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“For me, conferences are like little mental vacations: a chance to go visit an interesting place for a couple of days, and come back rested and refreshed with new ideas and perspectives.”

- Erin McKean

Respected Seniors & Dear Friends,

It was a pleasure to have an August gathering at the DOS Annual conference 2014. Various dignitaries not only from different corners of the country but also from across the globe met to exchange ideas and take ophthalmology to the next level. With halls full of dignitaries till the end of conference and discussion at the pinnacle, it was indeed a pleasurable sight.

The idea of having a sub-specialty meet had a great response where all the specialists had the opportunity to discuss, debate and interact with their respective counterparts under a single roof and take their specialty one step ahead. First ever Young ophthalmologist (YO) session was appreciated by one and all. Young ophthalmologists took the full advantage for having sessions specially customized for them while simultaneously enjoying meeting their counterparts from across the country and making new friends.

Ophthalmology seemed to have undoubtedly evolved to be one of the most developed specialties in modern medicine. A giant leap in technology was seen in the conference and live surgeries demonstrated a perfect example of the same. It surely felt great to be a part of the organizing committee of the annual conference and meet you all. I am sure we can improvise on this to have an even larger and more interactive gathering in the coming year.

Sincerely Yours

Rajesh Sinha
Secretary,
Delhi Ophthalmological Society
Guest Editorial

Pediatric Ophthalmology

The decision to remove a cataract in a child is a much more difficult proposition than in an adult. In adults, one does not need to consider the loss of accommodation, the possible implications for amblyopia, the long-term risk of glaucoma, and the availability of appropriate powered IOLs.

For dense cataracts in children, it is obvious that the eye will not see if the cataract is left in place- but the child may not see any better even if it is removed, if it has been present since early infancy and there is profound amblyopia. In considering the question “Why remove this cataract?” - the answer “Because it is there” is not sufficient. This is a potential pot of gold for the unscrupulous surgeon. Parents, who want the very best for their child, will willingly pay whatever they can, to restore sight to the cataractous eye. The surgeon however has the benefit of knowledge of potential complications, amblyopia, etc, and should discuss these things with the parents in an honest and unpressured manner, so that the parents can make an informed decision.

I recently heard of a case in which parents were asked to pay $5000 for cataract surgery in a unilateral and densely amblyopic case, in which there is no chance of visual rehabilitation. The whole extended family contributed so that the child could have the surgery. Of course there was not useful improvement in vision. The surgeon, no doubt could justify decision to operate, but my opinion is that it should have been made absolutely clear to the parents that there was no chance that the surgery would improve the child’s life.

The actual cataract surgery is the easiest part of the management of the case. After the surgery, comes the amblyopia therapy, the many visits to the ophthalmologist, the expense of contact lenses, or glasses, and the anxiety and stress of the whole process.

My plea is for surgeons to think of the whole child, and the child’s family, rather than just the cataract!

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Dear Friends & Colleagues,

Greeting from Delhi Ophthalmological Society!

The Delhi Ophthalmological Society is organizing the “6th DOS Teaching Programme” (DOST-6) on 14th & 15th June, 2014 (Saturday & Sunday) at Jawaharlal Auditorium, AIIMS, Ansari Nagar, New Delhi. It is aimed at teaching the Postgraduate MD/MS/DNB/DO Ophthalmology Students from all of India and will be a two day exhaustive program.

Entry for First 500 Students & Members.

The Last date of Registration is : 2nd June, 2014

Download Registration Form (www.dosonline.org)

Online Registration (www.dosonline.org)

All participates of the programme will get :

- Academic credit hours from DMC.
- Past MD/MS/DNB question papers will be discussed.
- Ample time will be given to clear doubts.
- Solve your problem and clarify your doubts from the Masters.

For any further information, feel free to drop a mail at dosrecords@gmail.com.

We look forward to welcome you to the DOS Teaching Programme.

Thanking you,

Sincerely yours,

Rajesh Sinha
Secretary, DOS
Several aspects of cataract surgery in children differ from adults. Ocular anatomy, cataract morphology, exaggerated response to surgical trauma, and the need for amblyopia therapy are major concerns in pediatric cataract surgery. Moreover, intraoperative differences such as location and type of incisions, management of anterior and posterior capsules, need for anterior vitrectomy and IOL power calculations are other important issues to be considered. Here we will discuss the practical pearls of pediatric cataract surgery with various experts in this field. The questions have been prepared by Dr. Tarun Arora (TA) Senior Resident Cornea, Cataract & Refractive Surgery Services, from R.P. Centre for Ophthalmic Sciences, All India Institute for Medical Sciences, Ansari Nagar, New Delhi.

**TA: What evaluation is important for bilateral congenital cataract and is there any genetic preponderance in cases of congenital cataract?**

**SH:** If the cataracts prevent a view of the fundus, then we arrange an ultrasound to demonstrate the posterior segment i.e. exclude retinal detachment, retinoblastoma etc. We see many more unilateral congenital cataracts than bilateral. In bilateral cases, there is an identifiable genetic cause, or a positive family history in about half the cases. Fortunately we rarely see rubella cataract these days.

**ARV:** Clinical examination of the child should include a complete examination of all systems, including respiratory, nervous, and cardiovascular systems. Supportive laboratory investigations should include hemogram, blood sugar, titres for antibodies to TORCH agents, echocardiography if required. Special tests to rule out metabolic diseases should be ordered whenever necessary.

Except in cases of hereditary cataract (either / both the parents having cataract, or a family history of cataract) congenital cataracts are not often found to have genetic preponderance. However, a detailed history of any visual disability or surgeries in the family pedigree should be documented. In these cases, genetic testing may be warranted.

**JR:** As a rule, systemic workup, if required, should be done only for bilateral cataracts. A tailored approach is recommended, based on the morphological findings of cataract. Oil droplet cataract or Christmas tree cataracts provide direct hints to the laboratory work up and diagnosis. Serum calcium, TORCH or rubella titters and urine for reducing substances are the most commonly employed tests. A large number of mutations and genetic polymorphisms...
have been shown to be associated with congenital cataract and our institute has a number of on-going research projects in this regard. In the future, we may succeed in identifying a large number of candidate genes.

**SKK:** We proceed with ocular investigations only in cases of congenital cataracts unless history or examination suggests certain pertinent features suggesting syndromic/ infectious malformation. For bilateral congenital cataracts, it is important to rule out systemic conditions such as TORCH, Down’s syndrome, galactosemia. Unilateral cataracts are not usually associated with occult systemic or metabolic disease, and laboratory tests are not warranted. Laboratory examination include urine for reducing substances and amino acids, TORCH titres (toxoplasmosis, rubella, cytomegalic inclusion disease, Herpes simplex), VDRL screening (if not done already in a newborn screen), blood for calcium, phosphorous, glucose, and perhaps red cell galactokinase levels.

Congenital cataracts are known to run in families. When inherited, familial cataracts are usually autosomal dominant and always bilateral but may be asymmetric. X-linked and autosomal recessive inheritances have been reported but are rare. Not all inherited cataracts are congenital.

**KK:** Recommended lab workup includes TORCH titers, VDRL, serum calcium and phosphorus levels and urine for reducing substance. Additional systemic workup should be done in coordination with the pediatrician. Some of the bilateral cataracts can indeed be hereditary, hence a family history should be sought.

**TA:** What all should be looked in the ocular examination of pediatric cataract and how does it define our surgical management?

**SH:** I no longer insert IOLs in babies, but if I did, the size the eye influences the IOL power. We may not have high enough powers available. The presence of PHPV, including retinal dysplasia and a persistent hyaloid artery would alter the surgical approach. The ultrasound Doppler would demonstrate blood flow in the hyaloid vessels, preoperatively, allowing us to have intraocular diathermy etc available.

**ARV:** A preoperative examination with fully dilated pupils, if necessary under anesthesia, is mandatory in both the eyes. It includes examination under the operating microscope or slit lamp biomicroscope to assess the type and extent of the cataracts. Particular attention should be paid to presence of a pre-existing posterior capsule defect or a total white cataract, as it is not advisable to perform hydrodissection in these cases. Also, one should look out for presence of an anterior or posterior capsule plaque as the surgical strategy will change if either is found. Other examinations that should be performed include tonometry to rule out glaucoma, corneal diameter measurement, posterior segment evaluation, keratometry, biometry, gonioscopy and ultrasound biomicroscopy. In cases of microphthalmos and microcornea, IOL implantation maybe deferred during the primary surgery.

Coexisting morbidities such as persistent fetal vasculature, mal developed anterior chamber angles, coloboma, etc should be carefully looked for as their presence changes not only the surgical strategy but also the prognosis.

**JR:** Ocular examination of a child requires patience and perseverance. Before taking up the child for surgery, we often perform examination under anesthesia to record every minute detail in young children. We must record visual acuity/grade of fixation, IOP, corneal clarity, laterality and type of cataract, fundus examination and ultrasound B-scan whenever in doubt. A clinical examination must rule out microphthalmos, spherophakia, strabismus, nystagmus or lenticular dislocation. One must remember that administering anesthesia to children is often a risk and the surgeon must be very meticulous in every step of examination.

**SKK:** From ophthalmic point of view, prognosticating a child is important. Visual assessment is important through Teller acuity or Cardiff whatever the child permits. In younger children less than 2 months of age, a fixation reflex is also usually not developed. Hence, it may be difficult to assess acuity in these.

A slit-lamp examination should be performed to classify the morphology of the cataract and to identify any associated abnormalities of the cornea, iris, lens, and anterior chamber in cooperative children. Careful observation of the optic disc, retina, and fovea should be obtained if possible. Distant direct ophthalmoscopy helps determine whether the cataract is significant enough to occlude the visual axis. The usual rule of thumb is cataracts smaller than 2-3 mm can be observed. If no view is obtained, posterior segment evaluation by means of ultrasonography can sometimes elicit closed funnel retinal detachment reminiscent of retinopathy of prematurity. In case of anechoic posterior segment, other tests such as electrophysiological tests may be performed if the child does not even follow light or has other poor prognosticating features such as oculodigital sign etc.

**KK:** It is important to rule out associated ocular conditions such as PHPV, glaucoma, retinal pathologies as they may have an adverse prognosis. Nystagmus and squint would also indicate a guarded visual prognosis. The size and location of the lens opacity would dictate the necessity and timing of surgical intervention.
Ta: What is your preferred site and location of wound incision and do you suture these incisions in all cases? What is your protocol for suture removal?

Sh: I sit at the top of the operating table and I’m right handed, so if I’m not planning to insert an IOL, I use a 20 gauge V-lance at 11 o’clock, and another for the infusion at 4 o’clock. These stab incisions are just in from the limbus, and aimed directly towards the centre of the pupil. Midway through the procedure I will swap the vitrector and infusion over. I use 10 nylon and I always close both wounds as I cannot rely on the child not to rub his eye with the same fingers that have just been inside his mouth. I did use an absorbable suture for a while, to avoid having to remove the sutures, but I had an instance of wound dehiscence and iris plugging of the wound, so I changed back to nylon.

Arv: Pediatric cataract can be removed through a relatively small incision, as the lens has no hard nucleus. If no IOL implantation is planned, two 1mm self sealing valvular paracenteses incisions are usually made at or near the limbus. When an IOL is being implanted, following removal of the lens material, a 3 mm single plane self-sealing valvular clear corneal incision with an internal entry length of at least 1.5 to 2mm should be made. All incisions are sutured with 10-0 nylon sutures due to the low scleral rigidity and also because children have a tendency to rub their eyes. We remove the sutures after 3-4 weeks.

Jr: In children, it is often preferred to take a scleral or limbal incision. In older children, one may go for clear corneal incisions depending upon the surgeon experience. It is not scientifically justified to leave the wound and paracentesis site unsutured in children as the incision may leak. We recommend closure of wounds using 10-0 nylon suture that can be subsequently removed at 4 – 6 weeks postoperatively.

Sh: I don’t do adult cataract surgery. I use the vitrector to perform both the anterior and later, the posterior capsulectomy. The surprising thing to many surgeons, is that six months after the surgery, the hole in the anterior capsule is perfectly round and cannot be differentiated from a CCC.

Arv: The anterior capsule in children is very elastic, and therefore it may be difficult to perform a controlled manual continuous curvilinear capsulorhexis (CCC). However, a manual CCC is the gold standard in terms of maintaining the integrity of the capsular edge.

For the beginners, my suggestion is that always aim for small size. ACCC should be performed under high viscosity OVDs (cohesive) with either microincision capsulorhexis forcep or Kraff-Utrata forceps. Capsular staining with trypan blue is a useful adjunct, especially in cases where there is a poor glow. Frequent grasping and regraping of the capsular flap allows better control of the ACCC. Further, the force on the anterior capsule should be centripetal and upwards towards the corneal endothelium to avoid peripheral extension.

