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DOS Times

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A Bulletin of Delhi Ophthalmological Society

Posterior capsule Rupture

IOL Master: Mastering the Master

Malignant Tumors of Lid

Amblyopia

Ectatic Corneal Disorders

Keratoprosthesis

Special National Issue

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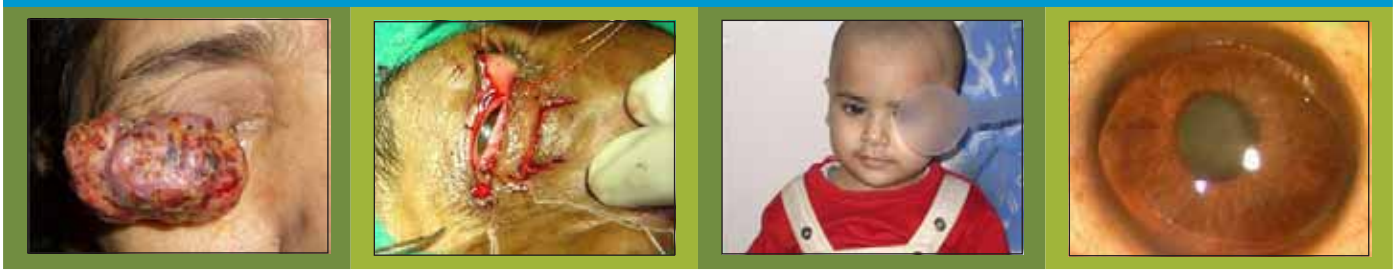

Attention DOS Member

DOS Times is not published in the month of May & June each year

Editorial

My Dear Friends and Colleagues,

As this year draws to a close and the next one blinks on the horizon, I look back with mixed feelings. My endeavour throughout the first year of my tenure, has been more connectivity. I thought it would be a breeze! But it has turned out to not so easy.

My focus was twofold:

Enhanced connectivity between the DOS office and members. Networking between the members themselves. I envisioned a society, where all the members are approachable to each other and can share their thoughts and discuss their problems in a jiffy. And this was workable, doable. I set out to do just that. And so DOS came on facebook. We have uploaded the material; to make a start. And we have send out invites to all the members whose emails we have. Many have joined. A beginning has been made. We have kicked off, but all of you have to take the ball forwards.

Forums and Discussions and Chatting –a big DOS family, all out there. Live conferencing, real time problem solving and leisure. The future is out there. And this is our own Pitch; our own game to play. This is our profession, our hobby, our passion. Let us make it big.

A Penny For Your Thoughts:

Progress occurs when energy and enthusiasm join hands with experience. For young and computer savvy residents; this is my request to you. Give us more ideas and help us make them a reality. Ophthalmology is a high technology speciality and let us explore, how we can use real time multimedia communication to enhance our skills and enrich our lives.

Conferences, Workshops and Skill Transfer.

We really need your feedback. I firmly believe that fresh ideas and innovative paradigms are urgently required. DOS must be on the vanguard of ground breaking protocols in conferences and suchlike. We must never be stagnant. Must always move forwards like the mountain spring. Fresh and Fast.

So put on your thinking caps friends and give us some new breaks. May be you can do it while watching World Cup Soccer. Watching Cricket has become such a bore!

Yours Truly.

Thanking you,

Dr Amit Khosla
Secretary,
Delhi Ophthalmological Society

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Management of Posterior Capsule Rupture

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Posterior Capsular Rupture, (PCR) continues to trouble phaco surgeons despite advancements in skill and improved fluid dynamics of the machines. Barring few instances like posterior polar cataract, surgeons cannot afford to counsel patients for PCR preoperatively. This makes PCR an unwanted and unpredictable accident during cataract surgery that takes away the "wow" factor from postoperative experience. Even if patient escapes CME, Astigmatism and other complications, the quality of vision is often suboptimal. All measures should be taken to reduce incidence of PCR, however the incidence can rarely be zero. So the issue frequently raised after PCR is whether it was managed appropriately. Here, by posing questions to experts in the field we make an effort to clear few controversies in the management of PCR.

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DVS: What is the first step you do once a PCR is noted?

ARV: First I would stop performing my surgical step and inject dispersive viscoelastic, Viscoat from the side port, before retracting the instrument out of the eye. Viscoat maintains the contours of the globe and prevents / reduces vitreous prolapse.

HL: The most important step on noting a PCR is *not to withdraw the phaco probe immediately* in haste. With the infusion on, one should take visoelastic in the non dominant hand and inject it through the sideport to *inflate the bag* and then only withdraw the phaco probe. If one withdraws the phaco probe in haste, the anterior chamber will collapse, the PCR may enlarge and the hyaloid phase which may not yet be disrupted will break leading to prolapse of the vitreous in the anterior chamber and/or prolapse of the nuclear fragments in to the vitreous cavity. The next step is to confirm the presence of the PCR and assess it's extent and the integrity of the hyaloid phase.

SKK: Stop the procedure, with draw the hand piece, fill the AC with Viscodisperssive OVD and assess

DVS: Your experience on converting a PCR into PCCC?

ARV: I would consider converting a PCR into a PCCC if the rent is small. Before I convert I make sure that there is no vitreous herniation through the PCR. I fill the chamber with Healon GV or Provisc and try to keep the central posterior capsule flat or slightly concave. I prefer using a Microforceps and crisp focusing and frequent grasping and regrasping of the flap is useful. I try to restrict to a small size.

HL: Converting a PCR into a PCC is ideal, but most of the times impractical. Conversion to PCC is possible only if the PCR is small and central and the hyaloid phase is nearly intact. I have found it difficult to convert peripheral PCRs into PCC and the presence of vitreous in the anterior chamber increases the difficulty. In linear PCRs one faces problems as first one has to create a flap and then do PCC. Also, if the

hyaloid phase is intact, risk of disrupting it while attempting PCC is very high. If one wishes to do a PCC in the presence of a disrupted hyaloid phase, then minimal controlled vitrectomy is essential prior to attempting PCC. *One should attempt conversion to PCC only when one has a small central/ paracentral PCR (not more than 3 mm), good viscoelastic, excellent capsulorrhexis forceps and a good operating microscope. However the failure rate is still very high.*

SKK: It can be done esily in case the vitreous face is still intact, do not try otherwise as you can pull on the vitreous. it is difficult in cases of high myopia, post VR surgery and pediatric cases. I use Utrata forceps, hold the tear at the base of the flap and slowly rhex after confirming the grip on PC. Try to lift the flap towards the centre of cornea and do not press down wards.

In case there is frank vitreous in AC, then the same can be done after performing vitrectomy

DVS: How do you confirm hyaloid phase is intact or broken?

ARV: I inject preservative free triamcinolone (Aurocort) to stain the vitreous. Vitreous in an intact face will appear as a prolapsing uniform surface as opposed to a broken face where the vitreous will be seen dispersed with white particles.

HL : If one observes the borders of the PCR and if they appear to be shiny/golden, or if they appear to be under stretch or rolled up in the presence of irregular chamber depth, then the chances are that the hyaloid phase has been disrupted. Also, restricted movements of the nuclear fragments and/ or entanglement of the nuclear pieces with vitreous will point towards a broken hyaloid phase. One can do the following test.

- ***Sponge test:*** One can sweep a sponge along the incision to detect the presence of vitreous.
- ***Sweep test:*** One may also try to sweep the spatula from the anterior chamber angle under the incision towards the PCR, vitreous, if present, will be seen dragging in (due to tendency of the vitreous to come towards the wound).
- ***Halo test:*** Put viscoelastic in the bag, to flatten it or keep it slightly convex but not concave to elicit the halo test. Try to look for the ring reflex by applying the pressure in the center of the capsule with the help of a rounded repositor. If the PC is intact, a halo will be seen which will vary in size depending upon the amount of pressure applied. This ring reflex will be broken at the site of PCT.
- ***Stain test:*** One can also inject Triamcilone into the anterior chamber just adjacent to the PCR (not above it as it will fall back into the vitreous cavity), vitreous will stain. However, when one is in doubt one should consider the hyaloids phase as disrupted.

SKK: The Ant Rhexis margin and pupillary shape gets distorted with sudden deepening of AC. The magins of the PC rent gets pushed and the PC rent size increases. In cases of fluid vireous, where you may not see all the above mentioned signs a gentle sweep by a fine spatula /iris retractor through the side port under the main incision can be helpfull

DVS: How to manage PCR with full nucleus or nucleus fragment?

ARV: Aim is to prevent the nucleus or nuclear fragment from dropping into the vitreous. I inject Viscoat over the PCR and then retract the instrument out of the eye. I then perform limbal vitrectomy, using the irrigation and vitrectome handpieces separately. Once the vitreous strands are severed, I resume phacoemulsification with very low bottle height. I emulsify the remaining fragments with a low bottle height, moderate aspiration flow rate and vacuum and supra-optimal ultrasound. I keep a constant check on the vitreous, in which case, aspiration hand piece is replaced by vitrectomy probe. Everytime I withdraw my irrigation, I inject Viscoat from the paracentesis to prevent forward bulge of the iris-lens diaphragm.

HL : If the hyaloid phase is intact, its safety and that of the cornea is the key. This depends upon the hardness of the cataract and the chamber stability. In the presence of non-leaky wound, good bottle height (don't lower the bottle height), adequate to high power settings with lower fluidics parameters (to avoid surge) are required.

Supra capsular or anterior chamber phaco should be done, after prolapsing the nuclear fragment from the capsular fornices mechanically or by phaco probe. One must coat hyaloids phase with dispersive viscoelastic like methyl cellulose or chondroitin sulphate. As phacoemulsification is done closer to the corneal endothelium the hard cataract may damage.

In cases of vitreous in the anterior chamber and particularly if the PCR is large, first step is to ensure that the nuclear fragments will not fall into the vitreous cavity. After inflating the bag with visco, I use the chopstick technique, two instruments, one sinsky and one chopper, both instruments can be put through the main port or one through the main port and the other through the sideport, then I hold the nuclear fragments between the two instruments and bring them out of the capsular fornices and place them near the anterior chamber angle. Once the nucleus has been stabilized, one can proceed with vitrectomy and if the nuclear fragment is small and soft, one can do anterior chamber phaco.

In case of a larger nuclear fragment, I enlarge the size of the incision to the size of the nucear fragments- if the size of the remnant is < ½ the nucleus, the incision size is enlarged to 4 mm, if the size of the remnant is ½ the nucleus, the incision size is enlarged to 5-6 mm, in case of a full nucleus, the incision size may be enlarged to 9-10 mm. In case of the whole nucleus, one should abandon the phaco incision, make an ECCE incision at another site and using the chopstick technique remove the nucleus manually. Breaking the nuclear fragments into smaller pieces manually between two instruments can also be done. I prefer to enlarge the incision to the size of the nucleus. However the chamber depth must be maintained throughout with viscoelastic.

SKK: With full nucleus I will convert to ECCE, and with nucleus fragment left and vitreous in AC I use visco disperssive ovd

for compatmentalization of the AC and plugging the rent and proceed with Phaco on low and slow mode. Reduce the bottle height to 80, the flow rate to 25. always look out for vitreous plugging the tip and causing occlusion. In case there is frank vitreous in AC, vitectomy is a must after securing the fragment in the AC or by removing it by forceps

DVS: How to manage PCR with epinucleus?

HL : Epinucleus is toughest to manage in cases of PCR & most often, I have seen epinuclear plate falling into the vitreous cavity. Nucleus being firm can be held mechanically by the instruments, or by the phaco probe. Epinucleus, being soft, cannot be held by any instruments. It cannot be held even with the phaco probe because it bites through.

If the hyaloid phase is intact then the epinuclear plate can be removed easily either by phaco or irrigation and aspiration. If the vitreous is present in AC, one needs to do limited vitrectomy (if vitrectomy is done excessively in a large PCR, the epinuclear plate may fall back). I usually try to prolapse the epinuclear plate out of the rrhexis margin by a rounded repositor. Once it is in the anterior chamber, I put my viscoleastic cannula under the epinuclear plate and start injecting methylcellulose by pressing the posterior lip of the wound, maintaining the chamber depth all the time; epinuclear plate flows out of the wound along with the viscoelastic. I call this method viscoexpression. Some of the epinuclear plate may also come out while doing IA for cortical matter.

In my experience the epinuclear plate causes less inflammation & post-operative complication in the capsular fornices than in the vitreous cavity. Therefore, I will rather leave the epinuclear plate, than be more aggressive & drop it into the vitreous cavity. If, however, the epinuclear plate is present superiorly, an attempt should be made to remove it, as it may slip into the pupillary space. Secondary vitrectomy with cortical removal can be done through parsplana if it obstructs the visual axis.

SKK: This will be same as nucleus fragment management. but with low phaco power

DVS: How to manage PCR with cortex?

ARV: The principle remains the same. I inject Viscoat over the PCR and then retract the instrument out of the eye. I perform bimanual limbal vitrectomy. Once there is no vitreous in the plane of the capsular bag I would use bimanual approach with the irrigation and aspiration handpieces separately introduced through the limbal paracentesis for cortex aspiration. I avoid using the main incision. Before withdrawing the hand pieces I inject Viscoat to maintain the anterior chamber depth.

HL : As for as possible, the cortex needs to be removed, it is not very difficult. In case of PCR, certain areas (cross-incisional) areas may not have any vitreous, that part of the cortex can be handled with ease. Best method for handling the cortex in presence of PCR is with vitrectomy cutter. Cut the vitreous & then go to suction mode & now it can be used as an irrigation / aspiration.

Secondly, if one does not have a good cutter / vitrectomy is not working then the I/A system can be used for cortex removal only in areas where there is no vitreous, another sideport may be made for this. If vitreous is in anterior chamber, *mechanical suck & spit* is a better controlled system than irrigation and aspiration. In a well-formed chamber with a visocleastic, with a 27 gauge cannula go underneath the capsulorrhexis margin, hold the cortical fibres & do not try to suck these, instead pull them out of the incision & then spit them out. Take the cannula back, & repeat the same process. Sometimes the spitting can be done in the AC & can be then removed later. While sucking if the vitreous if caught, spit the cortical matter within the eye, which can be washed afterwards. Sometimes, cortical matter may be removed after insertion of IOL with viscoelastic. Better to leave the cortex behind if vitrectomy cutter is not available.

SKK: Dry aspiration with multiple applications of OVD is my first option if vitreous face is intact.

In presence of vitreous I use my vitrectomy probe. the trick is to perform vitrectomy by using 'Cut –IA mode " and sucking out the cortex out while using 'IA-Cut mode'. This way I avoid the traction at the vitreous and also save my Anterior capsule.

DVS: Tips to insert IOL in presence of PCR & choice of IOL?

ARV: I prefer to use an injector over a holder-folder forceps. I prefer to use an AcrySof IOL for a small rent as it unfolds very gently into the bag. If the rent is large, I make sure there is a circumferentially uniform anterior capsule support. In such a case I would implant a three piece IOL with round optic edges in the sulcus. For sulcus fixation the IOL power has to be reduced by 0.5 diopters from the power calculated for in-the-bag fixation. If the anterior capsule support is inadequate I would prefer scleral fixation of a PMMA IOL.

HL : *If the hyaloid face in intact or the PCR is small with a disrupted the hyaloid phase is intact, place the IOL within the bag. When you are implanting the lens in the bag, some precautions need to be taken:* One can tie a 10-0 thread to the trailing haptic, which can be used for the retrieval of the IOL, if it starts sinking. Dilation of the IOL should be avoided, if it gets entangled with the vitreous, it can enlarge the PCR. IOL should be placed in such a way that even if the tear gets extended, the long axis of the IOL should be at 90^0 to the long axis of PCR / or axis of extension of PCR. The haptics should be away from site of extension of the tea

If in doubt, put the IOL in the sulcus over the CCC margin. Take a sinsky hook/dumbbell dialer, pull one of the haptics towards the center well beyond the CCC margin, then guide the haptic underneath the CCC margin, the haptic will move into the capsular bag, then remove the sinsky hook. If need be, second instrument can be used (rounded repositor) which goes on top of the haptic & underneath the rrhexis margin to guide the haptic into the bag.

I prefer to use the foldable lens. If centration of IOL is doubtful with CCC ideal in size 4-5 mm, shape and centration optic can be prolapsed out of the CCC while

haptics remain in the bag. (Reverse optic Capture). If the CCC is not very large the 3 piece foldable lens can be placed over the sulcus. If CCC is ideal – the optic can be prolapsed into the bag. Usually single piece lens should be avoided, as it can cause lot of iris chaffing.

If the PCR is associated with CCC margin tear, it is better to put the ECCE IOL (big optic IOL) for better centration Even doing secondary implantation of the IOL in these situations may be a good idea. After 6-12 weeks, the capsule really forms up, reassessment & exact IOL choice becomes easier. ½ dioptre less power is required if lens is to be placed in the sulcus.

SKK: In case PCCC was possible I'll use single piece Hydrophobic Acrylic IOL in the bag In such situation I will try to insert the leading haptic into the bag and then dial the trailing haptic in the bag. aim is to avoid pushing of leading haptic into the vitreous. But if PCCC is not available. I use 3piece acrylic or PMMA IOL in the sulcus, Here also try not to push the IOL in one go but put thr leading haptic over the ACCC and slowly dial the trailing haptic. The trick is to keep the IOL parallel to the Iris plane and vertical to it.

DVS: Indication for referral to posterior segment surgeon?

ARV: I routinely refer all cases of PCR to the retinal surgeon in the immediate postoperative period, i.e. within 1 week for a peripheral retinal and macular evaluation. They also have a repeat follow up after few weeks.

HL : Indications – lack of good vitrectomy machine, impending nucleus drop, nucleus/large fragment of nuclear/ epinuclear plate and/or IOL drop.

SKK: Any piece going behind the PCCC in absence of vitrectomy machine

DVS: Management of zonular dialysis without PCR?

ARV: I inject Viscoat over the area of dialysis in an attempt to avoid vitreous prolapse. I form the bag and anterior chamber by injecting sodium hyaluronate (Provisc). I then introduce a pre-threaded capsule tension ring to form the contours of the capsule bag. This stabilizes the bag and stretches it so that it does not get inadvertently aspirated. If the area of dialysis is $\geq$ 2 clock hours I prefer to use the Ahmed's segment, as it allows localised suturing of the bag to the sclera without compromising its integrity.

HL : In the management of zonular dialysis without PCR, Cionni's ring is better as compared to CTR ring as the pull on the scleral suture can be used to adjust & center the IOL. If Cionni's ring is not available, CTR can be used. It is better to use CTR in all case of zonular dialysis irrespective of the size of the dialysis.

If zonular dialysis is $>180^0$, double Cionni's ring should be used, if the zonular dialysis is between 180-90°, single Cionni's ring can be used, if the zonular dialysis is <90°, a CTR can be used. If zonular dialysis $<60^0$ (1 clock hour) placing the long axis of IOL at the site of zonular dialysis can be done. Large size multi piece PMMA lens (lens with good memory) is considered to be better option.

SKK: If the Dialysis ia less than 3 clock hours I use CTR. If it is more than that between 90 to 210 deegrees I prefer using Cionni with single scleral fixation. More than that but less than 300 degree I use Cionni with double scleral fixation using Prolene sutures.

DVS: Management of zonular dialysis with PCR?

ARV: I prefer scleral fixation of the IOL. I do not attempt to implant the capsule tension ring fearing its drop into the vitreous. An exception to this is when the PCR is small and central and has been converted to a PCCC.

HL : Presence of PCR with zonular dialysis complicates the situation. The chances of decentration and dislocation of IOL are very high. Secondary IOL may be a better option after 4-6 weeks of primary surgery. By that time, the vitreous can be identified better & the remaining capsule can be identified well. However, if the PCR is small, not >3-4 mm, then it is possible to consider this case as one of zonular dialysis – one can place CTR/cionnis & foldable IOL inside the bag & before placing the lens 10-0 suture can be tied to the haptics for retrieval in case of dislocation. If the PCR is large, & zonular dialysis is small, then lens can be placed in the sulcus with/without optic capture in the CCC, depending upon size and centration of CCC. If the PCR is large & zonular dialysis is large, then, large optic size lens can be placed into the sulcus. After vitrectomy, if lens is found decentered >1.5 mm, then it is advisable, to do scleral fixation of the lens or retroiris fixation of the ECCE IOL or specifically designed IOL. If one is not confortable with the above techniques, one can use ACIOL. Though in my opinion, if surgeon is not comfortable with scleral fixation of the IOL, he is better off with secondary implantation of the IOL.

SKK: Although I have manged few cases with CTR. It should be used only if you have intact ACCC and have been able to convert your rent to PCCC. In majority of cases if dialysis is more then 4 clock hours in presence of PCR I remove the entire bag and plan for AC IOL or SF IOL

DVS: What post-operative procedures are done as follow up in PCR cases?

ARV: Frequent retina evaluation for retinal breaks, detachment or cystoid macular odema, periodic IOP measurement as it could rise due to persistence of vitreous, inflammation or pigment dispersion especially in cases of sulcus fixated IOLs.

In case of nuclear drop do you insert iol or leave it for 2nd surgery?

Yes I would implant an IOL in all cases. Before implanting I ensure adequate anterior vitrectomy with no residual strands of vitreous in the pupil, anterior chamber or incision. With increased sophistication and advances in technology, posterior segment surgeons can fragment even a very dense nucleus through the pars plana approach.

HL : The chances of corneal oedema, AC inflammation, secondary glaucoma, CME, decentration of IOL & late RD, are comparatively higher after PCR.

For corneal oedema, one needs to give hypertonic saline. I prefer to keep the pupil mobile in case of uveitis, besides the standard treatment of steroids. In cases of PCR, as visco removal may not be complete, which can cause rise in IOP which can last for a week or two, so keep the IOP under control. If IOP is high after 1 week, it is a matter of concern, then steroid responsiveness of the patient should be looked into. If BCVA after 2 weeks is not good, then one needs to rule out CME, more so in diabetics & if vision starts deteriorating after 2-3 weeks, then consider chronic CME. Using investigations like FFA & OCT & suitable management strategies have to be adopted. If vision does not improve after 6-12 weeks, then intravitreal tricot & Avastin may be considered. If progression is very fast, one needs to rule out shallow RD. The patients need to be followed up regularly for development of any retinal tears. The patient should be educated about the 4 'Fs' flashes, floaters (sudden appearance), falling of vision & field loss for early detection and management of retinal detachment.

SKK: No special procedure is done.

DVS: In cases of nucleus drop do you insert IOL or leave it for 2nd surgery?

HL : In case of nucleus drop, one needs to refer the case to a posterior segment surgeon. Whether to put the IOL or not depends upon the competence of the VR surgeon, as majority of the cataract can be removed with a fragmentome except for the very hard cataracts which need to be removed from the anterior route. IOL can be placed in majority of the cases provided if there is enough of capsular support for the IOL to be well centered in the sulcus and/or bag. ACIOL should be avoided.Also, the anterior segment surgeon should have facility for anterior vitrectomy.

Thus, no IOL in cases where IOL cannot be centered, no ACIOL should be placed no vitreous in AC & if cataract is very hard (which cannot be removed with fragementome).

SKK: Since we have VR back all the time and do have phaco fragmatome available, I insert my IOL at the time of first surgery.

DVS: Parameters for anterior vitrectomy on a peristaltic machine?

ARV: With the Infiniti Phacoemulsifier, I use the CUT-ASPIRATE mode, with highest cut rate, low bottle height, modest vacuum and aspiration flow rate. On the Infiniti, I preset 2500 cut rates/second, 60-70 cms bottle height, 300 mmHg vacuum and 25 cc/minute aspirate flow rate.

SKK: I have been using 800 cuts with 250 suction with 25cc flow rate

PG: Donot know, not using peristaltic pump

DVS: Parameters for anterior vitrectomy on a venture machine?

ARV: I do not have enough experience with the venture machine, but I would consider a vacuum of 300 mmHg, 100 cms bottle height and maximum cut rate.

SKK: I do not use Venture system.

PG: Cutting 2500 /min aspiration low 50 to 100, irrigation minimum or dry vitrectomy with viscoelastic should be done. Use of 23 or 25 g cutter is better in such situations. [but the topic is post dislocation of lens, here since you are anyway doing pars plana vitrectomy so vitreous coming from behind is not a problem

JSG: Vitrectomy with Venturi machine is always preferable. Parameters are at least 1200 cut rate and suction of around 100 -150 mm. One must remember that any tug on the vitreous in AC is transmitted to vitreous base.

DVS: How do you ensure complete anterior vitrectomy?

ARV: I inject triamcinolone to ensure adequate anterior vitrectomy. I use it at several stages: first after performing an initial vitrectomy before moving on to phacoemulslfication, second if in a doubt before IOL implantation and lastly before closing the paracentesis after removing the residual OVD with a vitrector. There are subtle signs such as peaking / distortion of the PCR opening, peaking/distortion of the anterior capsulorhexis or the pupil that can also indicate presence of vitreous strands.

HL : Put the IOL & after constricting the pupil, put Tricot in the anterior chamber. One will be able to see each & every fiber of vitreous in the A.C. Even if one has done excellent vitrectomy pre-IOL insertion, this step is a must after placing the IOL.

Do not put the triamcilone before placing the IOL and constricting the pupil as it may seep into the vitreous cavity & can reduce the visibility.

If you do not have a vitrectomy machine, the vitreous in the A.C. is not as harmful as in the section. Even when the pupil constricts fully with pilocarpire, post-op, some vitreous strands may be seen going towards the section. So I mechanically sweep the iris repositor from the peripheral angle to the center. So all the vitreous can be pulled into the center. I actually put the vana's scissor at the pupillary plane to cut the vitreous in the center. If vitreous remains in the section, it causes lot more traction & associated complications. Many a times, in a smaller setup, if the vitrectomy machine/cutter is not working properly, this method may be followed.

SKK: The end point is when the pc margins falls back and shape of PCCC is not distorted. We regularly keep sweeping under the incission with help of fine spatula. Injection of Triamcilone is very help full in vsualisation of vitreous strands.

PG: Use triamcinolone inj in anterior chamber to stain the vitreous and pilocarpine injection can also be used to contsrict the pupil and observe the shape of pupil , it should be regular and round , vitreous strands coming from bhind will distort the shape of pupil.As you remove the vitreous completely the pupil will become round.

JSG: Staining with Triamcinolone acetonide may be used, or if one has slit illumination facility in the microscope that can be used.

