole can be tackled even later.

GP: Yes, surgery can be tried with explained guarded prognosis. The subretinal haemorrhage warrants removal and hence we will have to go in for surgery. And as we are going in for surgery, best to correct the macular hole along with it. Even with submacular haemorrhage, in this patient, vision of 1/60 is not completely explainable with the macular pathology. Probably there must have been an optic nerve injury also along with the macular damage due to the blunt trauma. The quantification of optic nerve injury can be done by doing a VEP. If the subretinal haemorrhage was not there in this case, I would have observed this hole as post traumatic holes can close spontaneously over the next 6 months.

What would be the surgical strategies that can be tried ?

AK: If the above strategy fails, the patient can be taken up for primary vitrectomy with PVD induction with submacular injection of recombinant tissue plasminogen activator. It can be injected subretinally after creating a self-sealing retinotomy using 41 G needle attached to tuberculin syringe. No attempt at ILM peeling may be required, as it is a case of recent onset traumatic macular hole which would close just by bleed displacement. Tamponade can be achieved using SF6 gas. Patient should be advised to maintain propped up position to facilitate displacement of bleed from the fovea.

SC: Another approach would be to just do an early vitrectomy with gas tamponade and see how this situation responds to displacement and hole closure. So the merits have to be weighed here on case-to case basis. Using an adjuvant like tPA is not indicated for a small amount of subfoveal bleed and presence of macular hole.

HT: First step is gas and tissue Plasminogen activator injection (tPA). Observe for 48 hours. If we are lucky, the blood displaces inferiorly and PVD gets induced with subsequent hole closure. Even if the macular hole does not close, we can always operate later if the blood has displaced away from the macula.

If displacement is unsatisfactory, then proceed to vitrectomy. Sequence of events: PVD, brilliant blue, reassess. If most of the

blood has self-evacuated during the previous steps due to the fluid currents, I would do an ILM peel and gas and close. If there is still blood and the perifoveal region is elevated, I would create a midperipheral detachment with a 41 g cannula, put PFCL and make a mid-peripheral retinotomy and drain (otherwise blood would migrate back to macula during FAE: fluid-air-exchange). Then I would do a normal ILM peel under PFCL followed by FGE: fluid-gas-exchange.

GP: The following strategies:

- Intravitreal TPA, followed by supine position for 1 hour and then vitrectomy ILM peeling and gas injection.
- Vitrectomy with subretinal r-tissue plasminogen activator (tPA)-assisted clot lysis. Using a 41 gauge cannula, subretinal TPA injection can be done and then after clot lysis the blood can be taken out through the macular hole itself. If it is a large haemorrhage, it is better to make another opening to drain the blood out, but in this case, we have a thin film of fluid which allows us to use the macular hole to drain it out.
- Intravitreal r-tPA-assisted pneumatic displacement without vitrectomy using an expansile gas and postoperative positioning.
- Pneumatic displacement alone can also be tried, but 12 days duration will make the blood more resistant to movement without tPA.

Take home pearls

In acute full thickness macular hole secondary to trauma complicated by subretinal haemorrhage due to a choroidal rupture, visual prognosis and surgical treatment will depend on duration and mobility of submacular haemorrhage and location of choroidal rupture. If the choroidal rupture is not visible due to large amount of immobile subretinal haemorrhage and the patient presents within 2 weeks then early surgical treatment is prudent. The goal of surgery is to displace or remove the subretinal haemorrhage. If there is massive amount of subretinal blood then the strategy is to use tPA to lyse the clot, remove the lysed blood via a paramacular retinotomy or push it out via the macular hole with a PFCL massage and then a gas tamponade. However if the submacular haemorrhage is a thin film, mobile and patient presents within 2 weeks of injury, then either we can wait for spontaneous closure of macular hole or do pneumoretinopexy to displace the sub macular blood. The gas injection may induce a vitreous detachment, relieving the attachment to the hole or tamponade it allowing the hole to close spontaneously. However if the hole persists then a conventional vitrectomy with ILM peeling and gas tamponade can be done. If there is a subfoveal choroidal rupture or the submacular haemorrhage is present for longer than 2 weeks or there is a subfoveal scar then surgical treatment can be deferred as the functional outcome is poor.

Case 4 - Myopic Foveoschisis with macular hole

CR : A 68 year old man has loss of vision for one year and has BCVA with -10.5 DS 6/24 Snellen's acuity, N10 in the Right eye and BCVA with -8.5 DS 3/60, N10 in the left eye.

Both eyes have myopic macular degeneration involving the fovea and the Optos image (**Figure 5**) shows a staphyloma of the macula with FTMH and subretinal fluid with underlying choroidal degeneration. The OCT (**Figure 6**) show a FTMH with schisis at the edges of the macular hole.

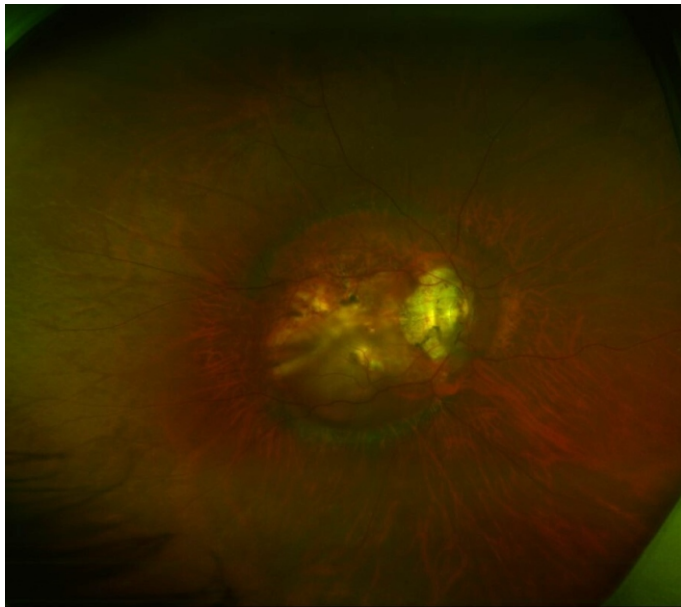


Figure 5 : Optos image showing right eye macular staphyloma with choroidal degeneration with a FTMH with detachment of the inferior macula within the staphyloma.

Ultrasound shows axial length of the right eye:29 mm and left eye:28 mm with deep staphyloma.

What would you suggest for the right eye?

LG: My approach would be to try vitrectomy and ILM peel. The staphyloma does not appear very deep. A large ILM peel and gas tamponade would be in order. Phaco-emulsification and intraocular lens implantation at the same time can be

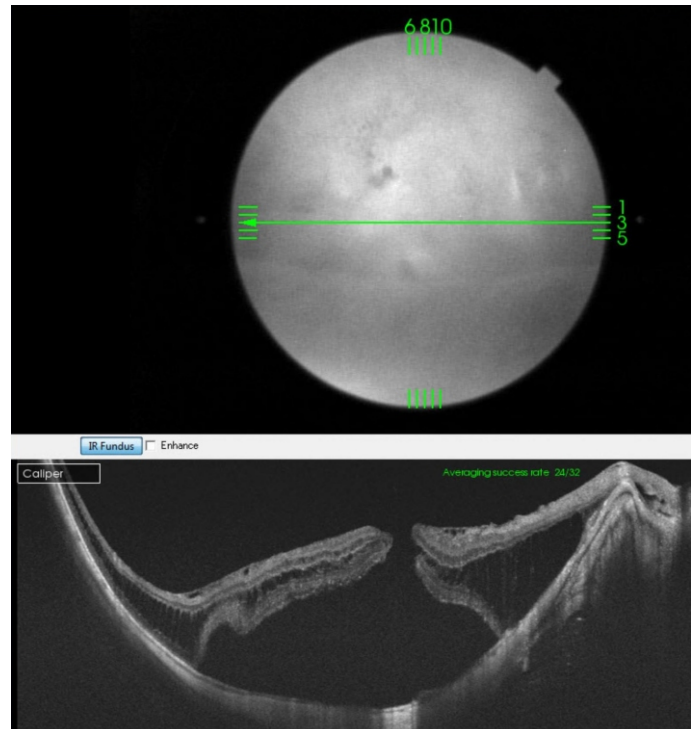


Figure 6 : Swept source OCT shows a FTMH with subretinal fluid and inner and outer retinal schisis at the edges of the macular hole with steep staphyloma margins.

considered if some degree of cataract is present.

Even if PVD appears to be present, I would use triamcinolone acetonide to look for strands or sheets of residual schitic vitreous before staining for ILM. Considering the macular hole and retinal detachment one would expect the dye to migrate sub retinally. If one is able to plug the macular hole with triamcinolone this may be prevented to some extent.

Trying to inject the dye under air may not be a good idea since the air under pressure can stretch the macular hole in the absence of ILM peel. Either way dye can still make its way into subretinal space even under air.

I am deliberately not using the technique of staining with PFCL bubble over the macula (as described for the next case), since in the presence of a staphyloma and macular hole, there is a risk of the PFCL bubble migrating under the retina.

If one finds the ILM forceps slightly short for comfortable ILM peel, removing the cannula will give the extra length needed to reach the retina.

AK: I would suggest going ahead with vitrectomy with PFCL assisted ILM peeling followed by placement of multilayered ILM flaps over the macular hole. I routinely do it using NGenuity visualization system and obtaining real time feedback through microscope integrated intraoperative optical coherence tomography (MIOCT). There is an indispensable role of MIOCT in assisting complete removal of posterior vitreous cortex and internal limiting membrane (ILM) peeling in the treatment of myopic macular hole retinal detachment.⁹ MIOCT helps to identify vitreoschisis and confirm the position of ILM flaps over the macular hole intraoperatively.

There are few things which need to be taken care during surgery. Complete removal of posterior vitreous cortex using triamcinolone should be attempted. Vitreoschisis should be identified and removed with the help of diamond dusted membrane scraper or forceps. The opening of the macular hole can be sealed by placing a drop of ophthalmic visco-surgical device (OVD) like Healon over it. It minimises the chances of subretinal migration of dye and PFCL. Newer dyes like ILM Blue can be used to enhance visualisation of thin ILM as they are heavy and the dye gets concentrated at the macula. Peeling should be gentle as ILM is thin and there is poor contrast. PFCL should be also injected gently ensuring that it remains as a single bubble. After peeling and placing of flaps, peripheral retinotomy can be made to drain the remaining fluid and lasered. Long-acting gases are usually preferred in staphylomatous eyes like 14% perfluoropropane (C3F8), which provides long term tamponade.

SC: First thing important is to document recent visual loss and symptoms here. I would like to review old records here to see what the vision was in this eye earlier. The patient we are dealing with has only eye with useful vision. Once progressive visual deterioration in the last few months has been elicited on history, surgery should be considered.

DS: I presume the left eye is inoperable/amblyopic, for which no details are provided. We have a perfect storm here: One-eyed, old-aged high myope with posterior staphyloma, myopic chorio-retinal degeneration, chronic macular schisis with detachment AND macular hole, in the better-seeing eye! In simple words: a little to gain with great difficulty and a lot to lose. The lens status is missing. I'd get it out of way even if clear (& no IOL), aphakia also helps to focus better intraoperatively on a myopic macula. I use Trypan Blue & BBG dyes together under air for 3-5 minutes to stain and loosen the ERM & ILM. I'd hinge the ILM flap at the edge of hole, trim it, and cover the hole with it just before fluid-

air exchange. I also try to peel ILM to the edge of the staphyloma if possible. This is about the only situation where I drain subretinal fluid through the macular hole. Even with good surgery, one must beware of flat but enlarged macular hole & atrophied macular retina, which could stump visual recovery in spite of anatomic success.

HT: There are definitely two schools of thought here and I would do a vitrectomy only, stain well with brilliant blue and peel under a flat contact lens. I would use PFCL to displace the SRF to the periphery and do a drainage retinotomy above the edge of the staphyloma.

GP: Recently Barbara Parolini had presented the MTM staging system based on OCT of the myopic retina. This system of classification takes into account the perpendicular as well as parallel forces on the surface of the myopic macular schisis.

They described four MTM retinal stages (1. Inner/Outer Maculoschisis; 2. Predominantly outer Maculoschisis; 3. Maculoschisis-Macular Detachment; 4. Macular Detachment) and three foveal stages (a. Normal fovea; b. Inner Lamellar-Macular-Hole; c. Full-Thickness-Macular-Hole).

Guidelines of management of MTM were proposed, but customized for each stage. Initial stages 1a and 2a, which define maculoschisis in the inner or inner-outer or only outer layers of the retina, should be observed. Stages 3a and 4a, defining macular detachment with and without associated schisis, should be treated with a macular buckle (MB). Stage 1b, which is a lamellar macular hole in a myopic eye, should be treated with pars plana vitrectomy (PPV) only in symptomatic cases. Stages 2b, 3b, and 4b should be treated with a MB and PPV should be added in a second step only if the presence of a lamellar macular hole requires intervention to improve visual function. Stage 1c, which is a full thickness macular hole in a myopic eye, should be treated with PPV. Stages 2c, 3c and 4c should be treated with a combination of simultaneous MB + PPV to treat both the retinal pattern of schisis or detachment and the full thickness macular hole.