Jr: Capsulorhexis in pediatric cataract is not an easy task as it may appear in surgical videos. Since the capsule is very elastic compared to adults. There is a high tendency of the rhesis to run away; so it is best to consciously attempt making a small arc of a circle. My best advice to beginners would be first to carefully watch and assist pediatric cataract surgeons. Aim for perfection but do not get disheartened. If your approach is scientific and if you apply common sense of physics, things should be fine. Using capsule staining dye helps in achieving perfect continuous curvilinear capsulorrhexis particularly in white cataract.

Sh: In case a child is planned to be left aphakic, e.g., in severe microphthalmos or infant less than 3 months of age, I create only two side port incisions (~1.3mm) which may left unsutured if intra-operatively they do not leak. However, if IOL is to be put, I always create a main port superior incision of 2.8 mm length which is sutured and is covered by the lids.

Sk: In case of aphakia the anterior capsule is perfectly round and cannot be differentiated from a CCC.

Sh: The anterior capsule being elastic tends to run towards the periphery. To achieve a good capsulorhexis I would advise using a high molecular weight viscoelastic, using a forceps to perform the capsulorhexis, frequent releasing and grasping of the capsulorrhexis edge, and maintain the pull toward the centre at all stages. Beginners should always aim for a smaller capsulorhexis.

Ta: How do you find doing pediatric anterior capsulorhexis different from those in adults and what advice would you give to beginners for achieving a perfect central circular curvilinear capsulorhexis in these patients?

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Sk: A pediatric lens capsule is more elastic than an adult one. It is to be performed under a cohesive viscoelastic fill to make the anterior capsule concave and completed with the help of Utrata/ or 23/25 G ILM peeling forceps. It should always be extended by applying tangential force at the edge of the capsule rather than applying the tearing force at the centre.

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Experts’ Corner

TA: What other techniques do you employ to create anterior capsular opening in pediatric eyes and which one do you prefer the most?

SH: I am not a high volume surgeon. For me, the least complicated and least risky method is to use the vitrector, with takes very small bites and does not put any tension on the capsule at all.

ARV: We prefer manual continuous curvilinear capsulorhexis which is usually performed with microincision capsulorhexis forceps.

JR: In children, high viscosity OVDs like sodium hyaluronate 1.4% is very helpful to facilitate anterior capsulorhexis. We prefer the manual anterior continuous curvilinear capsulorhexis the most as its strength and edge characteristics have been studied in great detail. Other techniques may not result in consistent results and increase the scope of surgical complications. Staining the capsule is a useful strategy to delineate the capsule better.

SKK: In case of anterior capsular fibrosis, Fugo plasma blade, 23/ 25G scissors or a vitrectomy cutter can be used to create the capsular opening. In routine cases, Utrata/ or 23/25 G ILM peeling forceps suffice.

KK: In infants and younger children, I occasionally use the vitreous cutter to create anterior capsular opening.

TA: What is the age limit until which you perform posterior capsulorhexis and does it depend on etiology of cataract?

SH: I do a vitrector capsulectomy in all children.

ARV: Our strategy is that all children under the age of 3 years are subjected to posterior continuous curvilinear capsulorhexis (PCCC) and anterior vitrectomy. Children between 3 to 6 years are subjected to PCCC but no vitrectomy. In children over 6 years, PCCC is not performed. However, even in older children, if the child is unable to cooperate for an Nd:YAG capsulotomy for any reason, a PCCC is performed. If there is plaque on posterior capsule, it is removed with either manual PCCC or with vitrector.

JR: Maximum age of primary posterior capsulotomy (PPC) is highly variable from one surgeon to another. In our practice, I perform PPCs in children less than 6 years of age. At times, the age is relaxed further, up to 8 years, but it is unusual to perform PPC beyond that. In children with trauma or uveitis or posterior capsular plaque or posterior polar cataract or posterior lenticulon, we perform it more often.

SKK: I perform primary PCCC in all children up to 8 years of age routinely. Sometimes even bigger children like post traumatic cataracts (increased risk of VAO), children uncooperative for Yag Cap (mental retardation, nystagmus, Down’s) can also be performed.

KK: I perform posterior capsulorhexis till the age of 7-8 years. If there is a primary posterior capsular opacity as in cases of posterior subcapsular or complicated cataracts, I would perform a posterior capsulorrhexis even in older children.

TA: What are the pearls for performing a good posterior capsulorhexis in pediatric eyes?

SH: If I am not implanting an IOL, I use the vitrector to perform the posterior capsulectomy and anterior vitrectomy. The most important thing is to do it slowly. The vitreous is quite solid and needs to be nibbled bit by bit, without any traction I don’t want to leave any strands of vitreous behind, that could become a scaffold for lens fibres to grow on. If I have implanted an IOL I perform the posterior capsulectomy and anterior vitrectomy, after the IOL is in place, via the superior limbal wound, by retracting the iris and capsule with an iris hook, and placing the vitrector head behind the IOL.

ARV: After refilling capsular bag with a heavy molecular weight cohesive viscoelastic, PCCC is initiated with the vertical element of a 26-gauge cystitome. Vertical element is held at a slant and with a swiping motion, the posterior capsule is engaged and simultaneously directed towards the left (in the direction of the 3 O’clock position) and anteriorly (towards the microscope). Additional viscoelastic is injected in the area surrounding the puncture to make the posterior capsule concave or flat. Viscoelastic substance is not injected through the puncture towards the vitreous face. The left of the inferior end of the puncture is grasped with Kraff-Utrata capsulorhexis forceps or microincision forceps fashioning the PCCC in a clockwise manner. The result should be a well-centered PCCC concentric to and smaller than the anterior capsulorhexis.

JR: I prefer manual posterior continuous curvilinear capsulorhexis (PCCC) with needle cystitome and forceps. It is imperative to perform this in children less than 6 years of age or in eyes with nystagmus in which future YAG capsulotomy may be difficult. Aim for a 3 – 3.5mm rhexis and if one is uncomfortable with forceps, a vitrector may be used for creating the opening. One must not forget to perform a limited anterior vitrectomy.

SKK: For performing posterior capsulorrhexis, anterior chamber should not be overfilled with OVD to avoid making the chamber too deep which pushes the posterior capsule back and may create difficulty creating the rhexis. Under good from retroillumination system, a Utrata or 23G forceps can be used to complete the rhexis. Contrary to anterior capsulorrhexis, 23G forceps are more useful than Utrata forceps for posterior capsulorrhexis.

KK: Aim to keep the size of the posterior capsulorrhexis about 1-1.5 mm less than the anterior rhexis. Try not to disturb the anterior vitreous face by injecting viscoelastic in the space of Berger.
Experts' Corner

**TA: What is the minimum age in which you would implant an IOL and does it depend on laterality of cataract?**

**SH:** I leave babies aphakic. Bilateral cases will wear glasses. Unilateral cases will wear a contact lens. I did insert IOLs in babies with unilateral congenital cataract, for a while, but as in the Infant Aphakia Treatment Study, I found there were more post-op complications and the visual outcomes were just as poor. We are fortunate to have an excellent contact lens service available.

**ARV:** IOL implantation is still controversial in children under 2 years, especially those under 1 year, as the safety of IOL implantation in these eyes is not proven. Although IOL implantation should be aimed at particularly in unilateral cataracts, in all eyes, an IOL should be implanted only if it can be safely implanted in the capsular bag. Ciliary sulcus fixation of IOL should be avoided in children.

We suggest that if you are starting out with pediatric cataract surgery, you should start operating children older than 5 or 6 years. Only after you have gained experience in older children, should you operate younger children. Think 100 times before implanting an IOL in children younger than 2 years of age.

**JR:** With the infant aphakia trial results, I am in the favor of implanting IOLs even in children less than 1 year of age. We have implanted IOLs in children in as young as one month of age if the eye does not have any contraindications for the same. Our unpublished data in this regard has exciting results with a large number of cases where we have implanted IOLs in infants. We advise against IOL implantation in microphthalmos.

**SKK:** We evaluate the child’s eye in terms of axial length, white to white diameter during EUA and other eye status. If ocular examination suggests normal structure, an IOL can be implanted in children as young as 1 month. In unilateral cases, we generally prefer to implant IOL compared to bilateral cases where spectacles can still be prescribed. We implant IOLs in all cases more than 6 months of age if there is no other contraindication such as PFV, microphthalmos etc.

**KK:** In cases of unilateral cataract, I prefer to implant IOL after 6 months of age and in bilateral cataracts after 2 years of age.

**TA:** What types of IOL do you implant in pediatric eyes and do you find any differences in results on follow up on the basis of IOL material and design?

**SH:** The problem is in getting the correct power IOL rather than the right design. For smaller eyes I would use a three-piece foldable lens.

**ARV:** We prefer only single piece foldable hydrophobic acrylic IOLs in pediatric eyes. However, in cases where optic capture of IOL is performed, a 3 piece hydrophobic acrylic IOL is implanted.

**JR:** It has been shown in a number of studies that foldable hydrophobic acrylic IOLs are the most bio-compatible and provide the most favorable results with fewer incidences of PCO. This is our obvious preferred choice; however, in a resource strained country like ours, we often are compelled to implant PMMA IOLs. After analyzing our patients, I believe that more research is required in this area to develop an ideal IOL material and design.

**SKK:** Putting a foldable acrylic hydrophobic IOL within the bag is the safest when done in pediatric eyes. Multifocal hydrophobic IOLs also show good outcomes in pediatric eyes. Rigid lenses also show good outcomes in pediatric age group except for the need to enlarge the incision size and suturing required. We don’t have any experience with hydrophilic lenses.

**KK:** My preferred choice is a single piece foldable hydrophobic acrylic IOL. They tend to fare much better in terms of long term inflammation, stability and PCO rates as compared to other IOLs.

**TA:** What is your protocol for choosing correct IOL power in different pediatric age groups and how often do you find IOL surprises?

**SH:** We measure the axial length and the keratometry whilst the child is under anaesthesia and then do the IOL calculation. I always aim to make the child myopic to try to lessen the amblyopia. In a young child, I will aim for -1.50D and in an older child -0.50D. In bilateral cases I try to leave one eye emmetropic, and the other myopic. Using this protocol, the children can usually manage well without glasses in primary school, and will be myopic and require glasses in high school. Often they end up with a refraction of -7D, but I think their amblyopia is less.

**ARV:** Our preferred approach is to undercorrect the IOL power to avoid huge myopic shifts as the child grows up. The amount of undercorrection depends primarily on age at surgery: younger the child, more the undercorrection. However, it also depends on other factors such as fellow eye status, family history of myopia / refractive errors, laterality and severity of amblyopia. Several normograms on IOL power selection have been published in literature. If the decision regarding IOL implantation needs to be changed, e.g. in cases of ciliary sulcus, appropriate adjustment may need to be done. However, even after undercorrection, we do see refractive surprises sometimes.

Our approach for undercorrection depending upon the age at the time of surgery is as follows:

<table>
<thead>
<tr>
<th>Age</th>
<th>Undercorrection (%)</th>
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<tbody>
<tr>
<td>0-3 months</td>
<td>35%</td>
</tr>
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</table>

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Experts’ Corner

3-6 months 30%
6-12 months 25%
1-2 years 20%
2-4 years 15%
4-6 years 10%
>6 years 5%

As an example, in a 7 months old child, if the IOL power is calculated at + 36.0 D, we undercorrect by 25%. So IOL power to be implanted would be 36 - 9 = + 27.0 D.

JR: In children, IOL power calculation is done using the guidelines by Dahan and Drusedau. Under-correction of 20% is applied for less than 2 years and 10% for children up to 6 years of age. However, now there is a trend to aim for emmetropia, especially in older children. For years, surgeons have used the SRK II formula but we now prefer SRK/T or Hoffer Q.

IOL surprises are common in complicated cases like children with Marfans where one cannot account for the pathological myopia. With better techniques to ensure accurate axial length and corneal curvature, we see lesser postoperative surprises. Fortunately, we are now better equipped to deal with surprises, if at all they occur.

SKK: For implantation of IOLs in pediatric age group, following calculation is used.

<3 months: 30%
3-6 months: 20-25%
6m-1 year: 15-20%
1-2 years: 10-15%
2-5years: 2-5%
5-6years: 2%

It is not rare to find IOL surprise. In few of our cases, due to error in biomey or erroneous, immediate IOL surprise was found. These cases were immediately planned for IOL exchange. Most cases of IOL surprise are encountered after few years of IOL implantation when eye undergoes natural myopization with time. In such cases, piggyback IOL can be implanted or IOL exchange can be performed. In bilateral cases, spectacles can be prescribed or IOL can be implanted or IOL exchange can be performed.

KK: I use the Dahan’s criteria for choosing correct IOL power in children, although tend to under-correct a bit less. The incidence of refractive surprise is pretty low and is limited to unco-operative children in whom axial length or keratometry could not be measured accurately or in children having very short or very long eyeballs.