DVS: Do you suture the wound in can be PCR?

ARV: Yes I suture all incisions, the main as well as the paracentesis incision in all cases of PCR.

HL : If I am able to place the foldable IOL & if the wound configuration is good, I do not suture the section. However, it is better to error on the side of caution & suture the wound. Also if the wound is enlarged for a PMMA lens, it needs to be sutured.

SKK: Not if it has been managed well on table, but if there is a second intervention needed by the VR guys I do put 1 suture even for my 2. 75 incission

DVS: Do you prefer same sitting vitreous surgery for nucleus drop?

ARV: Yes for a soft to medium density nucleus I would prefer to perform pars plana vitrectomy with lensectomy using a contact lens at the same sitting. If it's a dense nucleus I would refer to a retinal surgeon at the earliest possible.

HL : Yes, if primary vitrectomy is available, can save you from the embrassement & the patient from psychological trauma and also the cornea is clear & surgery is easier. Next day the patient may have keratitis & the surgery will have to be deferred for a week/or two. But the final outcome may not be different in the two scenarios.

SKK: Yes, I prefer same sitting since I have support of VR in my OR.

DVS: How do you evaluate a case with PCR and posteriorly dislocated nucleus/epinucleus lens matter?

PG: Time since cataract surgery

- Status of cataract wound - sutured or not
- Corneal transparency – cornea should be clear enough to perform complte and safe vitrectomy
- Inflammation
- Intraocular pressure – lens induced glaucoma
- Status of vitreous in anterior chamber, size of PCR, IOL is placed or not, support of capsule
- Size of dislocated lens nucleus
- Retinal exam to look for breaks, retinal detachment or hemorrhage because of the vitreous manipulations during cataract surgery.

JSG: The following points have to be evaluated:

- Time elapsed following cataract surgery
- Corneal status – superficial edema should not bother – epithelial scraping with cotton tipped buds often enables a clear view, since I use BIOM only 3 sq. mm clear area is required for a thorough vitrectomy of course one has to wait in case of severe striate keratopathy.
- Presence of inflammation – role of steroids.
- Secondary glaucoma – it is always advisable to control IOP for a couple of days before surgery as CRVO / expulsive hge have been reported in high risk eyes when surgery was done immediately after IOP control.
- Status of the posterior capsule/presence of IOL.
- Size of the dislocated fragment – I remove even the smallest fragments.
- Thorough retinal examination (usually without scleral depression) for breaks, RD, etc. If hazy media USG-B scan must be done.

DVS: Criterion for deciding vitreous surgery?

PG: Vitreous in anterior chamber and wound incarciration.

- Inflammation.
- Increased intraocular pressure.
- Size of nucleus.
- Retinal detachment.

JSG:
- Though there is a school which doesn't advocate vitrectomy for nucleus fragments less than 25% size – I do vitrectomy for even the smallest fragment.
- Associated detachment and/ or vitreous hge – early vitrectomy.

DVS: Do you agree that long term prognosis is not affected by delaying surgery for a week or two?

PG: Should be operated early if the cornea is clear enough to perform complete and safe vitreous surgey.

JSG:
- Ideally if VR surgeon is available, best results if operated on the same day.
- If there is a delay due to any reason, if cornea is clear vitrectomy should be done on the day of presentation to the VR surgeon.
- Beyond two weeks there is rise in incidence of long term complications.

DVS: How long can one wait for cortex to absorb if all other parameters including cornea, intraocular pressure and CME are very well controlled?

PG: If there are no associated conditions mentioned above one can wait for months if the floaters and not annoying to the patient.

JSG: If the above mentioned parameters are under control, one can wait indefinitely unless the intensity of floaters or visual loss becomes an indication for surgery.

DVS: What are common intraoperative problems encountered while managing such cases/precautions to be taken by vitreous surgeon?

PG: Corneal wound should be well secured ,if the cataract surgeon has not sutured the wound the first thing is to suture te wound before starting vitrectomy.

- Visualization of infusion cannula due to lens matter

- Scleral burn due to phaco needle – assistant should keep pouring ringer lactate /irrigasol on sclerotomy site while fragmentation is being performed
- Induction of PVD , a complete vitrectomy should be done

JSG:
- Corneal wound should be sutured with 10-0 nylon. I usually use a depressor to test the integrity of the wound.
- All vitreous tags to the corneal wound, cortex materials if any should be removed from AC.
- Often lens matter obscures the visualisation of the infusion canula – holding and depressing the canula with the forceps externally and using the cutter to clear the lens matter around the tip of the canula with the AC maintainer on usually solves this problem.
- Nucleus fragments should be freed of all vitreous attachments by the cutter before introducing the fragmetone. Of course initial PVD induction is essential.
- I use a bimanual technique using the fragmetome in one hand and the MVR blade in the other hand to remove the nucleus, always working in the mid-vitreous.
- A thorough examination of the peripheral retina using scleral depression under high magnification is absolutely essential to rule out any breaks (micro and macro), small nuclear fragments adhering to the peripheral vitreous skirt- the situation should be managed accordingly.

DVS: While most patients do very well, what are the common causes of poor recovery following vitrectomy for such cases?

PG: Persistant cystoid macular edema

- Persistant glaucoma inspite of complete removal of lens matter
- Retinal detachment is not uncommon

JSG: Cystoid macular edema, chronic glaucoma, retinal detachment(rare)

DVS: Your experience on Sutureless vitrectomy for such cases?

PG: Good option, I am using 23G phaco fragmentation needle for nucleus removal .The surgery is equally safe and remains sutureless for the patient.

- 23G or 25 G Cutter is better for performing vitrectomy in anterior chamber.

JSG: I often do a hybrid 23 – 20G as 23G phaco fragmetome is not yet available.

- 23G cutter is very useful for anterior vitrectomy.

DVS: Suggestion to your anterior segment colleagues on this issue under these headings?

(a): Suturing of the wound

PG: Always suture the corneal wound with 10 nylon

JSG: Always suture the corneal wound with 10-0 nylon

(b): Anterior vitrectomy

PG: Do anterior vitrectomy if you are sure and prepared to do it at the time of complication, Use triamcinolone injection for staining vitreous in anterior chamber, it helps in complete removal of vitreous

JSG:
1. Always do bimanual vitrectomy
2. Infusion directed away from the post capsule
3. High cut rate and low suction
4. Clear all vitreous anterior to PC

(c): IOL implantation

PG: Always try to implant IOL on rhexis and suture the wound and refer the case to VR Surgeon.

JSG: Always try to implant IOL on rhexis and suture the wound and refer the case to VR Surgeon. Do not attempt IOL if anterior capsule is not sufficient.

(d): Additional tips

PG: Do not try to remove the dislocating nucleus with various innivative methods if you are not trained to perform vitreous surgery. The safest method of saving these eyes is vitrectomy with fragmentation

Best is to inform the patient about the complication and reassure him that vitreous surgery can be done to improve his condition.

JSG: Never panic and never use the vectis to retrieve the nucleus or do any other manipulation. Clear the vitreous from the AC , put IOL on rhexis/ cilliary sulcus, suture the wound and refer to VR surgeon. Truth laced with reassurance is the best way to inform the patient.

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IOL Master: Mastering the Master

Neeraj Bhargava *MS,* ***Nitin Vichare*** *MS, DNB*

Phacoemulcification has become a routine surgical procedure and preferred method of cataract extraction in all grades of cataract. Newer machines and improved fluidics has made modern day cataract surgery easier and has increased the expectations of patients regarding the visual outcome following cataract surgery. However the most important part of the cataract surgery is not the actual surgery but the pre operative biometry and IOL power calculation. Determining the correct power of the IOL to be implanted in the eyes is most crucial step. Ability to predict IOL power with high degree of accuracy is the key for successful visual outcome.

All aspects of biometry are essential. Inaccurate keratometry by 0.50 diopters will cause final IOL power to be inaccurate by the same amount. Similarly variation in measurement of axial length by 0.1mm will cause 0.3 diopters of IOL power variation. Thus minor errors in measurement cause cascading effect leading to post operative refractive surprises.

IOL Master, an ultra high resolution optical biometry system, is in clinical practice for over a decade now[1,2]. It has ability to measure axial length with unprecedented degree of accuracy. This combined biometry instrument gives measurements of axial length, keratometry, anterior chamber depth (ACD) and option to calculate IOL power from different formulae. The most important use of IOL Master is in 'optimisation' of IOL power as per post operative visual outcome.

Principle

IOL Master employs the principle of Optical coherence Biometry (OCB). It uses partially coherent infrared light beams of 780 nm diode laser with coherence length of 130 nm[3]. Laser light emitted is split up into two beams in a Michelson Interferometer. One mirror of interferometer is fixed and other is moved at constant speed making one beam out of phase with other. Both beams are projected in the eye and get reflected at cornea and retina. The light reflected from the cornea interferes with that reflected by the retina if the optical paths of both beams are equal. This interference produces light and dark band patterns which is detected by a photo detector. The signals are amplified, filtered and recorded as a function of the position of the interferometer mirror. An optical encoder is used to convert the measurements into axial length measurements (Figure 1).

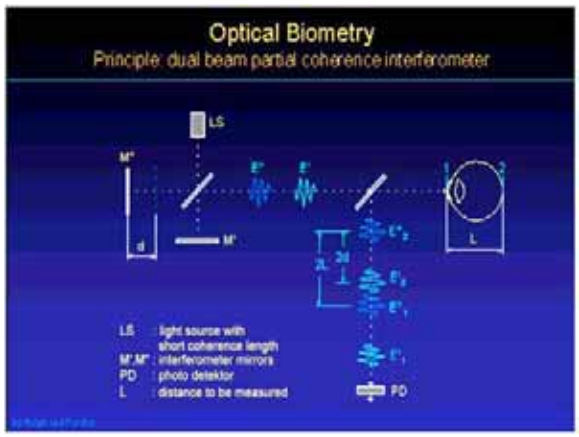

Figure 1: *Principle of optical biometry*

Special feature incorporated is use of dual beams. In interferometer eye needs to be absolutely stable so as not to disturb interference patterns. Use of dual beams makes IOL Master insensitive to longitudinal movements and measurements can be made with ease.

IOL master Vs Ultrasound A Scan

Commonly used ultrasound A scan uses 10 MHz probe which has a accuracy of 0.1 mm in best of hands. Applanation scan requires touching the cornea which causes indentation and gives falsely short axial length. Immersion A scan, though more reliable is time consuming and uncomfortable to patient.

OCB measures the axial length from corneal apex to retinal pigmentary epithelium while A scan measurers up to vitro retinal interface only. IOL Master thus gives the true refractive length than anatomical axial length. Accuracy of IOL Master is 0.02 mm which is operator independent. IOL Master is upright, non contact, ultra high resolution biometry. It is patient and user friendly. Highly ametropic patient can wear glasses while sitting on IOL Master which aids in fixation. This has advantage in measuring fovea in cases of posterior staphyloma. However significant media opacities limit the use of IOL Master.

Evaluation of axial length measurement results

With patient sitting comfortably he is asked to look directly in the small red fixation light. The operator adjusts the focus of the video image to get the reflection of the light in the centre of the cross hair (Figure 2). This alignment causes measurement of centre of macula. Measurement is initiated with pressing of push button. It is advisable that to get the best reflection nothing should have touched cornea just before starting the measurements. If patient is using contact lens its use should be discontinued at least 24 hrs prior to measurements.

Axial lengths measurements are displayed in the form of signal curves graphs. Ideal display of axial length graphs termed as valid signal curves characteristically shows very good signal-to-noise ratio (SNR > 10). It has narrow primary maxima with central terminal peak. Secondary maxima are present symmetrically on either side, separated by approximately 800 µm from primary peak. Tertiary and quaternary peaks are visible with low and even base line (Figure 3).

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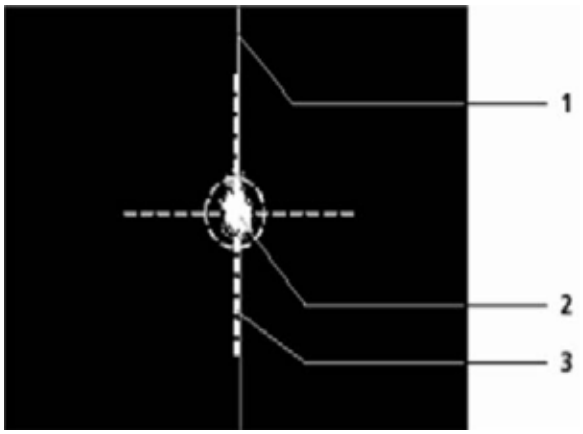

Figure 2: *Alignment for axial length measurement*

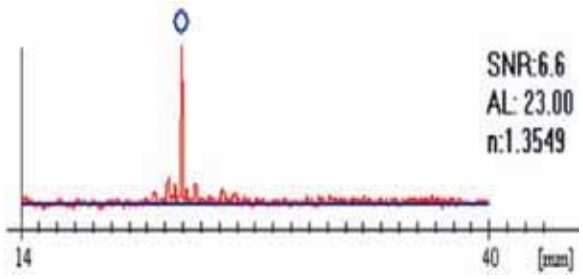

Figure 3: *Ideal display of axial length graph*

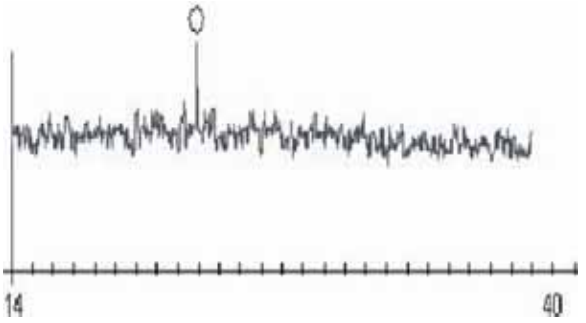

Figure 4: *Borderline SNR display*

Borderline SNR as displayed in graph (Figure 4) has SNR value between 1.6 -2.0. Graph display is uneven with primary maxima having short peak and indistinct secondary maxima. Such display is seen with moderate grade cataract and should be interpreted with caution.

Invalid signal displays are seen with SNR value less than 1.6 with indistinguishable primary maxima. Display graph is irregular (Figure 5) with high variations in axial length displayed. Also machine gives "error" message and these results need to be discarded. Unsteady (non fixating) patient, strong ametropia and dense medial opacity along the visual axis gives invalid signal display.

Intra test variations

During measurements of axial lengths certain measurements are away from mean results. Such intra test variations need to be recognized otherwise average axial length displayed will not be representative of true axial length. Such results are frequently due to strong reflection of optical beam from either internal limiting membrane or choroids. These reflections are sometimes as strong as PRE reflection causing particular axial length measurement to be off the mark. Signal curve graph shows primary maxima having steps or multiple peaks or bifid at the top (Figure 6). Post run editing helps the examiner to view the graphs and corresponding axial lengths measured. Such off the mark readings needs to be deleted to avoid errors in averaging the axial length.

Axial length modes

One of the advantages of IOL Master is able to measure axial length in different situations like phakic, pseudophakic or silicone filled eyes just by selecting the appropriate mode (Figure 7). By opening window of 'AL Setting' examiner can select the mode. In AL setting 9 different modes are available.

Best measurement technique

To get consistent readings which are within 0.02mm of each other the examiner can sample multiple areas around the central reticule

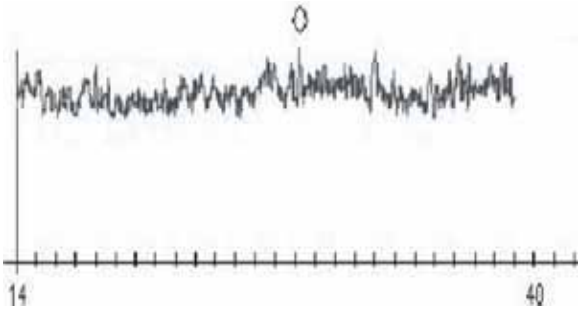

Figure 5: *invalid signal*

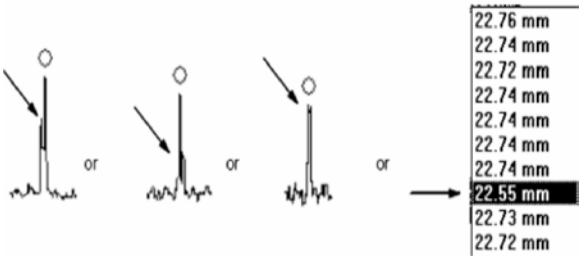

Figure 6: *Intra test variation*

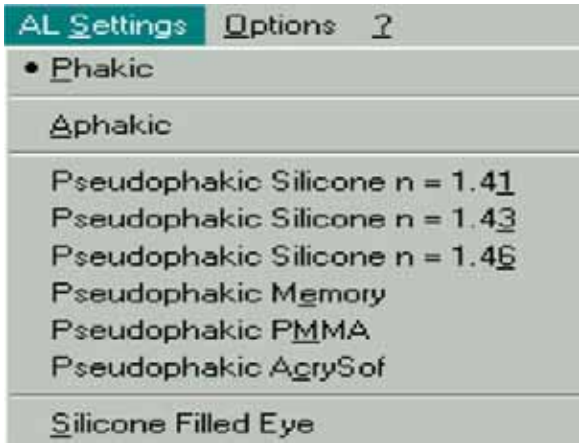

Figure 7: *AL setting*

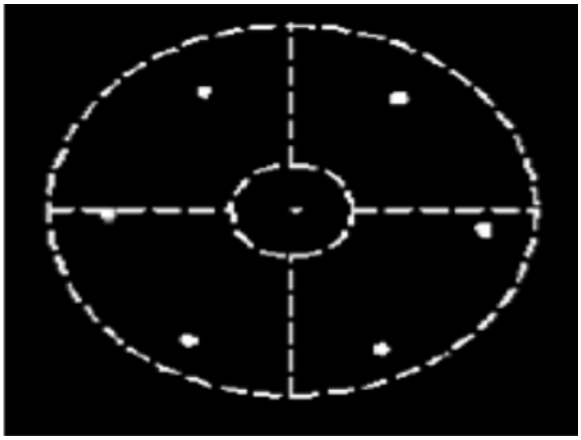

Figure 8: *Corneal curvature measurement*

and can also move slightly front and back while taking multiple readings. In this way the examiner can find out the area around central reticule to get the best results and subsequent readings can be taken at that spot. This technique is particularly helpful in cases of small corneal scars, anterior or posterior sub capsular cataract or other media opacities.

Keratometry

As a combined biometry instrument IOL Master measures other aspects also. Central corneal power is measured as in automated keratometry. Keratometry mode can be activated by clicking cursor on the icon or by pressing space bar after finishing axial length measurements. Patient asked to focus straight and blink several times to get even tear film.

On video screen examiner sees 6 peripheral points located in the field between the two auxiliary circles (Figure 8). The central point is usually not focused and is not evaluated. Five internal individual measurements are taken for a single keratometer measurement within 0.5 seconds. If a measurement point is not correctly identified, a blue flashing dot will appear.

The IOL Master requires THREE keratometer measurements to be taken! Only then will a mean value be passed on to the IOL calculation.

The corneal curvature results are displayed in mm or diopters along with corresponding axis. If cornea is completely spherical then only one radius or power is displayed. Knowing steeper axis helps in planning the incision during surgery.

It needs to be remembered that IOL Master measures central 2.5 mm diameter compared to manual keratometer which measures central 3 mm zone. Since IOL Master measures more centrally, keratometry readings show steeper K value than manual keratometry.

In pseudophakic eyes multiple images appear due to refection from anterior surface of IOL leading to measurement errors (Figure 9). Tip in such cases is to move the IOL Master approximately 1 mm away (defocusing). Corneal image becomes

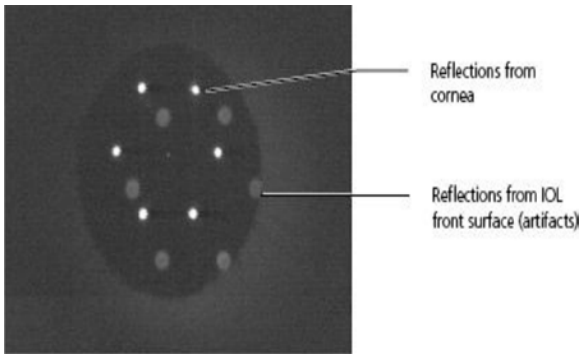

Figure 9: *Keratometry in pseudophakic eyes*

slightly larger, while the artifacts of the IOL become fainter. Measurement is then possible.

Presence of corneal scars, severe dry eyes, epithelial defects and partially concealed reflections due to half open lids or ptosis leads to measurement errors.

Other parameters

Anterior chamber depth (ACD)

Anterior chamber depth on the IOL Master is interpreted as the distance between the anterior vertex of the cornea and the anterior vertex of the lens. Hence, the displayed distance includes the thickness of the cornea. On activating ACD measurement lateral slit illumination will automatically be turned on. The fixation point is displayed in optimum focus in the rectangle on the screen (Figure10). Calculation of the anterior chamber depth requires the input of the corneal radius. If a valid keratometer measurement was performed prior to ACD measurement, the system will automatically use the measured radius for the calculation. ACD

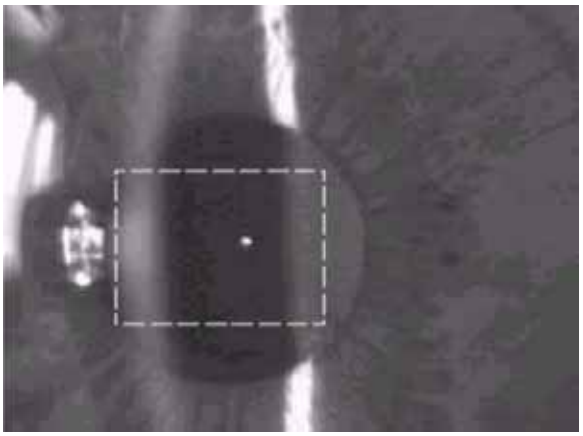

Figure 10: *ACD measurement*

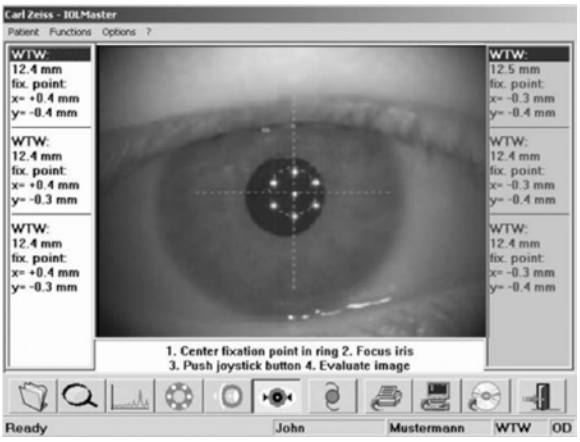

Figure11: *WTW measurement*

measurement values are required in IOL power calculation formulae like Hollady 2, and Heigis.

White-to-White Measurement (WTW)

IOL Master can give easy measurements of WTW distance. As operator activates the function, a cross bar appears. Trick is to focus on the iris rather than cornea. Align the IOL Master so that the six peripheral light spots are symmetrical to the cross hairs and the iris structures or the edge of the pupil appears optimally focused (Figure 11). Measuring WTW distance is useful in calculation of phakic IOL.

IOL power calculation

After taking axial length and keratometry readings, IOL power mode is activated. The values of mean axial length and keratometry are passed on automatically. Operator needs to select the surgeon to enable the activation of IOL power calculation. Surgeon must have entered the parameters of his preferred IOLs in database beforehand. Soft wear then calculates IOL power for the different types of lenses. Results for four different types of lenses are displayed at a time (Figure 12). Up to 20 surgeons name and correspondingly data of their preferred lenses can be entered.

IOL powers are displayed in the steps of 0.5 diopters or 0.25 diopters. Included in the standard IOL master soft wear package are five popular IOL power calculation formulae are Haigis, Holladay, SRK II, SRK/T and Hoffer Q. While calculating IOL power surgeon can choose his preferred formula from the task bar. Newer version of the machine has mode for phakic IOL and post refractive surgery patients.

Optimisation

Included in the IOL Master is the most important part that is ability to optimise or fine tune your IOL power as per your choice of IOL. It must be remembered that A constants which are provided by the manufacturer are based on contact A scan and manual keratometry. This value represents where we anticipate the IOL to sit in relation to cornea. Specifically how near or far from cornea. The "constant" will decrease with ACIOL as compared

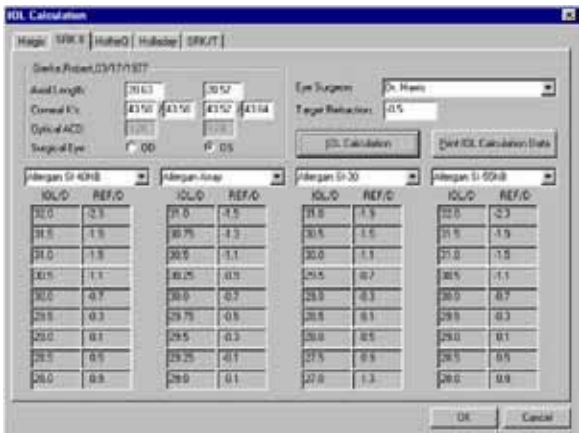

Figure 12: *IOL power calculation display*

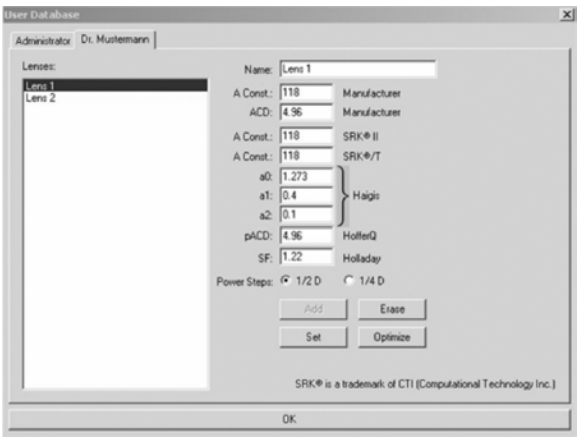

Figure 13: *Optimisation of IOL power*

to PCIOL. As ACIOL sits closure to cornea, less power is needed. The manufacturer estimates the A constant by approximating from similar lens models. The final lens position in capsular bag will be influenced by IOL style (optic - haptic design, flexibility, angulations, reaction to fibrosis) and surgical technique (rhexis, incision, phaco technique). There fore A constant provided by manufacturer needs to be optimised as per surgeon's technique.