In this case, I would prefer the macular buckle with vitrectomy.

When would you prefer macular buckle alone or in combination with vitrectomy?

AK: With the advent of modern MIVS which is safe and predictable, macular buckling is less preferred now-a-days as it can cause buckle slippage, injury to ciliary nerves, vortex veins and optic nerve. There are also chances of globe perforation,

muscle disinsertion, ocular motility disturbances, refractive error, dry eyes, etc.¹² We have obtained favorable outcomes using vitrectomy alone in majority of our patients.^{11,13} Complete release of anteroposterior and tangential traction is must. MIOCT and NGenuity are very helpful in achieving that goal.

SC: My choice would be a combination surgery if possible. As macular buckle is an ab externo surgery it can be tried first with the possibility of adding a vitrectomy with phacoemulsification and ILM flap and silicone oil injection.

DS: I have no experience with a macular buckle, but I guess it is not mandatory for good outcome in this case.

HT: I have no experience with macular buckle but have done vitrectomy for such myopic macular holes. The results have been satisfactory even with only vitrectomy. There is an MRI based classification of Staphylomas where the authors have recommended macular buckle for narrow staphylomas and vitrectomy for broad shallow staphylomas.

Take home pearls

A macular hole in a highly myopic eye presents a surgical challenge as it is secondary to tangential traction causing schitic changes in the retina, vitreo-schisis above it and a disproportionately large scleral to retina surface area ratio due to staphyloma. The axial length being long, conventional instruments may not reach the posterior pole, therefore upper sclerotomies are placed closer to the horizontal meridian, more posteriorly and in some cases without the cannula. Since the PVD is mostly anomalous, the posterior hyaloid is identified by staining with intravitreal triamcinolone and completely removed along with any residual cortical vitreous on the retinal surface. Due to the poor contrast from the underlying choroidal degeneration, good staining is achieved by dye staining under air and sometimes under PFCL or a cohesive visco-elastic agent is used to block the macular hole so as to avoid subretinal dye migration. However PFCL is generally avoided as it may migrate under the macular hole if care is not taken to achieve a single bubble large enough to cover the hole. ILM peeling is done all around with a hinge at the macular hole edge and then inverted over it. A retinotomy can be done at the edge of the staphyloma to drain the subretinal fluid or drain through the hole with low flow as there is a possibility of it getting stretched and enlarged. A long acting gas such as 14% C3F8 is preferable. A concomitant cataract surgery helps in clearing the media for visualisation. Use of intra-operative OCT to correctly identify ILM and vitreoschisis has been recommended.

Vitrectomy with ILM peeling is combined with macular buckling only for steep staphyloma to promote retina-scleral adhesion and relieve the traction.

Case 5 - Macular Hole in rhegmatogenous retinal detachment

CR : A 41 year old lady presents with recent loss of vision in the left eye.

She has essential hypertension and has inferior branch retinal vein occlusion.

Left eye BCVA is hand movements close to face.

There is a dry fibrovascular proliferative tissue(FVP) along the inferior arcade and detached hyaloid and a macular hole as is seen in **Figure 7**. The retina is totally detached with no peripheral breaks.

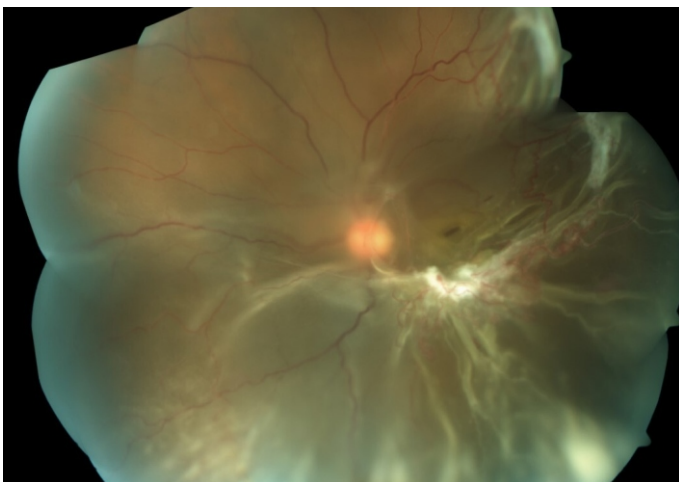


Figure 7 : A montage colour fundus photo shows total retinal detachment with a condensed Weiss ring over the ONH and a FTMH with inferior dry fibrovascular proliferation and distal sclerosed vessels.

How would you stabilize the retina for an effective ILM peeling?

LG: There is a possibility that the macula was already cystic due to BRVO before it opened up into a hole due to the traction at the inferior arcade caused by the Fibrovascular proliferation. If this were true, it would have repercussion on the hole closure as well as visual recovery.

The first step would be to remove the Fibrovascular proliferation. I would not hesitate to use chandelier illumination and bi manual surgery to facilitate quick dissection of the tissue. Once all traction is relieved, ILM peeling will be done.

AK: Use of perfluorocarbon liquid (PFCL) helps to flatten and stabilize the retina. Use of PFCL acts as a counter acting force. It pushes the retina back shifting the subretinal fluid to the periphery. Use of PFCL also provides a clear media for maneuvering.

SC: The recent visual loss indicates recent onset Retinal detachment. I would do a good vitrectomy with as much base removal as comfortably possible. Remove the fibrovascular proliferation after a PFO support for posterior retina making sure to avoid subretinal migration of PFO.

DS: Let's first clarify the diagnosis: this is NOT a macular hole in RRD; this is primarily a macular TRD due to BRVO, which developed a macular hole and converted into a combined-mechanism RD. Secondly, the age and BRVO don't match. I'd look for secondary causes of HT and assess renal status.

HT: I would stabilize with PFCL. I am wary of these proliferations as in my experience they are typically more adherent, and the underlying retina is usually thinner than diabetic proliferations. I would tackle the periphery first, the reason being that once I have done the ILM peel and placed the flap in position (if I use a flap) then I want no turbulence after this step and only want to have to remove PFCL and inject tamponade. Leaving the periphery un-tackled would result in a lot of manipulation subsequent to the ILM peeling and that can cause turbulence within the PFCL bubble even with a valved cannula system and this could dislodge the flap.

GP: Two methods of stabilization can be done for effective ILM peeling in this case

1 Make a retinotomy in the supero-nasal area, do a near complete air fluid exchange to make the retina flat and then shift to fluid. Using optic nerve as a stabilizing area, we can peel from nasal to temporal area and once we have the broad edge of the ILM, we can shift direction to complete the peel.

2 However in this case, it is better to remove the FVP fully and then have PFCL on top of the macula and do the staining under the PFCL and then peel under PFCL. Plan to remove the FVP fully before PFCL injection.

What is the most efficient way to stain with dye in a mobile retina with macular hole?

LG: For ILM peel I would place a bubble of PFCL over the macular hole and inject the dye around the bubble. By rotating the PFCL

bubble, the dye can be allowed contact with the retina around the macular hole without permitting the dye to migrate subretinally. Then I would remove the dye and peel the ILM with the PFCL bubble in place. The bubble is not too large and so it permits the bubble to roll to one side when ILM is actually being peeled but exerts enough counter pressure to facilitate the ILM peel.

AK: Staining should be attempted after complete removal of vitreous including posterior vitreous cortex after PVD induction. In a mobile retina, it can be stabilized using PFCL. After PFCL injection over the macula, dye can be injected under the PFCL bubble to achieve adequate staining. For effective staining, dye should be kept for adequate time and infusion should be switched off so that dye does not get washed.

SC: I would stain under PFO as stain takes up well with brilliant blue even under PFO.

DS: ILM can be peeled without stabilizing detached macula, after staining it under air with dyes as mentioned before.

HT: Prior to injecting PFCL. BBG Toxicity is minimal and even if does gain access to the subretinal space, it will be diluted with SRF and displaced to the periphery by the PFCL that is injected to stabilize the retina. BBG can also be injected under PFCL where I would start injecting at the disc edge since there is a change in PFCL contour there that allows the BBG to slip under the PFCL bubble with a lower risk of subretinal PFCL migration due to elevated injection pressure of BBG.

GP: PFCL assisted technique: PFCL is injected to cover the hole and dye injected under the bubble. We can also stain the ILM completely with BBG and then remove the excess and then use PFCL. However, this technique may allow BBG to enter into the subretinal plane. Hence in retinal detachment with macular hole, the preferred technique would be to use PFCL to flatten the retina and then use BBG below it.

If macular hole was not the primary break, would you still consider doing ILM peeling?

LG: I would perform ILM peel for the macular hole irrespective of whether it is the only break or there are other breaks.

AK: Yes, peeling ILM will lead to removal of tangential traction leading to closure of macular hole. It will also decrease the chance of postoperative occurrence of epiretinal membrane and other proliferative vitreoretinopathy changes like macular pucker formation in cases of old retinal detachment.

SC: Yes I would consider ILM peel as it would also decrease traction in the area from fibrovascular proliferation to macular area.

DS: I do not recommend ILM peeling as a primary procedure for macular holes secondary to macular tractional membranes (as here) or those co-existing with an RD with peripheral breaks. In my experience, most of these breaks close simply by peeling the tractional membrane and settling the RD. As I prefer silicone oil in many of these cases, I have a second chance to peel ILM during oil removal, though it is required in very few cases.

HT: Yes, it is unlikely though that a total RD with a proliferation has no peripheral break even upon searching during a vitrectomy. It would usually be a combined detachment. Nevertheless, even if it was a macular hole detachment, the hole would need to be closed and that would mean an ILM peel.

GP: Macular hole is usually not the primary break in these cases. The break is usually within the area of FVP or very close to it. If we are going in for a vitrectomy, it is better to close the hole as well, if visibility is good.

Take home pearls

A macular hole in a detached retina presents a challenge for ILM peeling manoeuvre. Whether the macular hole was the cause of the retinal detachment or developed secondary to degenerative changes following retinal detachment due to a peripheral retinal break, the consensus is to peel the ILM around it, to achieve closure in the primary surgery itself.

A thorough vitrectomy is done to remove all adhesions at the posterior pole whether due to fibrovascular proliferation, epiretinal membrane or partial PVD. Subretinal fluid can be drained from the peripheral break by a fluid-air exchange or displaced by using PFCL on the posterior pole to stabilize the macula and reduce the risk of subretinal migration of the dye. The ILM is stained under the PFCL or air and peeled around it following which fluid-air exchange is done through the macular hole or the peripheral break and a tamponade is used. ILM can still be peeled without using PFCL or draining the SRF, but peeling needs to be initiated from the temporal peripapillary retina using the ONH as the anchor that provides counter-traction and then proceed to the temporal perifoveal area.

Case 6 - Proliferative diabetic retinopathy with macular hole

CR : A 55 year old gentleman with 1.5 years history of blurred vision with scatter laser done previously presents with BCVA in the left eye of 6/18 Snellen's acuity and near vision of N18. Fundus examination reveals a fibrovascular proliferative tissue on the optic nerve head with ILM stria and dragging of the fovea nasally as in **Figure 8**.

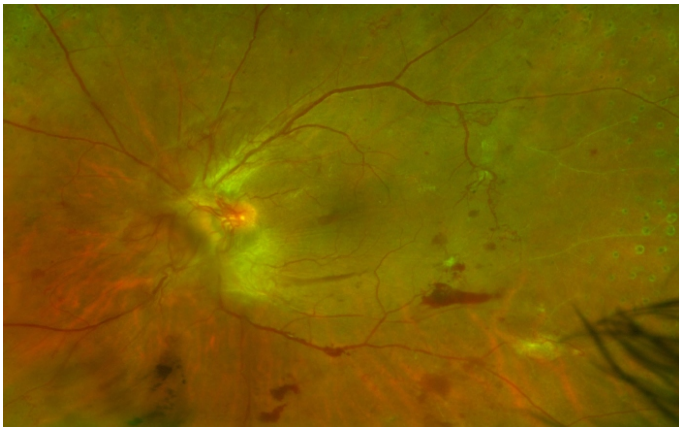


Figure 8 : Optos photo of the left eye shows a fibrovascular tissue at the optic nerve head causing contraction with ILM stria nasal to fovea with distortion of the foveal reflex and few new vessels temporal to the macula and peripheral laser scars.

There is vitreoschisis with a thin epiretinal membrane at one edge of the macular hole on OCT as in **Figure 9**.

In traction related to contraction of the posterior hyaloid in PDR or to the proliferative tissue itself, would you prefer to just remove the proliferative membranes for the closure of hole or do ILM peeling in all such cases?

LG: I would first remove the FVP from disc with its extensions across the retina. Then stain for ILM. In all likelihood one would find some tears in the ILM. I would peel ILM irrespective of presence or absence of ILM tears.