TA: What are the indications of optic capture in these cases and what are the salient points for carrying it out successfully?

SH: I have not aimed for optic capture, but sometimes it occurs without my effort.

ARV: In-the-bag fixation is the most preferred site of IOL implantation both in adults and children. An alternative technique to stabilize the IOL is optic capture. It is believed that optic capture would allow surgeons to avoid planned anterior vitrectomy and minimize the risk of PCO.

Optic capture fuses the anterior and posterior leaflets of the capsular bag almost completely in a 360° position, except at the haptic–optic junction. Theoretically, capsular fusion anterior to the IOL optic might reduce central lens epithelial cell migration or at least direct the cell movement anteriorly over the lens optic, which is presumably an unsuitable substrate for lens survival. Thus, capsular fusion may help in decreasing VAO. Another advantage of performing optic capture through is its ability to achieve a well-centered IOL.

Following manual anterior continuous curvilinear capsulorhexis, and irrigation/aspiration of lens material, a continuous manual PCCC is performed. Under a high viscosity ophthalmic viscosurgical device (OVD), the IOL is implanted in the capsular bag. Thereafter, the IOL optic is gently pressed backwards and is captured through the posterior rhexis using a spatula / Leister hook. The haptics are placed in the capsular bag fornix. If the round opening is stretched into an elliptical one, it indicates complete capture.

Optic capture is a technically challenging procedure. The key point is to achieve a “capturable” PCCC. Although there are several options for performing PCCC, a continuous manual PCCC is a prerequisite for optic capture of the IOL through the PCCC. The opening in the posterior capsule should not only be continuous, but also well centered and of an optimum size. If the opening is too small, it is difficult to capture the optic and there is the possibility of tearing the posterior capsule due to excessive stress to the PCCC edge. On the other hand, if it is too large, the optic may not remain captured. Thus creating an opening of an optimum size is a prerequisite for successful optic capture. The diameter of the PCCC opening should be approximately 1.0 mm smaller than the IOL optic. The principal behind the use of this technique is to avoid the need for vitrectomy. However, occasionally, the vitreous face is disturbed while performing primary PCCC or capturing the IOL optic and unplanned vitrectomy may be required.

JR: Optic capture remains our second choice of IOL fixation after preferred capsular bag implantation of IOL. In cases of posterior lenticonus or unusually large posterior
rhexis optic capture provides stable fixation of IOL. Trick to perform optic capture is to first implant IOL with its haptics into sulcus and the press optic edge on each side to capsular bag. Posterior optic capture where haptics are in the bag and optic is behind the posterior capsule is technically more difficult.

**SKK:** Optic capture is performed where either anterior or posterior capsulorrhexis is non-intact. It is generally required in traumatic cases/ or in eyes with pre-existing posterior capsular defect where it may not be feasible to put the IOL in bag by buttonholing the optic posteriorly through the PCCC. We prefer to implant IOL in the bag in routine pediatric cases rather than performing posterior optic buttonholing.

**KK:** In my hands, indications of Optic capture are limited to cases in which the posterior capsular support is inadequate like in posterior lenticous or traumatic cataracts. In such cases I prefer to implant a 3 piece hydrophobic acrylic IOL in the sulcus and capture the optic through the anterior capsulorrhexis. One very important point to be remembered is that the capsulorrhexis should be smaller than the optic size. Also optic capture is much easier with a 3 piece IOL than a single piece IOL.

**TA:** What is the incidence of PCO formation in your cases and how do you manage it subsequently?

**SH:** In all cases under four years of age, I will perform a primary posterior capsulectomy and anterior vitrectomy. In children older than four, I rely on them being able to cooperate for a YAG capsulotomy when the time comes, which it almost always does, eventually.

**ARV:** We have observed that age at surgery and management of posterior capsule are important factor for development of visual axis obscuration.

The Incidence is almost 8-10%. We manage it either with yag capsulectomy or pars plana membranectomy with vitrectomy as per child’s age and co-operation.

**JR:** PCO formation is a major concern in pediatric cataract surgery. There is no doubt that young eyes are prone to inflammation and PCO. Primary posterior capsulotomy with anterior vitrectomy helps to significantly reduce the occurrence of PCO to less than 20% in young children. Despite all surgical advances, PCO remains a challenge for all of us. I have operated a number of children with cataract secondary to uveitis who require aggressive anti-inflammatory cover. Postoperatively, some children are amenable to YAG capsulotomy. However, surgical membranectomy may be required in children with thick PCOs to ensure clear visual axis.

**SKK:** In our patients, the incidence of PCO is approximately 3-5%. In the initial post-operative period, generous use of strong cycloplegics (2% homatropine) is attempted in conjunction with topical steroids. If the PCO fails to respond to these, membranectomy is planned if the patient is uncooperative for Nd: Yag laser capsulotomy or if the child is cooperative enough for Yag laser, Yagcapsulotomy is performed.

**KK:** With the use of hydrophobic acrylic IOLs and performing posterior capsulorrhexis and anterior vitrectomy, the incidence of PCO has dropped down considerably to 5-10%. In case of PCO in an older co-operative child, Yag capsulotomy in the OPD can be done. In younger children or in thicker PCO cases, a membranectomy is needed in the OT.

**TA:** How early do you perform postop refraction and do you prescribe contact lenses or spectacles? What protocol do you follow for amblyopia management?

**SH:** For babies with unilateral congenital cataract, I perform the retinoscopy and order the contact lens, one week post-op. The parents are taught how to insert the lens etc, one week after that. The sound eye is patched for 50% of the waking hours, until the child has developed some visual curiosity and interest. This is usually at about 4 months of age- and then the patching is increased to 90% of the baby’s waking hours.

**ARV:** In pseudophakic patients, a fortnight after
suture removal, spectacles are prescribed with complete correction for distance. Thereafter, every 3 months, if refraction changes, appropriate spectacles are prescribed. At the age of 2.5 years, spectacles with near addition of + 2.5D are prescribed. However, executive bifocal ‘D’ is preferred.

Similarly in aphakic children, after suture removal, spectacles are prescribed with complete correction for distance. Aphakic spectacles of not more than + 20 D are prescribed. Every 3 months, if refraction changes, appropriate spectacles are prescribed. No addition of + 2.5 D is given till 2.5 years (bifocal aphakic eyes cannot be more than + 14.0 D). We preferred plastic spectacle frames with fiber glass.

**Contact Lenses:**

In all aphakic patients, particularly unilateral cataracts, on the second postoperative day, contact lenses are prescribed of + 25.0 D with help of pre-operative keratometry and corneal diameter data. The material used is silicon hydrogel. On every EUA – CL, the lens fitting is checked and power and parameters are re-confirmed.

**Patching Regimen**

When we find strabismus or in cases of unilateral cataracts, we start patching according to degree of amblyopia. Parents are instructed to have their child wear an adhesive occlusive patch. We usually advise partial occlusion for atleast 6 hours postoperatively. If an allergy developed to occlusive patches, a cloth patch could be worn over the spectacle lens of the pseudophakic eye.

**JR:** We perform postoperative refraction as early as possible – once the inflammation has subsided significantly usually within first two weeks. It is important to underline the importance of immediate use of refractive correction and we prefer glasses because most patients cannot afford contact lenses. Preschool children can be provided with near add and older children can be given bifocals. We must check refraction in every visit and correct if there is a change of 0.5 diopters. Amblyopia management is begun early and we recommend full-time occlusion initially and then scale it down to part-time in sort of a dose-dependent manner.

**SKK:** Post-operative refraction is done first during EUA when the child is taken up for cataract surgery for the other eye once one eye is operated in a case of bilateral cataract. If the biometry of the two eyes is similar, same power of glasses is prescribed to both both eyes provisionally. Final refraction is prescribed after suture removal under EUA at 4-6 weeks after surgery.

**KK:** In cases of aphakic children, I perform the refraction as early as 3 days and prescribe glasses. I prescribe contact lenses in all unilateral aphakes and in some cases of bilateral aphakes where the parents are extremely motivated to handle contact lenses. I follow the protocol for amblyopia management suggested by the ATS group.

**TA:** In which pediatric cataract cases do you observe exaggerated post-operative reaction and how do you prevent or manage the same in such cases?

**SH:** An exaggerated inflammatory response occurs particularly in eyes with a very dark iris. I such cases I prescribe hourly dexamethasone drops for the first few days. The rebound tonometer makes it a lot easier to check for steroid induced IOP elevations.

**ARV:** Exaggerated postoperative inflammation is expected in eyes with traumatic cataracts, uveitis, juvenile rheumatoid arthritis, small pupils and extremely small eyes of infants particularly those under the age of 6 months.

In these eyes, our regime is to treat inflammation aggressively by giving topical corticosteroids (1% prednisolone acetate) with an extended taper over 3 months, topical atropine as well as oral steroids. In these eyes, it is also a good idea to inject intracameral preservative free triamcinolone acetonide at the end of surgery to reduce the inflammatory response.

In eyes with chronic uveitis and complicated cataracts, intravitreal injection of ozurdex (depot preparation of steroid) is also useful as it combats the inflammation within the eye without causing systemic side effects associated with oral steroids.

**JR:** Cataracts secondary to trauma or uveitis are inherently notorious for higher levels of inflammation. Cases with higher intraoperative tissue handling, like those with lens coloboma or spherophakia may show increased flare. In certain cases of non-infectious uveitis, we have found great results with intraoperative dexamethasone implant, Ozurdex. In other cases, we recommend frequent intense topical betamethasone therapy, occasionally combined with oral Prednisolone. Steroids are tapered over 6 – 8 weeks and homatropine can be used twice a day to prevent irido-lenticular adhesions.

**SKK:** Exaggerated post-operative reaction is seen in certain conditions like previously operated eyes, eyes with Juvenile Idiopathic Uveitis (JIA), post-traumatic eyes, infants, eyes with rubella etc.

In case of excessive reaction, strong steroids and cycloplegics should be administered to the child after ruling out endophthalmitis. Pre-operative steroids and cycloplegics can also be considered in high risk conditions.

**KK:** Traumatic and complicated (uveitic) cataracts. I perform the surgery under steroid cover and only after the inflammation has been well controlled.
**Experts' Corner**

**TA: What postoperative complications do you observe most commonly on follow-ups and how do you deal with them?**

**SH:** There are a lot of possible complications. Amblyopia is the commonest, of course. Pupil distortion, is a concern for parents. Glaucoma occurs in about 30% of eyes over the child’s lifetime- so lifetime follow-up is recommended.

**ARV:** The most common complications seen following pediatric cataract surgery are glaucoma and visual axis obscuration. Younger the age, more the incidence of these complications. The best way to deal with these is to perform frequent postoperative examinations, under anesthesia if needed, so that early diagnosis can be performed. Visual axis obscuration is treated with Nd:YAG capsulotomy if the child is co operative, or with a pars plana membranectomy and vitrectomy. Glaucoma is treated medically with eyedrops, and if uncontrolled, is treated surgically.

**JR:** Complications are common after pediatric cataract surgery. I would list them as PCO, amblyopia, glaucoma, uveitis, IOL position defects and retinal complications in the order of importance. The most important way to deal with them is to recognize them early. We must suspect them at every follow-up visit. Spending time with the patients family, counseling them and building a support team of paediatric optometrists, paediatric glaucoma and retina specialists is important in effectively managing these patients.

**SKK:** The children if operated by standard surgical techniques and good post-operative care infrequently present with undesirable outcomes. However certain children are predisposed to complications and should be monitored carefully. They include infants less than 6 months, post-traumatic cataracts, previously operated eyes such as trabeculectomy, vitreoretinal surgery, keratoplasty etc. The most common complications seen in these groups include visual axis opacification (VAO), or IOL decentration in traumatic cases, and sometimes glaucoma.

In case of early VAO, we prescribe the infants with strong cycloplegics like atropine which may help to clear the VAO partially. In case it is dense and mature, it is best to re-operate the child.

**KK:** Visual axis opacification, aphakic glaucoma and amblyopia are the common complications that we encounter. Regular and vigilant follow up schedule and aggressive treatment in case any of the complications are detected is the best way to go about dealing with them.

**TA: Do you use toric IOLs /multifocal IOLs/ scleral fixated IOLs in pediatric cases and what are your suggestions for the same?**

**SH:** No, No and No!

**ARV:** We do not prefer multifocal IOLs in pediatric cataract surgery. The inherent limitation of a multifocal or a bifocal IOL design is that there is some loss of contrast and light energy loss. In children, the primary goal is to provide a focused and sharp image, and glasses is the secondary concern. Therefore, we aim at monofocal IOL implantation with correction of residual refractive error with spectacles.