Initially, surgeon can use optical lens constants available on the ULIB web page which gives lens constants for different types of lenses as per data pooled in from world wide IOL Master users. To derive his own optimized A constant surgeon needs to select minimum of 20-25 cases. All these cases should be operated by same surgeon with same surgical technique using same type of lenses. Post operative data and final post operative refraction at end of 6 weeks are noted. This data needs to be entered in IOL Master which then calculates the new optimised lens constant for that type of lens for that particular surgeon (Figure 13).

We feel this is the most important aspect of the IOL Master which helps the surgeon to get consistent targeted refraction post operatively.

Role in your practice

IOL Master has become a gold standard in biometry and IOL power calculation. It is the most important tool now a days available to refractive surgeons who aim for perfection in their every aspect of surgery. IOL Master gives you the most accurate and consistent IOL power for your choice of IOL[4].

Limitations

IOL Mater is an optical device so very dense cataract, thick posterior capsular plaque, mature lens, presence of significant media opacities like corneal scars, vitreous hemorrhage limits its use. In cases of nystagmus or patients with unsteady eyes IOL Master fails to measure the axial length.

References

1. Drexler W, Findl O, Menapace R, et al. Partial coherence interferometry: a novel approach to biometry in cataract surgery. Am J Ophthalmol 1998;126:524-34.
2. Kiss B, Findl O, Menapace R, et al. Refractive outcome of cataract surgery using partial coherence interferometry and ultrasound biometry: clinical feasibility study of a commercial prototype II. J Cataract Refract Surg 2002;28:230-4.
3. Fercher AF, Mengedoht K, Werner W. Eye length measurement by interferometry with partially coherent light. Opt Lett 1988;13:186-8.
4. H Eleftheriadis. IOLMaster biometry: refractive results of 100 consecutive cases. Br J Ophthalmol 2003 August; 87(8): 960-3.
5. www.doctor-hill.com

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Malignant Tumors of Lid

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Malvika Singh *MBBS,* ***Sonia Singh*** *DO*

Tumors of eyelid and ocular adnexa need a special attention because of their proximity to the eye, brain and paranasal sinuses. Malignancies can mimic a number of benign conditions. Usually a definitive diagnosis can be made only on histological examination of biopsied tissue.

Epidemiology

Approximately 5-10% of all skin cancers arise in periorbital skin.[1] Basal cell carcinoma (BCC) accounts for nearly 80% of all non-melanoma skin cancers and 90% of all malignant eyelid tumors.[2,3] Squamous cell carcinoma (SCC) is relatively uncommon in the eyelids, accounting for approximately 9% of all malignant eyelid tumors.[2] Sebaceous gland carcinoma (SGC) and Malignant melanoma (MM) of the eyelid are rare lesions accounting for approximately 5%[4] and 1%[5] cases respectively but have considerable potential morbidity and mortality.

Pathogenesis

Pre-existing lesions, heritable and sporadic genetic mutations, environmental factors, and immunologic factors contribute to carcinogenesis. BCC, SCC, MM, and Merkel cell carcinoma (MCC) share ultraviolet (UV) radiation as one contributing factor, although SCC is less directly related to UV radiation.[6]

The pathogenic mechanism of common tumors are listed in Table1.

Classification

Based on the tissue of origin malignant lid tumors can be divided into following categories:

- Epithelial tumors – Cutaneous – BCC, SCC, MCC
  - Sebaceous gland – SGC
  - Sweat gland – Mucinous, Nonmucinous (signet ring and histioid cell) and adenoid cystic carcinoma
- Vascular tumors - Hemangiopericytoma, Angiosarcoma, Kaposi sarcoma
- Mesenchymal tumors - Fibrosarcoma, Liposarcoma, Rhabdomyosarcoma
- Melanocytic - Melanoma (superficial spreading, nodular, lentigo maligna melanoma)
- Lymphoreticular - Non-Hodgkin's lymphoma, Burkitt's lymphoma, Kimura's disease, Lymphosarcoma, Mycosis fungoides
- Nervous tissue - Neurofibrosarcoma
- Metastatic - Breast (most common), GIT, Lungs, Skin, Genitourinary system

Gurunanak Eye Center
Maharaja Ranjit Singh Marg, New Delhi

Clinical Evaluation and Work Up of Eyelid, Periocular or Periorbital Skin Lesions

Evaluate and document the physical characteristics of the suspicious lesion including number, location, size in 2 or 3 dimensions, shape, color, contours, borders (discrete, un-defined, regular, irregular etc.) and (a)symmetry. Other features to be noticed are the superficial telangiectatic vessels, loss of eyelashes and lid architecture and position (ectropion and retraction).

On palpation fixation to the tarsus and orbital rim should be assessed.

Examination of the head and neck for premalignant and malignant skin lesions (especially actinic keratosis, lentigo maligna, Bowens disease etc.); enlarged lymph nodes; scars from prior biopsies, radiation therapy, surgery, or trauma.

Sensory motor alterations (Perineural invasion) – Decreased or altered sensation (Vth nerve), Pain, Ophthalmoplegia, Ptosis or Facial weakness.

Complete ophthalmic examination with precise attention to the caruncle and plicae, conjunctiva, inferior and superior conjunctival fornices, extraocular muscles, and naso lacrimal system. The eyelids must be everted and evaluated.

Orbital examination for globe proptosis, dystopia, Exophthalmometry and any palpable masses, should be noted.

Radiographic evaluation especially for larger tumor or those located near the medial canthus or the lacrimal apparatus. CT scanning may demonstrate bone alteration or destruction and extension into the nose, sinuses, orbit, or the cranial cavity. MRI imaging may show subtle tissue plane invasion or destruction and extension beyond the orbital septum or into the cranial cavity. It may also be useful for assessing perineural spread. Dacryocystography (DCG) with/without subtraction and computed tomography can clarify the anatomy and patency of the naso-lacrimal system.

Photography and drawing are required for documentation.

Methods of Spread

- Local extension into adjacent structures- includes intraepithelial spread into epidermis and conjunctival epithelium, Lacrimal secretory and excretory system, Orbital soft tissues and Cranial cavity. Table 2 shows the risk factors and signs of orbital invasion.
- Metastasis
  - *Lymphatics* - Upper eyelid (preauricular), Lower eyelid (submandibular or cervical). Sentinel lymph node (SLN) is the first node where metastasis occurs. Mapping and biopsy of SLN can be useful for detecting micro-metastasis.

Table 1.

Malignancy	History/ precursor	Environmental factors	Genetic factors	Infection/ immunosuppresion
BCC	Personal history of multiple BCC, male sex, tendency to sunburn, positive family history	UV radiation PUVA for psoriasis External beam radiation Chemical-arsenic	Phenotype-fair skin, blue eyes, red or blond hair, Specific mutations Nevoid basal cell syndrome Xeroderma pigmentosum Basex syndrome, Rombo syndrome	HIV, Pharmacologic
SCC	Keratoses-solar/ actinic, Bowen's disease, Burns (Marjolin's ulcer), Chronic ulcers, Osteomyelitis	UV radiation Arsenic, Tobacco, psoralens	Xeroderma pigmentosum Oculocutaneous albinism Upregulation of FGF and FGF-BP, EGF-MT P1-MMP and proMMP-2 activation, Elevation of TGF-alpha and EGF-R	HPV 16, Pharmacologic
SGC	Female sex, Asian origin	UV radiation Ionizing radiation, Diuretics	Muir Torre syndrome hMSHZ human Mut S homologue 2 gene, mutation of p53	HIV, HPV
MM	family history Precursor lesions - Congenital nevi, dysplastic nevus, Lentigo maligna	UV radiation PUVA therapy Tanning beds	Gorlin syndrome Xeroderma Pigmentosa FAMMM (Familial atypical multiple mole melanoma syndrome) Chromosome lp, 9p21, 1lq23 (melanoma suppressor gene), 17p11-13 (p53)	
MCC		UV radiation	Chromosome 1	Merkel cell polyomavirus, organ transplantation

- *Hematogenous* – Spread to Lung, Liver, Bone, brain, Gastrointestinal tract can occur. Serologic workup (e.g. complete blood count, LFT, KFT) CT scans of the Head, chest, abdomen, and pelvis; Upper GI endoscopy and colonoscopy; and Positron emission tomography scan can be useful.
- *Perineural spread (PNS)* - Most commonly in SCC » BCC » Melanoma. It is responsible for recurrence and can involve contents of superior orbital fissure, foramen rotundum, foramen spinosum.

Basal Cell Carcinoma

Eyelid, periocular or periorbital BCCs grow steadily, rarely metastasizes, and are usually not fatal. However, if treatment is delayed they may eventually destroy the eye, orbit, nose, sinuses, nervous system, and the face. Various data support the notion that the hair follicle stem cell is the progenitor of BCC.[16,17]

Topographic distribution in descending order of frequency is lower eyelid (44%), medial canthus (37%), upper eyelid (9%), lateral canthus (6%), glabella (2%), and more than one eyelid segment (2%).[20]

Clinicopathologic Types[3,7,8,9,10]

Circumscribed is the most (60%) common type with cystic and nodular subtypes. Often combination patterns arise such as nodulocystic, nodular-ulcerative (Figure 1), and a pigmented variant which contains melanin pigment. Clinically, these lesions present as a pearly or translucent papule often with peripheral telangiectatic vessels and central ulceration, and possibly variable brown pigmentation. Histopathology typically shows well-defined nests of small, hyperchromatic, basaloid tumor cells with peripheral palisading and retraction of tumor cells from the adjacent connective tissue.

Diffuse type – Accounts for 10-40%. It may be further divided into morpheaform (sclerosing), infiltrative and micronodular subtypes.

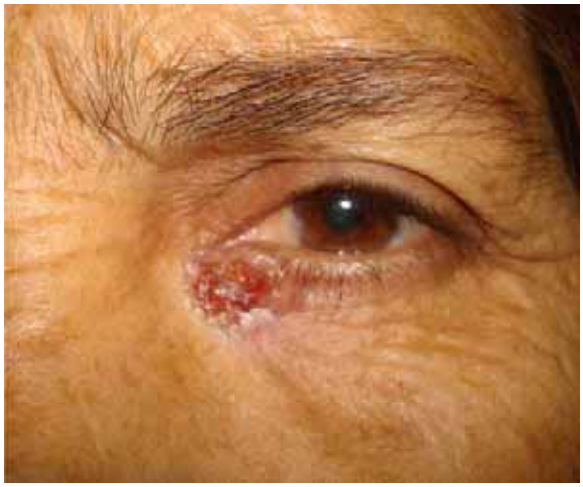

Figure 1: *Nodulo-ulcerative basal cell carcinoma*

Table 2

Risk factors for orbital invasion

- Primary periocular site – medial canthus (close to periosteal tissues and tissue planes)
- Recurrence
- Long duration of symptoms – neglected cases of delay in diagnosis
- Subtle clinical features particularly with infiltrative or morphemic/sclerosing types
- Perineural invasion

Clinical signs of orbital invasion

- Fixation of a mass to the orbital rim
- Limitation of ocular motility
- Globe displacement
- Brow and upper eyelid ptosis
- Visible or palpable mass

These can present clinically as inconspicuous flat lesions with some skin thickening and occasionally ulceration. Histopathology demonstrates irregular cords and smaller aggregates of tumor cells that are interspersed within a variably fibrotic or sclerotic dermis. Perineural involvement and microscopic extension is common.

Superficial BCC - These present clinically as erythematous, possibly scaly patches or thin plaques. Histopathology reveals multifocal islands of tumor cells budding from the epidermis with extension into the superficial dermis.

Basosquamous (metatypical) type - Characterized by areas within the lesion with squamous differentiation. A superficial biopsy of such a lesion may be difficult to distinguish from SCC.

Outcome and prognosis - BCCs are usually slow growing with excellent prognosis. They have high cure rates (95% to 99%) with extremely rare mortality.[2,7,8,11,12] They rarely metastasize. BCC can locally invade into the orbit necessitating some type of exenteration in about 4% of patients.[9,13,14] Retrospective studies show that the mortality rates from eyelid, periocular or periorbital BCC is <3%.[9]

Clinical risk factors for recurrence are prior recurrent tumor, large (>2 cm) size, medial canthus or upper eyelid location, involvement of multiple eyelid segments, clinically ill defined borders, multicentric tumors, immunosuppression, age less than 35 years, invasion of tarsus, and extension into the orbit, nose, sinuses, or nervous system.[2,3,11,12,14,15]

Squamous Cell Carcinoma

SCC is an invasive epithelial malignancy derived from epidermal keratinocytes. The wide variation in clinical appearances presents great difficulty in differentiating it from BCC. It can appear de novo or more frequently may arise from a pre-existing skin lesion: actinic keratosis (squamous cell dysplasia), intraepidermal carcinoma (IEC, carcinoma in situ) and keratoacanthoma.

Actinic keratosis – AK is far more frequent than IEC and SCC. A prospective study reported that 60% of SCC arose at the site of an AK that had been clinically diagnosed the previous year.[18] The distinction between IEC and AK is based on the partial epidermal involvement in AK. Clinically it presents as a rough, scaly, erythematous patches occurring on chronically sun-exposed skin.

The follow up is important since the risk of malignant transformation within one year is less than 0.001%,[18] but potentially increases between 5% and 20% in 10-25 years.[19]

Intraepidermal carcinoma – Histologically atypical keratinocytes completely fills epidermis respecting the basement membrane. Clinically it appear as poorly demarcated, erythematous crusted lesion with propensity to involve hair follicle and simulating chronic blepharitis. In one series 16% had focal areas of dermal invasion constituting evolution into SCC.[21]

Bowen's disease – It is a specific type or superficial squamous IEC initially described in the trunk and later involving sun-exposed skin of the head and neck. It differs from IEC from having well defined margins with velvety, scaly erythematous plaque and moist granular base.

Keratoacanthoma – presents as a solitary well defined nodule with a central keratotic crater surrounded by layers of well differentiated epithelial cells that may have cytological atypia. Clinically 25% undergo malignant transformation.[22]

Invasive squamous neoplasia - Clinically it appear as roughened scaly plaque, which tends to develop telangiectasia, central ulceration and/or keratinous crust or fissures over a period of time. If the crust is removed, commonly an ulcer will become apparent, showing a well defined erythematous base with elevated and firm margins. Lesions may initially resemble blepharitis and one needs to have a high index of suspicion and must look for signs of loss of normal tissue architecture and biopsy such lesions.[21]

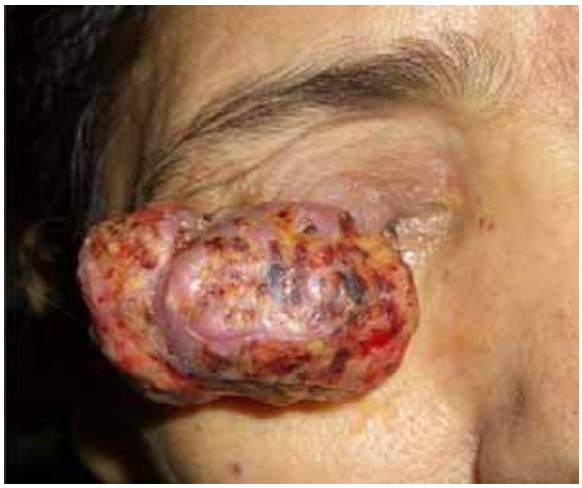

Figure 2: *Recurrent squamous cell carcinoma*

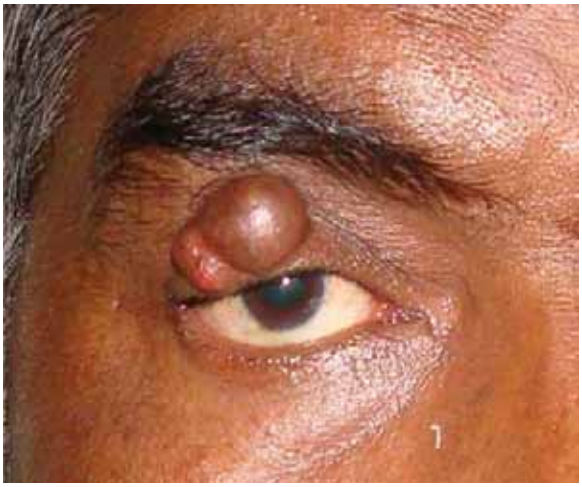

Figure 3: *Sebaceous cell carcinoma resembling chalazion*

For larger lesions (Figure 2) an incisional biopsy may be appropriate to allow surgical planning. Topographic distribution in Lower lid, Medial canthus, Lateral canthus and Upper lid is 60.8%, 17.6%, 11.8% and 9.8% respectively. [23]

Histology – Demonstrate nests, sheets and strands of malignant epithelial cells showing mitotic figures that arise from keratinocytes located in the spinous layer of the epidermis. The tumor cells have abundant eosinophilic cytoplasm and large vesicular nuclei. Keratinization is evident by the formation of keratinous cyst that are concentric layers of squamous cells. The well differentiated type has proper keratinized pearls and intracellular bridges. The majority of lesions have typical histopathological features discussed above. Rare variants include: spindle-cell, acantholytic, pseudovascular, adenosquamous and verrucous.

Outcome and prognosis - Lymph node metastasis rates vary between 1% and 21%, whereas perineural spread (PNS) has been found in 4% to 8%.[23,24,25] Hematogenous spread is possible but seems much less common than either lymphatic or perineural routes. Predisposing clinical factors to lymph node metastasis include rapidly growing, large (>20 mm), thick (>4mm), recurrent, or incompletely excised tumours.[26] Direct orbital invasion tends to occur with rapidly growing or neglected lesions and is seen in 6% cases.[23] Mohs micrographic surgery, or en face techniques should be planned, as these are associated with low recurrence rate of 3.64%[25] and 2%[23] respectively.

Sebaceous Gland Carcinoma

Sebaceous carcinoma is a malignant neoplasm that originates from cells that comprise sebaceous glands, particularly in the tarsus (Meibomian glands), in association with the cilia (Zeis glands), caruncle and eyebrow. In India, incidence of SGC seems to be higher accounting for 28%[27] and 40–60% in two series. It can exhibit aggressive local behavior and masquerade as benign lesions such as chalazion, blepharitis, keratoconjunctivitis or ocular phemphigoid, often resulting in delay in diagnosis and misdirected therapy. In a review, 63% occurred in the upper eyelid, 27% in the lower eyelid, and 5% diffusely involved both eyelids.[28]

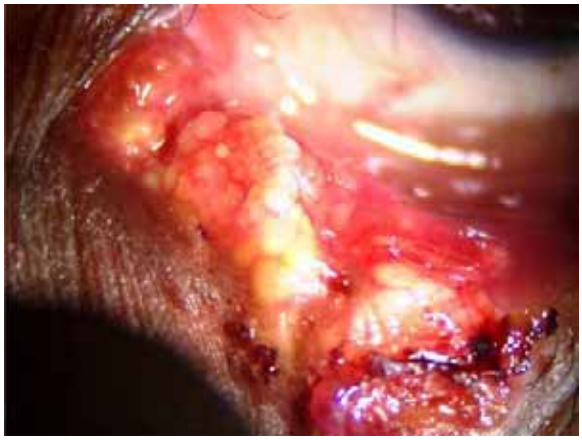

Figure 4: *Sebaceous cell carcinoma presenting as fungating lesion*

Clinical features[5]

Solitary Eyelid Nodule – It is the most common presentation with painless, firm, subcutaneous nodule in the eyelid that can resemble a chalazion (Figure 3). It is fixed to the tarsus. The skin over the lesion is generally smooth and fairly movable. The tumor assumes a yellow color because of the presence of lipid. Eversion of the affected eyelid can reveal a fungating lesion arising from the tarsus (Figure 4).

Diffuse Pseudoinflammatory Pattern – It is seen as diffuse unilateral thickening of the eyelid resembling blepharitis which is likely to extend into the epithelium of the forniceal or bulbar conjunctiva or cornea (Figure 5). SGC should be a diagnostic consideration in older patient with a unilateral "blepharitis" that does not respond to standard treatment.

Pedunculated lesion – It is usually seen as round to oval mass growing outward and becoming pedunculated. There can be keratinisation with appearance similar to cutaneous horn.

Figure 5: *Diffuse pseudoinflammatory pattern of sebaceous cell carcinoma*

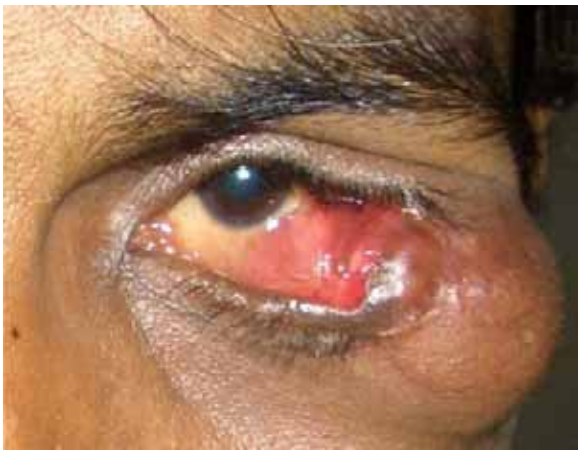

Figure 7: *Sebaceous gland carcinoma with orbital invasion*

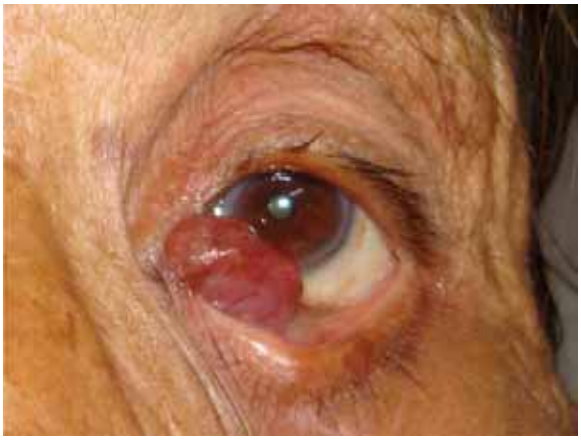

Figure 6: *Sebaceous gland carcinoma presenting as a caruncular mass*

SGC can also occur as caruncular (Figure 6), eyebrow or even lacrimal gland mass.

Histology – Four patterns are recognized. (a) The lobular pattern occurs most frequently and mimics the normal sebaceous gland architecture with less differentiated cells situated peripherally, and better differentiated, lipid-producing cells located centrally. (b) In the comedocarcinoma pattern, the lobules show a large necrotic central core surrounded by peripheral viable cells (c) The papillary pattern occurs frequently in small conjunctival tumors characterized by papillary projections and areas of sebaceous differentiation. (d) The mixed pattern can exhibit any combination of the three patterns.

SGC can exhibit intraepithelial spread into the eyelid epidermis and conjunctival epithelium in 44–80% of cases.[29,30] Incorrect initial histopathologic diagnoses have been reported in 40% to 75% of cases by an inexperienced pathologist.[5,31] Pagetoid invasion of the eyelid or conjunctival epithelium can simulate squamous cell carcinoma in situ. The presence of lipid in normal sebaceous glands and in sebaceous neoplasms can be demonstrated with the oil-red-O stain by which lipid has a red color.

Prognosis and outcome - Sebaceous carcinoma can locally invade the adjacent epithelia or the orbital soft tissues (Figure 7). The most common method of metastasis is to the regional lymph nodes in about 30%.[32,33] Advanced cases exhibit distant metastasis to lung, liver, bone, and brain.[34,35] Mortality reaches upto 18-30%.[36,37] Factors that are associated with a worse prognosis include vascular, lymphatic, and orbital invasion; involvement of both upper and lower eyelids, poor differentiation; multicentric origin; duration of symptoms greater than six months; tumor diameter exceeding 10 mm; a highly infiltrative pattern, and pagetoid invasion.[36,37] Local recurrence can develop in 18%, death from metastasis in 6% and exenteration necessary in 13%.[31] With more recently employed treatment methods, there is a tendency to avoid exenteration and to use more conservative methods of treatment.

Malignant Melanoma of Eyelid

Malignant melanoma (MM) results from the malignant proliferation of melanocytes. MM is the cancer with the greatest increase in incidence in recent years. It represents only 5% of malignant skin tumors but is responsible for >65% of deaths associated with skin cancer.[38] The majority originate de novo without history of precursor lesions. However, there are 3 described precursor lesions:

Dysplastic nevus – Clinically are asymmetrical, flat lesions with a slightly raised center. Borders may be irregular or poorly defined. Colors commonly observed are brown, tan, and blue; black is rare. About 28% of melanomas originate in a pre-existing dysplastic nevus.[39]

Congenital Nevus – Is composed of a collection of melanocytes in the epidermis, dermis, or both and is present at birth or identified by the end of the first year of life. These lesions may be giant, large, small or nevus of Ota. The association with MM is uncertain and varies according to study, but in general may be considered to

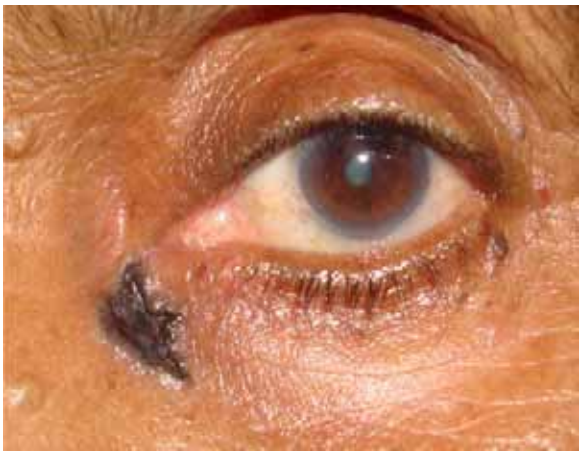

Figure 8: *Superficial spreading melanoma*

be 5% for giant nevi, 4% for large nevi,[40] 1% to 2% for small nevi,[39] and 4.6% for those with nevus of Ota.[41]

Lentigo Maligna - Is a slowly progressive, usually pigmented, irregularly shaped lesion. It is usually found on the face in older adults or individuals with actinic damage. It represents melanoma in situ. When there is invasion into dermis, the term "lentigo maligna melanoma" is used.[42,43]

White and Neil[40] clinically subdivided melanoma as follows:

- Melanoma in situ (atypical melanocytic proliferation)—neoplastic cells are present but have not penetrated the dermal – epidermal junction.
- *Superficial spreading melanoma*—accounts for about two-thirds of all cutaneous melanoma. In this type of melanoma, radial growth phase predominates during which melanocytic proliferation is intraepidermal. This lesion presents as a dark, flat with well defined borders (Figure 8). Lesions may be variegated and include tan, black, gray, or pink areas.
- *Nodular melanoma*—generally originates de novo and has only a vertical growth phase. It tends to arise in the papillary dermis and has a blue-black colour (Figure 9).
- Lentigo maligna melanoma originates as a lentigo maligna that has been present for 10 to 15 years in skin with severe sun damage. When the surface of the lentigo maligna presents an elevation or formation of nodules, this indicates transformation to malignancy.
- Amelanotic melanoma has lost its pigment. It is usually nodular but can be flat and is frequently misdiagnosed.
- *Acral lentiginous melanoma*—originates on the palms of the hand, soles of the feet, or extremities.
- *Desmoplastic melanoma*—Most frequently located on the head and neck, and frequently subjacent to a lentigo maligna melanoma. It is histologically comprised of melanoma cells between a dense desmoplastic stroma.