AK: In cases of vitreomacular traction with macular hole, I try to remove all the tractional fibrovascular membranes and adherent hyaloid which are responsible for causing traction at the macula after performing triamcinolone assisted vitrectomy. After that I would do ILM peeling to relieve the tangential traction with placement of flaps over the hole. It increases the chance of macular hole closure. It also helps to decrease the chances of postoperative occurrence of epiretinal membrane and other proliferative vitreoretinopathy changes. ILM peeling is difficult in cases of PDR as ILM is strongly adherent in some areas and there are chances of break formation due to avascular retina. Also, media clarity gets compromised due to presence of bleed around fibrovascular proliferation. Centre sparing ILM peeling can be done in cases where macula is thin and cystic.

In cases of Vitreomacular traction without macular hole, I would not initiate PVD induction from optic disc. I would release vitreous attachments around the macula then initiate the PVD from disc. This limits the chances of iatrogenic macular hole formation. MIOCT is helpful to confirm release of traction and to check for macular hole formation.

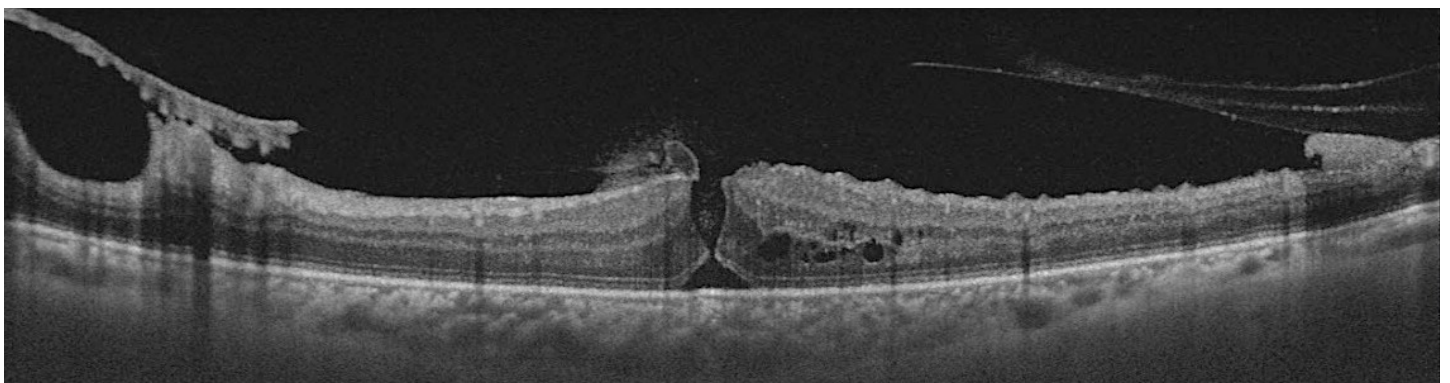


Figure 9 : Swept source OCT radial scan shows the thick fibrous proliferative tissue at the peripapillary area with a small macular hole and vitreoschisis temporal to it.

SC: The first step is to do a good posterior hyaloid removal and identify the vitreoschisis and take care of it. The fibrovascular proliferation on disc also needs to be removed. Once all traction is taken care off, I would like to stain and identify any residual membranes if present. The hole is very small and once all the traction is relieved closure should happen without ILM peeling.

DS: This is a textbook case to demonstrate redundancy for ILM peeling. This is anyway a tiny hole; even for a larger hole secondary to tractional PDR, ILM removal is redundant to close it, though one could argue in favor of ILM peeling for a better foveal anatomy and to reduce ERM recurrence. My technique for membrane peeling would be as described before. Trypan Blue dye is essential here to removal the ERM.

HT: I would do an ILM peel in all the cases. It has two benefits, firstly, it ensures that I have removed all the proliferation and the second membrane is removed too. Secondly this would avoid a second procedure for a step that I could have easily completed the first time itself. The only scenario in which I would postpone the ILM peel would be if the visualization deteriorates intraoperatively due to a bleed while removing the proliferation.

GP: In this case, there is only very mild traction at the macula, macula has neither detached, nor is there a hole. I would like to observe the patient since the vision is good (6/18) and there is no recent history of decrease of vision. The history of loss of vision is 1.5 years prior. This patient may remain at 6/18 for a long period of time as scatter laser was done prior to 1.5 years. Going in to do vitrectomy in this case may find us newer FVP and nonvascular complexes and removing hyaloid from these areas may cause more complications. Overall, in a patient with lasered PDR who has 6/18 vision, staying stable over 1.5 years, I would want to observe as risk versus benefit in improving vision is rather poor.

If surgery is performed, ILM peeling will need to be done. In PDR the ILM is very adherent and may come in a piecemeal fashion.

Can you suggest tips with regards to ILM peeling maneuvers in cystic macula?

LG: In a cystic macula one would resort to foveal sparing ILM Peel. In the present case however, it is not an issue due to two reasons- The macula has a very few cystoid spaces and should withstand ILM peeling well and there is already a full thickness hole.

AK: There are high chances of macular hole formation if peeling is tried over cystic macula. I prefer center sparing ILM peeling in cystic macula to avoid deroofting of thin retinal tissue over the fovea. I also confirm the extent of cystic retina and occurrence of accidental macular hole using intraoperative optical coherence tomography and modify my steps accordingly.

SC: If one desires to peel in an edematous macula it has to be a single flap foveal sparing peel away from edema gently, just a temporal flap is enough, or if nasal ILM is more accessible because of stria it can be removed remaining away from fovea .

DS: Over a cystic, schitic macula (\pm a large outer lamellar hole), it is PVD induction, which is the precarious procedure; not the ILM peeling. And anyway - not just in thin retina - in EVERY case, I have replaced pinch-and-peel with the safety of Finesse[®] forceps to lift an edge atraumatically.

HT: The peel would be slow and gentle and if I notice too much traction on the retina then I would stop the peel and peel around this area and allow some time for the retina to relax if the ILM peels smoothly then I would proceed, otherwise I would truncate the area and leave a patch of ILM disconnected from the surroundings. Ideally, using an intraoperative OCT would help but I have no experience with one.

GP: In a cystic macula, there is always the risk of de-roofing of the inner retina. Hence PVD induction should be very careful and we should not peel the PVD off the macula. We should lift carefully and use the shave mode with high cut rate and low suction to remove the posterior hyaloid.

Then we need to stain the ILM well and try to lift up the ILM from nasal or inferior part of the macula where ILM is thicker. Using the disc as a harnessing support we can peel nasal to temporal. Make sure that we stop close to the fovea and complete the peel from all areas near the fovea. Do not de-roof the ILM from the fovea. Final removal of ILM from the fovea region can be done by controlled shaving with cutter.

Take home pearls

When a macular hole develops in proliferative diabetic retinopathy it is due to a combination of contraction of the posterior hyaloid or tangential traction from a fibro-vascular proliferative tissue around the macula and is often complicated by the presence of a cystic macular thickening.

The goal of surgery is to remove all proliferative tissue around the macula, identify the vitreo-schisis or secondary membranes and detach the posterior hyaloid to relieve all traction. Decision to remove the ILM is a surgeon's discretion as it is believed to ensure the complete removal of the remnants of the posterior hyaloid and also remove scaffold for re-proliferation. However, ILM peeling over a cystic macula can cause retinal damage by deroofting the cysts. If a surgeon decides to remove the ILM, then, after staining with a dye, the ILM is peeled using a finesse loop or by pinch-and peel technique sparing a circular rim around the fovea followed by a tamponade. This technique of fovea- sparing ILM peeling reduces the risk of over-manipulation trauma as the underlying macula could be atrophic due to ischemia and ILM is generally friable in a diabetic eye and comes off piece-meal, necessitating multiple attempts.

Case 7 - Macular hole in chronic rhegmatogenous retinal detachment

CR : A 20 year old man with history of blunt injury 5 years back and loss of vision 3 months back in the left eye presents with BCVA 2/60 Snellen's acuity, 6/24 reduced Snellen in the left eye. On fundus examination there is macular hole with peripheral break inferiorly and temporal retinal detachment with superotemporal demarcation line (**Figure 10**).

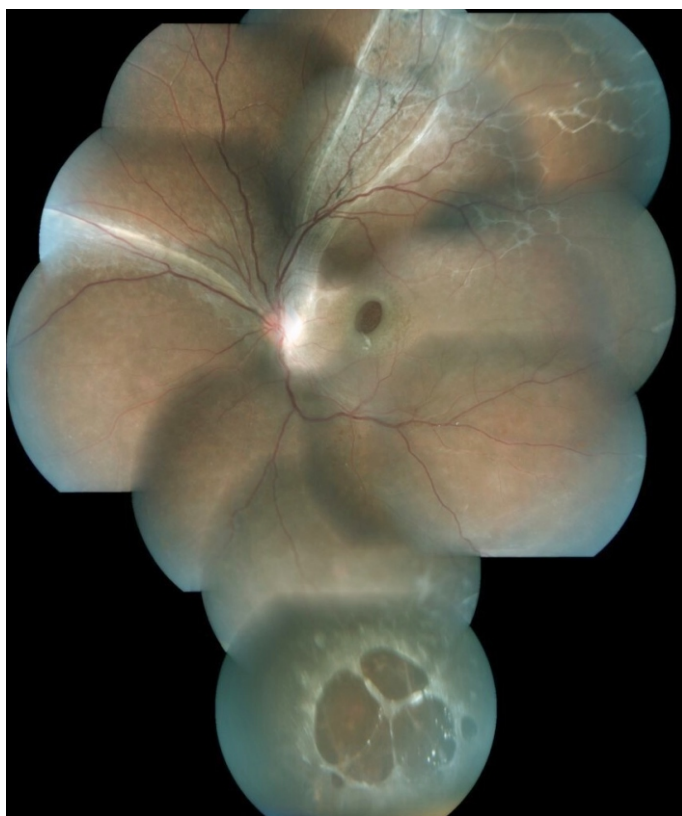


Figure 10 : Shows a montage photo of the left eye showing an atrophic break in the inferior periphery with a FTMH and supero-temporal demarcation lines.

OCT shows vitreous prolapse into the FTMH with adherence to the retinal pigment epithelium (**Figure 11**).

Considering that this is a chronic retinal detachment with possible chronic macular hole with a guarded functional outcome, what would be the choice of surgery: Scleral buckling

or primary vitrectomy with ilm peeling and tamponade?

LG: One wonders what is the cause of recent onset loss of vision? The fundus photo clearly shows demarcation lines and thin atrophic retina in between that seems to include all except an upper nasal wedge. There is significant sub retinal gliosis in peripapillary area that probably contributes to the large size of the macular hole. The purpose of any surgery in this case is to maintain the small area of the upper nasal wedge of retina attached.

In view of the same I would opt for first buckling the inferior break as the first approach. Since the macular hole appears plugged by vitreous gel, it is unlikely to be contributing to the rhegmatogenous retinal detachment.

AK: It is a case of chronic retinal detachment with demarcation line. An inferior peripheral break is also present. There is presence of vitreous prolapse into the full thickness macular hole with absence of posterior vitreous detachment (PVD). In this case, I would plan to go ahead with passing encircage followed by primary pars plana vitrectomy with PVD induction to remove the posterior hyaloid. After that, PFCL would be slowly injected over macula to stabilize the retina and Brilliant Blue G (BBG) dye would be injected beneath the PFCL bubble to stain the ILM. 360 degree ILM peeling would be done and ILM flaps will be placed on the macular hole. Peripheral shave vitrectomy will also be done to release all traction around the break. Fluid air exchange will be done. Peripheral laser of break and all suspicious lesions will also be done. Postoperative tamponade will be achieved by using silicone oil or 14% perfluoropropane (C3F8) gas.

SC: I would consider a buckling procedure in this scenario as the first choice . The prognosis of macular hole surgery is not good here, so the visual benefits of macular hole surgery have to be explained to patient. Moreover macular hole surgery will not benefit here.

DS: Vitrectomy is preferred here to attempt macular hole closure and improve retinal oxygenation to maximize visual outcomes in this chronic RD; remember, the patient is noticeably young. The macular hole appears to have an ERM on nasal edge, and has excessively cystic edges, due to chronicity of macular edema. As indicated before, I will not peel ILM first time (only ERM, without staining), use silicone oil; and peel the ILM SOS during oil removal, over an attached retina.

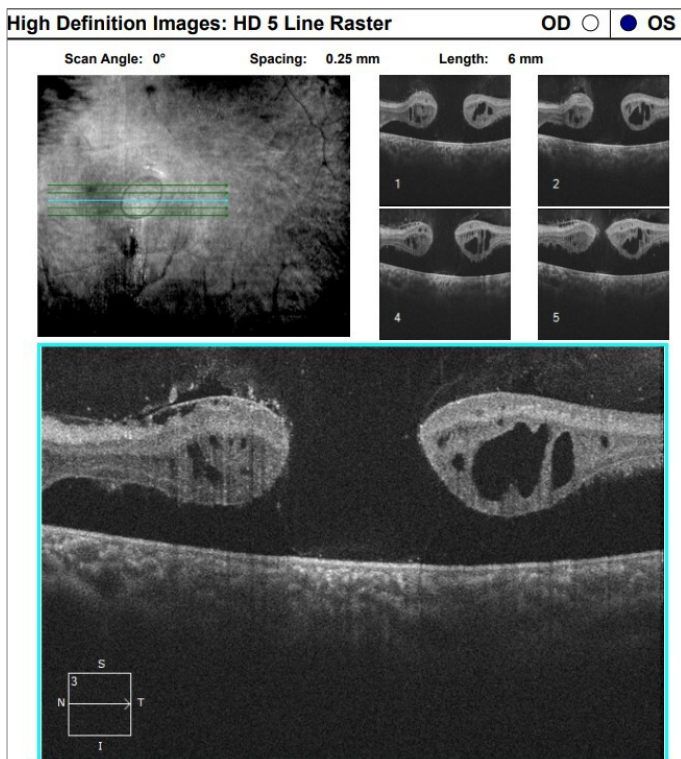


Figure 11 : OCT of the left eye shows a FTMH with subretinal fluid and prolapse of the vitreous into the macular hole with apparent adherence to the retinal pigment epithelium at the floor of the macular hole and cystic degeneration of the hole edges. There is no hyaloid separation.