However, in children older than 5 years of age and who have greater than 1.5 diopters of corneal astigmatism, we do implat Toric IOLs. In our series of more than 20 eyes, we have found the use of Toric IOLs beneficial in reducing corneal astigmatism, and thereby improve the quality of unaided vision for the patient. This is extremely important for rehabilitation of these children.

Scleral fixation of IOL is reserved for those children with ectopia lentis where the capsular bag cannot be preserved. This is performed only in children who are above the age of 3 years. 9-0 prolene or gortex sutures should be used to fixate the IOL to the sclera, since there are several reports of degradation of the suture material over a longterm followup with 10-0 nylon suture. In cases of ectopia lentis, however, my preference is to try and preserve the capsular bag and implant an in the bag IOL alongwith bag fixation, in order to maintain the natural compartmentalization of the eye.

**JR:** We believe that like in adult eyes, there is a scope for the use of next generation IOLs like toric, multifocal and scleral fixated IOLs in children. We recently published a series on bilateral implantation of multifocal IOLs which received encouraging comments from a number of scientific institutions. Another study comparing scleral fixated IOLs with lensectomy-vitrectomy and in-the-bag IOLs is underway. Today, we have much more in our armamentarium against pediatric cataract than what we had a few years ago. While only more research can shed light on this topic, a caveat is that appropriate case selection is a must before considering these options.

**SKK:** Scleral fixated IOLs: They should be used only once the scleral elasticity of the child’s eye reaches that of an adult eye. I prefer to use them only when a unilateral aphakic child crosses 10 years of age.

**Toric IOL:** Due to unreliability of biometric readings in children, I do not implant toric IOLs in children less than 10 years of age.

**Multifocal IOLs:** We have used multifocal IOLs in children (age group 10 years or more) with good outcomes for both near and distance vision. At this age, a child is neuro-adaptive and can achieve good outcomes.

**KK:** I have used scleral fixated (sutured with 9-0 prolene suture) IOL in aphakic patients with good outcomes. I do not use toric/multifocal IOLs in paediatric cases.
Experts’ Corner

**TA:** What are the future trends in pediatric cataract surgery and do you think femtosecond laser will have an important role in this field?

**SH:** I hope that future trends will include the ready availability of very high powered IOLs, sufficient to make the child see clearly for near, immediately after the surgery. I hope that rubella vaccination becomes universal so that rubella cataracts become historical reminiscences only. I don’t think there will be much of a role for the femtosecond laser.

**ARV:** Femtosecond laser has the distinct advantage of being able to perform an anterior and posterior capsulorhexis of desired size and shape. This is particularly crucial in pediatric eyes. I do believe that as the femtosecond technology evolves, it will be a great asset in pediatric cataract surgery. Currently, there are 2 concerns with its use in pediatric cataract surgery: one is that when performing posterior capsulorhexis in an already open eye, there may be a higher risk of contamination as the patient has to be shifted outside the operating room and docked to the laser machine. Also, the high cost of the procedure worldwide is an obstacle today.

However, I do believe that this technology will be very useful in pediatric cataract surgery in the times to come.

**JR:** Clearly, the future of pediatric cataract surgery is exciting. While there are a number of innovations in the pipeline, femtosecond laser is probably here to stay. It must undergo rigorous studies before it is an established technique in pediatric cataract management. It is important that we continuously update our surgical skills and contribute effectively in shaping a bright future for these children.

**SKK:** Femtosecond laser is being used in a big way for adult cataracts. It may prove to be a boon for pediatric cataracts as it has a role in creating anterior and posterior capsulorrhexis in pediatric eyes. However, due to logistic issues such as docking which is required twice and increased surgical times under general anesthesia, it may not be a practical approach for all patients.

**KK:** Use of premium IOLs like multifocal or accommodative IOL with better designs, newer instruments for capsulotomy and better formulae for more predictable IOL power calculations.

Femtosecond laser needs to undergo a lot more refinements and developments before it will be routinely used for paediatric cataract surgery.
IOL Power Calculation in Children

The placement of an intraocular lens (IOL) in children and infants undergoing cataract surgery is gaining wider acceptance. With improved surgical equipment and techniques, the acceptable age for IOL implantation is becoming progressively younger. However calculation and selection of an exact intraocular lens (IOL) power for the eye of a child which is in a process of growth presents varied challenges. The implantation of an IOL with a fixed power in an eye that is still growing makes it difficult to chose an accurate IOL power that best benefits the child’s eye. The younger the child, the more difficult is the problem.

This is a challenging task for any ophthalmologist but probably more so in the our settings due to the lack of instrumentation in the operating-room like hand held keratometers and the A-scan ultrasound which increases the difficulty of calculating the IOL power in pediatric cataract surgery. Even with the availability of these instruments in the operating room, small and developing eyes of children possess unique challenges when calculating an IOL power. Also the formulas that we use in these cases were primarily designed for adult eyes, hence their extrapolation in paediatric eyes can be disastrous.

IOL implantation after cataract surgery in children >1 years of age is now widely accepted, although the implantation of IOLs during infancy is still controversial. In these scenarios, it is important to have optimal approach for determining IOL power in children and infants. As implanting an IOL at the calculated emmetropic power in children risks significant myopia/ refractive surprise at ocular maturity. For each case, the IOL power needs to be customized based on many characteristics including the age at which surgery is planned, unilateral or bilateral, where ambylopia is present and likely compliance with glasses and occlusion therapy in the post operative period.

Here in again, ophthalmologist in our settings faces additional challenges. Many children present late and have dense ambylopia. In addition, parents compliance to ambylopia therapy and required regular follow-up may be poor. Thus, apart from calculation, selecting an IOL power and its implantation and its after effects are much more challenging to the ophthalmologist in the our setting compared to the west.

How to proceed

Before implanting an IOL in a child we need to be aware of physiological development of a normal, aphakic and a pseudophakic eye in children and their visual needs. We should know if a refractive shift is anticipated, and if so what is its nature and magnitude and at what age is it seen. Also we should be aware of target refraction that should be sought following the IOL implant.

Normal eye development

Maximal ocular growth occurs in the first few years of life with the changes as enumerated below.

The normal newborn eye has a mean axial length (AL) ranging from 16.6 to 17.0 mm and a mean keratometric power of 51.2 diopters (D). It reaches an appx mean adult value of 23.6 mm at appx 15 years of age. As the child’s eye develops, the refractive changes seen are primarily due to the growth in the AL. More than 50% of this change in AL occurs before 1 year of age and the maximum axial growth occurs during the first 2 years of life. O’Brien and Clark demonstrated a mean AL of 15.38 + 0.25 mm in preterm infants of 33 weeks. AL increased at a rate of appx 0.18 mm/wk until 40 weeks then slowed down to 0.15 mm/week until the age of 3 months. The mean AL of infants at 3 months was 18.23mm. The growth rate then slows...
down even further again to an appx mean adult value of 23.6 mm at 15 years of age.3,4,5.

The change in mean keratometric values occurs primarily within the first 6 months of life. The corneal curvature decreases from an average power of 51.2 D at birth to a mean of 43.5 D (adult value), as the AL increases from an average of 16.8 mm at birth to 23.6 mm in adulthood.

Due to above mentioned changes, during the first year of life, lens and total eye power decreases by more than 10 D but there after it decreases only 3-4 D from the age of 2yrs to 10 years of age when it stabilizes6.

Refractive shift after surgery

In pseudophakic eyes, the implanted IOL power does not change as in aphakia also it is static. If the AL grows normally as in an unoperated child, increasing myopia or reduction in hypermetropia would be expected. Axial growth after cataract surgery can be due to normal eye growth as in hypermetropia would be expected. Axial growth after 2 years at the time of surgery, amount and duration of visual deprivation, aphakia or pseudophakia, laterality and genetics7,8.

McClatchey and Parks followed a series of aphakic children into adulthood and found a that there was a decrease in the hypermetropic error which followed a logarithmatic regression curve 7 and was very similar to that predicted from Gordonand Donzis’s study data on phakic children9. They also calculated the long-term effects of pseudophakia on refraction in aphakic eyes on long-term follow-up and predicted an amount of 6.6 D mean myopic shift over a mean follow-up of 11 years.

Most of the children below 2 years at the time of the cataract surgery with or without IOL implantation have a significantly greater than predicted myopic shift and a greater variance in the refractive change than those older than 2 years at the time of surgery. Vanathi et al noted a mean myopic shift of 7.35 D in 12 children (mean age 6.7 years) post-uniocular cataract surgery followed for a mean of 7.8 years10. In a study of 52 eyes in 42 patients aged 12 months to 18 years undergoing cataract surgery with IOL implantation, Crouch reported a mean myopic shift of 3.66 D in children operated on at the age of 3-4 years, 2.03 D in children operated on at the age of 7-8 years, 1.88 D in children operated on at the age of 9-10 years, 0.97 D in children operated on at the age of 11-14 years and 0.38D in children operated on at the age of 15-18 years8 (Table 1).

Table 1: Amount of Myopic Shift

<table>
<thead>
<tr>
<th>AGE</th>
<th>Plager et al</th>
<th>Crouch et al</th>
<th>Wilson et al</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt; 1 yr</td>
<td>6.22D</td>
<td></td>
<td></td>
</tr>
<tr>
<td>2-3 yrs</td>
<td>4.6D</td>
<td>3.66D</td>
<td></td>
</tr>
<tr>
<td>3-4 yrs</td>
<td>2.68D</td>
<td>2.03D</td>
<td></td>
</tr>
<tr>
<td>6-7 yrs</td>
<td>1.25D</td>
<td>1.88D</td>
<td></td>
</tr>
<tr>
<td>7-8 yrs</td>
<td>0.61D</td>
<td>0.97D</td>
<td></td>
</tr>
<tr>
<td>9-10 yrs</td>
<td>0.38D</td>
<td></td>
<td></td>
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</tbody>
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Residual refractive error

It is very difficult to decide about what residual refractive error to be left after surgery in infants and children. Literature search also remains ambiguous in this question. Some surgeons are recently targeting emmetropia after surgery at all ages beyond infancy to counteract amblyopia, however most aim for hypermetropia up to 5 years of age when the consensus opinion is towards emmetropia.

Some surgeons have been known to choose adult IOL power in children younger than 2 years. This action is on the premise that with expected growth and the myopic shift, the child will have emmetropia and good vision in adulthood. However this strategy results in significant hypermetropia in the later years, a condition that need to be corrected. Near vision is very severely affected because the pseudophakic eye does not accommodate, and hence even mild hypermetropia may cause severe amblyopia in case of anisometropia.

Few authors recommend emmetropia postoperatively after IOL implantation in children< 2yrs of age especially in unilateral cases. This helps in easier amblyopia management in early postoperative period especially in patients with poor compliance and follow up. With later ocular growth, the resulting refractive shift towards myopia will require the use of a contact lens or a refractive surgery to correct the residual refractive error and minimize the resultant aniseikonia in such cases.

Some recommendations are therefore children between 2 and 4 years to have IOL implanted in a way to obtain a refraction equal to that of the fellow eye and then do a reduction in the IOL power by 2D to allow for myopic refractive shift. It is also suggested that children elder than 4 years of age be implanted IOLs with powers to match the spherical equivalent of the fellow eye. For older children, it is recommended to aim for emmetropia, and then adjusted depending on the other eye to avoid anisometropia greater than 3D.
Initial residual hypermetropia has the advantage that with the axial growth of the eye, the hypermetropia will reduce and the adult refraction will be closer to emmetropia. However, this advantage must be balanced with the fact that the uncorrected hypermetropia in children may cause exacerbation of amblyopia. To avoid this problem, some surgeons prefer to aim for emmetropia in the immediate postoperative period. It helps in the battle for preventing late myopia will be more and more apparent as the child’s eyes continue to grow. Thus, a better solution may be to find a compromise between these two extremes.

Enyedi et al recommended a postoperative refractive goal of +6 for a 1-year-old, +5 for a 2-year-old, +4 for a 3-year-old, +3 for a 4- year-old, +2 for a 5-year-old, +1 for a 6-year-old, plano for a 7-year-old and -1 to -2 for an 8-year-old and older.

In Wilson’s survey of ASCRS and JAAPOS members in 2001, there was wide variation in the aimed postoperative refraction for infants at 6 months of age, ranging from emmetropia to high hypermetropia (aprx 7D), with most aiming for moderate hypermetropia (>3D to <7D). For infants at 12 months of age, most surgeons aimed for moderate or mild >0D to <3D) hypermetropia. At 2 years of age there was a consensus to aim for mild hyperopia or emmetropia. This is consistent with the findings of Plager et al who postulated that a shift toward decreasing hyperopia i.e. myopic shift will occur at a decreasing rate throughout childhood and, hence, gives children a hypermetropic refractive error. The magnitude of the planned hyperopia increases with decreasing age and is modified by the AL and refractive error of the fellow eye.