Table 3: Tumor Node Metastasis Classification: Cutaneous Melanoma

pTis	Melanoma in situ (Clark I)
pT1a	≤ 1mm, level II or III, no ulceration
pT1b	≤ 1mm, level IV or V, or ulceration
pT2a	>1-2mm, no ulceration
pT2b	>1-2mm, ulceration
pT3a	>2-4mm, no ulceration
pT3b	>2-4mm, ulceration
pT4a	>4mm, no ulceration
pT4b	>4mm, ulceration
N1	1 node positive
N1a	Microscopic disease
N1b	Macroscopic disease
N2	2-3 nodes positive or satellite or in-transit metastases without positive nodes
N2a	2-3 nodes positive, microscopic disease
N2b	2-3 nodes positive, macroscopic disease
N2c	Satellite or in-transit metastases without positive nodes
N3	≥ 4 nodes positive; or matted; or satellite or in-transit metastases with positive nodes
M1a	Metastasis in skin, subcutaneous or non regional nodes
M1b	Lung metastasis
M1c	Metastasis in other sites or any site and elevated lactate dehydrogenase level

Table 4: Clinical Stage Groupings: Cutaneous Melanoma

Stage 0	Tis N0 M0
Stage IA	T1a N0 M0
Stage IB	T1b-2a N0 M0
Stage IIA	T2b-3a N0 M0
Stage IIB	T3b-4a N0 M0
Stage IIC	T4b N0 M0
Stage III	Any T N 1-3 M0
Stage IV	Any T Any N M1

In 2002, The American Joint Committee on Cancer published the most recent tumor node metastasis classification system for melanoma.[44] Application of this classification system requires histologic confirmation and takes into account 3 criteria: (1) Breslow thickness of the tumor, (2) Clark level, and (3) ulceration (Table 3). Patients with MM should be clinically staged to permit selection of the appropriate type of treatment and follow-up[45] (Table 4).

Prognosis and outcome - Prognostic factors for MM are classified into 3 groups on the basis of the disease extent:[46]

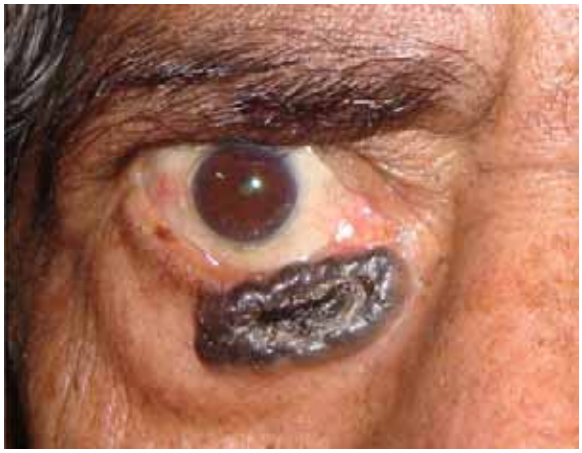

Figure 9: *Nodular melanoma*

- *No evidence of metastasis* – Tumor thickness, and ulceration are the most important predictors of survival. Other predictors are patient age (elderly patients), site of primary melanoma (trunk or head and neck), level of invasion, mitotic figures and sex (males).
- *Regional lymph node metastasis only* - Three most important predictors of survival are number of metastatic lymph nodes (poorer prognosis for higher number), tumor volume [(microscopic (nonpalpable) vs. macroscopic (palpable)] and ulceration. The positivity rate of sentinel lymph node has been reported to be 16%.[47]
- *Distant metastasis* - Anatomic site of distant metastasis (visceral vs. nonvisceral) is the most important predictor of survival. The Collaborative Eyelid Skin Melanoma Group (CESMG) reported distant metastases in 7%. The local recurrence rate of ~ 25% has been found.[48]

Merkel Cell Carcinoma (Neuroendocrine/Trabecular Carcinoma)

Merkel cell carcinoma (MCC) of the eyelid is a rare, highly aggressive skin malignancy. Friedrich Merkel in 1875 first described cells found in the basal layer of the epidermis that he believed had a function in the sensation of touch. These clear, oval, nondendritic, epidermal cells became known as Merkel cells.[49] In 1973, Winkelmann and Breathnach[50] hypothesized that malignant neoplasms may arise from the Merkel cells.[51] The typical clinical appearance of a MCC is a solitary, asymptomatic, rapidly growing, pink-red to blue-purple "violaceous" vascularized cutaneous nodule with occasional ulceration on a sun exposed area. The growth is rapid with duration of symptoms less than 6 months.[52] Histopathologically, MCC are poorly defined dermal masses often with infiltration of the subcutaneous tissue.[53] Compactly arranged in characteristic trabecular patterns, the cells are characterized by scant cytoplasm, numerous mitotic figures, a high apoptotic index, large prominent nucleoli and salt and pepper dense chromatin. The National Comprehensive Cancer Network classify MCC into the following stages: stage I (primary tumor <2cm), stage II (primary tumor ≥ 2cm), stageIII (regional nodal disease), and stageIV (distant metastasis).[54] On the basis of this staging system, the 3-year survival rates for all sites are approximately 90% for stage I, 70% for stage II, 60% for stage III, and 20% for stage IV.[55] A median survival of 9 months is reported for patients with metastatic disease.[56]

Biopsy of Eyelid, Periocular or Periorbital Skin Lesions

Any lesion that may possibly be malignant should be biopsied. It is better to biopsy a benign lesion than to miss or delay the diagnosis of a malignant lesion.

It can be either excisional or incisional. An excisional (total) biopsy is reasonable when a lesion is most probably benign, fairly small, easy to remove, or not involving the eyelid margin, canthi, or the lacrimal system. An incisional (partial) biopsy is useful if a malignancy is suspected or is highly likely and for masses on the eyelid margin, canthi, or the lacrimal system. This can be obtained via superficial shave, full thickness or punch biopsy. Some techniques can increase the accuracy of pathologic diagnosis: the tissue should be representative of the clinically suspicious lesion; the specimen size should be big enough for processing; the specimen should not be excessively traumatized, crushed, or cauterized; and the material should contain some normal tissue.

Sentinel lymph node (SLN) biopsy is a minimally invasive procedure for identifying microscopic metastasis for additional regional or systemic treatment. It involves injecting a vital blue dye and radio-colloid (usually technetium 99m) around a tumor or the site of its excision. Lymphatic channels stain blue, and these are mapped using lymphoscintigraphy and a gamma probe aids localization of radioactive sentinel nodes. If positive, then node dissection / parotidectomy or radiotherapy is required.

Conjunctival map biopsy is used for pagetoid spread of SGC. It includes taking multiple biopsies from bulbar and palpebral conjunctiva to form a map of extent of tumor spread across an ocular surface.

Treatment of Malignant Eyelid Tumors

Surgery is the treatment of choice for eyelid, periocular or periorbital lid tumors. It affords the advantage of total tumor removal and allows for the histo-pathological examination of the tissue. The two most effective modalities are Mohs' micrographic surgery (MMS) and excision with frozen section control. The surgeon marks the various margins with either suture or colour dye. A diagram explaining the orientation and location of the margins relative to the eyelids is then provided to the pathologist which can guide the surgeon regarding any location(s) of remaining cancer cells or incomplete or unclear ("positive") margins. When all histologic margins are tumor free, the surgeon can begin reconstruction. Advantages of MMS are:

- Highest available cure rate combined with most predictable reconstruction.
- Decreased morbidity, mortality and disability.
- Decreased time in the operating room by eliminating potentially long waits for frozen sections.
- Objective assessment of complete tumor removal by the micrographic surgeon.

- Conservation of tissues, providing for more simplified reconstruction
- Minimization of secondary procedures.
- Superior functional and cosmetic results.

Non surgical modalities are reserved for patient who are debilitated, fearful or refuse surgery, and who have unrealistic expectations about the cosmetic healing after surgery. Some methods may be beneficial for selected patients. For example, photodynamic therapy, carbon dioxide laser therapy, and therapy with retinoids or α-interferon may benefit patients with multiple tumors. Chemotherapy may be useful for extensive and infiltrative disease as co-adjuvant or for metastasis to distant sites. Systemic agents like Cisplatin, Doxorubicin and Capecitabine have been used for BCC and SCC and topical Mitomycin-C 0.4% four times a day for one week on therapy and one week off therapy been used for SGC.

Radiotherapy is useful for BCC and SCC, as fractioned doses between 5000-7000 cGy for patients with positive margins, perineural spread or extensive disease.

Principles of Lid Reconstruction

Despite utilization of tissue-sparing Mohs techniques, postresection defects are typically 4-6 times the original clinical tumor dimensions.[57,58] Specific evaluation of eyelid defect includes assessment of anterior and posterior lamellae. Reconstruction must attempt to restore a thin, elastic, cutaneous surface, a semi-rigid 'skeleton', and an internal keratinized mucous membrane lining keeping the following aims in mind:

- To restore physiologic functioning of the eyelids with respect to vision, lid closure, mobility & tear drainage
- To re-establish anatomic integrity
- To provide best cosmetic appearance

The *size of the defect* should be assessed as given below:

- For young patients (tight lids)
  - Small - 25-35%
  - Medium - 35-45%
  - Large - Greater than 55%
- For older patients (lax lids)
  - Small - 35-45%
  - Medium - 45-55%
  - Large - Greater than 65%

The choice of procedure shall depend on the extent of lid defect.

Small defects can be closed by direct approximation. A lateral canthotomy and lysis of inferior crus of lateral canthal tendon can give an additional 5-10mm in horizontal dimension. *Medium sized defects* are dealt by lateral semicircular rotational flap, the sliding tarsoconjunctival flap, and the free autogenous tarsoconjunctival graft. *Large lower lid defects* are managed by sharing tarsoconjunctival bridge flap from upper lid (Hughes procedure) with an overlying skin graft or myocutaneous rotational or

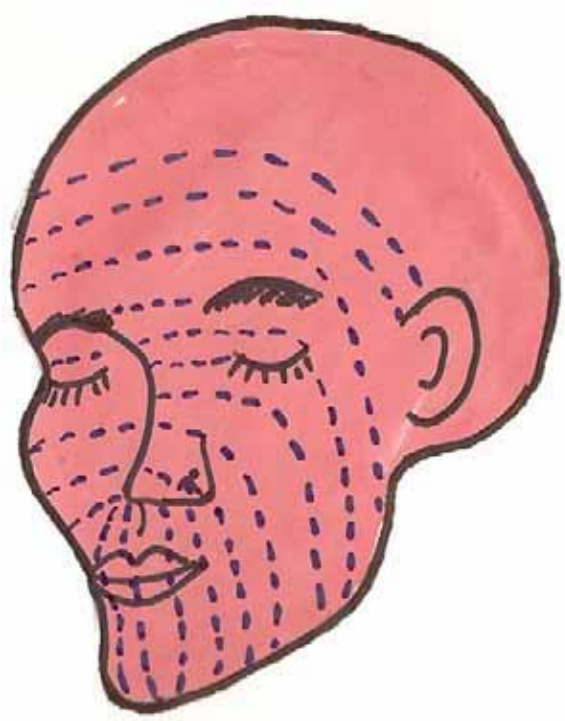

Figure 10: *Relaxed skin tension lines*

advancement flap.[59] The Mustarde semirotational cheek advancement flap[60] is another technique to repair large lid defects. *Large upper lid defects* are repaired by lid sharing techniques like Cutler-Beard[61], eye lid switch flap of Mustarde or a modified 'reverse' Hughes procedure[62]. If >3mm superior tarsus remains following tumor resection, it can be advanced inferiorly to reconstruct the upper lid margin in a single stage procedure. *Lateral canthal defects* that are <1/3rd of upper and lower eyelid margin are closed by suturing the upper and lower tarsal plates to the periosteum just inside the lateral orbital rim. Larger defects require tarsoconjunctival flaps. Tarsoconjunctival flaps in both the upper and lower lid are advanced and tarsal attachments are secured in the area of desired medial canthal tendon near the superior aspect of posterior lacrimal crest for full thickness *medial canthal defects*. Recognition and repair of canalicular injury is important in all medial eyelid defects.

Infilterative anaesthesia should always be used with lignocaine and epinephrine 1:100000, irrespective of whether local or general anaesthesia is used.

Skin flaps are preferred to skin grafts because they maintain the original colour of local tissue, shrink less (10% vs 25-50% for grafts[63]), provide inherent blood supply, subcutaneous tissue provides volume and maintains surface contour. Factors limiting blood flow to local cutaneous flap are extensive previous surgery/ irradiation, lines of tension, aggressive pressure dressing, hematoma formation and infection. Incisions should be placed within or parallel to the various facial 'lines' that develop from specific orientation of collagen and elastin fibers modified by the effects of gravity and facial muscle contraction. *Relaxed skin tension lines (RSTL)* correspond to lines of facial expression.(Figure 10)

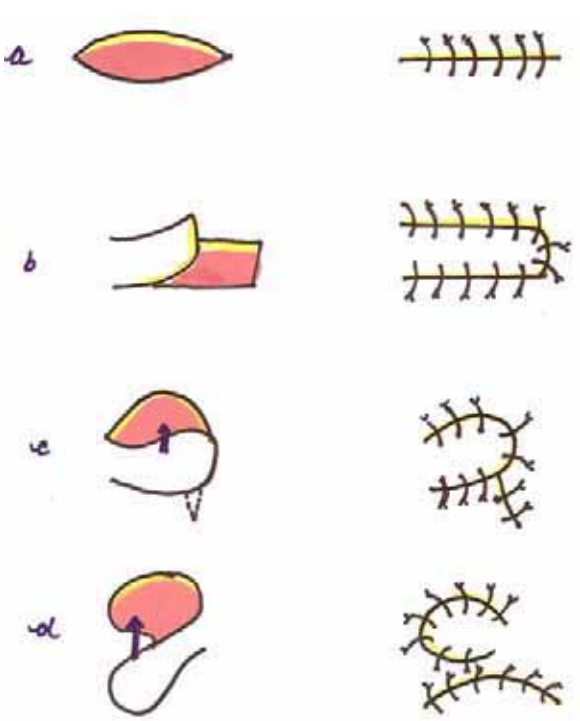

Figure 11: *Local cutaneous flaps-a)Sliding b) Advancement c) Rotation d) Transposition*

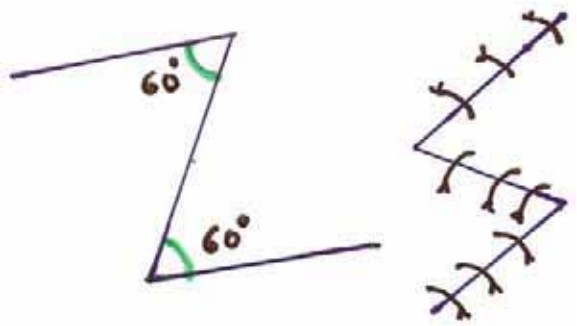

Figure 12: *'Z' plasty*

Skin incisions should be parallel to the lid margin in the upper lid & perpendicular to the margin in the lowerlid to avoid lid laxity & ectropion formation. Local cutaneous flaps can be classified as sliding, advancement, rotation, or transposition.[64] A *sliding flap* is an area of skin, undermined and moved over its subcutaneous base to primarily close a skin defect. An *advancement flap* is three sided, undermined, sliding skin flap that is advanced to cover an adjacent skin defect. A *rotation flap* is undermined strip of skin that is rotated into an adjacent defect. The *transposition flap* is undermined skin and subcutaneous tissue that is elevated and transposed to a non adjacent skin defect. (Figure 11)

Z Plasty is a transposition flap technique where the flaps can increase the length of the skin in a desired direction, change the direction of the scar so that it will lie in the same direction as the skin lines, and rotate the axis of the tissue included in the Z plasty. The scar is excised in an elliptical fashion, which then forms the common central member. Two interdigitating triangular flaps are outlined with a common central member in the line of contracture and the two arms of equal length are extended 60^0 from opposite ends of the central member. After transposition, the central member of the resultant Z will be at right angle to the original central member (scar). (Figure 12)

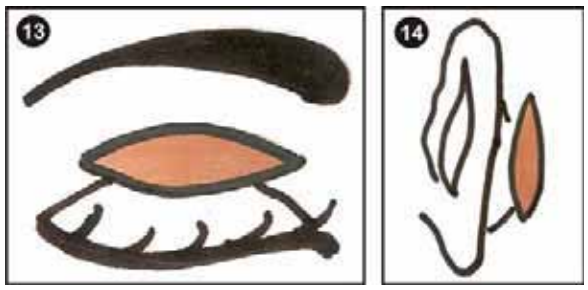

Figure 13: *Full thickness skin graft from upper lid*
Figure 14: *Post-auricular full thickness skin graft*

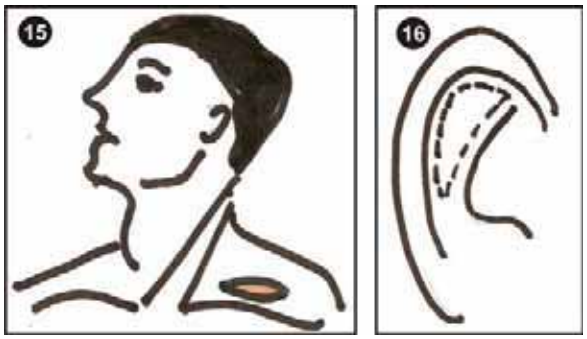

Figure 15: *Supraclavicular full thickness skin graft*
Figure 16: *Auricular cartilage graft-surface marking*

Grafts required for lid reconstruction can be as follows:

Full thickness skin grafts are composed of epidermis and the entire dermis. They can be obtained from ipsilateral/ contralateral upper lid skin (in case of lower lid defects), skin from retroauricular area, supraclavicular area, inner aspect of arm or nasolabial fold. (Figure 13,14 &15)

For construction of tarsus, upper 2/3rd of superior tarsal plate, auricular cartilage (Figure 16,17), hard palate mucosa, nasal septal cartilage or nasal alae can be used.

Conjunctiva is substituted by superior bulbar conjunctiva, oral mucous membrane, nasal mucous membrane or amniotic membrane.

Commonly used Surgical Techniques

Direct closure

A full thickness, vertical, pentagon-shaped defect with point near the conjunctival fornix is created so as to minimize tarsal buckling and eyelid margin notching. 6,0 vicryl sutures with atraumatic

Figure 17: *Auricular cartilage graft-dissection*

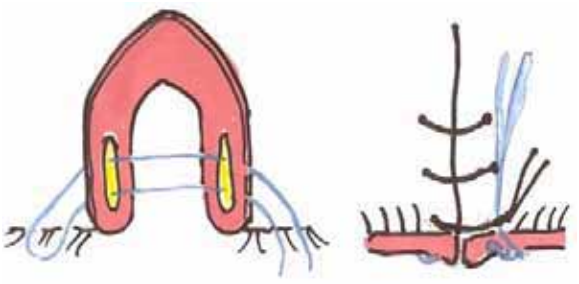

Figure 18: *Direct closure using vertical mattress suture*

spatula needle is used for suturing. Vertical mattress sutures are passed at gray line, posterior lid margin and anterior lid margin. Initial bite of the suture is placed at the gray line about 3mm from the cut end. This is the deep bite. The needle emerges at 3mm from the cut end on the other side at an equal depth. A return bite 1.5 mm from cut end is taken to finally emerge 1.5mm from cut end on the side the suturing was initially started. On tying the suture, slight eversion of the lid margin after repair must be achieved so as to prevent wound contracture once edema subsides. The ends of the gray line and posterior lid margin suture should be left long so that they can be included in the in knot of the anterior lid margin suture to prevent accidental corneal rub. The tarsal plate is closed with partial thickness bites using vicryl suture and finally skin is sutured using 6,0 silk. (Figure 18)

Tenzel's semicircular flap

It is used for both upper and lower lid defects. Rotation of a semicircular musculocutaneous flap, 18mm horizontally and 22 mm vertically beginning at the lateral canthus, extending upward for lowerlid defect & downward for upper lid defect can be done. Cantholysis of the corresponding crus of canthal tendon is performed. (Figure 19)

Tarsoconjunctival bridge flap (modified Hughes procedure)

It is used for lower eyelid defects > 50% of the horizontal length of the eyelid .Tarsoconjunctival bridge flap from the upper eyelid is used for the posterior lamella.It is covered by a full thickness skin graft. (Figure 20) The flap is left in place for 4-6 weeks prior to second-stage separation. It is not suitable for one eyed/ amblyogenic age.

The Cutler Beard Procedure

It is used for large central defects of the *upper eyelid.* A full-thickness segment of lower eyelid tissue is passed under an intact bridge of the lower eyelid margin and is sutured into the defect in the upper eyelid. (Figure 21,22) In the second stage done 6-8 weeks later, the flap is divided. It is not suitable for one eyed/amblyogenic age.

In Inverse Cutler Beard upper lid is utilized to repair the lower lid colobomas.

Median Forehead flap

It is used for massive tissue loss in upper lid. (Figure 23) The base of the flap is separated at 8 weeks. The disadvantages are a thick donor skin which is less mobile.

Temporal forehead (Fricke) flap

Temporally based flap from suprabrow is used to supply tissue to upper lid and sometimes lower lid. (Figure 24,25)

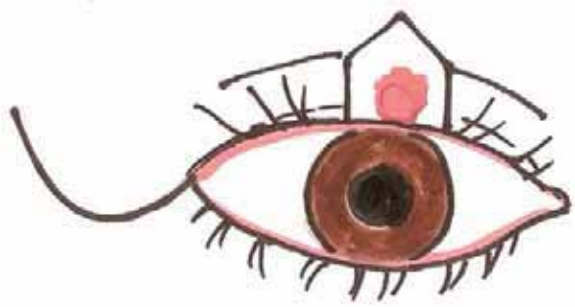

Figure 19: *Tenzel's semicircular flap*

Figure 20: *Tarsoconjunctival bridge flap(Modified Hughe's procedure)*

Figure 21: *Cutler–Beard procedure: A full thickness of lower lid is dissected to cover the upper lid defect*

Figure 24: *Dissection of temporal forehead flap*

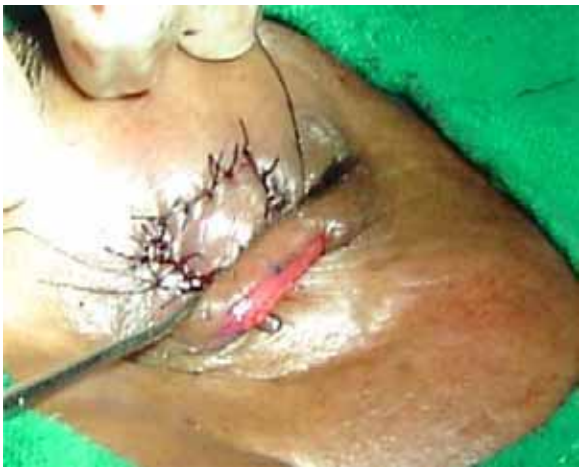

Figure 22: *Cutler-Beard procedure: A bridge of intact lower lid margin acts as a bucket handle below which the flap passes*

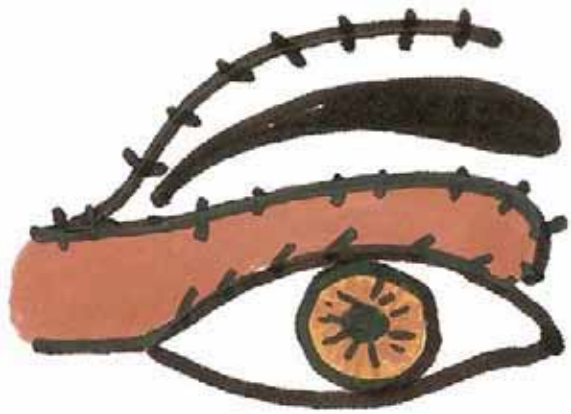

Figure 25: *Sutured temporal forehead flap*

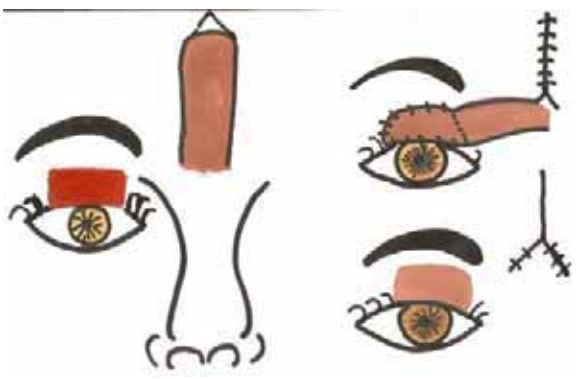

Figure 23: *Median forehead flap*

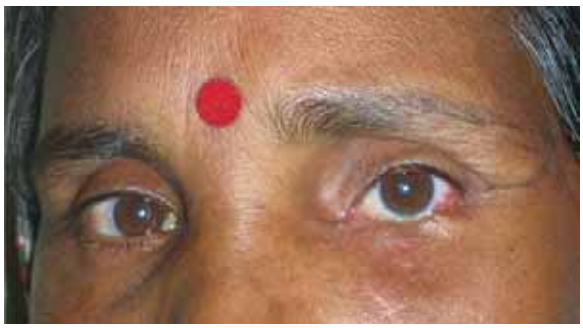

Figure 26: *Postoperative appearance after Mustarde's cheek rotational flap showing a long vertical scar and absence of eyelashes in the lower lid*

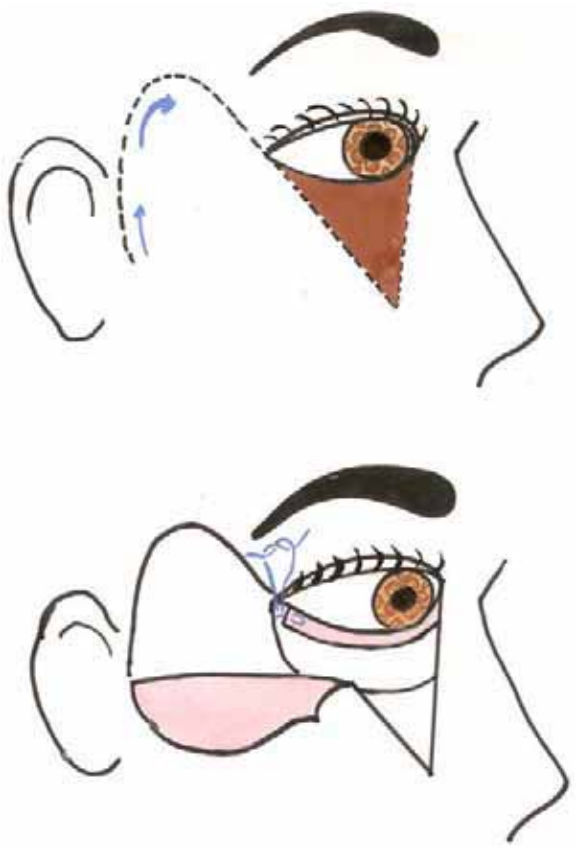

Figure 27: *Technique of dissection in Mustarde's cheek rotational flap*

Mustarde rotational cheek flap

It is used for vertically deep lower lid defects. The demerits are excessively long scar line on face, excision of a large triangle of normal skin and the adynamic nature of the reconstructed lower lid. (Figure 26) A large superiorly based triangle with medial edge at the nasolabial fold is outlined. A semicircular flap beginning at the lateral canthus and extending to the area immediately adjacent to the auricular tragus is dissected with the superior limit of the semicircle at or above the brow to prevent sagging of the lateral lid margin. (Figure 27) The medial triangle is excised. The posterior lamella graft is placed and the rotational cheek flap is mobilized nasally.