HT: In this scenario, it is unlikely that we would be able to restore central vision. Aim of surgery would be to restore field and normal attached retinal status. I would do a buckle first and reassess. Vitrectomy for macular hole only if the patient insists that I try, I would be very hesitant to enter this eye unless forced to.

GP: The amount of macular detachment and the hole does not completely explain the BCVA of 2/60 in the eye. Hence there may be chronic degeneration of the macular region and/ or optic nerve damage which is irreversible. Hence the prognosis after surgery is guarded. In view of the large inferotemporal hole, which is broad and the contractions around the demarcation lines in the superior quadrant, associated with a macular hole, we may do a combined surgery in this young man with a buckle to support the inferotemporal retinal break and a vitrectomy, PVD induction and ILM peeling to close the hole.

Would you consider staged procedure: first SB and then macular hole surgery?

LG: If the retina is attached following scleral buckling, I would leave the macular hole alone. Only if the retinal detachment persists (despite the inferior break having been closed) will I consider revision surgery in form of vitrectomy, ILM peel.

AK: I will go ahead with primary vitrectomy. I will use an encircling band as it is an old retinal detachment with inferior retinal break. Recently, we have obtained similar successful outcomes in old retinal detachments without the use of encircling band using NGenuity 3D visualization system.¹⁴ It helps in complete removal of peripheral vitreous by providing a magnified and high resolution view of the retinal periphery.

SC: If we are able to achieve a level of visual acuity which is close to pre-detachment level more surgery is not indicated. Moreover, macular hole surgery will not benefit here.

GP: Staged procedure can also be tried, first to attach the retina by doing a buckle and then tackle the macular hole. However, I would prefer the combined approach as there are degenerations in the edges of the macular hole and it would be better to attach the edges back early itself.

If in the setting of recent injury with retinal detachment due to dialysis with a secondary macular hole, what would be the procedure of choice?

AK: I would go ahead with primary vitrectomy as we can tackle retinal dialysis and macular hole in a single surgical sitting, releasing all the vitreoretinal traction and treating the primary pathology. It will help in decreasing postoperative discomfort to the patient and achieving early visual rehabilitation.

DS: In a fresh traumatic RRD with macular hole and peripheral dialysis in a young patient, I would prefer scleral buckling: re-attachment of a fresh RRD could itself close a coexisting macular hole, more so when traumatic.

HT: I would do a buckle for the dialysis RD (I assume this would be a young phakic patient). Then I would wait for spontaneous macular hole closure. If no closure, then I would do a vitrectomy and ILM peeling provided there is a perceived potential for visual improvement. This strategy has two benefits, firstly I don't need to approach the periphery in a young.

GP: In a recent injury with dialysis, it is better to take up the patient for a buckle. Many of the tractional macular holes close spontaneously in 6 months.

Take home pearls

If a macular hole develops in a chronic retinal detachment due to a peripheral break, then it is more likely due to degenerative changes along with subretinal or pre retinal membrane traction. The goal of surgery is to re-attach the retina and improve the visual field. If posterior vitreous detachment is absent and the macular hole is plugged by the vitreous then Primary Scleral buckling can be attempted for the peripheral break to re-attach the retina. In this instance, closing a macular hole will not improve re-attachment rates or visual prognosis due to the chronic nature of the detachment. However if the retinal detachment persists then a subsequent vitrectomy with ILM peeling and tamponade can be done. The other option is to do Primary Vitrectomy with PVD induction, peripheral encirclage, meticulous removal of epiretinal and subretinal proliferative tissue, ILM peeling with or without a flap and tamponade. If there is a history of recent injury and a rhegmatogenous retinal detachment due to retinal dialysis, no PVR and a macular hole, either Scleral buckling or Primary vitrectomy with or without ILM peeling and tamponade can be done. Both approaches are designed to close the primary break with possibility of spontaneous closure of macular hole with successful re-attachment of retina. ILM peeling can be done with flap or without flap but attention is paid to meticulously remove epiretinal membranes.

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EYLEA™ SOLUTION FOR INTRAVITREAL INJECTION IN VIAL. Approved name(s) of the active ingredient(s) One ml solution for intravitreal injection contains 40 mg aflibercept. Each vial provides a usable amount to deliver a single dose of 50 µl containing 2 mg aflibercept. Indication EYLEA™ is indicated for the treatment of neovascular (wet) age-related macular degeneration (wAMD). Dosage Regimen wAMD The recommended dose for EYLEA™ is 2mg aflibercept, equivalent to 50µl EYLEA™ treatment is initiated with one injection per month for three consecutive months, followed by one injection every 2 months. There is no requirement for monitoring between injections. Long term (after the first 12 months of treatment), it is recommended that patients continue to be treated with EYLEA™ every 2 months. Method of administration Intravitreal injections must be carried out according to medical standards and applicable guidelines by a qualified physician experienced in administering intravitreal injections. Following intravitreal injection patients should be instructed to report any symptoms suggestive of endophthalmitis (e.g. eye pain, redness of the eye, photophobia, blurring of vision) without delay. Each vial should only be used for the treatment of a single eye. Contraindications Know hypersensitivity to aflibercept or to any of the excipients, active or suspected ocular or periocular infection, active severe intraocular inflammation. Special warnings and special precautions for use Endophthalmitis and retinal detachments may occur following intravitreal injections. Patients should be instructed to report any symptoms suggestive of endophthalmitis or retinal detachment without delay and should be managed appropriately. Increases in intraocular pressure have been seen within 60 minutes of an intravitreal injection. There is a potential risk of immunogenicity and arterial thromboembolic events following intravitreal use of VEGF inhibitors. EYLEA™ should not be used in pregnancy unless the potential benefit outweighs the potential risk to the foetus. Women of childbearing potential have to use effective contraception during treatment and for at least 3 months after the last intravitreal injection of aflibercept. Undesirable effects Very common: Conjunctival hemorrhage, eye pain. Common: Retinal pigment epithelial tear, detachment of the retinal pigment epithelium, cataract, cataract cortical, cataract nuclear, cataract sub capsular, corneal erosion, corneal abrasion, intraocular pressure increased, vision blurred, vitreous floaters, vitreous detachment, injection site pain, foreign body sensation in eyes, lacrimation increased, eyelid edema, injection site hemorrhage, punctate keratitis, conjunctival hyperemia, ocular hyperemia. For full listing of undesirable effects, please refer to the full product insert. For further prescribing information, please contact Bayer Zydus Pharma Private Limited, Bayer House, Central Avenue, Hiranandani Estate, Thane, Maharashtra, India Pin-400607. Email: medicalinfo.india@bayerzidyuspharma.com. Source: Based on CCDS / Version 10 / 19 Apr 2016; PI Rev 03 Nov 2016. Date of revision of API, 17 Nov 2016. For the use of healthcare professionals only.



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INNOVATOR'S ISLE - I

Chromovisco assisted ILM flap placement technique for large macular holes

Authors :

Dr. Atul Dhawan, MS, FERC¹
 Dr. Janani Sreenivasan, MS²
 Dr. T. R. Kanishka Devi, DNB¹



Author Affiliations :

Retina Service,
 Dr. Agarwal's Eye Hospital, Chennai
²Shri Bhagwan Mahavir Vitreoretinal Services,
 Medical Research Foundation, Sankara Nethralaya, Chennai

Corresponding Author :

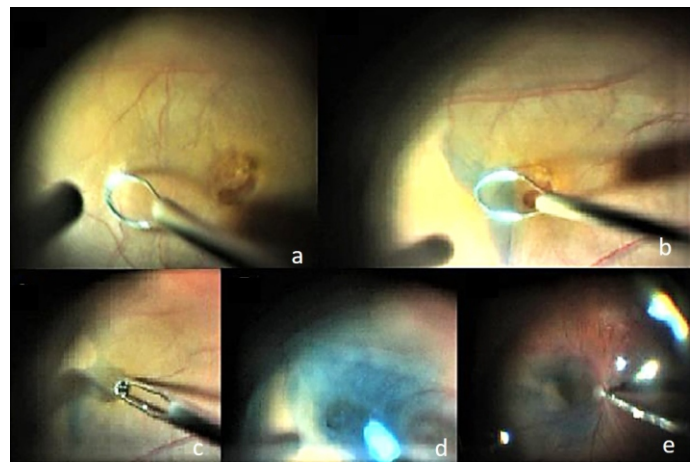
Dr. Atul Dhawan, MS, FERC
 Senior Consultant, Retina Service, Dr. Agarwal's Eye Hospital, Chennai

Introduction

Vitrectomy combined with internal limiting membrane (ILM) peeling and endotamponade is the standard of care for macular holes (MHs), with excellent anatomical success rate (90%).^{1,2} The inverted ILM flap technique has been associated with type 1 closure and a better functional prognosis, with temporal inverted flap having fewer complications.³⁻⁵ Many authors have used viscoelastic as an adjunct in MH surgeries.⁶⁻⁸ In this technique temporal inverted flap was combined with the application of coloured viscoelastic. The viscoelastic material helps to retain the flap during fluid air exchange and the dye helps to identify viscoelastic thereby ensuring complete removal. The anatomical and functional outcomes were good with reasonable restoration of foveal morphology.

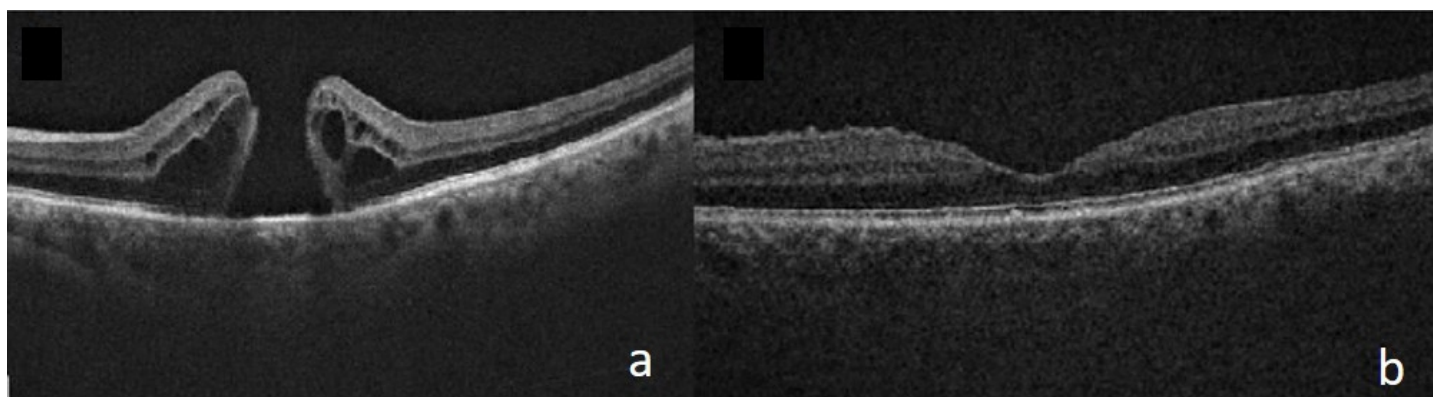
Methodology

This was a retrospective study conducted at a tertiary eyecare in which we included 30 eyes of 30 patients with idiopathic large MH (stage 3 and 4) with a minimum hole diameter $\geq 400\mu\text{m}$. Exclusion criteria include idiopathic MH stage 1&2, minimum hole diameter of $<400\mu\text{m}$, history of previous retinal surgery,



Intraoperative image: a- Finesse loop for creation of flap by pinch and peel method; b-180° temporal flap of one disc diameter; c- Radial inversion of flap covering the MH; d- ILM after staining with chromo-visco solution; e- SF6 gas tamponade.

and secondary MHs. Routine pre-op and postoperative assessment was done including visual acuity, Intraocular pressure, Anterior and Posterior segment evaluation. SD-OCT



SD-OCT-Foveal morphology comparative image. a- preoperative image of a patient with MH with BCVA logMAR 1.47; b- Postoperative 6-month image of the same patient with complete re-establishment of ELM and EZ (U type closure) with BCVA logMAR 0.77.

was used to measure preoperative MH parameters and assess postoperative anatomical outcomes.