In the Infant Aphakia Treatment Study, the target refractive error after IOL implantation is 8D for infants 4-6 weeks of age and 6D for infants 6 weeks to 6 months of age. There is no study that demonstrates a visual advantage of one approach over the other.

Biometry in children

In addition to problems already enumerated, the measurements of AL and keratometry in children are difficult to measure and less reproducible, accurate and predictable than for adults. Measurements of AL and keratometry in OPDs and office are very difficult and practically impossible in most young children and infants. Biometry is usually done under anesthesia in a small child who is unable to cooperate with precise fixation and centration (Figure 1a,b).

A-scan ultrasound biometry is the conventional method for measurement of AL in all age groups. It can be performed using either traditional applanation or immersion techniques. There may be a measurement error with applanation technique in recorded AL. It is important to make sure that the tip does not indent the cornea in applanation technique. It has been seen that the AL measurements made with this technique were, on the average, 0.28 mm less than measurements made using an immersion technique.

Figure 1(a): A-Scan under anaesthesia (b): Keratometry using handheld autokeratometer under anaesthesia
With the immersion technique, the probe does not come into direct contact with the cornea, a coupling fluid between the cornea and probe prevents inadvertent corneal indentation. When the probe is properly aligned with the optical axis of the eye the sound beam is perpendicular to the retina, the retinal spike is displayed as a straight, well defined steeply rising spike. When the probe is not properly aligned, the ultrasound beam is not perpendicular to the retinal surface and the retinal spike is displayed as poorly defined slow-rising spike.

We preferentially use the immersion technique in paediatric patients. Repeated measurements are taken until three equal measurements are obtained with sharp well defined retinal spikes. In addition to the problems enumerated above, in pediatric patients, AL measurement is frequently done in the operating room under anesthesia where a trained person may not be available.

The most appropriate ultrasound velocity for AL measurement is different at different ALs. Most machines employ a single average ultrasound velocity. For example, in a patient with axial myopia of 28.00 mm, AL is best measured at an average velocity of 1,550 m/sec, while in a patient with axial hypermetropia of 19.00 mm, AL is best measured at an average velocity of 1,560 m/sec. The human eye primarily comprises of aqueous and vitreous, of which both have an ultrasound velocity of 1,532 m/sec. Cornea and the lens have different ultrasound velocities. If the AL is measured at an average ultrasound velocity of 1,532 m/sec, a corrected axial length factor (CALF) of 0.32 mm is added to the measured AL to obtain the exact AL. This method is more accurate than using an average ultrasound velocity. In aphakia, an ultrasound velocity of 1532 m/sec is recommended.

Error in axial length (AL) measurement cause the most optically significant refractive errors in IOL power calculation and is appx 2.5 D per mm of error, which is very significant. This error increases to 3.5 D/mm in very short eyes (aprx 20 mm). Thus, it is crucial that we take every possible step to minimize error in AL measurement.

Mittelviefhaus et al in their study concluded that due to lack of fixation in children who have keratometry under general anesthesia, inaccurate keratometry readings may be measured16. For keratometry measurement, we avoid using a speculum and measure with a hand held Nidek AutoKeratometer. Measurements should be taken as soon as possible after induction of anesthesia to avoid the problems associated with corneal dryness. Balanced salt solution should be instilled on to the cornea to maintain a smooth corneal surface for accurate readings. We need only refractive power measurement with an auto keratometer, hence the problem of inaccurate axis measurement has got no bearing in these cases.

Partial coherence interferometry (PCI) has been recently used extensively in cooperative children. This technique relies on using a laser to measure the echo delay and intensity of infrared light reflected back from tissue interfaces. Advantages of PCI over conventional ultrasound techniques include high reproducibility, contact-free measurement, and observer independence of the measurements17. Now a days ray tracing in a I-Trace is also being used for IOL power calculation in cooperative children18.

**Intraocular Lens Power Calculation**

Once the plan has been made to do a cataract surgery and implant an IOL in the child. The desired postoperative refractive goal is determined according to the factors elaborated above. Next issue is to decide which power calculation formula should be used to reach that refractive goal. A number of formulas can be used to predict the IOL power needed to achieve the desired refractive goal. Primarily the formulas used in children for IOL lens power calculation have been largely derived from studies in adults. These intraocular lens power calculation formulas fall into two major categories; empirically determined regression formulas and theoretical formulas.

The regression formulas, such as the Sanders-Retzlaff- Kraff (SRK) formula, are based on analysis of a large samples of postoperative results in adults. The formula does not work well for unusually long (>25 mm) or short (<22.5 mm) eyes. The formula generally undercorrects short eyes and overcorrects eyes with long ALs, because it attempts a linear fit to a hyperbolic relationship. Hence these are notoriously very unreliable for small paediatric eyes.

The first-generation theoretical formulas assume that the IOL will be at a constant position or postoperative anterior chamber depth (ACD) will be similar in all eyes, regardless of their AL. Since the measured postoperative ACD was found in various studies to be directly proportional to the AL of the eye (longer eyes had larger ACDs), these formulas were found to be less accurate for long or short eyes. Several second-generation theoretical formulas have emerged, such as the Hoffer Q formula, that have replaced the constant ACD with one that included a correction for AL of the specific eye.

In adults, the Holladay formula is considered to be most accurate for eyes with an axial length between 22 and 26 mm. The Hoffer Q formula is considered to be most accurate for short eyes (<22mm) and the SRK/T formula is considered optimal for long eyes (>26mm).

In recent work by Eibsibitz et al, an analytical comparison of predicted implant power using keratometry values up to 55D and axial length values as short as 16 mm was performed for two different refractive goals using the SRK II, SRK/T, Holladay I, Hoffer Q, and Haigis equations. Significant differences in intraocular lens power prediction were found among the Hoffer Q, Holladay-I, and SRK...
II formulas in the pediatric range of axial length and keratometry values. The Holladay-I and Haigis formulas were found to be similar in their IOL prediction. The SRK/T was comparable to the Holladay I and Haigis formulas but still differed in the high keratometry values.

When determining individual ACD constants in the Hoffer Q formula for short, medium, and long eyes, the results in the short eye (<22.0 mm) series are less accurate using the personalized pACD derived from the 36 short eyes examined than when using the pACD derived from the entire 450 eye series. The same is true for the long eye (> 24.5 mm) series. This illustrates that developing a personalized ACD for AL subgroups at the extremes is of no value for the Hoffer Q formula and actually makes the results clinically less accurate in short eyes. A similar analysis performed for the Holladay and SRK/T formulas in short eyes showed no statistically significant benefit to a sub group of shorter long ALs using personalized SF or A-constant compared with using the overall 450 eye personalized SF or A constant.

Although no formula has been proven to have an advantage, it is preferable to use the theoretical formulas (e.g., SRK-T, Holladay I and Holladay II, Hoffer I and II, Hoffer Q and Haigis) because they are generally more accurate for small eyes, and in the pediatric studies they appear to be slightly more accurate overall.

**Author’s recommendations**

A one stop solution cannot be found for a complex problem of IOL power selection for paediatric patients. All of us have to formulate our own guidelines based on above mentioned facts and studies. Following is the protocol we follow at our centre which is working well for us. We take the final decision about the IOL based on following factors:

- **Whether to implant an IOL or not** - We donot routinely implant IOLs below 1yr of age in bilateral disease. In infants with unilateral cataract we aim for IOL implantation in all cases. However in some cases where AL<19mm, we may have to implant an IOL in the sulcus and in extreme cases <17mm AL, sometimes we have to leave the child aphakic also.

- **Age at the time of cataract surgery** - When an IOL is implanted in infancy, marked axial growth must be expected over the first 1 to 2 years after surgery. Therefore, IOLs implanted in infancy are usually selected to produce a 20% or more undercorrection. The closer to birth, the more marked this undercorrection will need to be. In children between 1-2 yrs we undercorrect by 10%. Beyond 2 yrs we leave the child emmetropic.

- **Status of the fellow eye** - More hypermetropia can be left when the surgery will be done bilaterally since non-compliance with glasses is less amblyogenic in these children. In an eye with monocular cataract, if the fellow eye is pseudophakic – it is important to look for refractive status in the fellow eye. Attempts should be made to minimize the anisokonia in the eyes with unilateral surgery.

- **Visual acuity** - Presence of dense amblyopia may influence towards a decision to have less hypermetropia (or even emmetropia) in an effort to help recover vision by using the occlusion therapy and minimizing the need for glasses. In this instance, late myopia is acceptable. Most of the studies show that it will be difficult to treat dense amblyopia. Myopia can probably be more easily handled with contact lenses or refractive surgery later in life.

- **Expected compliance of child/family to glasses/contact lens/occlusion therapy** - If poor compliance is expected – it is better to leave the least possible refractive error, otherwise dense amblyopia may result.

- **IOL power** - In general, the smaller is the AL, higher is the IOL power and more is the undercorrection needed. For example, at age 1 month, if one child has an emmetropic power of 50 D and another child at same age has an emmetropic power of 40 D, the first child will necessitate a higher residual refraction. In other words, we may use approximate expected refraction in the first child as +10 D, while in second child we would use +8 D. On an average following is the undercorrection applied age wise:

  - 0-6 Months +7.5
  - 6 Mon-1 yr + 5
  - 1 – 2 years + 2.5
  - > 2 years emmetropia

- **Implantation in-the-bag/ sulcus** - If a decision regarding the site of fixation needs to be changed after entering an eye and before IOL implantation – an appropriate adjustment is needed to be made. A plus-power IOL that is more anterior in the eye will have a greater refractive effect than if it were relatively posterior. Sulcus fixation of IOL produces a relative myopic shift from the estimated refraction. The IOL intended for capsular bag placement should be decreased by 0.75 D to 1.00 D (depending on the IOL power) when planned to be placed in the ciliary sulcus.

- **Secondary IOL Implantation** - keratometry reading errors have been known to occur after wearing RGP contact lenses which can persist up to 4 wks. Since the effects of RGP contact lens wear on corneal curvature and resultant IOL calculations is variable and unpredictable, it is important that IOL calculations be made at least 4 weeks after discontinuation of contact lens wear.
Pediatric Ophthalmology

Table 2: The mean predicted refraction, observed refraction and prediction error between SRK II and Pediatric IOL Calculator

<table>
<thead>
<tr>
<th></th>
<th>SRK II</th>
<th>Pediatric IOL Calculator</th>
<th>*p value</th>
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<tbody>
<tr>
<td>Mean Predicted Refraction (Diopter, SD)</td>
<td>-0.06 (1.17)</td>
<td>1.19 (2.35)</td>
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<tr>
<td>Mean Observed Refraction (Diopter, SD)</td>
<td>-0.84 (1.31)</td>
<td>0.10 (1.74)</td>
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<tr>
<td>Mean Prediction Error (Diopter, SD)</td>
<td>1.03 (0.69)</td>
<td>1.14 (1.19)</td>
<td>0.74</td>
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* Independent-Samples T Test
p value < 0.05 (significant)

Table 3: Accuracy of predictability between SRK II and Pediatric IOL Calculator

<table>
<thead>
<tr>
<th>Observed refraction (Spherical Equivalent)</th>
<th>SRK II (n = 16)</th>
<th>Pediatric IOL Calculator (n = 15)</th>
<th>Overall (n = 31)</th>
<th>* p value</th>
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<tr>
<td>≤ ± 2.0 Diopter</td>
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<tr>
<td>Accurate</td>
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<tr>
<td>0.0 D to ± 0.5 D</td>
<td>3 (18.75%)</td>
<td>7 (46.67%)</td>
<td>10 (32.26%)</td>
<td>0.097</td>
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<tr>
<td>Inaccurate</td>
<td></td>
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<tr>
<td>&gt; ± 0.5 D to ± 1.0 D</td>
<td>7 (43.75%)</td>
<td>0 (0.00%)</td>
<td>7 (22.58%)</td>
<td></td>
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<tr>
<td>&gt; ± 1.0 D to ± 2.0 D</td>
<td>4 (25.00%)</td>
<td>5 (33.33%)</td>
<td>9 (29.03%)</td>
<td></td>
</tr>
<tr>
<td>&gt; ± 2.0 Diopter</td>
<td>2 (12.50%)</td>
<td>3 (20.00%)</td>
<td>5 (16.13%)</td>
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* Pearson chi-square test
p < 0.05 (significant)

• **Parent’s refractive error** - it is also important to ask about high refractive error in parents. It has been noted that if both parents are myopic, 30% to 40% of their children become myopic, whereas if only one of the parents is myopic, 20% to 25% of their offspring will become myopic. If neither of the parents is myopic, fewer than 10% of their children will become myopic. Keeping these genetic factors in mind and anticipating more eye growth in these cases, these children may be left with more hypermetropia than stated earlier.