Medial canthal reconstruction

The most important aspect of medial canthal reconstruction is the attachment of the edge of the lid remanants to the posterior reflection of the medial canthal tendon to restore the direction of medial canthal angle. Defect should be fashioned in the shape of diamond. The skin defect can be closed by one of the following methods:

- *Laissez faire:* Excised medial canthal area left to heal by granulation, especially when periosteum is intact and the excision takes an equal amount or more above, rather than below the medial canthal ligament

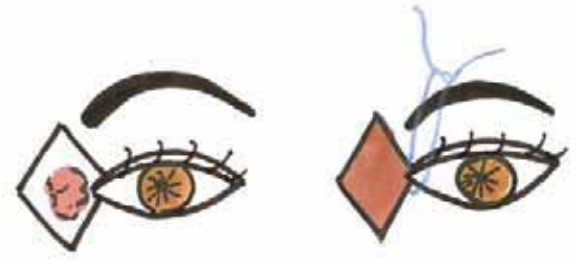

Figure 28: *Glabellar forehead flap*

- A full thickness post auricular skin graft can be sutured in place.
- A glabellar fore head flap can be used. (Figure 28)

To summarize, successful management of malignant lid tumors demands biopsy of any suspicious lesion, complete metastatic work up in cases with a positive report, appropriate treatment and a careful follow up. Minimizing sun exposure and use of protective clothing, sunglasses, and sunscreens help in prevention of Basal cellcarcinoma and Squamous cell carcinoma. Education campaigns targeting the medical officers and patients can encourage early consultation and diagnosis thereby reducing mortality rates. Close follow up is important especially for recurrent, highly inflammatory, or large neoplastic lesions that occur in the 'H zone' (upper lip, nose, periauricular, periocular) of face where there is a great risk of incomplete removal.

References

1. Cook BE, Bartley GB. Treatment options and future prospects for the management of eyelid malignancies: an evidence-based update. Ophthalmology. 2001;108:2088–2100.
2. Cook BE Jr, Bartley GB. Epidemiologic characteristics and clinical course of patients with malignant eyelid tumors in an incidence cohort in Olmsted County, Minnesota. Ophthalmology 1999;106:746–50.
3. Margo CE, Waltz K. Basal cell carcinoma of the eyelid and periocular skin. Surv Ophthalmol 1993;38:169–92.
4. Font RL. Eyelids and lacrimal drainage system. In: Spencer WH, ed. Ophthalmic Pathology: An Atlas and Textbook, 4th ed. Philadelphia: W.B. Saunders, 1996; v4, chap. 11, 2270–8.
5. Jerry JA, Demirci H, Marr BP Sebaceous Carcinoma of the Ocular Region: A Major review. Survey of ophthalmology: 2005; 103-122.
6. Miller SJ: Biology of basal cell carcinoma: 11. J Am Acad Dermatol 1991; 24:161.
7. Holds JB. Basic and Clinical Science Course: Orbit, Eyelids, and Lacrimal System. San Francisco: American Academy of Ophthalmology; 2007–2008:158–176.
8. Older JJ, Grostern RJ. Eyelid Tumors: clinical diagnosis and surgical treatment. 2 ed. New York: Thieme; 2003.
9. Weber RS, Miller MJ, Goepfert H. Basal and Squamous Cell Skin Cancers of the Head and neck. Baltimore: Williams & Wilkins; 1996.

10. Spencer WH. Ophthalmic Pathology: an atlas and textbook. 3 ed. Philadelphia: W.B. Saunders; 1986:2169–2178.

11. Francis IC, Benecke PS, Kappagoda MB. A ten-year survey of eyelid cancer. Aust J Ophthalmol. 1984;12:121–127.

12. Mannor GE, Hybarger DP, Meecham WJ, et al. Epidemiology of 888 Cases of Periocular Skin Cancer. New Orleans: American Society of Ophthalmic Plastic & Reconstructive Surgery; 2001:113

13. Howard GR, Nerad JA, Carter KD, et al. Clinical characteristics associated with orbital invasion of cutaneous basal cell and squamous cell tumors of the eyelid. Am J Ophthalmol. 1992;113:123–133.

14. Leibovitch I, McNab A, Sullivan T, et al. Orbital invasion by periocular basal cell carcinoma. Ophthalmology. 2005;112;717–723.

15. Mannor GE, Hybarger CP. Factors Associated With Recurrence of Cutaneous Periocular Basal Cell Carcinoma. Anaheim: American Academy of Ophthalmology; 2003.

16. Tumbar T, Guasch G, Greco V et al: Defining the stem cell niche in skin. Science 2004; 303:359-363.

17. Tilli CM, Van Steensel MA, Krekels GA et al: Molecular aetiology and pathogenesis of basal cell carcinoma. Br J Dermatol 2005; 152: 1108-1124.

18. Marks R, Rennie G, Selwood TS: Malignant transformation of solar keratoses to squamous cell carcinoma. Lancet 1988: 1:795-797.

19. Johnson TM, Rowe DE, Nelson BR, Swanson RA: Squamous cell carcinoma of skin (excluding lip and oral mucosa). J Am Acad Dermatol 1992; 26:467-484.

20. Mannor GE, Hybarger CP, Meecham WJ, et al. Basal Cell Carcinoma of the Eyelids: an analysis of 841 cases. New Orleans: American Academy of Ophthalmology; 2001.

21. Sullivan TJ, Boulton JE, Whitehead KJ: Intraepidermal carcinoma of the eyelid. Clin Experiment Ophthalmol 2002; 30: 23-27.

22. Chow TK, Chacko E, Cleary C et al: Keratoacantoma of the lower eyelid. Eye 2005; 19: 689-690.

23. Donaldson MJ, Sullivan TJ, Whitehead KJ et al: Squamous cell carcinoma of the eyelids. Br J Ophthalmol 2002;86:1161–1165.

24. Reifler DM, Hornblass A. Squamous cell carcinoma of the eyelid. Surv Ophthalmol.1986;30:349–365.

25. Malhotra R, Huilgol SC, Huynh NT, et al. The Australian Mohs database: periocular squamous cell carcinoma. Ophthalmology. 2004;111:617–623.

26. Veness MJ. Time to rethink TNM staging in cutaneous SCC. Lancet Oncol. 2008;9:702–703.

27. Abdi U, Tyagi N, Maheshwari V, et al: Tumours of eyelid: a clinicopathologic study. J Indian Med Assoc 1996: 94; 405–9, 416,418.

28. Kass LG, Hornblass A: Sebaceous carcinoma of the ocular adnexa. Surv Ophthalmol 1989; 33:477–90.

29. Chao A, Shields CL, Krema H, Shields JA: Outcome of patients with periocular sebaceous gland carcinoma with and without pagetoid conjunctival epithelial invasion. Ophthalmology 2001; 108:1877–83.

30. Jakobiec FA, To K: Sebaceous tumors of the ocular adnexa. in Albert DM, Jakobiec FA (eds): Principles and Practice of Ophthalmology, vol 4. Philadelphia, PA, WB Saunders Co, 2000, ed 2, pp 3400–1.

31. Shields JA, Demirci H, Marr BP, et al. Sebaceous carcinoma of the eyelids. Personal experience with 60 cases. Ophthalmology. 2004;111:2151–2157.

32. Tan KC, Lee ST, Cheah ST: Surgical treatment of sebaceous carcinoma of eyelids with clinico-pathological correlation. Br J Plast Surg 1991; 44:117–21.

33. Rao NA, Hidayat AA, McLean IW, et al: Sebaceous carcinomas of the ocular adnexa: A clinicopathologic study of 104 cases, with five-year follow-up data. Hum Pathol 1982; 13: 113–22.

34. Khan JA, Doane JF, Grove AS: Sebaceous and meibomian carcinomas of the eyelid. Recognition, diagnosis, and management. Ophthal Plast Reconstr Surg 1991; 7:61–6.

35. Dzubow LM: Sebaceous carcinoma of the eyelid: treatment with Mohs surgery. J Dermatol Surg Oncol 1985; 11:40–4.

36. Boniuk M, Zimmerman LE: Sebaceous carcinoma of the eyelid, eyebrow, caruncle, and orbit. Trans Am Acad Ophthalmol Otolaryngol 1968; 72:619–42.

37. Doxanas MT, Green WR: Sebaceous gland carcinoma. Review of 40 cases. Arch Ophthalmol 1984; 102:245–9.

38. Kane WJ, Yugueros P, Clay RP, et al. Treatment outcome for 424 primary cases of clinical stage I cutaneous malignant melanoma of the head and neck. Head Neck. 1997;19:457–465.

39. Kopf AW, Rivers JK, Friedman RJ, et al. Dysplastic Nevi. In: Friedman RJ, et al, eds. Cancer of the Skin. Philadelphia: W. B. Saunders Company; 1991:125–147.

40. White GM, Neil HC. Melanocytes, Nevi, and Melanoma. In: Disease of the Skin. St. Louis: Mosby; 2002:425–444.

41. Baroody M, Holds JB. Extensive locoregional malignant melanoma transformation in a patient with oculodermal melanocytosis. Plast Reconstr Surg. 2004;113:317–322.

42. Malhotra R, Chen C, Huilgol SC, et al. Mapped serial excision for periocular lentigo maligna and lentigo maligna melanoma. Ophthalmology. 2003;110:2011–2018.

43. Barnhill RL. Tumor of melanocytes. In: Barnhill RL, Crowson AN., eds. Textbook of Dermatopathology. 2nd ed. New York: McGraw-Hill; 2004:635–707.

44. Balch CM, Buzaid AC, Soong SJ, et al. New TNM melanoma staging system: linking biology and natural history to clinical outcomes. Semin Surg Oncol. 2003;21:43–52.

45. NCCN Clinical Practice Guidelines in Oncology. Melanoma V.2.2009. http://www.nccn.org/professionals/physician_gls/PDF/melanoma.pdf.

46. Balch CM, Soong SJ, Gershenwald JE, et al. Prognostic factors analysis of 17,600 melanoma patients: validation of the American Joint Committee on Cancer melanoma staging system. J Clin Oncol. 2001;19:3622–3634.

47. Morton DL, Thompson JF, Cochran AJ, et al. Sentinel-node biopsy or nodal observation in melanoma. N Engl J Med. 2006;355:1307–1317.

48. Esmaeli B, Youssef A, Naderi A, et al. Margins of excision for cutaneous melanoma of the eyelid skin. Ophthal Plast Reconstr Surg. 2003;19:96–101.

49. Merkel F. Tastzellen und Tastkorperchen bei den Hausthieren und beim Menschen. Arch Mikroskopisch Anat (Bonn). 1875;11:636–652.

50. Winkelmann RK, Breathnach AS. The Merkel cell. J Invest Dermatol. 1973;60:2–15.

51. Buck CB, Lowy DR. Getting stronger: the relationship between a newly identified virus and Merkel cell carcinoma. J Invest Dermatol. 2009;129:9–10.

52. Liao P. Merkel cell carcinoma. Dermatol Ther. 2008;21:447–451.

53. Nicoletti AG, Matayoshi S, Santo RM, et al. Eyelid Merkel cell carcinoma: report of three cases. Ophthal Plast Reconstr Surg. 2004;20:117–121.

54. National Comprehensive Cancer Network. NCCN clinical practice guidelines in oncology: Merkel cell carcinoma. V.1.2008. Available at: www.nccn.org.

55. Allen PJ, Browne WB, Jaques DP etal. Merkel cell carcinoma: prognosis and treatment of patients from a single institution. J Clin Oncol. 2005; 23:2300–2309.

56. Swann MH, Yoon J. Merkel cell carcinoma. Semin Oncol. 2007; 34:51–56.

57. Anderson RL, Ceilley RI: A multispeciality approach to exicision and reconstruction of lid tumors. Ophthalmology. 1978;85:1150-1163.

58. Linberg JV: Periocular basal cell carcinoma in young adults. W V Med J. 1985;81:241-244.

59. Hughes WL. Total lower lid reconstruction:technical details. Trans Am Ophthalmol Soc. 1976; 74:321-329.

60. Mustarde JC. Repair and reconstruction in the orbital region. Edinburgh: Churchill Livingstone;1971.

61. Cutler NL, Beard C. A method for partial and total upper lid reconstruction. Am J Ophthalmol. 1955;39:1-7.

62. Mauriello JA Jr., Antonacci R: Single tarsoconjunctival flap(lower eye lid) for upper eyelid reconstruction (reverse modified Hughes procedure). Ophthalmic Surg. 1994;25:374-378.

63. Stephenson AJ, Griffiths RW, La Hausse-Brown TP: Patterns of contraction in human fullthickness skin grafts. Br J Ophthalmol. 2000;53:397-402.

64. Klapper SR, Patrinely JR: Eyelid and periorbital reconstruction. in Albert DM, Jakobiec FA (eds): Principles and Practice of Ophthalmology, vol 3. Philadelphia, PA, WB Saunders Co, 2008, ed 3, pp 3323-3342.

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Amblyopia Treatment Studies: What We Learn from them

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Amblyopia is defined as a unilateral or bilateral decrease of visual acuity caused by form vision deprivation and /or abnormal binocular interaction for which no organic causes can be detected by physical examination of eye and which in appropriate cases is reversible by therapeutic measures. It is the most common cause of monocular visual impairment in both children and young adults affecting 1-5% of the world population.

Causes of amblyopia include visually significant conditions causing stimulus deprivation eg. monocular or bilateral cataract, vitreous haemorrhage, ptosis, prolonged occlusion of one eye or conditions leading to abnormal binocular interaction eg. strabismus, anisometropia, aniseikonia, asymmetric or monocular cataract during the period of developmental plasticity of the visual system. Clinically, a difference of 2 or more lines between the normal and amblyopic eye in unilateral amblyopia or a reduction in best corrected central visual acuity to less than 6\12 is without any visible organic cause, is amblyopia.

Based on etiology, amblyopia may be classified into the following types -

- Strabismic
- Refractive
  - Iso-ametropic
  - Anisometropic
- Stimulus deprivation
- Organic- due to subclinical foveal malfunction or malorientation of cones
- Nystagmus related

Amblyopia is diagnosed by detecting any or a combination of the above amblyopiogenic conditions, preference of fixation by one eye or resistance to occlusion of the dominant eye in the absence of any alternate explanation for visual loss. Assessment of amblyopia is made by testing for visual acuity using preferential looking tests, optokinetic nystagmus and visual evoked potential in pre-verbal children and by using E chart, figure charts or alphabet charts like the Snellens or the ETDRS in older children.

Management of amblyopia aims at: –

- Providing the affected eye as clear foveal image as possible and as early as possible
- Providing visual advantage to the amblyopic eye compared to the better eye

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Treatment options include: –

- Refractive correction
- Occlusion therapy
- Penalization
- Drug therapy
- Pleoptics
- CAM stimulator
- Home vision therapy
- Surgery to treat the underlying cause of amblyopia

Refractive correction: The most fundamental part of the treatment of amblyopia is correction of refractive error so as to produce a clear retinal image at all times for both distance and near. A proper cycloplegic refraction is done in every case of amblyopia. The ATS-5 (eye glass phase study) was conducted by the PEDIG (Pediatric Eye Disease Investigator Group) to study the role of refractive correction alone in the treatment of amblyopia. 84 children with untreated anisometropic amblyopia aged 3 -7 years were given optimal refractive correction and visual acuity was measured at 5-week intervals until visual acuity stabilized or amblyopia resolved.Amblyopia improved with optical correction by 2 or more lines in 77% of the patients and resolved in 27%.

Occlusion: Occlusion therapy is the most effective means for treating amblyopia. Occlusion can be partial or total, full time or part- time, conventional or inverse. Though the amblyopia treatment study has concluded that penalization with atropine is as effective as patching and part time occlusion has similar efficacy as full time occlusion, we recommend totally opaque full-time occlusion therapy of the good eye alternated with a similar patching of the amblyopic eye. The ratio of patching the amblyopic eye and the sound eye depends on the age of the child.

Pediatric Eye Disease Investigator Group: (PEDIG) is a collaborative network dedicated to facilitating multicenter clinical

Table 1: Ratio of patching – dominant eye: amblyopic eye

Age (Years)	Occlusion of dominant eye (Full Time)	Occlusion of amblyopic eye
Up to 2	2d	1 day
Up to 3 Up to 4 Up to 5 Up to 6 and above	3d 4d 5d 6d	Alternating with conventional occlusion
At no time are both eyes open		

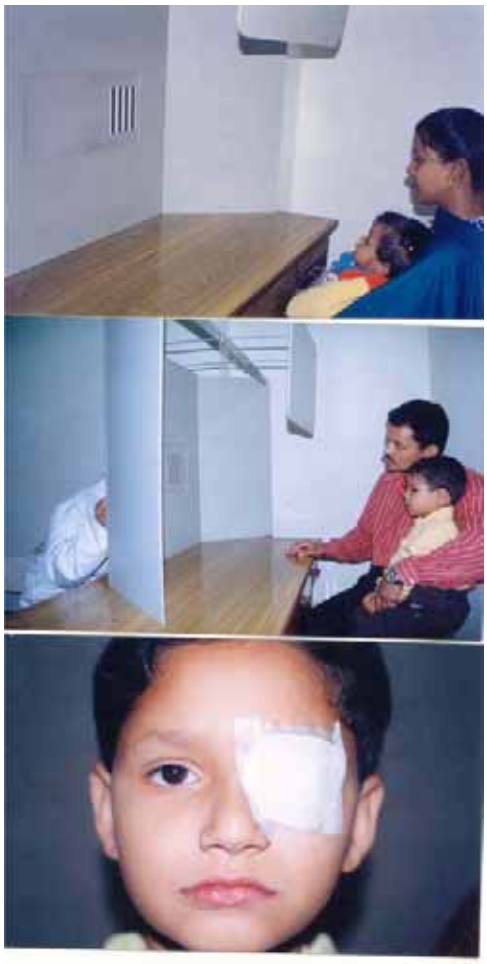

Figure 1: *(top, middle) Testing teller acuity. (bottom) Low cost occlusion patches using chart paper and 3-M micropore tape*

research in strabismus, amblyopia and other eye disorders that affect children. The network, which was formed in 1997, is funded by the National Eye Institute (NEI). In ATS- 1, 419 children of moderate amblyopia aged 3 to 7 years from 47 clinical centers in USA were randomized to patching or atropine. The study used isolated surrounded HOTV optotypes and the protocol was evaluated using Bayler-Video-Acuity-Tester (BVAT). Improvement was initially faster in the patching group, but after 6 months the difference in visual acuity between treatment groups was insignificant. (3.16 lines in the patching group vs 2.84 lines in the atropine group).

In a study done from our centre in patients aged 8-20 years with anisometropic amblyopia, faster recovery was seen in the patching group compared to the atropine group.

The ATS 2 study was conducted to compare 6 hours of patching with full-time occlusion for severe amblyopia (20/100 to 20/400) in children 3 to 7 years old. The cause of the amblyopia was strabismus in 27% patients, anisometropia in 34%, and both strabismus and anisometropia in 38%. At 4 months, there was no difference in amblyopic eye acuity between *groups* (mean difference at 4 months: 86% of patients in the 6-hour *group* and 82% of patients in the full-time *group* had acuity in the amblyopic eye that had improved from baseline by ≥3 lines from baseline. Another protocol compared 2 vs 6 hours of daily patching of the sound eye as treatments for moderate amblyopia (20/40 to 20/80) in children 3 to 7 years old. At 4 months: 62% of patients in each *group* had acuity in the amblyopic eye that was 20/30 or better and/or had improved from baseline by 3 lines. There did not seem to be a benefit to the greater number of hours of prescribed patching on the rate of improvement.

The patients during occlusion therapy are advised a regular follow up after every one month to look for visual acuity, fixation pattern and presence of occlusion amblyopia. Infants are usually followed up more frequently, after every fortnight to prevent occlusion amblyopia.

Possible problems with occlusion therapy

- Non-compliance- rate varies (49-87%)
- Functional problems especially in severe amblyopia
- Cosmetically unacceptable
- Occlusion amblyopia
- Constant manifest deviation in a child who did not have deviation or had an intermittent deviation
- Allergic skin rash
- Diplopia
- Increased angle of deviation
- Psychological problems

When to stop occlusion?

- When the visual acuity becomes equal in both eyes
- True alternation of fixation
- No visual improvement after 3-6 months of occlusion despite good compliance.

Following completion of amblyopia treatment, one may switch to part-time patching and finally weekly patching till nine years of age along with periodic follow-ups to prevent regression.

The prognosis depends on

- *Compliance* – The single most important factor determining the success of amblyopia therapy is patient compliance.
- *Age of the patient*- While improvement occurs rapidly below 3 years of age, chances of improvement decrease significantly beyond 6 years. Recent studies have shown that though reduced, the success rate of treatment of anisometropic amblyopia even beyond 12 years of age is good.
- *Type of amblyopia*- Strabismic amblyopia shows maximum recovery following occlusion while stimulus deprivation amblyopia fares the worst.
- *Pre-treatment visual acuity*- Poor pre-treatment visual acuity is an independent risk factor for poor prognosis.

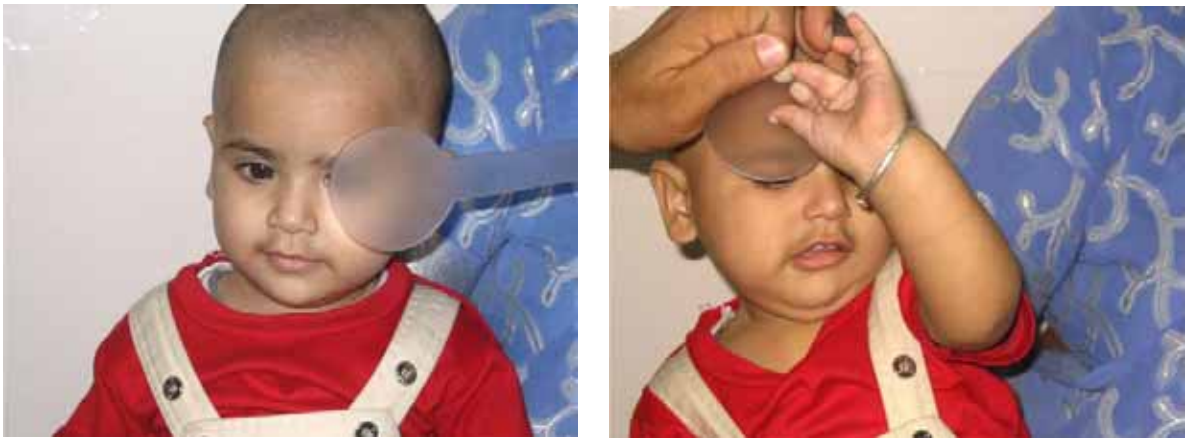

Figure 2: *Left amblyopia. Child is comfortable on covering the left eye bot does not allow covering of the right eye*

- *Type of occlusion* - Conventional full time total occlusion gives the earliest and most effective results. The rate of improvement depends on the monocular fixation pattern of the amblyopic eye.
- *Type of fixation* - Foveal fixation responds quickly to therapy while eccentric fixation has a poor prognosis. Though inverse occlusion has been tried in amblyopia with steady eccentric fixation, the results have not been satisfactory.
- *Near exercises* - Exercises such as threading beads, tracing pictures and reading fine prints augment the effect of occlusion therapy and should be encouraged.
- *Previous treatment* -Although some studies show that presence or absence of previous treatment had no effect on the amount of improvement in visual acuity, ATS-3 showed that patching 2 to 6 hrs\day with near visual activities may improve visual acuity when amblyopia has not been previously treated but appears to be of little benefit if amblyopia was previously treated with patching. (47% vs 20%). However failed previous attempted treatment should not prevent attempt to retry therapy.

Penalization: Penalization, though being cosmetically more acceptable, does not inhibit abnormal binocular interaction which had resulted in amblyopia. So, it has limited indications in our setup, despite the recommendations of ATS. Its main role is as maintenance therapy and also in patients of moderate amblyopia, in children uncooperative for occlusion, occlusion failure and possibly in Manifest-Latent nystagmus (Fusion Maldevelopment syndrome).