Surgical technique:

A standard 25-gauge, 3 port Pars Plana Vitrectomy (PPV) was performed by a single surgeon. Initial core vitrectomy was performed followed by Triamcinolone Acetonide assisted PVD induction and completion of vitrectomy. Then ILM was stained with 0.05% Brilliant Blue G (BBG) dye for 30 seconds. Initial part of the flap created using Finesse loop. The flap was extended superiorly and inferiorly 180° at the temporal side of about one disc diameter area. The ILM was inverted. The chromo-visco solution was prepared by taking 0.6ml of Hydroxy Propyl Methyl Cellulose solution 2% w/v (Aurovisc) in a 1ml syringe which was the LMW viscoelastic used for anterior segment surgeries. In the same syringe, 1 to 2 drops of 0.05% BBG dye was taken and mixed well. The prepared chromo-visco solution was placed over the flap drop by drop to maintain the flap in position. The infusion was stopped before addition of chromovisco solution. Then fluid air exchange was done, followed by Air-Sulphur hexafluoride (SF6) exchange (Figure 1). Thus, at the end of the procedure viscoelastic can be completely removed because of the blue colour. By this technique, surgeon can be sure that flap was maintained in the desired position at the end of the surgery. Postoperative prone positioning was advised for 5 days.

Results

There were 30 patients in the study. The mean minimum hole diameter (μ) was 616.43 μ m and mean base hole diameter (μ) was 1280.66 μ m. The mean preoplog MAR BCVA was 1.06 \pm 0.27. MH closure was observed in all patients, excepting 1 patient

who needed repeat surgery for hole closure. In this study, U type of closure was observed in 23 patients. Functional improvement in the visual outcome was analysed by keeping the preoperative mean logMAR as a reference. During follow-up visits i.e., 1 month, 3 month and 6 months, there was definite improvement in mean logMAR and it was statistically significant with the final mean logMAR BCVA 0.63 \pm 0.28. Final postoperative gain in visual acuity of >2 Snellen lines was noted in 27 patients, and in remaining 3 patients, it remained stable, or 1 Snellen line improvement noted. Dissociated optic nerve fibre layer (DONFL) was noted in 7 patients in temporal side of fovea without visual implications.

Discussion

In 2010, Michalewska and associates introduced the inverted ILM flap technique and found that the development of a U-shape closure type was most prevalent after it and had a better functional prognosis.³ Michalewska et al compared temporal inverted flap with classic inverted flap and concluded that both techniques have similar anatomical and functional closure rates, with temporal flap having fewer cases of DONFL.⁵ Flap displacement remains a concern in this technique.^{3,9} Ocular viscoelastic device (OVD) has been noted to be safe and effective to stabilize ILM flaps.⁶ Thus, we combined temporal ILM flap technique with coloured OVD to prevent flap displacement and ensure complete OVD removal.

The anatomical success rate achieved was comparable to the previous studies of temporal inverted ILM flap.^{5,14,10} 'U' type closure was the most common type of closure in this study. There was a statistically significant improvement in the mean logMAR observed from 1-month postoperative visit onwards. There was a positive correlation between change in logMAR BCVA and

difference in minimum hole diameter. These two observations are consistent with the literature.^{5,4,10.}

The limitations of the study include lack of assessment of change in retinal sensitivity (microperimetry) after surgery, retrospective nature, smaller sample size and short follow-up, which can be viewed as future scope of the study. Regarding the technique, the inverted ILM flap technique cannot be applied when ILM around the MH has been completely removed. Randomised controlled studies involving larger patients are needed to assess long-term outcomes.

Conclusion

The limited ILM peeling technique “**Chromovisco assisted ILM flap placement technique for large macular hole**” is a safe and latest alternate for traditional temporal inverted flap techniques for large MHs. U type closure was seen in majority of patients with visual recovery of >2 Snellen's lines noted in 90% of patients. The rapid visual recovery seen in this procedure is probably due to minimal ILM handling, with minimal disruption in foveal morphology and better retention of flap aided by coloured OVD.

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INNOVATOR'S ISLE - II

Novel rescue technique of rotational tongue transposition for lost internal limiting membrane flap during primary macular hole surgery

Rescue Rotational ILM Flap for Macular Hole

Dr. Ashok Nataraj, MS FMRF¹

Dr. Jay U Sheth, MD²

Dr. Unnikrishnan Nair, MS FRCS²

Dr. Manoj Soman, FRCS DNB²

Affiliations

¹Senior Retina consultant, Jyothis Eye Care, Kannur, Kerala

²Department of Vitreo-retina, Chaitanya Eye Hospital and Research Institute, Trivandrum, India

Corresponding Author:

Dr. Jay Sheth

Clinical Research Lead,
Chaitanya Eye Hospital and Research Institute,
Trivandrum.

None of the authors have any proprietary interest



Summary Statement:

We illustrate an innovative surgical technique of rotational flap translocation when faced with intraoperative loss of internal limiting membrane flap during primary surgical management of large macular holes. This rescue procedure can be successfully performed under saline as well as air tamponade, with good anatomical and visual outcomes.

Abstract:

Purpose: Management of large macular holes (MH) is challenging and many innovations in internal limiting membrane (ILM) peeling techniques have been described. Here we illustrate a novel surgical technique of rescue rotational tongue transposition of ILM flap in cases with intraoperative loss of primary ILM flap for management of large MHs.

Methods: The method involves taking an elongated tongue-shaped ILM flap from the margins of ILM peel and rotating it to be placed inside the MH similar to an inverted ILM flap technique.

Patients: In our case series, the rescue procedure was undertaken under saline for three cases and under air for one case. The timing of transposition was dependent on the surgical step when ILM flap loss was encountered, namely while performing the ILM peel or during the fluid-air exchange.

Results: At two months, three cases showed type-1 closure while one case showed type-2 closure of MH.

Conclusion: Rotational tongue transposition is a very simple surgical technique with good outcomes for management of intraoperative lost ILM flaps. Since one end of flap remains

attached to the main ILM bed, it potentially obviates the disadvantages associated with alternative techniques such as free ILM flap, retinal graft, the use of viscoelastic, glue or perfluorocarbon liquids and so on.

Introduction

Full-thickness macular hole (FTMH) consists of an anatomic defect in the fovea with the interruption of neural retinal layers extending from the internal limiting membrane (ILM) to the retinal-pigment epithelium (RPE).¹ The prevalence of idiopathic MH in general population varies from 0.2 to 3.3 per 1000.^{2,3} The standard of care in MH management remains pars plana vitrectomy with ILM peeling, and gas tamponade. Literature has shown macular hole closure rate of 85-90% after primary surgery.^{4,5} Secondary macular holes are related to pathological conditions such as trauma, high myopia,⁶ macular schisis,⁷ macular telangiectasia⁸ and uveitis⁹ may have variable surgical outcomes.

In 2010, an innovative technique called the “Inverted ILM flap technique” (IIFT) was described by Michalewska et al wherein the ILM is placed into the MH without completely removing it.¹⁰ The authors reported that this technique increased the rate of MH closure to 98% for large idiopathic MH while reducing the chances of having a flat open configuration of the hole. However, this technique has a learning curve with a possibility of losing the ILM flap either during peeling or during fluid air exchange (FAE), leading to compromised surgical outcomes.

Here we describe a modified procedure based on IIFT which can be performed when faced with intraoperative loss of ILM flap.

Methods

Four patients who underwent surgery for FTMH and had intraoperative loss of ILM flap during a planned IIFT were included in this study. All subjects underwent complete ophthalmic evaluation including best-corrected visual-acuity (BCVA) assessment using Snellen's chart, intraocular pressure measurement (IOP) by Goldmann applanation tonometer, anterior-segment evaluation using slit lamp biomicroscopy, fundus evaluation using +78 D lens and indirect ophthalmoscopy and also spectral-domain optical coherence tomography (SD-OCT) using Spectralis (Heidelberg Engineering, Heidelberg, Germany) respectively. MH measurements were done on SD-OCT. Written informed consent was obtained from all patients for the surgery.

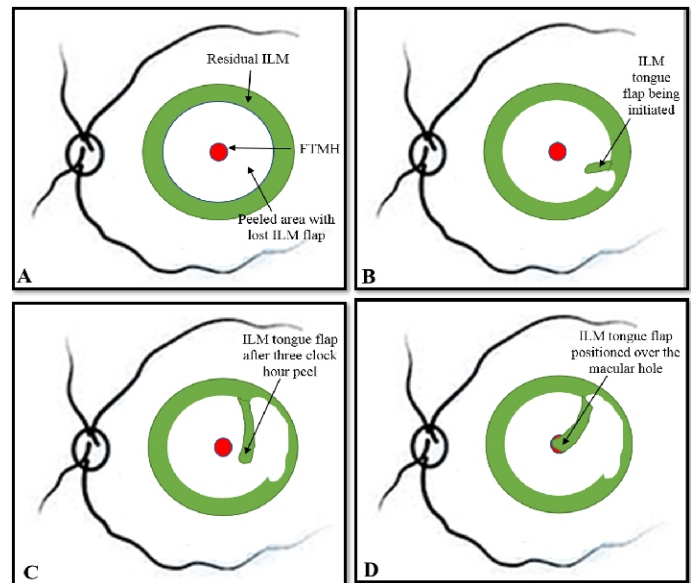


Figure 1: Diagrammatic illustration of surgical technique for performing rotational tongue transposition of internal limiting membrane (ILM) flap in a situation of intraoperative lost ILM flap (A). A strip of ILM peel is initiated using pinch and peel technique from the already peeled ILM margins temporal to the full thickness macular hole (FTMH) (B). The peel is continued for three-clock hours to get a long tongue shaped strip of ILM (C) which is then rotated at its base and gently nudged into the macular hole (D).

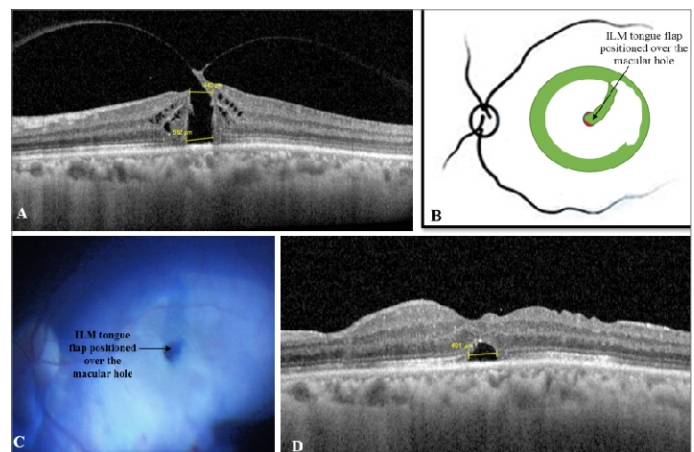


Figure 2: Spectral-domain optical coherence tomography (SD-OCT) of first patient showing presence of full-thickness macular hole (FTMH) with base diameter (BD) of 552 μ m. The patient underwent rotational tongue transposition of the internal limiting membrane (ILM) flap for intraoperative loss of ILM flap, under saline tamponade which is illustrated graphically (B) along with intraoperative photograph (C). At two months post-operatively, the patient demonstrated a Type-1 closure of FTMH.

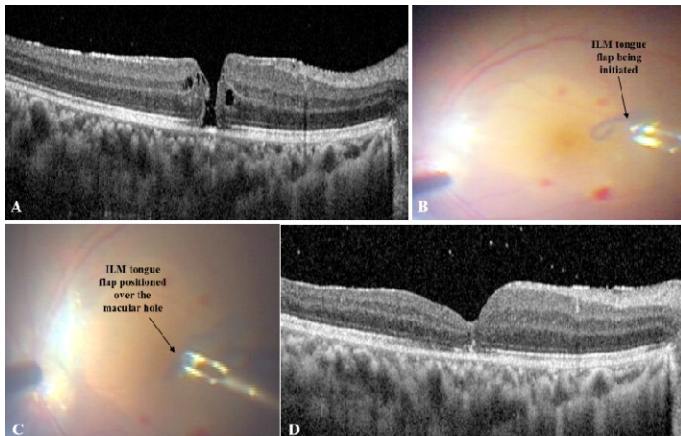


Figure 3: Spectral-domain optical coherence tomography (SD-OCT) of second patient showing presence of full-thickness macular hole (FTMH) with base diameter (BD) of 509 μm . The patient had intraoperative loss of internal limiting membrane (ILM) flap while performing fluid-air exchange. A rotational tongue transposition of the ILM flap was performed under air tamponade which is in the intraoperative photographs (B, C). At two months post-operatively, the patient demonstrated a Type-1 closure of FTMH.

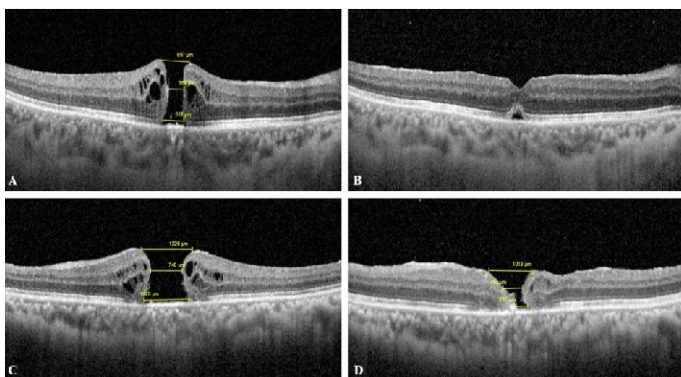


Figure 4: Spectral-domain optical coherence tomography (SD-OCT) of third patient showing presence of full-thickness macular hole (FTMH) with base diameter (BD) of 518 μm (A). At two months post-operatively, the patient demonstrated a Type-1 closure of FTMH (B). The fourth patient had an extra-large FTMH with a BD of 1070 μm (C) which showed a Type-2 closure at two months post-operatively (D).