• **The Pediatric IOL Calculator** - It can be used for IOL power calculation in both primary and secondary IOL implantation (Figure 2). It needs the patients age, A constant of the intended IOL to be implanted, AL and K values. Desired post operative refraction can also be entered to get the optimum IOL power value (Table 2&3). The pediatric IOL calculator can be downloaded from the AAPOS website: http://www.aapos.org/proinfo/downloads.html[21].

**Conclusion**

Despite all our efforts and safeguards in IOL selection, it is not uncommon to see refractive surprises in children undergoing cataract surgery and IOL implantation. Besides these, several other factors (e.g., gender, race, etc.) have been reported to affect growth of the normal eye, and may also influence eye growth after cataract surgery.

Refractive growth and shifts after IOL implantation in infants and children cannot be predicted accurately due to large standard deviation and current IOL formulas vary in their predictive outcomes. If the target refraction goal is emmetropia, amblyopia treatment will be easier but may result in myopia later in life. If the target refraction goal is hypermetropia, amblyopia treatment may be more difficult but emmetropia later in life is more likely. Although placement of an IOL in children has gained universal acceptance and placement of an IOL in infants is gaining favor among many ophthalmologists, there remains no IOL power calculation formula derived primarily on the basis of specific peculiarities of the child’s eye or the studies comparing outcomes from IOL implantation in children. With the trend leaning towards implanting IOLs in infants with shorter ALs, there certainly will be a greater need to understand the applicability and differences between the

![Figure 2: View of a page of paediatric IOL Calculator](image)
various formulas at the lower ends of AL and keratometry values.

Using currently available formulas and fine tuning the A-constant and surgeon factor may reduce postoperative refractive surprises, but unlike adults, volumes of surgeries are not there with most Ophthalmologists and with a wide range of AL and K values rendering adjustment of A-constants and surgeon factors very difficult, the one stop solution is unlikely in near future. Any modern IOL formula can be used on children but more error should be expected. Using immersion A-scan instead of contact and repeating K-readings to make sure they are reproducible may be a way to reduce errors. In children, given the need for highly accurate biometry, astigmatism control, and no refractive growth, caution should be used in considering the use of multifocal IOLs in infants and children.

Surgeons, especially who implant IOLs in infants and young children must be prepared for wide fluctuation in the long-term myopic shift. Both the amount of the myopic shift and the variance in this shift are likely to be greatest in children having surgery in the first few years of life. Anticipation of this refractive phenomenon and appropriate correction or compensation will help achieve better functional outcomes of these young eyes undergoing cataract surgery. We must also remember that an IOL implanted in a child’s eye must stay there for many years, ie his whole life time. Long-term outcomes will certainly remain unsure for years to come till a less ambiguous solution is found. Long-term follow up and evaluation of outcomes of refractive error in these eyes will help us to develop formulas specially-suited for IOL power calculation for childhood cataract.

References

www.deorderline.org | 29
Congenital corneal opacities (CCO), by definition, are present in the newborn. The prevalence of CCO is approximately 3 per 1 lac newborns. However, this increases to 6 per 1 lac if congenital glaucoma is also included. CCO is either unilateral or bilateral and the cause could be hereditary, developmental, metabolic or infectious. Accurate and early diagnosis is required for correct prediction of the natural history of the disorder, to look for associated ocular and systemic disorders, appropriate genetic counseling and for establishing a proper management plan.

Congenital corneal opacities (CCO) have been classified traditionally by a mnemonic ‘STUMPED’ (Figure 1). However another classification system has been recently proposed which may be better considered from a perspective of pathogenesis, surgical intervention and prognosis. The authors believe that though the ‘STUMPED’ classification may be helpful in remembering the aetiologies involved, it is not of much help in understanding possible pathogenesis. Nischal et al have proposed that CCO is either primary or secondary. While primary CCO includes corneal dystrophies and choristomas presenting at birth, Secondary CCO could either be kerato-irido-lenticular dysgenesis (KILD) or other secondary causes including infection, iatrogenic, developmental anomalies of the iridotrabecular system or lens or both, and developmental anomalies of the adnexa. The authors believe that this classification may be more appropriate in determining prognosis of any surgical intervention (Figure 2).

Accurate diagnosis and management of congenital corneal opacities begins with a detailed and complete maternal, paternal, obstetric and family history and a thorough systemic examination. Gross ocular examination could be initiated in the clinic, however a complete examination often requires an examination under general anesthesia (EUA). EUA kit is exhaustive and includes instruments for measurement of corneal diameter, intra-ocular pressure, accurate refraction and dilated fundus examination. (Figure 3) A-scan, B-scan and ultrasound biomicroscopy (UBM) could also be often required. Gonioscopy in a neonate is done using a Koepppe lens.

<table>
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<tr>
<th>Figure 1: STUMPED classification of Congenital Corneal Opacities</th>
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Congenital corneal opacity is an emergency and requires management by a paediatric corneal specialist. If not treated early, these would lead to permanent visual deprivation amblyopia. In this communication we describe the salient clinical features of common etiologies of congenital corneal opacities which would help the clinician in accurate diagnosis, differentiation and management. For the ease of our readers, we follow the ‘STUMPED’ classification.

1. **Sclerocornea**: Sclerocornea is the primary CCO present at birth. It is unilateral or bilateral usually asymmetrical scleralization of the peripheral or total corneal tissue. It is usually occurs sporadically but could also be familial or autosomal dominant.

   The corneal opacity is usually non-progressive and is an extension of the sclera on the cornea with presence of fine superficial vessels and loss of limbal landmarks. (Figure 4) Histologically, there is an irregular arrangement of the collagen fibres, loss of the lamellar arrangement of the corneal stroma with presence of vessels. Four variants of sclerocornea have been described:

   I. **Isolated Sclerocornea**: No other ocular abnormalities
   II. Sclerocornea plana
   III. Sclerocornea associated with Peter’s anomaly
   IV. Total Sclerocornea

   Management plan should be made after a UBM examination.

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**Figure 2**: Newer classification system based on pathogenesis, surgical intervention, and prognosis

**Figure 3**: Examination under anesthesia kit
to know the status of the anterior segment and the presence of a posterior Descemet membrane defect. The treatment is only surgical but the prognosis is guarded. Hence the decision to operate is difficult if the condition is unilateral and the visual acuity of the other eye is good. However, bilateral sclerocornea warrants early intervention to prevent amblyopia (Figure 5).

2. Congenital Glaucoma: Perhaps the most commonly seen and the easiest to diagnose of all the congenital corneal opacities is congenital glaucoma. While early and accurate diagnosis and successful treatment involving intraocular pressure control to a level where progression is unlikely would reverse the effect and preserve vision, a delayed diagnosis results in irreversible visual loss. Childhood glaucoma is a rare disease with an incidence of 1 in 10,000–18,000 births\(^5\). It is seen more frequently in males\(^6,7\) and is bilateral in 70% to 80% cases\(^8,9\) as:

a. *Primary*: isolated idiopathic developmental abnormality of the anterior chamber angle

b. *Secondary*: reduced aqueous outflow – congenital/ acquired ocular or systemic disorder

The children are brought by the parents with the complaints of watering, photophobia and blepharospasm. Examination reveals an elevated intra-ocular pressure, enlarged and clouded cornea due to breaks in the Descemet membrane and optic nerve cupping. An important sign of increased IOP is an enlarged eyeball due to an elastic cornea and sclera. The normal corneal diameter of an infant is 10-10.5 mm. A horizontal corneal diameter more than 11 mm is suggestive and more than 13 mm is pathognomonic of congenital glaucoma\(^11,12\) (Figure 6).

The diagnosis of congenital glaucoma is based on an accurate history and clinical examination including examination under anesthesia. Management is purely surgical and should be done by a glaucoma specialist. The choice of surgery could be goniotomy, trabeculotomy or trabeculectomy and depends on the clarity of the cornea. The most common surgery performed is combined trabeculectomy with trabecolotomy, sometimes with adjuvant mitomycin \(C\)\(^11,12\).

3. *Birth Trauma*: During an assisted forceps delivery during child birth, pressure induced by the forceps’ blade kept across the head might lead to blunt trauma to the eye and rupture of the Descemet membrane\(^13,14\). Evidence of other peri-orbital injuries might be co-existant at birth. Left eye is more commonly affected
due to left-occipito-anterior being the most common presentation\textsuperscript{13,14}. The Descemet tear is usually unilateral, vertical and leads to transient corneal edema at birth which usually clears due to resurfacing of the young corneal endothelium\textsuperscript{13,14} (Figure 7). This leads to high residual corneal astigmatism requiring urgent correction to prevent amblyopia. The most important differential diagnosis is congenital glaucoma which can be easily differentiated based on high IOP, large corneal diameter, corneal edema which occurs weeks after birth and clears when IOP is lowered, Descemet tear which is horizontal than vertical or oblique and an abnormal optic nerve head as seen on fundus examination.

Rigid gas permeable lenses along with occlusion therapy are the mainstay of treatment. Traumatised endothelium might show evidence of decompensation in future requiring penetrating keratoplasty\textsuperscript{15}.

4. \textit{Ulcer:} Corneal ulcers though rare are an important cause of congenital corneal opacity. Any fluorescein stained epithelial defect should be suspicious and examined for a corneal ulcer, commonly bacterial, viral or neurotrophic\textsuperscript{13}.

\textit{Herpes Simplex Keratitis:} Congenital Herpes simplex virus (HSV) is contacted after a birth through an infected birth canal. Neonatal HSV is acquired either prenatal or peri-natal from the mother. HSV is an oculo-systemic disease and diagnosing it early is important to prevent mortality\textsuperscript{16,17}.

Conjunctivitis, purulent or muco-purulent, is the most common finding of pediatric HSV infection. Ulcerative keratitis is usually epithelial and could be in the form of macro-dendrites, geographical epithelial defects or punctuate keratopathy. Isolated stromal keratitis is rare. Complications like cataract, chorioretinitis, optic neuritis and strabismus are also reported\textsuperscript{16,17}. Diagnosis is usually clinical but could be substantiated with laboratory testing of corneal epithelial scrapings.

The treatment of neonatal HSV is intravenous acyclovir keeping in mind the fatality of disseminated HSV. Therapeutic levels are reached in the aqueous with iv administration. Besides, mothers at high risk of HSV should be administered prophylactic antiviral treatment and delivery in such cases should always be through a caesarian section\textsuperscript{18,19} (Figure 8A).

\textit{Bacterial Keratitis:} Bacterial infections are rarely present at birth and are almost always acquired. The etiology could be the infectious status of maternal birth canal, prolonged duration of exposure of the child in maternal birth canal, integrity of the ocular surface, etc. Of all the many
organisms postulated to cause infection, the most serious infection is caused by Neisseria gonorrhoea. It presents with an incubation period of hours to few days with unilateral or bilateral excessive chemosis, conjunctivitis with copious purulent discharge often with a pseudomembrane. Unless treated it usually progresses to central ulcer, ring abscess, progressive corneal melt and corneal perforation. Emergency management with systemic penicillin is required. Supportive treatment includes topical antibiotics, cycloplegics and vitamin A prophylaxis (Figure 8B).

Bacterial Keratitis of other origin can be effectively diagnosed by corneal smear examination and culture reporting and treated with topical antibiotics accordingly. Topical corticosteroids can be administered with an aim to limit the area of the corneal scar only after the antibiotic sensitivity profile of the microbial agent is known; child is on sensitive topical antibiotics for at least 48 hours and is showing clinical recovery (Figure 8C).

Neurotrophic Keratitis is a degenerative disease characterized by decreased corneal sensitivity and poor corneal healing which decreases reflex tearing and leaves the cornea susceptible to injury. Epithelial breakdown can lead to ulceration, infection, melting, and perforation secondary to poor healing. Congenital corneal anesthesia (CCA) is a rare clinical entity in which the sensory deficit may be confined to the cornea, or extend to other divisions of the trigeminal nerve. The sensory deficit may occur as an isolated abnormality, as part of a complex neurological syndrome, or it may occur in association with multiple somatic abnormalities and congenital insensitivity to pain. This condition usually presents between the ages of 8 to 12 months. Children present with poor vision, photophobia, conjunctival injection, and corneal ulceration in the absence of pain and distress. A simple bedside clinical test to diagnose CCA which we follow is to administer one drop of betadine 2.5% eye drop in the conjunctival sac which would cause irritation to the child with normal corneal sensations and make him uncomfortable.

In most patients, conservative approaches such as copious lubrication, prevention of self-harm and cautious use of bandage contact lenses are effective in preventing progressive corneal damage. Tarsorrhaphy is effective in promoting epithelial healing and permanent lateral tarsorrhaphy may prevent further development of epithelial defects. A corneal graft carries a poor prognosis (Figure 8B).