The ATS 4 was conducted to compare daily atropine to weekend atropine as prescribed treatments for moderate amblyopia in children younger than 7 years. Weekend atropine provided an improvement in visual acuity of a magnitude similar to that of the improvement provided by daily atropine in treating moderate amblyopia (53% vs 47% had vision e" 20\25 at the end of 4 months). Stereoacuity outcomes were similar in the 2 groups.

Surgery: Surgery to treat the amblyopigenic factors promote visual recovery However, early surgery doesn't prevent against amblyopia.

Occlusion should always be done prior to squint surgery in cases of strabismic amblyopia since

- A prior surgery with alignment makes the task of assessment of dominant eye by preference of fixation difficult
- Motivation of parents get slackened after cosmetic correction and their commitment for occlusion gets lax
- An occlusion later may induce squint again.

Combined therapy

Combined optical and atropine penalization (COAT) is an effective treatment when occlusion therapy fails initially, and it might have a more rapid effect than single modality penalization therapy, but there is an increased risk of reverse amblyopia. Its effect may be particularly useful in anisometropic amblyopia.

Medical Treatment

Levodopa: Levodopa either extends or reactivates the visual system's sensitive period of neural plasticity thus leading to an increase vision in the amblyopic eyes (1 or 2 Snellen lines) for short periods (about a month), even in adults. The recommended dosage is 0.48mg\kg body weight. However, there can be side effects following drug administration like nausea, vomiting, diarrhoea, heart burn, abdominal pain, sleepiness, mood change, decreased heart rate and respiration, tiredness, hallucinations. Although some studies have shown beneficial effect of this treatment in combination with occlusion therapy, and that it might prolong the critical period during which occlusion is effective, its effect is thought to be temporary. In a study conducted in our centre, levodopa supplementation did not offer any advantage over occlusion alone. Moreover, the risk of occlusion amblyopia could increase with the use of drugs like levodopa that might affect the plasticity of the visual cortex. Recently, there is an ongoing

Table 2: Randomized trials of amblyopia treatment study					
	Aim	**No of patients**	**Age**	**Follow up**	**Recommendations**
ATS-1	To compare atropine and patching treatments for moderate amblyopia	419	3-7 yrs	6 mts-2 yrs	Improvement was initially faster in the patching group, but after 6 months the difference in visual acuity between treatment groups was insignificant. 3.16 lines (patching group) vs 2.84 lines (atropine group)
ATS-2A	To compare 6 hrs vs full time daily occlusion for severe amblyopia	175	3-7 yrs	3-7 yrs	6 hrs of patching equally effective as full time patching.86% pts in the 6-hour group and 82% pts in the full-time group had improved by 3 lines from baseline.
ATS-2B	To compare 2 versus 6 hours daily patching for moderate amblyopia	189	3-7 yrs	4 mts	6 hrs of patching as effective as 2 hrs of patching. At 4 months, no difference in amblyopic eye acuity between groups
ATS-3	To evaluate the effectiveness of optical correction alone vs 2-6 hrs\day of patching combined with near visual activities plus atropine sulfate for children aged 7 to 12 years.	507	7-13 yrs 13-17 yrs	6 mts	Amblyopia improves with optical correction alone in 1\4th pts (7 to 17 yrs), although most pts initially treated with optical correction alone will require additional treatment for amblyopia. Patching 2 to 6 hrs\day with near visual activities may improve visual acuity when amblyopia has not been previously treated but appears to be of little benefit if amblyopia was previously treated with patching.
ATS-4	To compare daily atropine to weekend atropine as prescribed treatments for moderate amblyopia	168	3-7yrs	4 mts	Weekend atropine provides an improvement in VA of a magnitude similar to that of the improvement provided by daily atropine in treating moderate amblyopia.47% in the daily group had vision e"20/25 at 4th mt compared to 53% in the weekend group. Stereoacuity outcomes similar.
ATS-5	Eye glass phase study	84	3-7yrs	30 wks	Ambylopia improved in 77% by optical correction and resolved in 27%.
ATS-5	To evaluate 2 hrs of daily patching for amblyopia moderate amblyopia	180	3-7 yrs	5 wks	Increase in visual activity was more if near activities were combined with patching. After five weeks, the patching group had improved by an average of 0.6 lines more than the control group(optical correction alone)
ATS-6	Comparing near and distance activities while patching	425	3-7 yrs	17 wks	No difference in visual acuity improvement between children performing near activities and distance activities during patching.
ATS-7	To determine improvement in binocular visual acuity during treatment of bilateral refractive amblyopia	113	3-11 yrs	1 yr	Bilateral refractive amblyopia improves with spectacle correction. Binocular visual acuity of 20/25 or better was achieved by 73%
ATS-8	To compare weekend atropine augmented by a plano lens with weekend atropine alone for moderate amblyopia	180	3-7 yrs	18 wks	Augmentation of weekend atropine with a plano lens does not substantially improve amblyopic eye acuity.
ATS-9	To compare patching with atropine eyedrops in the treatment of moderate amblyopia	193	7-12 yrs	17wks-10mts	Atropine and patching achieve similar results among older children with unilateral amblyopia.

study ATS -14, designed to evaluate two doses of levodopa (0.51 mg/kg/tid levodopa and 0.17 mg/kg/tid) as an adjunctive treatment to 2 hours of patching for residual amblyopia (20/50 or worse) in older children and teenagers between 8 and 17 years of age. Subjects will be randomized to receive 0.51 mg/kg/tid levodopa or 0.76 mg/kg/tid levodopa along with 0.17 mg/kg/tid carbidopa for a period of 8 weeks.

Citicholine (Cytidin-5-diphposphocholine): It is similar in action to levodopa. When administered in adult patients with strabismic amblyopia (1gm\day i.m for 15 days), it has showed improvement lasting over 6 months including improvement in contrast sensitivity and VEP.

Role of near activities

PEDIG conducted a multicenter pilot study to determine if children randomized to near or non-near activities would perform prescribed activities, and to estimate the effect of near activities in visual acuity of the amblyopic eye combined with two hours of daily patching (ATS-6). Sixty-four children aged 3 to less than 7 years old, with strabismic and/or anisometropic amblyopia (20/40 to 20/400) were randomly assigned to receive either 2 hours of daily patching with near activities or 2 hours of daily patching without near activities. After 4 weeks of treatment, there was a suggestion of greater improvement in amblyopic eye visual acuity in those assigned to near visual activities (mean 2.6 lines versus 1.6 lines). The treatment group difference in visual acuity was present for patients with severe amblyopia but not moderate amblyopia.

Other modes of treatment

Liquid crystal glasses -Liquid crystal glasses have recently been developed as a new treatment for amblyopia. They provide an electronic, controlled, intermittent occlusion of the sound eye allowing for visual stimuli input to the amblyopic fellow eye. A liquid crystal glass in the sound eye is used as an intermittent flickering shutter switched between "on", or occlusion, and "off", or light transmission. The flickering sequence can be adapted to the depth of amblyopia, the length of treatment, and the patient's age.

Opaque (occluder) contact lenses- Occlusive contact lenses can be used in treating amblyopia in children who are patch-intolerant and resistant to conventional treatment. However, patients should have close follow up to monitor any anterior segment complications from contact lens use and to identify the patients with occlusion amblyopia.

Penalizing filters: Ryser or Bangerter foils, which come in successive graduated densities, may be used to reduce visual acuity of the sound eye to less than the amblyopic eye, or to a poor level of visual acuity in all cases. Sometimes, adhesive tape or nail polish was used as a readily available procedure to produce fogging in the sound eye. These methods are used in mild amblyopia or as maintenance therapy, in school age cooperative children.

Macular stimulation with telescopic magnification – Older children (7-18 years) with anisometropic amblyopia not amenable to any further medical or surgical treatments have been tested with some improvement in vision.

CAM Vision Stimulator - Amblyopic eye is stimulated by slowly rotating, high contrast, square wave gratings of different spatial frequencies for 7 minutes but the results are not much promising.

Pleoptics Therapy - Active stimulation of the macula is done using Pleoptophore (Modified Gullstrand's ophthalmoscope), till fixation becomes central.

Red Filter Treatment - Red filter which excludes wavelengths <640 nm is used to stimulate cones at the fovea. Red light is ineffective in stimulating the eccentric point as compared to fovea, due to the lack of cones.

Recurrence of amblyopia after treatment

Recent studies suggest that approximately 20% to 25% of patients suffer amblyopia recurrence after successful treatment during the first year without therapy. PEDIG found that the risk of recurrence was higher in those with better visual acuity at the time of cessation of treatment, a greater number of lines improved during the previous treatment and a prior history of recurrence. Orthotropia or excellent stereoacuity at the time of patching cessation did not appear to have a protective effect on the risk of recurrence.

Occlusion amblyopia occurs when visual acuity decreases in the sound eye during amblyopia treatment. Clinically, occlusion amblyopia can come up from excessive administration of treatment by patching or penalization, but is not frequent and when it arises, it is generally transient and reversible. Treatment of suspected occlusion amblyopia consists of checking refraction and vision, stopping active treatment, and finally treating the previously sound eye.

In the ATS, visual acuity in the sound eye at 6 month examination was decreased by 1 line in 7% of patients in the patching group and 15% in the atropine group. A two or more lines decrease was seen in 1% of the patching group and 9% of the atropine group. Only 1 patient (from atropine group) was actively treated for a presumed occlusion amblyopia, with a return of visual acuity to its baseline level.

In conclusion, though the ATS studies have provided a lot of understanding of the options available for the treatment of amblyopia, full time total occlusion still remains the gold standard in the treatment of amblyopia. However the alternatives do appear to be successful and may be tried in certain cases. Also as many studies have shown that older children also improve with treatment, amblyopia therapy should be advised in all patients willing to try.

Ongoing trials-

- ATS-10 Bangerter filters for amblyopia
- ATS-11 Final Ramp-up for residual amblyopia
- ATS-12 Vision Therapy Pilot Study
- ATS-13 Current Spectacles for Strabismic Amblyopia
- ATS-14 Levodopa Pilot Study
- ATS-15 Increasing Patching for Amblyopia
- ATS-16 Augmenting Atropine Treatment for Amblyopia

References

1. Noorden GK Von. Practical management of amblyopia. International Ophthalmology. 1983;6:7-12.

2. Ehrlich MI, Reinecke RD, Simons K. Preschool vision screening for amblyopia and strabismus. Programs, methods, guidelines, 1983. Surv Ophthalmol. 1983;28:145-63.

3. Von Noorden GK. Classification of amblyopia. Am J Ophthalmol 1967;63:238-44.

4. Treatment of Anisometropic Amblyopia in Children with Refractive Correction-Pediatric Eye Disease Investigator Group. Ophthalmology. 2006 ; 113(6): 895–903.

5. A Randomized Trial of Atropine versus Patching for Treatment of Moderate Amblyopia: Follow-up at 10 Years of Age.Pediatric Eye Disease Investigator Group. Arch Ophthalmol. 2008 ; 126 : 1039–1044.

6. Menon V, Shailesh G, Sharma P, Saxena R. Clinical trial of patching versus atropine penalization for the treatment of anisometropic amblyopia in older children. J AAPOS. 2008 Oct;12(5):493-7.

7. [PEDIG] Pediatric Eye Disease Investigator Group. The course of moderate amblyopia treated with atropine in children: experience of the amblyopia treatment study. Am J Ophthalmol. 2003;136:630–9.

8. [PEDIG] Pediatric Eye Disease Investigator Group. A randomized trial of patching regimens for treatment of moderate amblyopia in children. Arch Ophthalmol. 2003;121:603–11.

9. Loudon S. E,Simonsz B,Joosse M.Vet al. Electronic Recording of Patching for Amblyopia Study: Predictors for non–compliance. Invest Ophthalmol Vis Sci 2004;45:

10. Scheiman MM, Hertle RW, Beck RWet al.Randomized trial of treatment of amblyopia in children aged 7 to 17 years. Arch Ophthalmol. 2005;123:437-47.

11. Repka MX, Cotter SA, Beck RW et al.A randomized trial of atropine regimens for treatment of moderate amblyopia in children. Ophthalmology. 2004;111:2076-85

12. Kaye SB, Chen SI, Price G et al.Combined optical and atropine penalization for the treatment of strabismic and anisometropic amblyopia. J AAPOS. 2002 Oct;6(5):289-93.

13. Bhartiya P, Sharma P, Biswas NR et al. Levodopa-carbidopa with occlusion in older children with amblyopia. J AAPOS. 2002 ;6:368-72.

14. Holmes JM, Edwards AR, Beck RW et al. A randomized pilot study of near activities versus non-near activities during patching therapy for amblyopia. J AAPOS. 2005;9(2):129-36.

15. Eustis HS, Chamberlain D. Treatment for amblyopia: results using occlusive contact lens. J Pediatr Ophthalmol Strabismus. 1996;33:319-22.

16. Pediatric Eye Disease Investigator Group Risk of amblyopia recurrence after cessation of treatment J AAPOS 2004; 8:420-8.

First Author
Ruby Misra MD

Congratulations

- **Prof. Jeewan S. Titiyal** performed Live Surgery at American Society of Cataract & Refractive Surgery (ASCRS) meeting held on 9th - 14th April, 2010 at Boston (USA). First Indian Ophthalmologist to perform Live Surgery in this prestigious ASCRS International Conference.

Surgical Management of Ectatic Corneal Disorders

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Ectatic disorders of the cornea are characterized by progressive corneal thinning, irregular astigmatism and decreased visual acuity.[1] They comprise of primary conditions such as keratoconus and pellucid marginal degeneration, as well as iatrogenic corneal ectasia, which may occur after refractive procedures such as laser in-situ keratomileusis (LASIK) surgery.

Over the last decade there has been a paradigm shift from penetrating keratoplasty to lamellar keratoplasty for the management of primary as well as acquired corneal ectasia. Several treatment modalities have emerged including collagen cross-linking, intrastromal implants, laser vision correction, and recent techniques in central and peripheral lamellar keratoplasty. This article reviews important surgical approaches to the management of ectatic corneal diseases.

Deep Lamellar Keratoplasty (DLK)

Penetrating keratoplasty (PKP) is the most commonly performed solid organ transplanatation and has enjoyed a relatively high success rate compared with transplantation of other tissues. However, endothelial graft rejection is observed in approximately 20% of cases undergoing PKP.[2] Although the results of PKP in cases of keratoconus are very good, variable number of keratoconus patients experience one or more endothelial rejection episodes, causing graft decompensation. Deep lamellar keratoplasty (DLK) is a surgical technique that can eliminate the risk of corneal endothelial graft rejection, with comparable optical results to PKP. DLK has been successfully used to treat various corneal pathologies that spare the corneal endothelium.[3,4]

The concept of a 'true' deep anterior lamellar keratoplasty (DALK) extending down to Descemet's membrane was proposed not very long ago. Older literature does not expand on the actual depth of 'deep' lamellar keratoplasty. Gasset reported a series of keratoconus patients in the late 1970s who underwent 'conectomy' and received full-thickness grafts stripped of Descemet's membrane transplanted into relatively deep lamellar beds. Dissection of host tissue 'close to' the Descemet's membrane and the term 'deep lamellar keratoplasty' was first introduced by Archilla in 1984 with the use of intrastromal air injection to facilitate removal of diseased host corneal tissue. The first study on the results of DLK compared with PKP in keratoconus was reported by Sugita and Kondo in 1997.[5] They showed that postoperative visual acuity was similar after DLK and PKP in cases of keratoconus. Recently DALK has gained due credit due to improvements in surgical techniques, and the availability of new surgical instruments and viscoelastics that have helped to improve surgical success and reduce surgery time.

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Techniques of DLK

The classical technique for DLK involves the removal of host tissue layer by layer until the deep stroma or the Descemet's membrane is bared. While stromal fibers are difficult to visualize when the amount of tissue becomes minimal, injection of irrigation fluid causes swelling of stromal fibers that can then be manipulated. The two techniques that have become popular in recent times are Melles technique and "big bubble" technique of DLK described by Anwar.

Melles Technique

Melles[6] *technique involves injection of* air into the anterior chamber that creates a mirror reflex to guide surgical instruments directly into the space between Descemet's membrane and the posterior stroma. The difference in refractive index between air and corneal tissue creates a reflex of the surgical knife, and the distance between the instrument and reflex can be used to judge the amount of stromal tissue. The blunt end of a microsurgery knife is used to dissect the stroma down to DM, using the reflection of the knife observed at the air-to-endothelium interface as a guide. After creation of a small DM detachment with BSS, viscodissection is performed to further extend DM detachment. After complete dissection of DM, the overlying stroma is removed to expose the smooth surface of DM.

Modifications in Melles technique

Shimmura *et al*[9] modified the Melles technique by performing anterior lamellar keratectomy prior to air injection. Senoo *et al*[10] *have used* a sclerolimbal approach for performing DLK. The method uses trabeculectomy to detach DM. A flap is made, as in trabeculectomy, and the region directly above Descemet's membrane is reached under direct vision. DM is detached by hydrodelamination and viscoelastic material is used to maintain the supra DM space. Parmar *et al*[11] used a 5-mm-long scleral incision for corneal dissection close to the level of the Descemet membrane. Using this technique, a scleral pocket incision is created with a crescent knife and dissection is carried into the clear cornea. Viscoelastic is injected into the scleral pocket for facilitating the separation of DM from the corneal stroma.

Funnell *et al*[7] compared the outcomes and complications of deep lamellar keratoplasty using Melles technique and penetrating keratoplasty for keratoconus. There was no significant difference in the proportion of patients achieving 6/9 or better between the PK and DLK groups. The study found that DLK causes less astigmatism and also has the advantage of no endothelial graft rejection. In another study, Watson *et al*[8] compared the DLK and PK using Melles technique in patients with keratoconus. They found that best-corrected visual acuity, refractive results, and complication rates were similar in both groups.

Big Bubble Technique

Archila introduced the technique of air injection between the DM and the overlying corneal stroma. In this technique a 26-guage

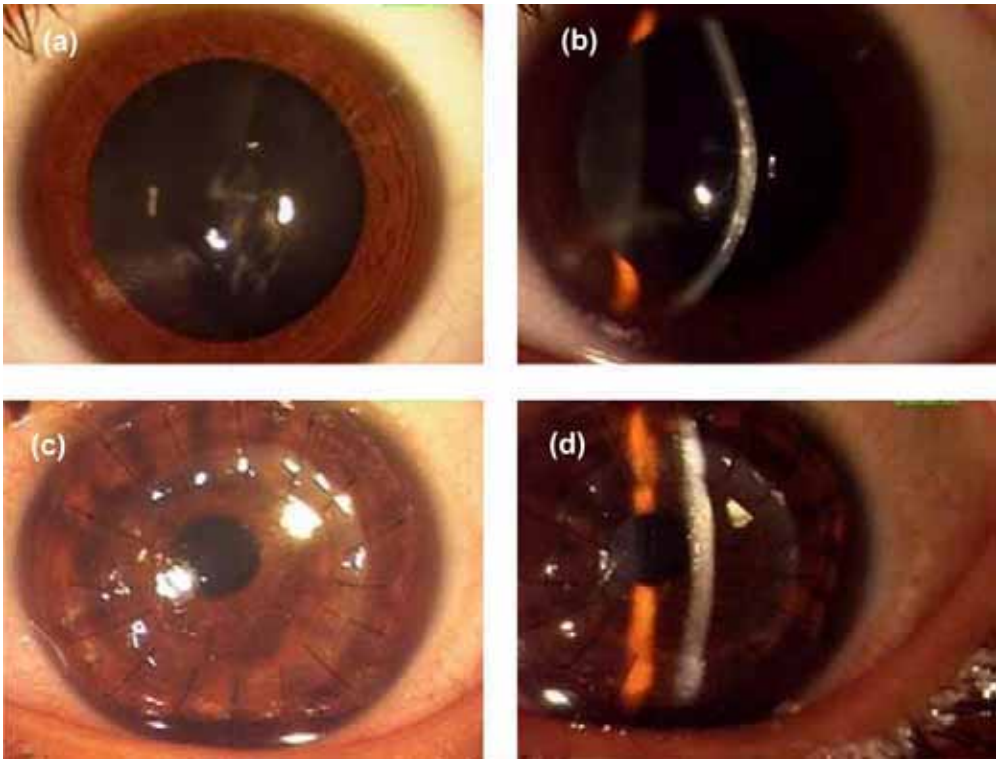

Figure 1(a): *Preoperative keratoconus (diffuse),* ***(b):*** *Preoperative keratoconus (slit)*
(c): *Post DLK-2 weeks (diffuse),* ***(d):*** *Post DLK-2 weeks (slit)*

needle connected to a tuberculin syringe filled with air is inserted obliquely into the stroma up to the corneal midperiphery. Air is injected and corneal stromal trephination is done. Dissection of corneal stroma is facilitated with a spatula to separate the Descemet's membrane from the deeper stromal layers.[12]

Anwar *et al*[13] modified Archila's technique by performing corneal trephination before air injection. About 60%-80% of the corneal stroma is trephined with the help of a suction trephine. A 27-guage needle attached to an air-filled syringe is bent at about 60° angulation 5 mm from its tip. The plunger of the air-filled syringe is depressed in order to form the big bubble between the Descemet's membrane and the deepest stroma. A partial thickness keratectomy is done with the help of a Beaver blade leaving a layer of corneal stroma in place. Using a sharp-tipped blade, held tangentially to the cornea, a small nick is made in the corneal stroma. Dissection can be carried out in this plane with the help of a spatula and long scissors.

Since DLK does not involve replacement of DM or endothelium, the donor quality criteria are not stringent. DM and endothelium is stripped off from the donor button which is then sutured over the host bed using 10-0 monofilament nylon sutures. The disparity between host cut and donor button is usually between 0.25 mm to 0.5 mm, the diameter of the graft button being larger[16]. Some corneal surgeons prefer to use a same size or an undersized donor button in patients with keratoconus[4] (Figure 1).

Several studies have shown good results with DALK technique (Table 1).

Modifications in the technique of DLK by air injection

Fournie *et al*[14] have described a modification of Anwar's big bubble technique. The initial air injection is made in the superficial corneal stroma. The aim of the first air injection is to induce corneal emphysema and facilitate superficial lamellar keratectomy. Subsequent dissection is carried out with the help of viscoelastic. Recently, Parathasarathy *et al*[15] reported a method of using a small air bubble in the anterior chamber to help determine if a successful big bubble has been achieved. The small bubble helps the surgeon to assess the extent of the big bubble in cases where the cornea is opaque or when air diffusion into the peripheral cornea prevents direct visualization into the anterior chamber.

The major complication encountered during DLK is intraoperative perforation of the DM. The incidence of Descemet's membrane perforation during DALK depends on the surgical technique and the expertise of the surgeon. Reported incidences vary in different studies.[16,18,19] Keratoconus patients are more prone to Descemet's membrane ruptures than patients with other disease, either due thinner corneas or due to an intrinsic property of the disease.[20]

DM perforation can be in the form of a microperforation or a macroperforation. Microperforations in the peripheral cornea can be manged by carewful streomal dissection and air injection at the end of the surgery. If a microperforation occurs in the central cornea, there is a risk of formation of double anterior chamber in the postoperative period. Injection a mixture of SF6 with air, or a mixture of C3F8 with air into the anterior chamber can also be

Table 1: Results of Big Bubble Deep anterior lamellar keratoplasty						
Authors	**Year**	**Number of eyes**	**Indication**	**BCVA e"20/40**	**Perforation rate**	**Rejection**
Anwar and Teichmann	2002	181	Keratoconus	Not reported	9%	None reported
Al-Torbak et al	2006	127	Keratoconus	75%	13%	3%
Fogla et al	2006	13	Keratoconus	100%	15%	None
Bahar et al	2008	17	Keratoconus	100%	7.6%	7.6%
Fontana et al	2007	81	Keratoconus	100%	13%	2%
Feizi et al	2010	129	Keratoconus	78%	4%	14.3%

used for temporarily sealing microperforations, or for flattening the secondary anterior chamber that tends to form after perforation[21].

Conversion to a PKP may be required in some cases. In such a scenario, a complete dissection of the host cornea should be carried out. 0.01% Trypan blue dye may be used to delineate any retained pieces of DM as well as to facilitate dissection of the trephined host cornea[22].

A rather rare complication after DLK is corneal stromal graft rejection that is characterized by sudden-onset decreased vision, subepithelial infiltrates, with or without stromal edema or anterior segment activity.[23] These cases are treated with prednisolone acetate 1% drops gradually tapered over 4 to 6 weeks.

Automated Lamellar Therapeutic Keratectomy (ALTK)

Microkeratome assisted lamellar keratoplasty is another novel technique used for surgically treating keratoconus and other corneal pathologies sparing the endothelium[24,25]. The major advantage of ALTK is that the donor cut is smooth which eliminates the risk of interface haze that can otherwise result in poor visual quality. Also, the dissection is easy to perform, and shortens surgical time considerably. The surgical technique offers more control in the depth of dissections and can be fairly standardized. The surgery does not require dissection up to the level of DM therefore reducing the chances of perforation of DM.