Surgical Technique

Three-port pars-plana vitrectomy was performed by a single surgeon (AN) using 25G valved -cannula system (Alcon), Constellation vitrectomy machine (Alcon, Fort Worth, Texas) and

non-contact wide-angled viewing system (OCULUS-BIOM; OCULUS Surgical-Inc., FL, USA) for all patients. Proportional vacuum vitrectomy with a cut-rate of 10,000 cuts per minute was utilised. Posterior vitreous detachment induction was performed after staining with triamcinolone acetonide and vitrectomy completed. Peripheral retina was depressed and evaluated; any retinal breaks or lattice degeneration if noted were treated by laser photocoagulation. Brilliant-Blue G (BBG) dye (Ocublue-Plus; Aurolab, TN, INDIA) was injected into the vitreous cavity after switching off the infusion. After two minutes of contact period, the infusion was switched on to wash out the excess dye. Using the high magnification lens of the non-contact OCULUS-BIOM viewing system, ILM peel was initiated by pinch and peel technique and was done covering two disc-diameter area around hole. 360° partial ILM was left attached to edges of the MH. Excess ILM was trimmed using cutter and an attempt was made to invert residual ILM into the MH (IIFT). At this point the ILM flap came free off the retina and was lost during subsequent manipulation in three cases (Figure 1A). In the fourth case, although this manipulation was successful, the flap was lost while performing the FAE.

In all four cases, the ILM was re-stained using BBG and edges of the peeled ILM clearly identified. A strip of ILM peel was initiated using pinch and peel technique from the already peeled ILM margins either inferior or temporal to the MH (Figure 1B). The peel was continued for three-clock hours to get a long tongue shaped strip of ILM (Figure 1C). This flap was then rotated at its base and gently nudged into the macular hole using the closed jaws of ILM peeling forceps (Figure 1D; Supplemental Digital Content 1). FAE was done in three cases and the air was replaced with 12% perfluoropropane (C3F8) gas. In the case 2, since the transposition was performed under air, a direct gas exchange with 12% C3F8 was done (Supplemental Digital Content 2). Post-operatively, the patients were advised prone position for 5 days.

Results

Of the four patients, two each were males and females, respectively. Age of the subjects ranged from 58 years to 69 years with a mean of 62.75 ± 3.96 years. The mean base diameter of the MH in 4 eyes was 662.25 ± 272.46 μm (Median: 535 μm ; Range: 509 μm - 1070 μm). Duration of the macular hole based on history and clinical records was more than six months in all cases. At two months follow up three cases showed type 1 closure while one case showed type 2 closure of the macular hole with improvement in visual acuity seen in all eyes (Figure 2, 3 and 4 respectively).

Table-1

Clinical profile of the patients

Patient Number	Age (years)	Gender	MHBD	Baseline BCVA	Intraoperative tamponade under which RTF transposition was performed	Type of MH closure	Two-months post-operative BCVA
1	69	Male	552	6/60	Saline	1	6/18
2	62	Male	509	6/24	Air	1	6/18
3	62	Female	518	6/36	Saline	1	6/12
4	58	Female	1070	CF 3M	Saline	2	6/60

MHBD- Macular hole base diameter in microns; BCVA- Best corrected visual acuity; RTF – Rotational Tongue Flap; MH – Macular hole; CF – Counting fingers.

Discussion

Inverted ILM flap technique during FTMH surgery facilitates improved anatomical and functional results in complicated cases, including large macular holes (>400 microns) and secondary macular holes, respectively.¹¹ Yamashita and co-authors categorised extra-large MH as those with diameter >550 microns.¹² In these patients, they compared results using conventional ILM peeling versus IIFT. They reported a closure rate of 88.4 % (38/43 eyes) with conventional ILM peeling and 100% (41/41 eyes) by IIFT, respectively. In our case series, since 2 patients each had large MH and extra-large MH respectively, with a mean diameter of $662.25 \pm 272.46 \mu\text{m}$, it was imperative to have an ILM flap which can be inverted into the MH for better anatomical and functional results.

A lost ILM flap is not an expected occurrence during FTMH surgeries and managing such situations is challenging for the surgeon. Under such circumstances, the usual course of action would be to mobilise another ILM flap which can subsequently be tucked into the hole. Some surgeons additionally use viscoelastic, perfluoro-n-octane liquid, or tissue glue for flap stabilization.^{11, 13} Keeping a free ILM flap inside the MH during FAE is tricky since it is prone to be lost or get displaced, as we encountered in the fourth case. A retinal patch graft covering the macular hole is, yet another technique reported in literature.¹⁴ It involves removing a patch of healthy retinal tissue and placing it in MH using perfluoro carbon liquids. However, it can lead to potential complications such as retinal detachment

and intra-operative bleed as a part of the procedure. The rotational tongue flap (RTF) technique does not involve removal of retina tissue or use of adjuvants such as viscoelastic, glue or perfluorocarbon liquids. The RTF can be easily mobilised during the surgery with minimal learning curve. As the RTF is hinged to ILM bed at one end, the possibility of lost flap or a free flap during FAE is negligible. Additionally, since the ILM inversion is achieved by rotating the flap at its base, there is a greater ILM stability as it is tucked inside the MH. Even in scenarios where flap loss occurs during a FAE, this technique can safely be executed under air tamponade as illustrated in case 4 of our series. A flap hinged temporal or superior to MH is preferable since initiation of flap mobilization is away from the papillomacular bundle and hence safer and less traumatic. Also, RTF fashioned this way is more stable during FAE since it is further from the tip of suction canula. Secondly, it is better to initiate flap mobilisation away from the papillomacular bundle since it is less traumatic. A similar technique has been described by Gekka et al in two cases with refractory MH.¹⁵ Nonetheless, our series focuses on performing this procedure as a rescue technique during accidental flap loss in primary MH surgery. Additionally, we exclusively illustrate creation and successful placement of RTF into the MH under air tamponade. Our preliminary experience suggests that fashioning and manipulation an ILM flap with its transposition is comfortable and undemanding under air tamponade.

Limitations of this technique would include situations where a sufficient length of RTF cannot be mobilised. Secondly, in cases

with a large initial ILM peel, a longer RTF would be needed so that it can reach the centre of the macular hole. Lastly, in recurrent or persistent MH, the residual ILM is usually very friable and raising a RTF could be challenging.

There are few potential indications where this technique can be useful, including persistent (failure to close) MH and recurrent MH where the RTF could be mobilised from the edge of already peeled ILM. Additionally, there is a possibility that this may be considered a primary surgical technique by using one long flap and rotating it into the MH. This would potentially eliminate performing large areas of ILM peel and subsequent chances of disassociated optic nerve fibre layer defect (DONFL) with suboptimal visual results.

In our series, three patients had Type-1 closure and one patient had Type-2, with good functional outcome at 2 months in all eyes. However, a larger series with diverse MH aetiologies and dimensions, and longer follow-up may be needed to confirm the safety and efficacy of this technique.

In conclusion, RTF translocation is a promising technique in a situation of lost ILM flap during IIFT or during a FAE in a FTMH surgery. This procedure is technically simple, requires no additional armamentarium and gives good anatomical and functional results comparable to conventional MH surgery.

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RETINA TECH

Robotic Surgery for Vitreoretinal Diseases – and beyond

Dr. Marc D. de Smet, MDCM, PhD^{1,2}

¹Microinvasive Ocular Surgery Center (MIOS sa), Lausanne, Switzerland

²Preceyes BV, Eindhoven, The Netherlands



The near future of eye care is one where a lot of time and resources will be devoted to treat age-related eye disease, for the simple fact that in general, people are living longer. Turning a medical student into an ophthalmologist takes many years and lots of resource, and then getting them to become one of the most-needed surgeons in the battle against age-related eye disease, a vitreoretinal surgeon, takes longer and costs even more. Clearly, retina surgeons reach the peak of their experience later in their career but then aging calls and starts to add hand tremor, and so the surgeons with the greatest experience have to hang up their gloves. A tragedy, when their skills are so much in demand.

Currently retinal surgery is further pushing the limits of even the steadiest of surgeons' hands. Small tremors or unintended movements in thin, friable retinas (like those with geographic atrophy) can very quickly cause damage, and subretinal gene therapies require relatively large volumes of therapy to be delivered through a small lumen needle to create the bleb in which the therapy goes. Get the wrong location, or have your hands tremor slightly, or even tire when slowly injecting the therapy, and you can have big problems in achieving the desired effect including reflux, formation of a macular hole, retinal tear or hemorrhage. These are all situations where a highly precise, tremor-free robot can assist.

So how might a robot for VR surgeons (or any eye surgeon)

work? You would want it to be compact, able to reach all appropriate targets in the eye while avoiding others such as the lens if we want to preserve them. You could have the robot filter out any tremor in the surgeon's hands, and you could program boundaries preventing an instrument's tip from moving beyond, thus adding additional safety. Most medical robots can provide this, but in ophthalmology, we ultimately need a high level of accuracy and precision and that's something that we've not really seen in other medical robots so far. For example, the Da Vinci system has a precision of around 01 mm.¹The precision we require in the eye is two to three orders of magnitude higher.

Let's discuss accuracy and precision. If we take an analogy from the shooting range, accuracy means how close the shots are to the target, and precision means how close the shots are together (**Figure 1**). The best outcome at the shooting range is when you have both together, and in the eye, when the robot micromanipulator made by Preceyes (Eindhoven, the Netherlands), can achieve an accuracy and precision of around 1-3 μm which is around two orders of magnitude better than surgeons can achieve.²

If robots hold all of this potential, and are at least ten times more accurate and precise than the most stable of human hands, how could we use them in the OR? We, at Preceyes, designed a system that is non-intrusive, where the surgeon remains in control, and which presently only uses a single arm (**Figure 2**).

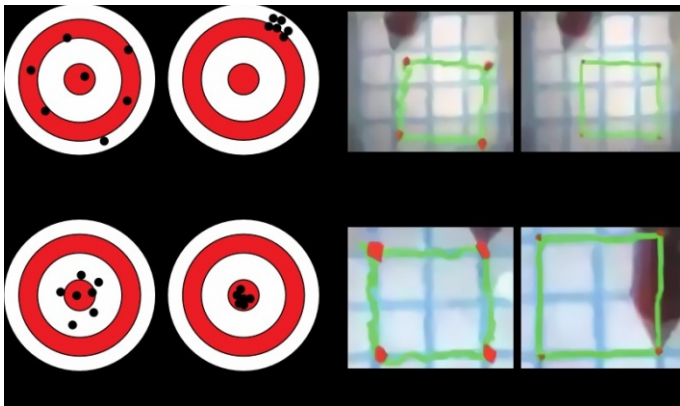


Figure 1 : Schematic example of accuracy and precision (left) and an example of human (top right) and robot-assisted (bottom right) surgical dexterity tests.

The robotic assistant fits on existing tables in the OR, and it uses the existing equipment. Beyond this base setting, increasingly complex systems will be added that will allow us to progress to full robotic assistance over the next decade. Surgeons should view it as a precision instrument that assists them when they really need it for very precise, specific tasks.



Figure 2 : The Preceyes robot affixed to the operating table in use during an internal limiting membrane peel procedure.

To give you an example of how precise the Preceyes robot can be, we have show it cannulating a porcine branch vein with a fine glass cannula (video: <https://youtu.be/rVkhtVsqqO4>).^{3,4} When you watch the video, you will see something new: when the saline is injected, you can see a small air bubble going in

retrograde direction inside the vein, and saline filling up of the whole vascular system. We've done this for up to 20 minutes in pig eyes without having the catheter fall out the robot is solid, and even in patients, can move relative to the patient, again with micron precision, keeping the instruments in the same position in the eye throughout. We have performed over 60 of these procedures without having the catheter break and without requiring vitrectomy. The reason the catheter does not break is that everything from the periphery down to the vessel is performed in a straight line. No torque is being created in the eye, so there is no reason for the cannula to break.