Figure 9: Metabolic Disease: Composite showing bilateral mucopolysaccharidoses (A,B) and after both eyes penetrating keratoplasty has been done (C,D)

The inheritance pattern for all mucopolysaccharidoses is autosomal recessive for all except Hunter’s syndrome which is X-linked recessive. Severe corneal clouding within a few years of birth is seen only in Hurler (I-H) and Maroteaux-Lamy (VI) syndrome. The general set of clinical findings in a child with corneal clouding suspicious of mucopolysaccharidoses is dwarfism, facial and skeletal deformities, hepatosplenomegaly and sometimes mental retardation and sometimes mental retardation. The detailed description of all these diseases is beyond this article. Mucolipidosis type IV also presents with severe corneal clouding at birth and is complicated by corneal epithelial irregularities and recurrent corneal erosions.

Management includes a detailed systemic evaluation by a pediatrician. Ocular management is done early to prevent amblyopia and is usually in terms of penetrating keratoplasty though deep anterior lamellar keratoplasty has also been reported (Figure 9).

6. Peter’s anomaly: Peters’ anomaly is a rare, congenital, unilateral or commonly bilateral malformation characterized by central corneal opacity of variable size and density associated with a defect in the posterior stroma, Descemet membrane and endothelium in the area of the opacity surrounded by relatively clear peripheral cornea. Also seen are iris strands that arise from the collarette and extend to the periphery of the corneal leukoma. Though Nischal KK et al consider this as an imprecise diagnosis in an era of a UBM, it is still the most commonly used term to explain
such a condition among the ophthalmologists and cornea surgeons. Incomplete formation of the anterior chamber angle is complicated by a high incidence of congenital glaucoma. Peter’s anomaly could be of 2 types:

I. Type 1: Corneal opacity with irido-corneal adhesion
   - Usually unilateral
   - Central stromal opacity with peripheral clear cornea
   - Normal lens and posterior segment—good prognosis

II. Type 2: Type 1 + involvement of iris or lens
   I. Usually bilateral
   II. Dense corneal opacity with irido-lenticular adhesions

III. Oculo-systemic involvement

IV. Poor prognosis

Histologically, there is a central concave defect in the posterior stroma with disorderly arrangement stromal lamellae and deficient Descemet membrane and endothelium. Management should be based on an examination under anesthesia including a UBM examination to know the status of the anterior segment. Peter’s anomaly could be sporadic or hereditary in origin and management plan must include a genetic counseling. Mutations in genes PAX6, PITX2, CYP1B1 and FOXC1 have been noted in Peters’ anomaly (Figure 10).

7. Posterior Keratoconus: Rare, sporadic, non-progressive, unilateral, conical protrusion of the posterior corneal curvature. This represents the mildest variant of Peter’s anomaly. Focal abnormalities of Descemet...
membrane and endothelium could be present. Corneal topography measuring the posterior corneal curvature is of paramount importance. The vision in the affected eye could be reduced due to significant astigmatism or refractive error and early management is required to prevent amblyopia33-35.

8. **Congenital hereditary endothelial dystrophy (CHED):** CHED exists in 2 variations with similar history and clinical features (Figure 11). Children would typically present with diffuse, limbus to limbus corneal clouding, epiphora and photophobia. Slit lamp examination reveals a 2-3 times thick corneal stroma which prevents a clear view of the anterior segment which is usually normal. CHED 2 patients might also present with nystagmus (Figure 12 A,B). Histological examination of the excised cornea reveals a roughened epithelium, 2-3 times thick corneal stroma with a diffuse blue-grey ground glass appearance, multiple layered and thick Descemet membrane (posterior collagenous layer) and an atrophic, irregular or absent endothelium36-38.

The most common misdiagnosis of CHED is congenital glaucoma which could be easily avoided based on a classical history, buphthalmos, increased horizontal corneal diameter, presence of Haab’s striae and a glaucomatous optic nerve head. Though these two conditions have been rarely known to co-exist,3 it is very common to see patients of isolated CHED been operated for congenital glaucoma. Early treatment is advocated to prevent amblyopia. Treatment is only surgical and is either penetrating keratoplasty36-38 (Figure 12 C) or Descemet’s stripping endothelial keratoplasty (DSEK) depending on the patient’s age and the status of the corneal edema39.

9. **Congenital stromal corneal dystrophy (CSCD):** First described by Witschel in 1978, corneal opacity in CSCD is present at birth, stationary, centrally dense and causes amblyopia and nystagmus. The condition is limited to the stroma which shows disorderly arrangement of the corneal stromal fibres. Management is surgical and requires urgent penetrating keratoplasty36.

10. **Corneal Dermoid:** Limbal Dermoids are benign congenital tumours that contain choristomatous tissue (normal tissue in abnormal location). Though rarely present in the entire cornea or conjunctiva, these are most commonly seen at the limbus in the inferotemporal cornea. These may contain tissues originating from all 3 germ layers including hair, nail, skin, fat, sweat or lacrimal glands, muscle, teeth, cartilage, etc.40-42 Malignant degeneration is very rare. Dermoids are categorized based on their location into:

I. **Limbal Dermoid:** usually superficial but may involve the deeper structures. (Figure 13 A).

II. Only involves superficial cornea sparing the limbus (Figure 13 B).

III. Involves the entire anterior segment including iris, ciliary body and lens.

Most limbal dermoids are sporadic and isolated findings, though 30% are associated with Goldenhar syndrome. Other abnormalities associated with dermoid are lid coloboma, aniridia, microophthalmos, cardiac and neurological abnormalities43.

Management of a dermoid is surgical excision but requires a prior UBM to know the extent and depth of the lesion. Limbal dermoids (Figure 14A) are excised and the base is either left bare, covered with an amniotic membrane (Figure 14B) or a lamellar corneal graft (Figure 14C) is
sutured depending on the thickness of the underlying stroma. Central dermoids require penetrating keratoplasty.

The article above represents a brief description of the major causes of congenital corneal opacity. The management is tricky and decisions are made taking into consideration a host of other ocular and systemic factors. Difficulties in the management include the high incidence of amblyopia and the frequent need of examination under anesthesia. The article could serve as a guide to the clinicians in accurate and prompt diagnosis of children with congenital corneal opacities. However, the importance of an urgent referral of these kids to a cornea surgeon at the first diagnosis cannot be underestimated.

References


Lacrimal drainage system begins to develop at the 10mm stage from a solid column of ectoderm which later canalizes. Canalization starts at about 3 months of intrauterine life and gets completed at birth or during first few weeks after birth (Figure 1).

Causes of watering eyes in an infant are:

**Eyelid problems:**
- Blepharitis
- Distichiasis
- Poor lid closure
- Entropion

**Surface Abnormalities:**
- Conjunctivitis
- Foreign body

**Corneal abrasion**
- Keratitis

**Congenital Glaucoma**

**Lacrimal outflow Abnormalities:**
- Punctal agenesis/Stenosis
- Canalicular obstruction
- Congenital dacryocele
- Nasolacrimal duct obstruction

All other causes must be ruled out before making a diagnosis of congenital nasolacrimal duct obstruction.

**Congenital nasolacrimal duct obstruction**

2-4% of infants present with obstructed NLD at birth which may manifest by 2 to 4 weeks of age. 80 – 90% of these resolve spontaneously by 2-4 months of age.

---

**Figure 1:** Lacrimal drainage system

**Figure 2:** Membranous block at valve of Hasner
Causes of NLDO in a child are:

- Membranous block at valve of Hasner (Figure 2)
- Stenosis of opening from narrowed NLD
- Hypertrophied Inferior turbinate

Clinically the child will present with:

- Epiphora
- Discharge
- Matting of eyelids (Figure 3)
- Skin maceration
- Regurgitation may be elicited on pressure
- Acute episodes are rare

On examination, positive regurgitation on pressure over the lacrimal sac confirms the diagnosis. Bacterial culture and sensitivity may rarely be done for cases with purulent discharge not responding to antibiotics.

Management of CNLDO

**Conservative Management:** should be done up to 9 months to 1 year of age. 80-90% of congenital nasolacrimal duct obstructions will resolve with conservative management. It includes:

- **Hydrostatic massage:** it is important to educate the mother about the proper technique of sac massage. The child's head needs to be stabilized and adequate pressure should be applied over the sac area directed inferiorly. The common canalicular opening must be blocked at time of massage to prevent regurgitation. Proper massage done 2-3 times a day (4-5 times each time) is adequate. If done too frequently, there may not be enough secretions in the sac to provide an adequate pressure to help open the obstruction.

- **Topical Antibiotics after massage**

**Probing for congenital NLD Obstruction**

If the obstruction is not relieved by conservative management, a therapeutic probing should be done by 9 months to 1 year of age. However, a proper massage must be attempted for 4-6 weeks before decision to probe is taken.

There are studies advocating early probing (<6 months) in an office setting with topical anaesthesia. However, what is usually recommended and practiced is a late probing (around 1 year of age) under general anaesthesia.

**Technique for probing**

- **Under General anesthesia.**
- **Child should not have URI as this will decrease the success rate of probing**
- **Puncta is dilated (Figure 4a)**
after feeling for the hard stop, the probe is passed inferiorly along the nasolacrimal duct. Patency of the passage should be confirmed (Figure 4c-d).

Post operatively, the child is given topical antibiotic drops/along with mild steroid drops and nasal decongestant nasal drops. Sac massage should be continued post operatively. Probing done in the first year has success rate of upto 95%. The success rate decreases with increasing age and number of probings.

Complications of probing are:
- Bleeding
- False passage
- Inability to pass the probe down the NLD

**Failed probing**

Older children and history of previous probing are factors for probe failure. Moreover, in around 2% of patients, anatomical variations can cause probe resistance.

Causes of probe failure are:
- Improper Technique
- Anatomical Variations
- Mucosal Hypertrophy
- Inferior Turbinate Hypertrophy
- Associated Nasal Pathology

**Management of failed probing**

- Repeat probing should be attempted.
- Inferior turbinate infrastructure when done adjunctively with probing helps to maintain patency of the duct in cases of failed probing. In our experience, this technique works very well in cases of failed probing and majority of these children do not need any additional procedure. However, it is not recommended along with primary probing (Figure 5).

It should be done under general anaesthesia after nasal packing.
Other options in cases of failed probing are:

- Silicone intubation
- Balloon dacryoplasty (Figure 6)
- DCR (> 3 years of age)

Silicone Intubation

Success rates of up to 70-85% have been described for intubation in congenital nasolacral obstructions. However, most of these are described in younger children and in primary/ previously once probed cases.

It can be done in cases where repeat probing has failed/ older children where stenosis or scarring is more. The tubes are left in place for 6 months.

Types of intubations described are:

- Monocanalicular:
  - Mini Manoka
  - Mono-Crawford

- Bicanalicular:
  - Self Retaining Bicanalculus intubation
  - Silicone
  - Crawford
  - Riteling

Problems of Silicone Intubation:

- Operative difficulties in passing the tubes
- Post-operative epiphora / mucoid discharge
- Partial / Total extrusion of the tube

Balloon Dacryoplasty

The procedure has been described for:

Figure 5: wider area is created at the lower opening of NLD by rotating the inferior turbinate

Figure 6: balloon dacryoplasty a) pre-dilatation
b) intraoperative c) balloon post dilatation.

Figure 7: inferior end of nasolacrimal duct after probing seen on nasal endoscopy
As a primary procedure in children at or after 12 months of age
Children in whom lacrimal system probing or silicone intubation has failed
It allows greater dilatation of the lacrimal duct. It is less invasive than silicone intubation and less likely to produce tears/scarring of the lacrimal system. It can be repeated if necessary.

Role of Nasal Endoscopy in probing
Wherever available, nasal endoscopy should be used during probing.

- It permits the direct visualization of the lower end of the Nasolacrimal duct (Figure 7).
- Can rectify the problems with syringing and probing and avoid false passages
- Associated nasal pathologies can be diagnosed

Dacryocystorhinostomy (DCR)
For unresolved nasolacrimal duct obstructions. DCR in a child should not be done before the age of 3 years. External/endonasal route can be selected. However, poorly defined landmarks, changing anatomy and tendency for scarring makes the surgery more challenging in pediatric cases. Intubation though described for pediatric DCRs does not seem to have any additional benefit.

Suggested reading

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Nystagmus is defined as a rhythmic involuntary to-and-fro movement of the eyes. It may be accompanied by unusual head position and head nodding in an attempt to compensate for the condition. Although various types of nystagmus in children are observed, the prevalence of Infantile Nystagmus Syndrome (earlier called congenital nystagmus) is about 1 in every 1000\(^1\). It is therefore likely to be encountered in a comprehensive or pediatric ophthalmology practice. It is important to recognize, correctly diagnose and follow up nystagmus in an infant, since it may be a manifestation of a serious neurological disease. Appropriate treatment for nystagmus can cause significant improvement in confidence and help in overcoming vision-related learning disability in children.