ALTK has been primarily developed to treat cases of keratoconic corneas with a minimum corneal thickness of 380 microns[25]. A 250

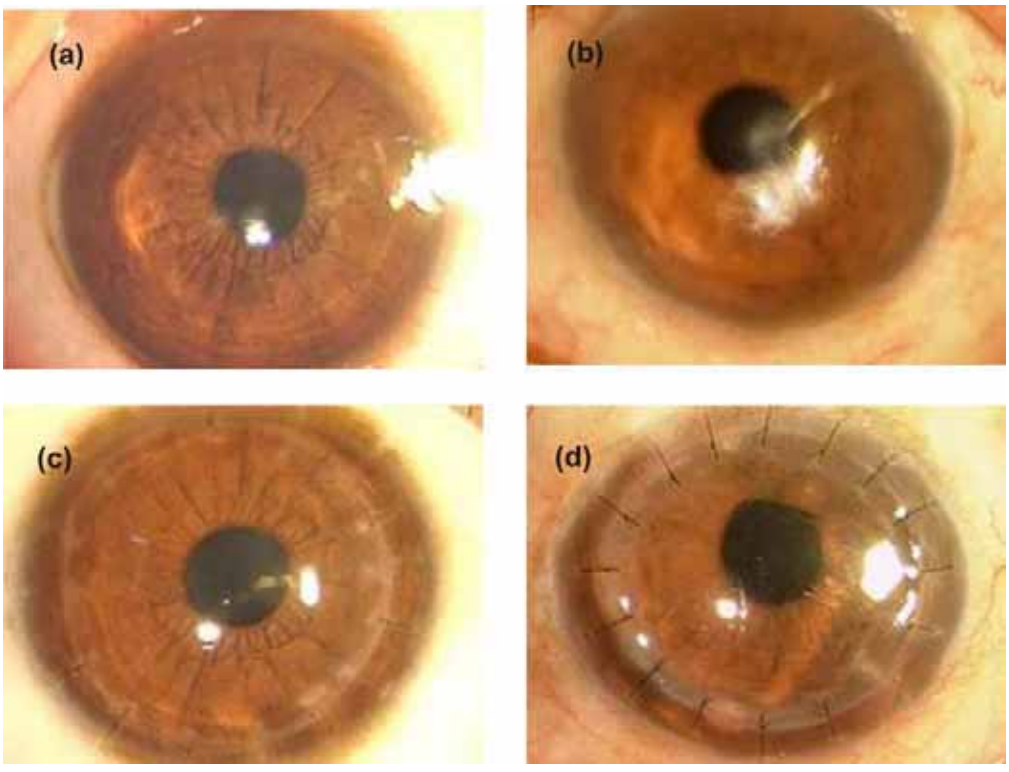

Figure 2(a): *Preoperative keratoconus (diffuse),* ***(b):*** *Preoperative keratoconus (slit)*
(c): *Post ALTK-6 months (diffuse),* ***(d):*** *Post ALTK-4 weeks (diffuse)*

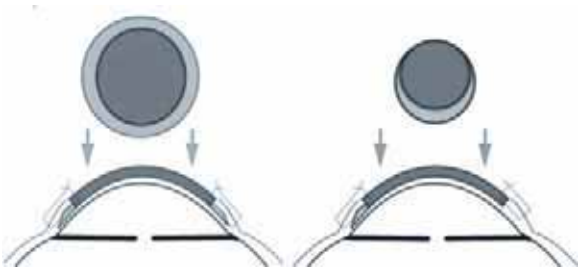

***Figure 3**: Schematic representation of a 360 degree tuck for keratoglobus (left), and an inferior Tuck for peripheral marginal degeneration (right).*

microns anterior corneal disc of host is excised by a microkeratome and a 350 microns thick donor corneal disc is transplanted. The donor lenticule is also harvested using a microkeratome and an artificial anterior chamber. The desired diameter of the donor lenticule is achieved by using different suction rings. (Figure 2)

Busin *et al*[25] evaluated the visual and refractive results of ALTK in patients with keratoconus with minimal corneal thickness of 380 microns. All patients underwent a standard ALTK surgical procedure. At the end of one year best spectacle corrected visual acuity e"20/40 and refractive astigmatism d" 4 diopters was achieved in majority of patients. The major complications reported in this study included irregular astigmatism (22%), high-degree astigmatism requiring secondary intervention (12%), epithelial interface in growth (2%), and cataract formation (2%).

"Tuck in" Lamellar Keratoplasty (TILK)

Tuck in lamellar keratoplasty is a special technique of partial thickness corneal transplantation that has been described for cases of advanced peripheral corneal thinning disorders like keratoglobus, pellucid marginal degeneration (PMD) or cases with a combination of keratoconus and PMD[26,27].

Surgical Technique

The surgery involves the creation of partial thickness groove of 180–240 µm on the host cornea using a Hessburg Barron vacuum trephine and excision of a central anterior stromal disc. Subsequently, a peripheral intrastromal pocket is created circumferentially in the corneal periphery up to a point 0.5 mm farther away from the limbus. The donor preparation involves fixing a corneoscleral donor button in an artificial chamber. An initial partial thickness incision is made up to a depth of 300 µm and superficial corneal tissue is excised leaving a central full-thickness graft with a peripheral partial thickness flange of about 2.5–3 mm. The tissue is punched from the endothelial side with hand-held trephines. The DM of the donor lenticule is stripped after staining with 0.1 ml of 0.06% Trypan Blue. The flange of donor lenticule is tucked into the peripheral intrastromal pocket of the host created previously, and sutured with sixteen 10-0 monofilament sutures. (Figure 3) FIn the presence of inferior thinning in cases of PMD with keratoconus only an inferior 180° peripheral intrastromal pocket is created instead of a circumferential pocket.

The central full thickness graft provides tectonic support to the central cornea while the thin peripheral flange tucked into the intrastromal pocket integrates into the host and provides tectonic support to the peripheral cornea. Moreover, there is no damage to the recipient's limbal stem cells as the dissection of limbal region is avoided that subsequently promotes healing of epithelium at the graft-host junction. (Figure 4)

Intacs

Intacs are polymethylmethacrylate segments which are designed to be surgically inserted into the deep corneal stroma to flatten the central cornea. An important advantage of using Intacs is that the prolate shape of the cornea is preserved over the central optical zone unlike laser and incisional procedures that plays a role in the maintenance of contrast sensitivity and improved visual acuity outcomes. Intacs were initially approved by the FDA in 1999 for correction of myopia from –1.00 to –3.00 D, with 1.00 D or less of astigmatism. However, with the advent of the excimer laser at about the same time, Intacs were not popular for refractive correction. Intacs have now been approved for use in patients with mild to moderate keratoconus who have clear visual axis, upper limit of keratometry readings in the range of 55 to 57 D, and minimum corneal thickness of 400 microns.

Intacs come in many different sizes and there are potentially many different combinations that can be used to achieve both flattening of the central cornea and reduce the astigmatism. In pure nipple cones it is best to use 2 symmetrical Intacs. If the patient is not severely myopic, lesser size symmetrical Intacs could be used so as not to overcorrect and induce hyperopia.

The surgical procedure involves creation of corneal tunnels at about 70% corneal depth using two Sinskey hooks and a mechanical spreader. Intacs segments are implanted in the respective corneal tunnels, maintaining a space of approximately 2.0-mm between their ends. The incision site is sutured using a single 10/0 nylon stitch[28]. Recently, femtosecond laser has been used to create channels. Besides being quick, femtosecond results in a high degree of certainty of the depth of placement of rings.

The technique may be associated with the occurrence of small epithelial defects that are evident on first postoperative day. Deposits surrounding the ring segments are occasionally seen, may increase over time, but are not associated with effects on visual acuity. Infection most commonly occurs as a result of a loose stitch or as a result of wound gape from migration of the Intacs to the site of the wound.[29-31]

Results

Colin *et al* in 2001 published the first series of 10 patients with 1-year follow-up. Intacs insert of 0.45-mm thickness was placed in the inferior cornea and 0.25-mm thickness was inserted superiorly. Postoperative month 12 UCVA was significantly better than preoperative UCVA[32].

In 2003, Boxer Wachler reported the results on 74 eyes of patients with keratoconus using asymmetrical Intacs. The study concluded that asymmetric Intacs implantation can improve both uncorrected and best spectacle-corrected visual acuity and can reduce irregular astigmatism[33].

In 2005, Alio *et al* performed a prospective study to evaluate the effect of implanting 1 versus 2 intracorneal rings in patients with keratoconus. A single Intac was placed inferiorly in cases where topographic pattern did not cross the 180-degree meridian,

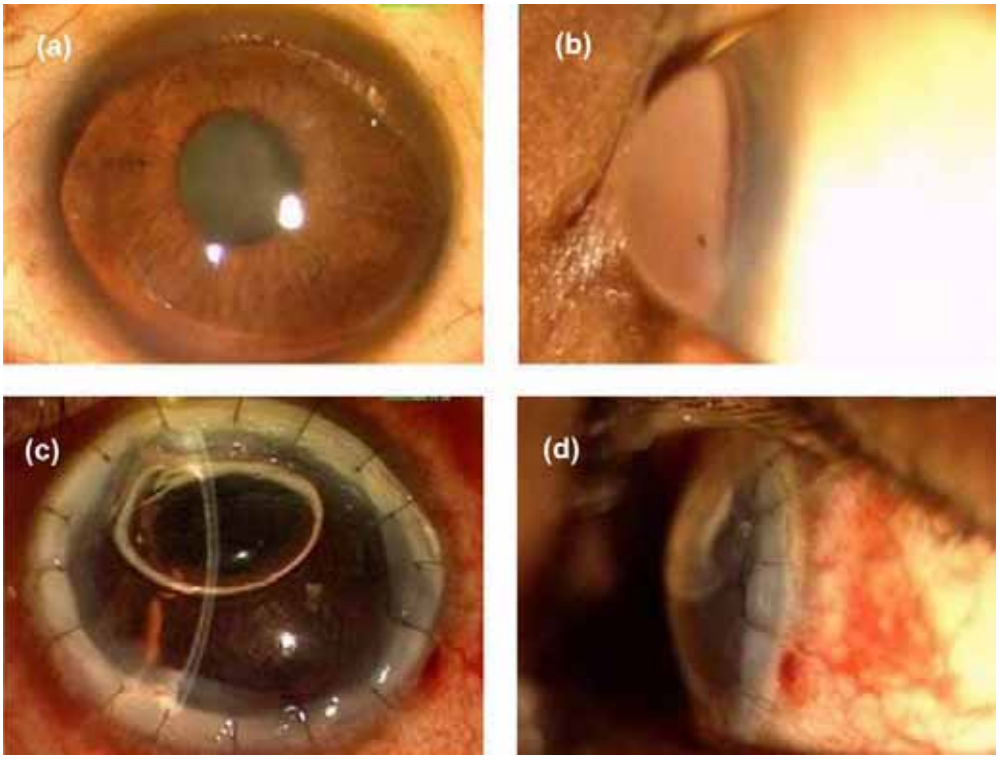

Figure 4(a): *Preoperative keratoglobus (diffuse),* ***(b):*** *Preoperative keratoglobus (diffuse)*
(c): *Post TILK - day 1 (diffuse),* ***(d):*** *Post TILK - day 1 (diffuse)*

whereas 2 Intacs were placed in cases where topographic pattern crossed the 180-degree meridian. At 1-year, the mean UCVA improved from 20/100 to 20/32 in first group and from 20/400 to 20/63 in the second group[34].

In 2005, Rabinowitz *et al* presented compared the results of femtosecond laser with that of the mechanical spreader for inserting Intacs is patients with keratoconus. Both groups showed significant reduction in average keratometry, spherical equivalent, BCVA, UCVA, surface regularity index (SRI), and surface asymmetry index (SAI). The laser group performed better in all parameters except change in SRI[35].

References

1. Tan DT, Por YM. Current treatment options for corneal ectasia. Curr Opin Ophthalmol. 2007 Jul; 18(4):284-9.
2. Kirkness CM, Ficker LA, Steele ADMcG, Rice NSC. The success of penetrating keratoplasty for keratoconus. *Eye* 1990;4:673-688.
3. Vajpayee RB, Tyagi J, Sharma N, Kumar N, Jhanji V, Titiyal JS. Deep anterior lamellar keratoplasty by big-bubble technique for treatment corneal stromal opacities. Am J Ophthalmol. 2007 Jun;143(6):954-957. Epub 2007 Apr 16.
4. Noble BA, Agrawal A, Collins C, Saldana M, Brogden PR, Zuberbuhler B. Deep Anterior Lamellar Keratoplasty (DALK): visual outcome and complications for a heterogeneous group of corneal pathologies. Cornea. 2007 Jan;26(1):59-64.
5. Sugita J, Kondo J. Deep lamellar keratoplasty with complete removal of pathological stroma for vision improvement. *Br J Ophthalmol* 1997; 81:184-88.
6. Melles GRJ, Lander F, Rietveld FJR, et al. A new surgical technique for deep stromal, anterior lamellar keratoplasty. *Br J Ophthalmol* 1999; 83: 327-33.
7. Funnell CL, Ball J, Noble BA. Comparative cohort study of the outcomes of deep lamellar keratoplasty and penetrating keratoplasty for keratoconus. Eye. 2006 May; 20(5):527-32
8. Watson SL, Ramsay A, Dart JK, Bunce C, Craig E. Comparison of deep lamellar keratoplasty and penetrating keratoplasty in patients with keratoconus. Ophthalmology. 2004 Sep;111(9):1676-82.
9. Shimmura S, Shimazaki J, Omoto M, Teruya A, Ishioka M, Tsubota K. Deep lamellar keratoplasty (DLKP) in keratoconus patients using viscoadaptive viscoelastics. Cornea. 2005 Mar;24(2):178-81.
10. Senoo T, Chiba K, Terada O, Mori J, Kusama M, Hasegawa K, Obara Y. Deep lamellar keratoplasty by deep parenchyma detachment from the corneal limbs. Br J Ophthalmol. 2005 Dec;89(12):1597-600.
11. Parmar P, Salman A, Kalavathy CM, Thomas PA, Jesudasan NC. Simplified technique for deep anterior lamellar keratoplasty. Cornea. 2007 Jul;26(6):707-8.
12. Archila EA. Deep lamellar keratoplasty dissection of host tissue with intrastromal air injection. Cornea. 1984-1985;3(3):217-8.
13. Anwar M, Teichmann KD. Big-bubble technique to bare Descemet's membrane in anterior lamellar keratoplasty. J Cataract Refract Surg. 2002 Mar;28(3):398-403.
14. Fournie P, Malecaze F, Coullet J, Arne JL. Variant of the big bubble technique in deep anterior lamellar keratoplasty. *J Cataract Refract Surg* 2007; 33:371-5.

15. Parthasarathy A, Por YM, Tan DT. Use of a "small-bubble technique" to increase the success of Anwar's "big-bubble technique" for deep lamellar keratoplasty with complete baring of Descemet's membrane. Br J Ophthalmol. 2007 Oct;91(10):1369-73.

16. Al-Torbak AA, Al-Motowa S, Al-Assiri A, Al-Kharashi S, Al-Shahwan S, Al-Mezaine H, Teichmann K. Deep anterior lamellar keratoplasty for keratoconus. Cornea. 2006 May;25(4):408-12.

17. Fontana L, Parente G, Tassinari G. Clinical outcomes after deep anterior lamellar keratoplasty using the big-bubble technique in patients with keratoconus. Am J Ophthalmol. 2007 Jan;143(1):117-124. Epub 2006 Oct 20.

18. Michieletto P, Balestrazzi A, Balestrazzi A, Mazzotta C, Occhipinti I, Rossi T. Factors predicting unsuccessful big bubble deep lamellar anterior keratoplasty. Ophthalmologica. 2006;220(6):379-82.

19. Leccisotti A. Descemet's membrane perforation during deep anterior lamellar keratoplasty: prognosis. J Cataract Refract Surg. 2007 May;33(5):825-9.

20. Jhanji V, Sharma N, Vajpayee RB. Intraoperative perforation of Descemet's membrane during "big bubble" deep anterior lamellar keratoplasty. Int Ophthalmol 2009 Dec. [Epub ahead of print].

21. Den S, Shimmura S, Tsubota K, Shimazaki J. Impact of the descemet membrane perforation on surgical outcomes after deep lamellar keratoplasty. Am J Ophthalmol 2007 May; 143(5):750-4. Epub 2007 Mar 23.

22. Sharma N, Jhanji V, Titiyal JS, Amiel H, Vajpayee RB. Use of Trypan Blue Dye for identification of remnants of host corneal tissue during conversion of Deep Anterior Lamellar Keratoplasty to Penetrating Keratoplasty. J Cataract Refract Surg (in press).

23. Watson SL, Tuft SJ, Dart JK. Patterns of rejection after deep lamellar keratoplasty. Ophthalmology 2006 Apr;113(4):556-60.

24. Vajpayee RB, Vasudendra N, Titiyal JS, Tandon R, Sharma N, Sinha R. Automated lamellar therapeutic keratoplasty (ALTK) in the treatment of anterior to mid-stromal corneal pathologies. Acta Ophthalmol Scand. 2006 Dec; 84(6):771-3.

25. Busin M, Zambianchi L, Arffa RC. Microkeratome-Assisted Lamellar Keratoplasty for the Surgical Treatment of Keratoconus Ophthalmology 2005;112:987–997.

26. Vajpayee RB, Bhartiya P, Sharma N. Central lamellar keratoplasty with peripheral intralamellar tuck: a new surgical technique for keratoglobus. Cornea. 2002 Oct;21(7):657-60.

27. Tuck In" Lamellar Keratoplasty (TILK) for corneal ectasias involving corneal periphery. Br J Ophthalmol. 2008 Feb; 92(2):286-90.

28. Rabinowitz YS. INTACS for keratoconus. Int Ophthalmol Clin. 2006 Summer; 46(3):91-103.

29. Chalasani R, Beltz J, Jhanji V, Vajpayee RB. Microbial keratitis following Intracorneal Ring Segment implantation. Br J Ophthalmol 2009 Jun 9. [Epub ahead of print].

30. McAlister JC, Ardjomand N, Ilari L, Mengher LS, Gartry DS. Keratitis after intracorneal ring segment insertion for keratoconus. J Cataract Refract Surg 2006 Apr; 32(4):676-8.

31. Bourcier T, Borderie V, Laroche L. Late bacterial keratitis after implantation of intrastromal corneal ring segments. J Cataract Refract Surg 2003 Feb; 29(2):407-9.

32. Colin J, Cochener B, Savary G, et al. INTACS inserts for treating keratoconus: one-year results. Ophthalmology 2001; 108:1409-1414.

33. Boxer Wachler BS, Christie JP, Chandra NS, et al. Nepomuceno Intacs for keratoconus. Ophthalmology 2003; 110:1031-1040.

34. Alio JL, Artola A, Hassanein A, et al. One or 2 Intacs segments for the correction of keratoconus. J Cataract Refract Surg 2005; 31:943-953.

35. Rabinowitz YS, Li X, Ignacio TS, Maguen E. INTACS inserts using the femtosecond laser compared to the mechanical spreader in the treatment of keratoconus. J Refract Surg 2006 Oct;22(8):764-71.

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Keratoprosthesis

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Keratoprosthesis is a surgical procedure where a severely damaged or diseased cornea is replaced with an artificial cornea. It is usually the last option for the surgeon and the patient who has visual potential in an eye with severely compromised cornea.

1789: Pellier de Quengsy - glass lens in silver ring for leukomatous cornea[1]

1853: Nussbaum - published the first human trial using quartz crystal implant[2]

1860: Heusser – inserted a glass plate in the cornea of a 19 year old patient[3]

1898: Dimmer, Salzer – Celluloid plates and egg membranes were used to stabilize glass lenses in the cornea[4,5]

1981: All these all Keratoprosthesis failed within weeks or months.

1930: Verhoeff – use of Quartz button[6]

1935: Filatov – implanted penetrating glass device into leucomatous cornea and covered it with a double conjunctival flap[7]

1950: Stone & Herbert[8], Cardona[9] - PMMA prosthesis designs in rabbits

1963: Strampelli B- Osteo - Odonto – Keratoprosthesis[10]

1979: Pintucci Keratoprosthesis

1998: AlphaCor Prosthesis

History

Indications for Keratoprosthesis

Disease or trauma can cause loss of corneal transparency and loss of vision. When this process becomes irreversible, it is treated surgically by Penetrating Keratoplasty (PK).

Major Limitations of PK

- Depends on availability of healthy human corneas
- Requires a technically sophisticated Eye Banking System
- Grafted corneal button may itself not remain transparent
- It is subject to immunologic rejection
- Rarely but donor corneal tissue can transmit devastating disease (Hepatitis, Jacob- Creutzfeldt disease)
- In certain disorders (mentioned below), technically successful grafts fail repeatedly.

Recent advances in our understanding of the ocular surface physiology and surgical technique have increased our ability to deal with complex problems of these structures. These include stem cells, amniotic membrane, lid reconstruction techniques, and improved pharmacological interventions.

When all of these fail, as they often do (as mentioned below) then one has to take recourse to a keratoprosthesis. A clinical pointer to the need for these devices is the presence of keratinization on the ocular surface.

Indications[11]

Bilateral blindness in severe cases of

- Stevens – Johnson syndrome
- Ocular cicatricial pemphigoid (stage 3 & 4)
- Chemical injury
- Trachoma (stage C0 according to WHO)
- Vascularized corneas with complete stem cell loss and dryness
- Multiple failed penetrating Keratoplasty/ Amniotic membrane or stem cell grafting

For these patients with complicated corneal blindness, a keratoprosthesis seems to be a logical and obvious alternative.

Prognostic Categories in Keratoprosthesis

Prognosis is best in the top group, then in falling order, with the last category the worst

- Noninflammatory conditions- Graft failure in corneal edema, dystrophies
- Graft failure after Herpes simplex Keratitis, Zoster, Infectious ulcer.
- Chemical Burns
- Pemphigoid
- Stevens Johnson Syndrome

Designs and Materials

Consists of Optical cylinder and Supporting flange

Non Biointegrated(Optical cylinder materials)

- Polymethyl methacrylate (PMMA) – most commonly used material
- Glass, Ceramic, Quartz , Silicon

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Non Biointegrated(Supporting flange)

- Methacrylate
- Teflon
- Dacron mesh
- Polycarbon

Biointegrated Supporting Flange (Autologous tissue)

- Tooth and bone (osteo-odonto-keratoprosthesis)
- Cartilage (chondro-keratoprosthesis)
- Nail (onycho-keratoprosthesis)

Keratoprosthesis designs have primarily been variations of 3 main types.

First Type PMMA stem with skirt embedded within the cornea

Second Type Transparent membrane with porous edges inserted into the cornea

Third Type PMMA 'collar button' with cornea between the plates

First type: It is most commonly used. The optical core is stabilized by a permanently attached supporting plate or skirt implanted in a pocket created in the collagen lamellae of the corneal stroma.

Consists of

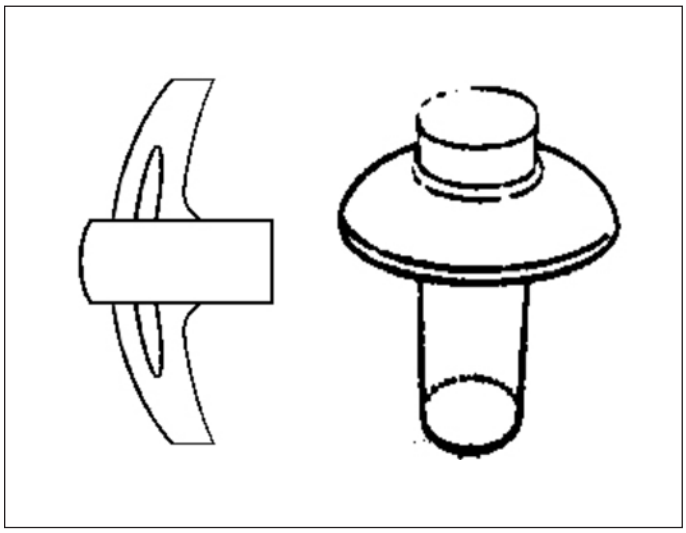
- *Central Optical Cylinder*
- *Supporting Flange*
  - PMMA stem
  - Skirt placed intralamellarly in the stroma(made of perforated grids of PMMA, nylon, Dacron, proplast/ covered by transplanted autologous tissue/ lid skin

PMMA- Advantages

- Excellent transmission of light
- Completely Transparent
- Biologically Inert
- Can be shaped to produce High Quality images
- Retains its shape and clarity with age

PMMA Disadvantages

- Rigid and Hydrophobic
- At points of attachment to the stroma, lack of elasticity creates stress that contributes to stromal necrosis and melting
- The elastic mismatch between the rigid plastic and the adjacent flexible tissue prevents formation of a tight interface , creating gaps that allow epithelial downgrowth and leakage of aqueous humour.
- Nutrients are unable to diffuse through PMMA, so an optical center of this plastic may not be able to support epithelium on its anterior surface.
- Cells and fibrous material do attach to the posterior surface, requiring surgical removal to restore vision.



Second Type: Consists of transparent plate with a porous periphery, allowing tissue ingrowth into the pores. Such designs have been made of polytetraflouroethylene, polyurethane, or modified gels. By using suitable pore size, these devices have been well colonized with tissue elements which might help to anchor the device and prevent future extrusion.

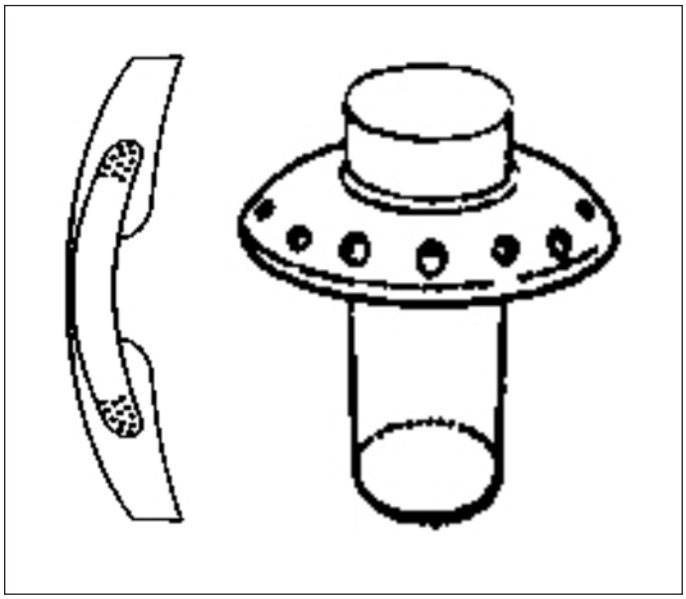
Third Type: Less common type has a collar button shaped device consisting of two plates joined by a stem, which constitutes the optical portion. This is inserted into the patient's cornea or a donor cornea so that the plates sandwich the corneal tissue between them. The optical stem is short, allowing a generous field and wide lates stabilize the stem so that it cannot easily deviate from the axis to the macula.