One of the best in vitro assessments of a surgeon's dexterity and skills these days can be shown in simulated subretinal injections (**Figure 3**) using a gelatin model and a Zeiss intraoperative OCT (iOCT). As the video shows, there's always some motion present in the human hand trying to inject, whereas with the robot, you're able to remain in the exact same position without motion

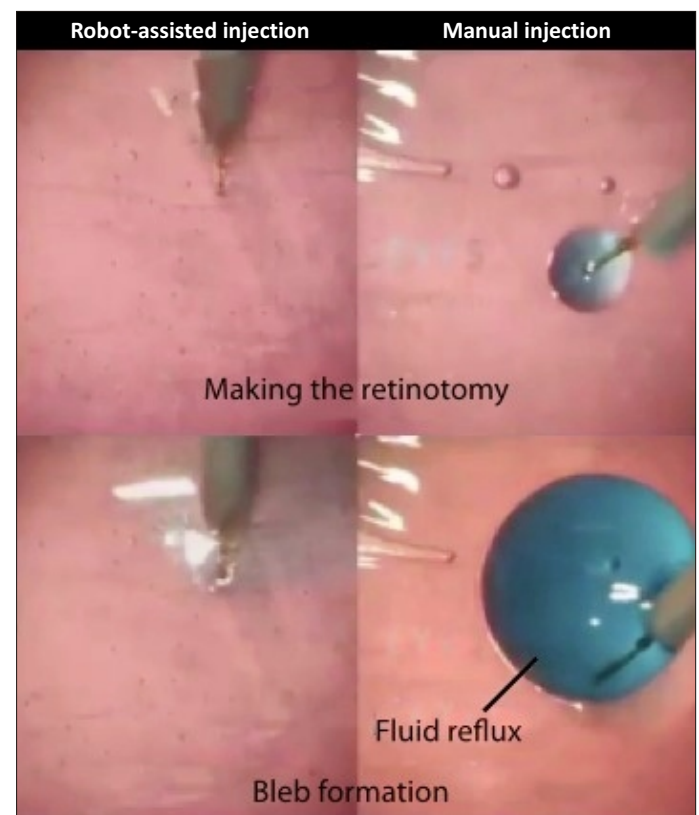


Figure 3 : Representative examples of simulated subretinal injections. The robotic assistant (left panels) makes a leak-free bleb, whereas the surgeon operating manually experiences leakage and fluid reflux at the site of injection.

Table 1. EURETINA 2019 trial of robot vs. surgeon in a simulated subretinal injection model.

	Robot	Manual
Injection time (s)	18–84	12–35
Steadiness of injection (μm) 1–5	16–121	
Drift (μm)	4–58	122–383

and create a bleb. Manually, the procedure was successful in only 44% of attempts, while it was achieved in 88% with the robot. We also found an increased injection time when the injection was performed with the robot (Table 1). Subjects were asked to inject for 30 seconds, rarely achieved manually, but in most attempts with the robot.

The robot was far steadier, less jerky, and induced far less drift that was seen with a manual approach, and the robot was far superior in terms of positioning in the Z-axis too, and a lot less tremor.

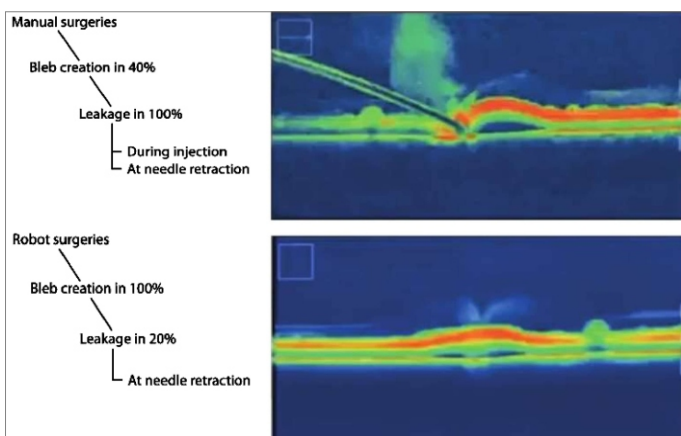


Figure 4 : Zeiss iOCT imaging of bleb creation in live animal porcine eyes: the robotic assistant consistently succeeded in creating blebs (as opposed to a 60% failure rate with the manual approach) and unlike the manual approach, where leakage occurred in all cases during injection and needle retraction, leakage was observed with the robotic approach was only 1 in 5 injections. This approach may have particular advantages with high-cost gene therapies, minimizing wastage (of both gene therapy and costs), while at the same time, maximizing efficacy.

The next step in evaluating the robot was to look at subretinal bleb creation during *in vivo* surgeries in porcine eyes, and to use an iOCT to visualize the degree of leakage during bleb creation and needle retraction (**Figure 4**). In the study, as in real life, manual bleb creation results in leakage, both during injection, and at needle retraction. With the robot however, we saw no leakage at injection, and leakage on needle retraction only 20% of the time. This improvement in precision and delivery has clear benefits for subretinal gene therapy administration: less wasted dose, and more targeted delivery of the therapy, and better localization of gene therapy to the region desired and required to manifest the therapeutic effect. No perforation of Bruch's membrane was observed with the use of the robot (we can stay well away from this layer), while this is less clear manually.

Rotterdam Eye Hospital evaluated the use of the Preceyes robotic assistant in adult patients with vision better than 0.5 logMAR, who required epiretinal membrane surgery. Ten patients underwent robotic surgery; five were operated manually. Multiple surgical steps were performed robotically: dye staining, dye removal, creation of the epiretinal membrane (ERM) flap, ERM grasping and peeling with forceps, and gas and fluid exchange. All steps were successfully completed robotically. Six months after the procedure, the visual acuity in the robot-operated patient group was statistically no different between the two groups

No peri- or post-operative complications were observed in either patient group, although it is worth noting that there was a learning curve associated with the Robot the ten surgeries took longer to perform on average than manual surgery. However, with time, as the study progressed, it took increasingly less time for the robot-assisted surgery to be performed. Some of this was to do with the setup, the level of assistance from the theatre

nurses. It is worth noting that early on in the study, we had placed a boundary on the robot that would prevent the surgeon from moving too close to the retina, at least at his first attempt. However, as the surgeon became more confident in using the robot, by the fourth case, the surgeon decided to remove this boundary, as he felt comfortable in getting close to the retinal surface safely.

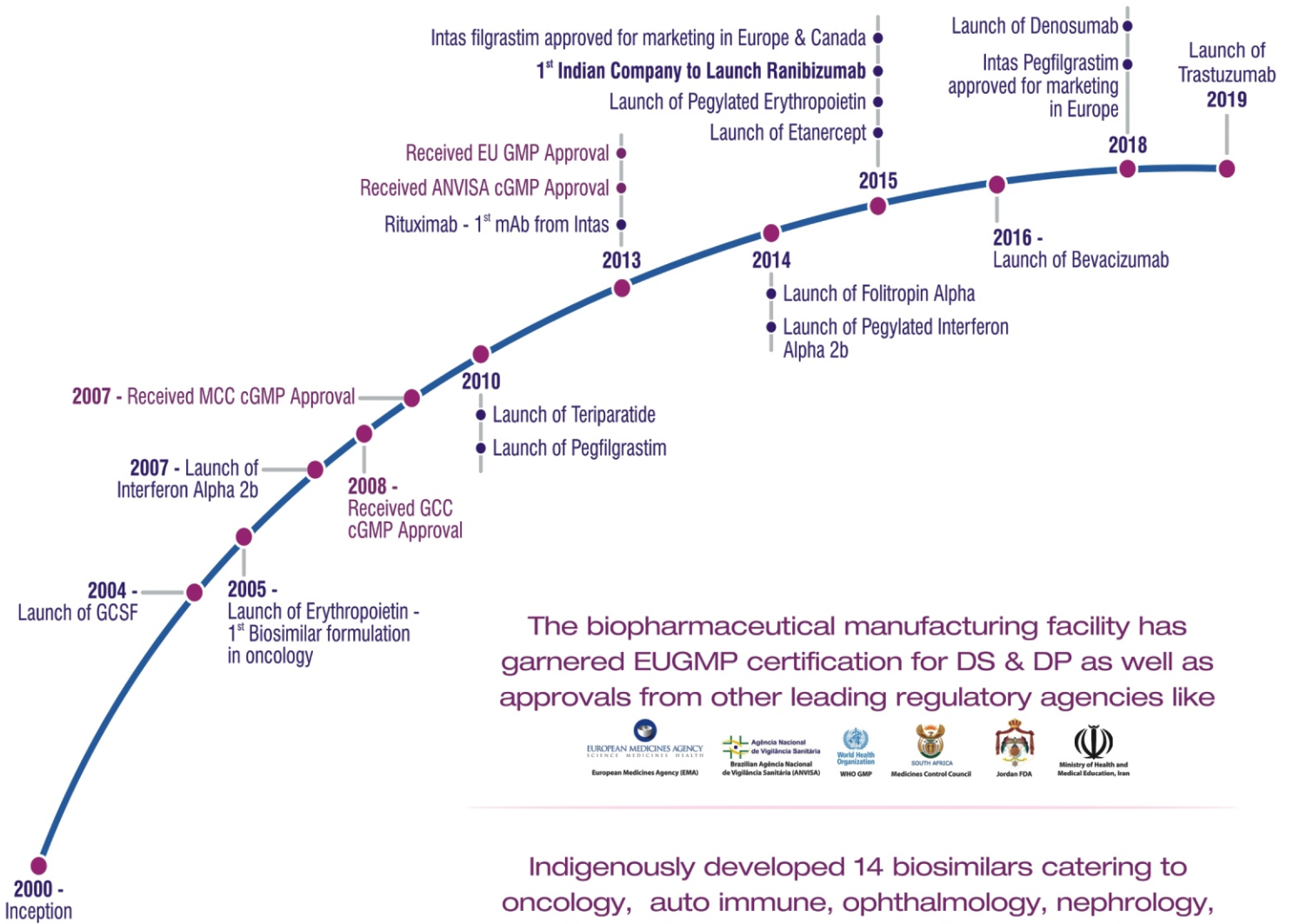
Where are we today? We can confidently state that robotic micro manipulators provide high accuracy and precision, which is essential for retinal surgery, and that the robot can be successfully used a number of retinal surgical procedures. We should emphasize that robotics offers time independence: the robotic arm does not get tired or move, which means that surgeon are under no pressure to complete a task quickly during any procedures. The next steps are to take the robot into ocular surgery applications of course conventional, but given what the instrument is capable of, novel applications too, not limited to the retina. We hope that many of you will contribute to what is certain to become the future of ophthalmic surgery.

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CASE REPORT

Overlapping features in Pediatric retinal disorders presenting as a diagnostic dilemma

Dr. Parveen Sen, M.S.¹
Dr. Krishna Kanta Roy, M.S.¹
Dr. Janani Sreenivasan, M.S.¹

¹Shri Bhagwan Mahavir Vitreoretinal Services,
Medical Research Foundation
Sankara Nethralaya, Chennai

**Corresponding author:****Dr. Parveen Sen**

Senior Consultant, Shri Bhagwan Mahavir Vitreoretinal Services,
Sankara Nethralaya, Chennai

Abstract:

Pediatric retinal disorders like neonatal Familial Exudative Vitreoretinopathy (FEVR), Retinopathy of prematurity and Persistent fetal vasculature can have overlapping features. Neonatal FEVR especially can mimic ROP and PFV due to the variegated clinical presentations. Accurate diagnosis of FEVR is necessary due to less predictable clinical course and progressive nature of disease requiring life-long follow-up. This can be achieved by meticulous examination under anesthesia with/without fluorescein angiography

Introduction

Familial exudative vitreoretinopathy (FEVR) is a rare inherited disorder of retinal angiogenesis, first described by Criswick and Schepens in 1969¹. Most FEVR patients have an avascular peripheral retina but phenotypic expression may be asymmetric and is highly variable, ranging from asymptomatic disease with peripheral retinal avascularity alone to moderate to severe disease with neovascularisation (NV), exudation and fibrosis causing macular traction and tractional retinal detachment (TRD) leading to severe visual dysfunction. These patients can

present at any age depending on the severity of the disease¹. Neonatal FEVR can especially present with retinal folds or complete retinal dysplasia resulting in very poor vision.¹⁻⁴ Phenotypic variability is very common in FEVR, and thus frequently masquerades other paediatric retinal disorders, notably retinopathy of prematurity (ROP) and persistent fetal vasculature (PFV).¹⁻⁵ In this study, we report two cases, one referred as ROP and other PFV; later unmasked as FEVR by high index of suspicion and careful examination under anaesthesia (EUA).

Case reports**Case 1**

A 4-month-old male child born via normal delivery at 28 weeks of gestation with a birth weight of 0.6 Kg, was referred for ROP management. The child was admitted in a neonatal intensive care unit (NICU) for 45 days after birth, due to very low birth weight and received supplemental oxygen. EUA revealed shallow anterior chamber, posterior synechiae, early ectropion uveae in both eyes (OU) with peripheral lens opacity in right eye (OD). Fundus examination revealed midperipheralsubretinal

exudates and a peripheral retinal fold going anteriorly upto the nasal lens causing a localised retrolental fibroplasia in OD. Left eye (OS) fundus revealed minimal vitreous haemorrhage, temporal dragging of disc and macula, straightening of temporal vessels, and flat NV at the ridge in temporal quadrant with fibrosis (**Fig-1**). Presence of disc drag and peripheral straightening of vessels in a 4-month-old child specially with midperipheralsubretinal exudates raised a suspicion of FEVR even though there was a history of prematurity. Parents also gave a history of consanguineous marriage. Both eyes underwent indirect laser photocoagulation (ILO) and intravitreal Bevacizumab (0.025ml) and Belt buckle (#240) application to

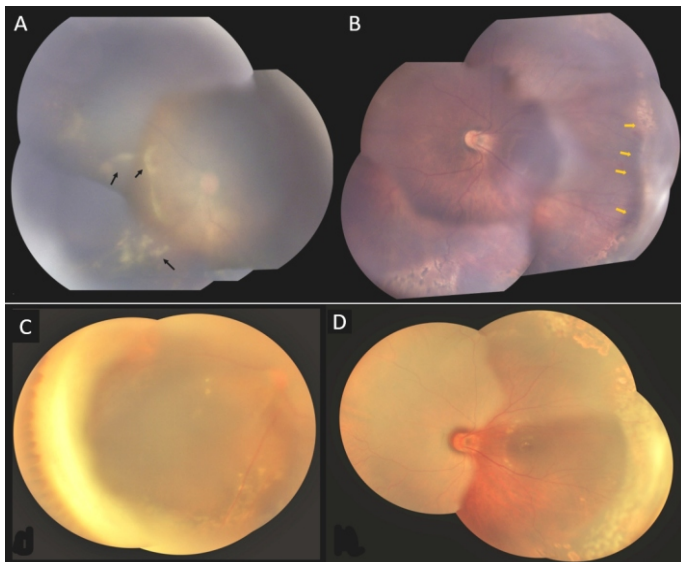


Figure 1 : Fundus images of a 4-month-old male child referred for management of ROP. Right eye fundus examination (Retcam, Clarity MSI, California) revealed avascular retina and hard exudates (black arrow) at mid periphery (A). Left eye had temporal dragging of disc and macula, straightening of temporal vessels, flat NVE at the ridge in temporal quadrant (yellow arrow) with fibrosis and peripheral avascular retina (B). Post treatment images showed reduced hard exudates in right eye (C) and resolved neovascularization in left eye (D).

reduce the peripheral traction. At 1 month post surgery the child was seen to have regression of NV and stabilisation of the TRD. Child was seen at 2 month follow up when new areas of exudation (suggestive of disease progression) were seen and further laser was done. In keeping with the progressive nature of the disease a diagnosis of FEVR in a premature child or “ROPER” was kept in mind.

Case 2

An eight-month-old male child born at 34 weeks of gestation with birth weight of 2.5 kg and stormy neonatal period including need for ventilator support, was referred as a case of OS PFV for further management. Parents noticed leukocoria in OS since the third month of age. EUA revealed bilateral normal corneal diameter with a refraction of -9.00 DS in OU. OD had early peripheral lenticular opacity not involving the visual axis and OS had localized nasal posterior synechiae with a thick retrolental fibrous membrane (RLF) obscuring fundus view. Fundus examination of OD revealed healthy disc and macula, minimal straightening of arcade vessels, hard exudates at mid periphery and attached retina (**Fig-2**). OS retinal details were hazy due to the RLF. The child underwent lensectomy, excision of RLF and vitrectomy in OS. After removal of the RLF a nasal falciform fold with severe dragging of disc and macula and few mid peripheral subretinal hard exudates were seen. Fluorescein angiography (FFA) under general anaesthesia using Retcam (Clarity MSI, California) revealed 360-degree peripheral avascular retina in OD with leaking NV at inferotemporal and superotemporal quadrant pointing towards a possible diagnosis of FEVR. In keeping with presence of proliferative retinopathy, ILO was done in OD to the peripheral avascular retina. OS was rehabilitated post surgery with contact lens and patching of OD. After 2-months, OD had regression of NV and OS had nasal falciform fold and rest of retina attached.

Discussion

The diagnosis of FEVR is straightforward when fundus examination reveals peripheral retinal avascularity and radial fold with subretinal hard exudates in a person born at full term with a positive family history.² This diagnosis may be confused with ROP if the clinical and FA features show peripheral avascularity, NV with leaking retinal new vessels in a premature child. It can mimic PFV if there is presence of retinal fold extending from optic disc radially to the periphery and anteriorly to the ciliary processes in a very young infant.² ROP tends to follow a predictable timeline and disease itself does not progress or recur in childhood or adulthood though sequelae may be seen.² By contrast, FEVR has an unpredictable course and it can progress/recur at any age, thus necessitating life-long follow up and treatment.¹⁻⁴ A small subset of premature infants who exhibit retinal findings more characteristic of FEVR than ROP with a progressive course have been described as ROPER (ROP vs. FEVR), by Berrocal et al.⁷ These eyes behave more like FEVR, with less predictable and long-term progressive disease requiring

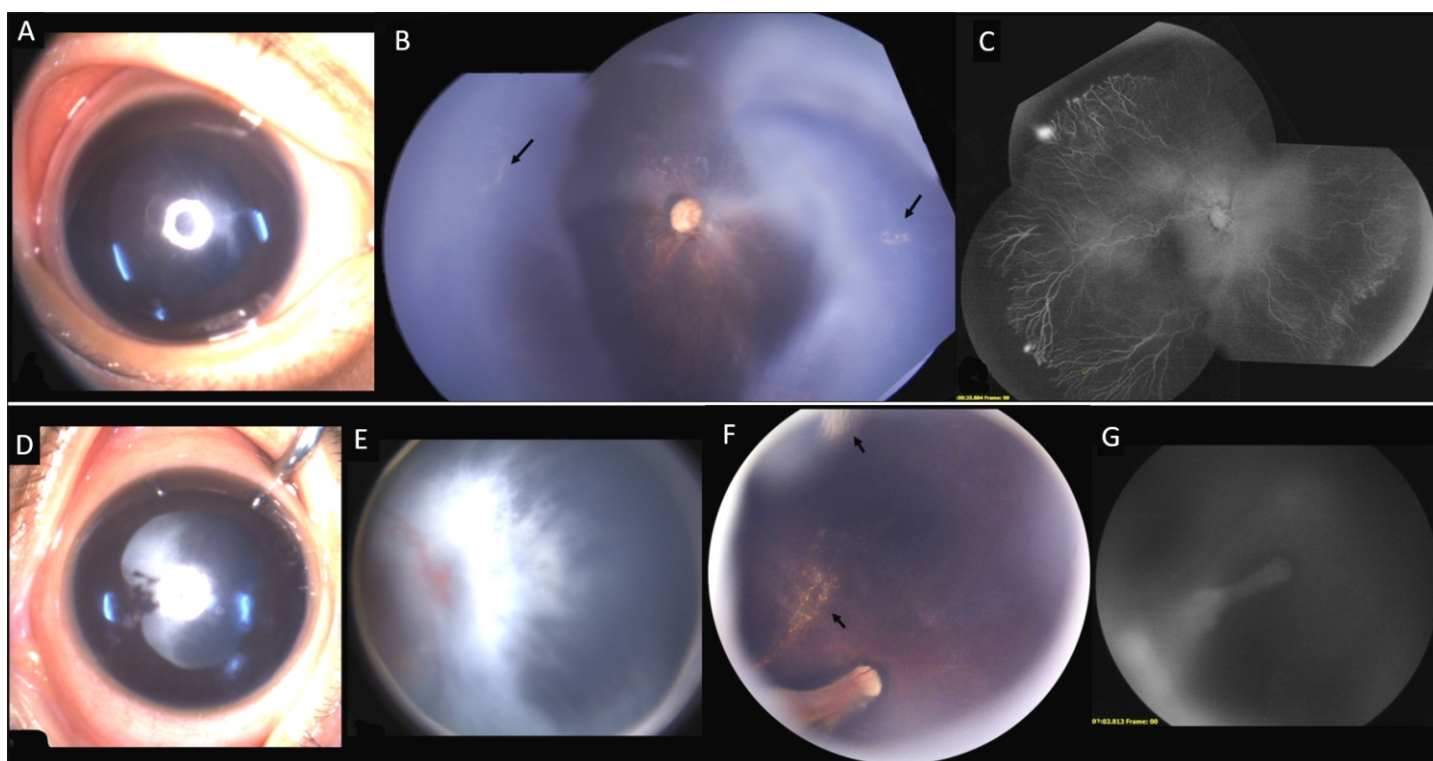


Figure 2 : Retacam images (Clarity MSI, Pleasanton, California) of a 8-month-old male child who presented with leukocoria. Right eye had peripheral lens opacity (A) and fundus showed straightening of arcade vessels, hard exudates (black arrow) at mid periphery (B). Fluorescein angiography revealed 360-degree peripheral avascular retina with pruning of the retinal vessels and leaking neovascularization at inferotemporal and superotemporal quadrant(C). Left eye had lenticular opacity (D) and thick retrolental fibrous membrane(E). After lensectomy and retrolental fibrous membrane removal, fundus examination revealed nasal falciform fold and hard exudates (black arrow) at mid periphery (F,G).

close continuous follow up and multiple treatment sessions⁷. In the first case described here, parental consanguinity, asymmetric disease in both eyes, presence of hard exudates, straightening of vessels, dragging of disc and macula along with mismatch between the clinical findings and age of the patient at presentation, all point to a diagnosis of FEVR. ROP in a premature child by 4 months would have presented as Stage 5 or cicatricial disease and not as a progressive retinopathy. Here, prematurity was a confounding factor and probably these patients belong to the “ROPER” subset.

PFV is a congenital ocular anomaly in which the embryonic hyaloid vasculature network fails to regress partially or completely.¹⁰ It can be anterior PFV presenting as cataract or retrolental opacity or posterior PFV presenting as a thick vascular stalk arising from the optic nerve, a retinal proliferative membrane, a retinal fold, RD, or optic nerve hypoplasia¹⁰ or combined anterior and posterior types. Unilaterality (bilateral in very rare cases) and microphthalmos are important associated

findings especially of posterior PFV.¹⁰ Due to the phenotypical variation and asymmetry, it's quite possible for a patient to have advanced FEVR findings in one eye and grossly normal other eye, mimicking PFV.¹⁻⁵ Unlike the falciform fold in FEVR, the stalk in PFV is usually not a retinal fold but a hyaloid stalk of persistent tissue that extends from optic nerve to posterior lens surface with varying degrees of retinal dysplasia.⁵ In the second case, bilateral asymmetric disease, subretinal hard exudates, clinical and typical angiographic findings in the less affected eye points more towards the diagnosis of FEVR. This case highlights the importance of FA, which confirmed the peripheral avascularity and picked up subtle NVEs in the less affected eye, thereby guiding laser treatment; crucial for maintaining vision in the better eye of the child.

Conclusion

These two cases presented here, bring to forefront the varied presentation of FEVR as well as overlapping clinical

presentations of pediatric retinal diseases like PFV, FEVR and ROP. It is important for the treating clinician to be aware of these varies presentations and do a meticulous examination under anaesthesia with or without FA wherever possible of both eyes and guide the patients accordingly. This is especially relevant in countries like India where, genetic testing of all cases may not be possible due to financial constraints as well as lack of universal availability of these tests. Along with lifelong follow up and treatment of retinal pathologies, examination of family members, and discussion regarding genetic testing and counselling should be a part of management of FEVR.

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Notes

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The CONSTELLATION® Vision System is an ophthalmic microsurgical system that is indicated for both anterior segment (i.e., phacoemulsification and removal of cataracts) and posterior segment (i.e., vitreoretinal) ophthalmic surgery. The ULTRAVIT® Vitrectomy Probe is indicated for vitreous cutting and aspiration, membrane cutting and aspiration, dissection of tissue and lens removal. The valved entry system is indicated for scleral incision, cannulae for posterior instrument access and venting of valved cannulae. The infusion cannula is indicated for posterior segment infusion of liquid or gas. **Warnings and Precautions:** The infusion cannula is contraindicated for use of oil infusion. Use of disposables and handpieces other than those manufactured by Alcon may affect system performance and create potential hazards. Attach only ALCON® supplied consumables to console and cassette luer fittings. Do not connect consumables to the patient's intravenous connections. Mismatch of consumable components and use of settings not specifically adjusted for a particular combination of consumable components may create a patient hazard. Vitreous traction has been known to create retinal tears and retinal detachments. The closed loop system of the CONSTELLATION® Vision System that adjusts IOP cannot replace the standard of care in judging IOP intraoperatively. If the surgeon believes that the IOP is not responding to the system settings and is dangerously high or low, this may represent a system failure. To ensure proper IOP Compensation calibration, place infusion tubing and infusion cannula on a sterile draped tray at mid-cassette level during the priming cycle. Leaking sclerotomy may lead to post-operative hypotony. Refer to the CONSTELLATION® Vision System Operators Manual for a complete listing of indications, warnings, and precautions.

DME*



Ozurdex[®]

(dexamethasone intravitreal implant) 0.7 mg



*For the treatment of adult patients with visual impairment due to Diabetic Macular Edema (DME) who are considered unsuitable for, or insufficiently responsive to, non-corticosteroid therapy or are pseudophakic

Allergan India Pvt Ltd

Level 7, Prestige Obelisk No 3; Kasturba Road; Bengaluru; Karnataka; INDIA 560 01
Tel: +91-80-40707070; Email: IN-Allergan@allergan.com; Website: www.allergan.co.in

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