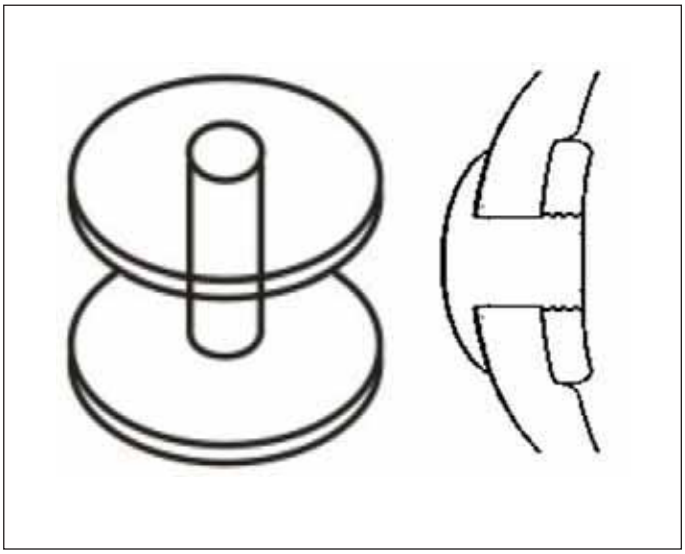
Special Types

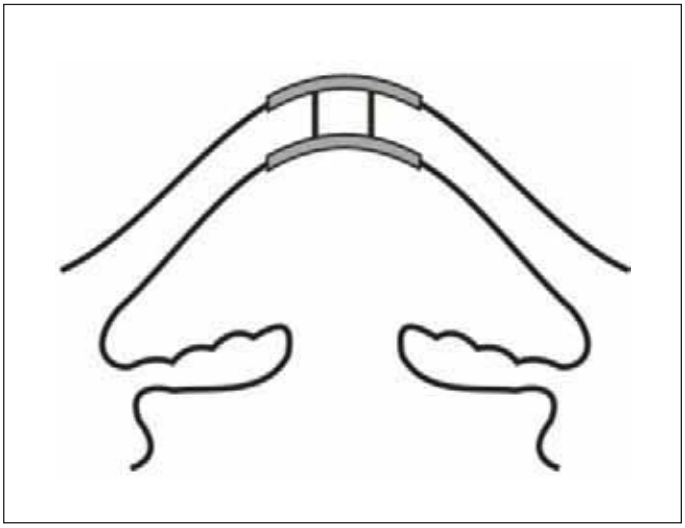
Boston Keratoprosthesis/Dohlman – Doane

Keratoprosthesis

The Boston Keratoprosthesis was first described in 1974 by Dohlman et al., however, it failed to gain popularity at that time







due to complications and imperfections in design12. The device was approved by the FDA for use in the United States in 1992. Reports from the literature support high success rates based on visual acuity outcomes and retention rates of the device.

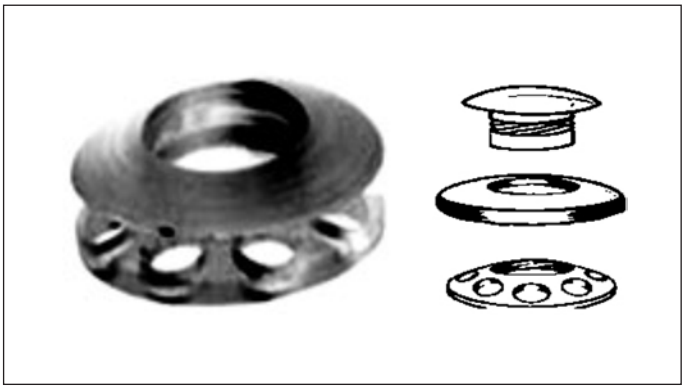
Indication

- Two failed grafts, with poor prognosis for further grafting
- Vision less than 20/400 in the affected eye
- Minimum vision of Light Perception
- Lower than optimal vision in the opposite eye

Advantages

- Long-term (many years) stability and safety.
- It is also known for having excellent optics.
- Its optical system can provide excellent vision if the rest of the eye is undamaged

Design: The Boston Keratoprosthesis consists of two plastic parts that clamp a corneal graft. The graft is then sutured into the patient's cornea like a standard graft.



There are 2 types of Boston KPros

The Boston Type I KPro – most commonly used

- Indicated in eyes with reasonable blink and tear production mechanism
- Optic/front plate – Diameter 6.0 to 7.0 millimeter. Its refractive power can be selected based on axial eye length and phakic vs aphakic status of the patient.
- Stem - This front plate also has a 3.35 mm stem that connects to a back plate.
- Back plate -7.0-8.5 mm in diameter made of PMMA. The back plate has 16 holes to facilitate direct communication with the aqueous for nutrition and hydration of the cornea.
- The donor corneal tissue is placed between the front plate and the back plate, with the plate being snapped or screwed onto the stem (newer designs are threadless and can be snapped together).
- The whole assembly is locked together with a titanium locking ring.
- The anterior-posterior length of the whole assembly is 3.7 mm, allowing or a visual field of 60 degrees13.
- The whole assembly can then be sutured to the recipient eye like a typical corneal transplant.

The Multicenter Boston Type 1 Keratoprosthesis Study (MBTKS), a prospective case series of 141 cases from 17 centers with an average follow up of 8.5 months, report retention rates of 95% with visual acuity outcomes of greater than 20/40 in 23% of patients and greater than 20/200 in 57% of patients. Failure for visual acuity to improve from the Boston Keratoprosthesis was attributed to underlying ocular disease such as advanced glaucoma, macular degeneration, or retinal detachment[14].

The Boston Type II KPro is similar in design to the Type I. Reserved for patients with extreme dry eyes, Symblepharon, Sequelae of some autoimmune and inflammatory disorders, Stevens-Johnson Syndrome, OCP15.

- However, this design is used only for patients with severe ocular surface disease where there are no fornices to support the device.
- This design has a 2 mm long anterior nub off the front plate for through the rub penetration.

- Front plate diameter of 6mm
- Posterior plate diameter of 8.5mm with 8 holes
- 4.7mm in length
- 40 degree field of vision

Contraindications

- Unilateral vision loss
- End-stage glaucoma or uncontrolled glaucoma
- Posterior segment pathology
- Presence of a functioning KPro in the fellow eye

Complications

Necrosis of tissue around the Keratoprosthesis (which if unchecked can lead to leak, infection, extrusion)

- Melt in the wet eye (Type I device)-now a minor problem
- Skin retraction in the dry eye (Type II device)- still a problem

Postoperative Uveitis –can lead to the following:

- Retroprosthetic Membrane
- Vitreous Opacities
- Retinal Detachment
- Macular Oedema,Epiretinal Membrane,etc

Glaucoma – especially in stevens Johnson Syndrome, pemphigoid,chemical burns.

Infection – Endophthalmitis –now rare

Osteo-odonto Keratoprosthesis

The use of osteo-dental tissue for Keratoprosthesis was first described by Strampelli in the early 196010 . There is long term retention of the implant. The surgery is multi-staged and requires cross speciality experience.

Design

Surgical Technique[11]

Stage I

- Preparation of globe with buccal mucous membrane graft.
- Preparation of Osteo odonto acrylic lamina (OOAL)

Stage 1A

- Preparation of Globe With Buccal Mucous Membrane Graft.
  - including Bowman membrane removed
  - 360° limbal peritomy
  - Oral mucosa anchored

Stage 1B

- Implant is fashioned from Osteo-dental tissue
- A monocuspidate tooth is removed along with the adjacent maxillary bone and a thin section is cut from the tooth.
- An optical cylinder made up of PMMA is inserted through a hole made in the section.
- A pocket is created in the lower eyelid into which the entire prosthesis is inserted and left for 3 months
- During this time soft tissue grafts to the bone to which the tooth is attached

Stage II

- Implantation of OOAL
  - Part of the oral mucosa is stripped off the cornea and sclera to make space for the final implantation of prosthesis
  - The prosthesis is detached from the eyelid pocket and implanted, with the optical cylinder protruding through a hole in the mucosa.

Glaucoma is the major complication.

Chirila (AlphaCor) Prosthesis

It is the newest keratoprosthetic device

It was FDA-approved in August 2002 for patients at high risk for donor penetrating keratoplasty (PKP).

Design

The implant is a 7-mm diameter, one-piece, non-rigid synthetic cornea.

It is composed of an outer skirt, that is an opaque, porous, high-water PHEMA (poly[2-hydroxyethyl methacrylate]), with a transparent central optic core of gel PHEMA .

Surgical Procedure

In Stage I

- An intrastromal trephine is used to remove the central posterior corneal lamellae for insertion of the device
- A corneal incision is made and dissection instruments are used to continue the corneal dissection throughout the circumference of the corneal graft, thereby creating an intralamellar pocket
- An AlphaCor sizer, used to test the size and centration, is inserted into the intralamellar pocket followed by removal of the posterior disc via a 3.5 mm intrastromal trephine.
- After insertion of the device and closure of the limbal incision, the surface is often covered with a Gundersen conjunctival flap
- If the Gundersen flap is inadequate to cover the cornea an amniotic membrane graft may be required
- In Stage II performed approximately 2 months after Stage I
- The overlying conjunctiva created by the Gundersen flap is removed.

- Trephination of the central 4mm of the conjunctival flap and anterior corneal lamellae.

Complications

- Corneal Melt
- Retroprosthetic Membranes

Pintucci Keratoprosthesis

The Pintucci KP can be implanted in thinned or perforated corneas, in corneas with stromal melting, and in eyes that have undergone several procedures including penetrating keratoplasty, other KP implantations, and glaucoma, cataract and vitreoretinal surgery

Design

- The supporting element of the Pintucci KP is made of a biointegrated Dacron fabric skirt that allows three-dimensional colonization by newly formed vascularized connective tissue.
- This fabric is soft and pliable, can be easily cut into the desired shape and sutured, and is chemically inert and not subject to resorption.
- The Dacron fabric support is fixed to the PMMA optical cylinder with a specific reliable method (international patent pending).

Keratoprosthesis still carries a somewhat greater burden post-operatively than standard keratoplasty. Successful outcome requires patient compliance, more frequent follow-up and more demands on physician time. However, in cases where further Keratoplasty appears futile, keratoprosthesis can be most rewarding.

References

1. Pellier de Quengsy G: Precis ou cours d' operations sur la chirurgie des yeux, Paris, 1789, Didot.
2. Nussbaum N : Cornea Artificialis, ein Substitut fur die Transplantatio Cornea, Deutsche Klinik.1853;34: 367.
3. Heusser J: Die Einheilung einer Cornea artificialis, Oesterr Ztschr Pract Med.1860;26:424.
4. Dimmer F: Zwei Falle von Celluloidpattern der Hornhaut, Klin Monatsbl Augenh.1891;29:104.
5. Salzer F: Uber den kunstlichen Hornhautersatz, Behrmann, Wiesbaden, 1898.
6. Verhoeff FH : Cited in Cardona H: Keratoprosthesis,Am J Ophthalmol.1962;54:284.
7. Filatov VP: Alloplastik bei vollstndig ,'hoffnungslosen' Leukomen, Sov Viest Opht.1936;9:400.
8. Stone W Jr, Herbert E: Experimental study of plastic material and replacement for the cornea: preliminary report, Am J Ophthalmol. 1953;36:168.
9. Cardona H: Keratoprosthesis, Am J Ophthalmol. 1962;54:284.
10. Strampelli B: Osteo Odonto Keratoprosthesis, Ann Ottalmol Clin Ocul.1963;89:1039.
11. Hille et al Cornea 2005;24:895-908.
12. Dohlman CH, Schneider HA, Doane MG. Prosthokeratoplasty. Am J Ophthalmol. 1974;77:694-700.
13. Ilhan-Sarac O, Akpek EK. Current concepts and techniques in keratoprosthesis. Curr Opin Ophthalmol. 2005 Aug;16(4):246-50. Review
14. Zerbe BL, Belin MW, Ciolino JB; Boston Type 1 Keratoprosthesis Study Group. Results from the Multicenter Boston Type 1 Keratoprosthesis Study. Ophthalmology. 2006 Oct;113(10):1779.e1-7. Epub 2006 Jul 26.
15. Doane MG, Dohlman CH, Bearse MG Coenea 2002;21:400-404

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Forthcoming Events: National

July

10th-11st CHENNAI

Annual Conference of
Intraocular Implant & Refractive Society
Venue: Hotel Taj Coromendel,
Chennai Tamil Nadu
Chairman Scientific Committee
Dr. Agarwal's Eye Hospital
19, Cathedral Road, Chennai – 600 086, India
Tel : +91-44-28112811 / 2811 6233; ***Fax :*** +91-44-28115871
Email : dragarwal@vsnl.com
Website: http://www.iirsi.com

July & August 2010

30th-1st TAMIL NADU

ICON 2010 : 58th Annual Meeting of
Tamilnadu Ophthalmic Association
Venue: Hotel Cennys Gateway and IMA Hall
Saradha College Road, Near Five Roads, Salem 636016
Conference Secretariat:
JB Eye Hospital, 76/19-D, Saradha College Road,
Salem - 636 007, ***Phone:*** 0427 - 2316124, 6505656

October 2010

22-24 UTTARAKHAND

NZOS & Uttara Eyecon 2010
(Combined Annual conference of North Zone
Ophthalmological Society and Uttarakhand State
Ophthalmological Society)
Venue: Hotel Park Plaza, Mall Road, Mussorie,
Uttarakhand
Organizing Secretary,
Dr. B.K.Oli
57, Haridwar Road, Dehradun-248001
Phone: 0-99971-22222, 0-94123-19035
Email: dr.bkoli@gmail.com

28-31 NEW DELHI

10th Annual Meeting of Uveitis Society of India
Venue: Advanced Eye Centre, PGI Chandigarh
Contact Person & Address
Dr. Vishali Gupta
email : vishalisara@yahoo.co.in
www : http://www.usi2010.in

December 2010

2-4 MYSORE

19th Annual Conference of Vitreo Retina Society - India 2010
Organizing Secretary,
Retina Institute of Karnataka
#122, 5th Main Road
(Next to Venlakh Hospital)
Chamarajpet, Bangalore - 18
Ph: +91-80-22410106 / 536 (Hospital),
Fax: +91-80-26607811
e-mail: retinainstitute@sify.com

Forthcoming Events: International

September, 2010

16-20 BEIJING, CHINA

APAO-AAO Joint Congress
China National Convention Centre, Beijing
APAO Central Secretariat
Secretariat, Asia Pacific Academy of Ophthalmology
C/o. The Chinese University of Hong Kong,
Dept. of Ophthalmology & Visual Sciences, Hong Kong
Eye Hospital, 3/F, 147K Argyle Street, Kowloon,
Hong Kong
Email: secretariat@apaophth.org
Tel: (852) 2762-3042, ***Fax:*** (852) 2715-9490,
Website: www.apao2010beijing.org

December, 2010

9-11 DUBAI, SAUDI ARABIA

14th Emirates Ophthalmic Conference 2010
Venue: Dubai International Convention and Exhibition
Centre (DICEC)
Conference Chairperson
Dr. Manal Taryam, MD
EMAOS President
C/o Meeting Minds - Experts
Dubai Media City, Shatha Tower - Office Suite 3113
P. O. Box 502464, ***Tel :*** +9714 4270492, ***Fax:*** +9714 3270493
Email: pco@eoc2010.com ◆ Web: www.eoc2010.com

9-12 MACAU, CHINA

International Symposium on Ocular Pharmacology and Therapeutics – ASIA
Venue: Venetian Macao-Resort-Hotel, Macau, China
Conference Coordinator
Dr. Lyat Shahal
Paragon Conventions
18 Avenue Louis-Casai, 1209 Geneva , Switzerland
Emal: isopt@isopt.net ◆ Web : http://www.isopt.net

Classification of Anti Fungal Drugs

DRUG	MODE OF ACTION	MODE OF ADMINISTRATION & DOSE	EFFECTIVE AGAINST	COMMENTS
POLYENE ANTIFUNGALS				
1. NYSTATIN	BINDS TO ERGOSTEROL, A MAJOR COMPONENT OF FUNGAL CELL MEMBRANE	TOPICAL 3.5 % EYE OINTMENT, POOR INTRAOCULAR PENETRATION	FUNGISTATIC CANDIDIASIS ASPERGILLUS	FUNFISTATIC, LOW EFFICACY, NOT USED COMMONLY
2. AMPHOTERICIN B	BINDS TO ERGOSTEROL, A MAJOR COMPONENT OF FUNGAL CELL MEMBRANE	TOPICAL 0.15% EYE DROP, INTRAVITREAL & INTRACAMERAL DOSE 5 μ gm IN 0.1 ml. LIMITED PENETRATION IN VITREOUS CAVITY,	BOTH FUNGISTATIC & FUNGICIDAL EFFECTIVE AGAINST ASPERGILLOSIS, CANDIDIASIS, MUCORMYCOSIS, CRYPTOCOCCUS, ENDOGENOUS FUNGAL INFECTION. FOR CANDIDA DRUG OF FIRST CHOICE.	EFFECTIVE AGAINST CANDIDA, RETINAL TOXICITY NEPHROTOXICITY NON TOXIC TO ENDOTHELIUM OR LENS.
3. NATAMYCIN	BINDS TO ERGOSTEROL, A MAJOR COMPONENT OF FUNGAL CELL MEMBRANE	TOPICAL 5% SUSPENSION, NOT RECOMMENDED FOR INJECTION. PENETRATION IS GOOD	BROAD SPECTRUM ANTIFUNGAL EFFECTIVE AGAINST CANDIDA, ASPERGILLUS, FUSARIUM AND CEPHALOSPORIUM. DRUG OF CHOICE FOR FUSARIUM SOLANI KERTITIS. AND FUNGAL KERATITIS.	FIRST LINE THERAPY IN FUNGAL KERATITIS, POOR CORNEAL PENETRATION
IMIDAZOLE ANTIFUNGALS				
4. MICONAZOLE	BINDS TO ERGOSTEROL, A MAJOR COMPONENT OF FUNGAL CELL MEMBRANE	TOPICAL 1% EYE DROP SUBCONJUNCTIVAL (5 TO 10 mg OF 10 mg / ml SUSPENSION) CAN BE GIVEN INTRAVENOUS (600–1200 mg/day)	BROAD SPECTRUM ANTIFUNGAL AND FUNGICIDAL EFFECT. EFFECTIVE AGAINST CANDIDA, ASPERGILLUS, FUSARIUM, CRYPTOCOCCUS, CLADOSPORIUM, TRICHOPHYTON.	LESS SYSTEMIC TOXICITY FEVER, CHILLS, NAUSEA. SOMETIMES ANAPHYLAXIS, COMBINED ORAL KETOCONAZOLE AND TOPICAL MICONAZOLE ARE USEFUL
5. CLOTRIMAZOLE	EXACT MECHANISM OF ACTION OF THIS ANTIFUNGAL IS NOT UNDERSTOOD	TOPICALLY 1 TO 2 % SUSPENSION AND OINTMENT.	FUNGISTATIC, EFFECTIVE AGAINST CANDIDA AND ASPERGILLUS	CORNEAL TOXICITY WITH LONG-TERM USE OF TOPICAL PREPARATION
6. ECONAZOLE	INHIBIT ENZYME CYTOCHROME P450 14 α - DEMETHYLASE REQUIRED IN FUNGAL CELL MEMBRANE SYNTHESIS	TOPICAL 1 TO 2% ECONAZOLE NITRATE OINTMENT. ORALLY 200 - 400 mg / DAY FOR SEVERAL WEEKS.	BROAD SPECTRUM ANTIFUNGAL FOR FILAMENTOUS FUNGI, POOR INTRAOCULAR PENETRATION SO EFFECTIVE ONLY IN SUPERFICIAL INFECTIONS OF EYE .	HEPATOTOXIC, TOPICAL ADMINISTRATION CAUSES OCULAR IRRITATION, LESS EFFECTIVE AGAINST CANDIDA SPP
7. KETOCONAZOLE	INTERFERES WITH THE FUNGAL SYNTHESIS OF ERGOSTEROL, A CONSTITUENT OF CELL MEMBRANE.	TOPICAL 1-2%, ORAL ADMINISTRATION 200 - 400 mg/d	ADJUNCTIVE SYSTEMIC ANTIFUNGAL AGENT IN FUNGAL KERATITIS, COMPLICATED BY ENDOPHTHALMITIS, DEEP KERATOMYCOSIS.	ALLERGIC REACTION, GYNECOMASTIA MILD LIVER DYSFUNCTION, VARIABLE EFFECT AGAINST ASPERGILLUS OR FUSARIUM. REQUIRES ACID PH FOR ABSORPTION

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TRIAZOLE ANTIFUNGALS				
DRUG	**MODE OF ACTION**	**MODE OF ADMINISTRATION & DOSE**	**EFFECTIVE AGAINST**	**COMMENTS**
8. FLUCONAZOLE	INHIBITS FUNGAL CYTOCHROME P 450 ENZYME 14 α DEMETHYLASE.	TOPICALLY 2% EYE DROP. ORALLY 200 MG/DAY, INTRAVENOUS PREPARATION AT 2 MG/ML (DOSE IS 100 MG/DAY)	GOOD EFFICACY AGAINST CANDIDA, POOR AGAINST FILAMENTOUS FUNGI	CARDIAC ARRHYTHMIAS, HEPATOTOXICITY , GOOD INTRAOCULAR PENETRATION
9. ITRACONAZOLE	INHIBITS FUNGAL CYTOCHROME P 450 OXIDASE MEDIATED SYNTHESIS OF ERGOSTEROL	TOPICAL 1-2 %, ORALLY 200-400/ DAY, GOOD ORAL BIOAVAILIBILITY	EFFECTIVE AGAINST ASPERGILLUS, CULVULARIA, CANDIDA, LESS AGAINST FUSARIUM	CARDIAC AND HEPATOTOXICITY
10. VORICONAZOLE (SECOND GENERATION TRIAZOLE)	LIPOSOMAL	TOPICAL 1%, ORALY 200 mg B.D. DAILY. INTRAVENOUS 200 mg B.D. DAILY INTRAVITREAL & INTRACAMERAL & INTRACORNEAL 50 µg IN 0.1 ml	SUPERIOR TO MOST OF THE OTHER ANTIFUNGALS AVAILABLE, GOOD CORNEAL PENETRATION, BROAD SPECTRUM AGAINST FILAMENTOUS FUNGI AND CANDIDA	TOPICAL 1% VERY EFFECTIVE AGAINST RESISTANT FUNGAL KERATITIS, FEW S/E WITH SYSTEMIC USE
PYRIMIDINE ANTIFUNGAL				
11. FLUCYTOSINE (5-FC)	INHIBIT FUNGAL DNA SYNTHESIS	TOPICAL 1% OPHTHALMIC SOLUTION. ORALLY 150 mg/kg/d (DIVIDED EVERY 6 HOURS).	ACTIVE AGAINST CANDIDA & CRYPTOCOCCAL INFECTIONS, NOT USED AS A SINGLE AGENT DUE TO EARLY RESISTANCE	APLASTIC ANEMIA, HEPATOTOXICITY, NEPHROTOXICITY
ECHINOCANDINS ANTIFUNGALS				
12. CASPOFUNGIN	INHIBITING THE ENZYME β (1, 3) - D- GLUCAN SYNTHASE AND THEREBY DISTRUBING INTIGRITY OF FUNGAL CELL WALL	TOPICAL 0.5%, INTRAVENOUS 70mg INITIALLY FOLLOWED BY 50mg IV DAILY. SLOW INFUSION OVER 1 HOUR.	REFRACTORY ASPERGILLOSIS, CANDIDEMIA, SYSTEMIC MYCOSIS.	HEPATOTOXIC
13. MICAFUNGIN	INHIBIT SYNTHESIS OF GLUCAN IN THE CELL WALL.	INTRAVENOUS 100mg/d AND 50mg FOR PROPHYLAXIS	ASPERGILLUS, SYSTEMIC MYCOSIS, CANDIDA	HEPATOTOXIC
OTHERS				
14. MYCOSTATIN	FUNGISTATIC. NOT EXACTLY KNOWN	TOPICAL DOSE 1,00,000 UNITS / ml	CANDIDIASIS	

DOS Credit Rating System Report Card
(November, 2009 - March, 2010)

DCRS November, 2009 – Venu Eye Institute & Research Centre

Total No. of Delegates as per Attendance Register	70
Total No. of form received from Delegates	47
Delegates from Out side (N)	31
Delegates from Venu Eye Institute & Research Centre (n)	15
Overall assessment by outside delegates (M)	224.5
Assessment of case presentation-I (Dr. Pawan Gupta) by outside delegates	208.5
Assessment of case presentation-II (Dr. Archana Sood) by outside delegates	214.5
Assessment of clinical talk (Dr. Subodh Sinha) by outside delegates	206.5
Cancelled Forms	1

DCRS December, 2009 – Safdarjung Hospital

Total No. of Delegates as per Attendance Register	85
Total No. of form received from Delegates	60
Delegates from Out side (N)	45
Delegates from Safdarjung Hospital (n)	13
Overall assessment by outside delegates (M)	366
Assessment of case presentation-I (Dr. Shivani Kochhar) by outside delegates	338
Assessment of case presentation-II (Dr. Aniket N. Shastri) by outside delegates	373
Assessment of clinical talk (Dr. B.P. Guliani) by outside delegates	371
Cancelled Forms	2

DCRS January, 2010 – Bharti Eye Hospital

Total No. of Delegates as per Attendance Register	102
Total No. of form received from Delegates	91
Delegates from Out side (N)	81
Delegates from Bharti Eye Hospital (n)	6
Overall assessment by outside delegates (M)	620
Assessment of case presentation-I (Dr. Dharitri Samantaray) by outside delegates	421.5
Assessment of case presentation-II (Dr. Meetu Bansal) by outside delegates	435.5
Assessment of clinical talk (Dr. Sudhank Bharti) by outside delegates	648
Cancelled Forms	4
Guest Lecture : Mr. Shishir Jha	533.5

DCRS February, 2010 – Centre for Sight

Total No. of Delegates as per Attendance Register	81
Total No. of form received from Delegates	66
Delegates from Out side (N)	50
Delegates from Centre for Sight (n)	14
Overall assessment by outside delegates (M)	395
Assessment of case presentation-I (Dr. Kanak Tyagi) by outside delegates	309
Assessment of case presentation-II (Dr. Avnindra Gupta) by outside delegates	362.5
Assessment of clinical talk (Dr. Mahipal S. Sachdev) by outside delegates	418
Cancelled Forms	2

DCRS March, 2010 – Guru Nanak Eye Centre

Total No. of Delegates as per Attendance Register	94
Total No. of form received from Delegates	74
Delegates from Out side (N)	62
Delegates from Guru Nanak Eye Centre (n)	12
Overall assessment by outside delegates (M)	483.5
Assessment of case presentation-I (Dr. Neha Rathi) by outside delegates	459
Assessment of case presentation-II (Dr. Neha Goel) by outside delegates	509
Assessment of clinical talk (Dr. B. Ghosh) by outside delegates	526
Cancelled Forms	0