**Clinical presentation**

Nystagmus in children is reported by their parents either as “wriggly eyes” or due to the presence of a head posture. Not infrequently it is associated with subnormal vision. The following may be seen:

1. Head held in a turned position (head posture), to take advantage of the null position
2. Poor vision in patients with ocular abnormality
3. Neurological symptoms like weakness, sensory loss and cranial neuropathies are seen in patients with central nervous system lesions

Nystagmus can be described as:

1. **Types:** Pendular / Jerky (has a fast component and a slow component)
2. **Direction:** According to fast phase
3. **Plane:** Can be horizontal, vertical, rotatory or their combinations

4. **Amplitude:** Can be small (<5\(^\circ\)), moderate (5\(^\circ\)-15\(^\circ\)), large (>15\(^\circ\)).

5. **Frequency:** Total number of complete to & fro movements/sec.: Can be slow(1-2Hz), medium(3-4Hz), fast(≥5Hz).

Intensity of nystagmus is the amplitude x frequency.

**Null zone:** This is the field of gaze in which the intensity of the nystagmus is minimal. In most individuals with nystagmus, the severity of the eye movements can be reduced by positioning their eyes in a particular gaze. At this point the nystagmus is the least with best visual acuity.

**Classifications of childhood nystagmus- forms of nystagmus**

There are various methods of classifying nystagmus.

**Acquired nystagmus\(^4\)**

This form of nystagmus is associated with intra-cranial space occupying lesions (iCSON) and neurological disorders like Arnold Chiari malformation or cerebellar disease (Table 1).

<table>
<thead>
<tr>
<th>Table 1: Cogan’s classification of nystagmus</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Sensory</strong></td>
</tr>
<tr>
<td>Visual acuity</td>
</tr>
<tr>
<td>Head posture</td>
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<tr>
<td>Management</td>
</tr>
</tbody>
</table>
**Congenital nystagmus**

This form of nystagmus may (sensory nystagmus) or may not be associated with poor vision (motor nystagmus). It is found in association with pathologies like optic nerve hypoplasia, retinal diseases and anterior segment diseases (cataract, corneal opacity, aniridia).

Cogan divided nystagmus into two groups- sensory and motor. Sensory nystagmus is related to vision loss and motor related to the defective control of muscle function (Table 2).

The above classification is however no longer considered an adequate classification.

**CEMAS classification**

The Classification of Eye Movement Abnormalities and Strabismus Working Group have recommended new names for nystagmus that begins during infancy. Three categories have been defined:

- **Infantile Nystagmus Syndrome (INS)** - It corresponds to what had previously been termed as motor or sensory forms of congenital nystagmus.

  This type usually develops by 2 to 3 months of age. The trajectory is usually horizontal in direction though it may also have a vertical or torsional vector. It is often associated with other conditions such as albinism, congenital absence of the iris, underdeveloped optic nerves, and congenital cataract.

  The important features of INS are:
  - Binocular and associated (similar in both eyes)
  - Uniplanar
  - No oscillopsia
  - Abolished in sleep
  - Dampped by convergence
  - Increase on fixation effort
  - Distinct waveform: ascending slow-phase

- **Fusional Maldevelopment Nystagmus Syndrome (FMNS)** – It includes the forms of nystagmus earlier called manifest latent and latent nystagmus. This occurs in association with amblyopia and strabismus and is always congenital. This is a jerk nystagmus with either a linear or decreasing velocity slow phase identical to that of gaze-paretic nystagmus. Classically, “pure” or “true” latent nystagmus (LN) occurs only with unicuslar fixation. There is no nystagmus with both eyes viewing, but when one eye is occluded, nystagmus develops in both eyes, with the fast phase toward the uncovered eye. True latent nystagmus is rare. Important features of FMNS include –
  - Congenital, conjugate horizontal jerk nystagmus.

**Table 2: Types of acquired childhood nystagmus and their causes**

<table>
<thead>
<tr>
<th>Causes of acquired nystagmus</th>
<th>Examples</th>
</tr>
</thead>
</table>
| Periodic alternating nystagmus | 1. Arnold-Chiari malformation  
2. Encephalitis  
3. Posterior fossa tumors  
4. Head trauma |
| Upbeat nystagmus | 1. Cerebellar degenerations  
2. Medullary mass lesion |
| Downbeat nystagmus | 1. Cranio cervical anomalies- 
Arnold-Chiari malformation  
2. Brain stem encephalitis  
3. Trauma |
| Seesaw nystagmus | 1. Midbrain mass lesion |
| Pendular nystagmus | 1. Visual deprivation |
| Spasmus nutans | 1. Chiasmal, suprachiasmal, or third ventricle gliomas |

- Increases under condition of monocular fixation.
- When one eye is occluded, a jerk nystagmus develops in both eyes with a fast phase toward the uncovered eye.
- Reverses direction with change in fixation.
- Linear or decreasing velocity exponential slow phase.
- Unknown cause.
- Binocular vision better than uniuscular vision.
- Noted in early childhood in patients of congenital esotropia, DVD.

**Spasmus nutans** - It is a rare constellation of ocular oscillation, head nodding, and torticolis that begins in infancy (usually between 4 and 18 months of age) and disappears in childhood (usually before 3 years of age). The nystagmus is generally bilateral (but it can differ in each eye and may even be strictly monocular), and it oscillates in horizontal, torsional, or vertical directions. It may sometimes be mimicked by tumors of the optic nerve, chiasma, or third ventricle therefore neuro-imaging is necessary to rule them out. This type of nystagmus usually does not require any treatment.

**Workup**

**Patient history** - we should ask for the age of onset, general health problems, and any neurological symptoms in the child. If the onset is acute, then usually the nystagmus is acquired. Children with albinism and oculocutaneous albinism complain of photophobia.

The idiopathic nystagmus syndrome is present in the first 2 months of life while the one associated with ocular disease usually appears after this age.
Family history and examination of relatives - Congenital motor nystagmus has a strong hereditary component. The sex-linked dominant form is the most common pedigree; the sex-linked recessive form is also relatively frequent.

A comprehensive eye examination is needed to identify the cause of poor vision which is often associated with the nystagmus.

Check visual acuity in a child old enough to respond. Assess monocular and binocular visual acuity using charts/Cardiff acuity cards in children below 3yrs. of age. Do fogging of the other eye using either high plus lens or polarizing lens to determine v/a in older kids using picture charts/ Snellen acuity chart. Also try to assess visual acuity with and without face turn.

Anterior and posterior segment examination - This is needed to find out the ocular associations and any cause for low vision. Iris and choroidal hypopigmentation is seen in Ocular/oculocutaneous albinism patients. They also have associated optic nerve and macular hypoplasia.

Some of the direct causes of nystagmus are:

1. Heredity.
2. Poor vision during infancy (eg), congenital cataracts, severe glaucoma, Peters anomaly.
3. Foveal hypoplasia - aniridia, albinism.
4. Optic nerve disorders - hypoplasia, atrophy.
5. Retinal disease-, Leber congenital amaurosis, achromatopsia, macular toxoplasmosis.
6. Retinal detachment - severe retinopathy of prematurity, familial exudative vitreoretinopathy.
7. Cortical visual impairment from perinatal insult or structural CNS abnormality.
8. Inner ear problems, such as infections or irritation.
9. Injury to the head or involving the body’s motor system.
10. Early (usually bilateral) visual deprivation.

Multiple neurological causes are increasingly being recognized as a cause of acquired nystagmus. None of the above causes may be found in a significant subset of patients who are known to have idiopathic congenital nystagmus.

Measurement of head posture: Goniometer (an orthopedic instrument) or a combination of a scale and a protractor can be used to measure the amount of face turn in older kids. This helps to decide the amount of surgical correction required.

Eye movement tracings- this is difficult in small children but when done can be quite helpful. They can be used to verify the type of nystagmus and determine the details of the movements (electonystagmography and videonystagmography). They are especially useful in suspected PAN (periodic alternating nystagmus) cases. The nystagmus amplitude, frequency, intensity and velocity can be measured in various positions of gaze. This helps in localizing the null zone and documenting the changes during the post operative period.

Neuroimaging is important and is indicated in a case of nystagmus in the following situations:

- All cases of acquired nystagmus
- Periodic alternating nystagmus
- Seesaw nystagmus
- Spasms nutans syndrome
- INS with poor vision and disc pallor

Treatment

It is important to remember that congenital nystagmus cannot be cured. However certain interventions can help to dampen the nystagmus and help to improve visual acuity.

Optical correction

This is done to achieve the best corrected visual acuity. A refraction is done to determine any refractive error (nearsightedness, farsightedness, or astigmatism). Correction of significant refractive error is the single most powerful therapeutic intervention for improving vision.

Over minus lenses stimulate accommodation and so can dampen the nystagmus to some extent.

Amblyopia therapy

Penalization/ over plus spectacle lens can be given in better eye (manifest latent nystagmus) for the treatment of amblyopia.

Contact Lenses

Contact Lenses help to improve vision in high refractive errors. They slow down eye movements, and because the optical center of the prescription is always centered on the eye with the contact lens, vision improves. Another theory is that the tactile feedback of feeling the contact lenses on the eyes may lead to better control of the movement and allow better vision. They may be used in older kids.

Botox injections

These have been given either retrobulbar or directly in all 4 horizontal recti. They are rarely used.

Medical Management

Baclofen has been used in the treatment of Periodic Alternating Nystagmus (PAN) with reported good results.
No drug has been found to be effective in Infantile nystagmus syndrome.

**Nystagmus Surgery**

Aims of surgical correction in a patient of nystagmus are:

1. To shift the null position from an eccentric position to primary gaze
2. To diminish the amplitude and frequency of nystagmus
3. To correct associated strabismus if present
4. To induce artificial divergence

Surgery is aimed to improve the waveform, increase foveation time and broadens null Position.

**Timing of surgery**

There is no optimal time for surgery in patients with congenital nystagmus. It is best to wait till 4 or 5 years of age to allow maturation of binocular visual system. This will reduce the likelihood of permanent iatrogenic strabismus and loss of binocularity after surgery. In acquired nystagmus, one should wait for at least one year for evolution of nystagmus waveform and any resulting head posture.

**Surgical options**

**Treat Anomalous Head Positions**

a) *Anderson surgery* This involves recession of yoked rectus muscles

b) *Kestenbaum Surgery*-This includes surgery on all four horizontal rectus muscles (recess-resect procedures in each eye) to move the eyes away from the “null position” of the nystagmus.

At our centre, we do Augmented Anderson procedure where recession of the yoke muscles is preformed (Figure 2). For a face turn to the left, the lateral rectus
muscle of the right eye is recessed and the medial rectus muscle of the left eye is recessed. Its advantage is that a second surgery can be done on the remaining two recti in children with missed PAN/residual head turn.

**Improve vision**

In patients without a null position or null in primary position the aim of surgery is largely directed at dampening the eye movements. Among the commonly used surgical methods are-

- Surgically induced artificial exotropia - Bimedial recession is done to stimulate fusional convergence and thereby dampen the nystagmus. This has the risk of the patient having a manifest exotropia and constant diplopia.
- Anterior tenotomy of horizontal rectus muscles (Hertle-Dell’Osso procedure)\(^\text{13}\). The surgery consists of a simple tenotomy, dissection, and suture of the involved extraocular muscles in place, with neither recession nor resection. The putative mechanism responsible for the damping effects of tenotomy is alteration of a proprioceptive tension-control loop.
- Large recession of all horizontal rectus muscles 14. By retro placing the muscle insertion behind the equator and thus posterior to the tangential point, the leverage is decreased and given amount of muscle innervations will have less rotational effect on globe.

Horizontal medial rectus muscle recession with or without posterior fixation sutures is done in patients of nystagmus blockade syndrome.

**Low Vision aid – one can provide**

1. Print size recommendation- large-print books
2. Reading glasses/bifocals
3. Incorporation of magnifiers
4. Increased lighting

**Implication in school** - Congenital nystagmus has several implications for School going kids. They have to be given the best optical correction including low vision aids. They should be allowed to prefer head posture/correct it surgically to have the best vision. Albinism kids are given tinted glasses. The parents need to be educated that congenital nystagmus is not a progressive condition and child should be sent to school.

**Prognosis**

Congenital nystagmus is usually a benign condition. It is not curable, but its symptoms can be diminished with spectacles or contact lenses. The best corrected vision for most individuals with congenital nystagmus is between 20/40 and 20/70, but correction to 20/20 is possible for some. Nystagmus associated with spasmus nutans (without ICOS) usually resolves spontaneously before the child reaches school age.

The prognosis for an acquired nystagmus depends on its cause.

**Summary**

Childhood nystagmus is still a partially understood entity. It has variable genetic transmission and usually no identifiable cause. CEMAS has broadly classified infantile into three types- infantile nystagmus syndrome, fusional maldevelopment nystagmus syndrome and spasmus nutans. Optical treatment remains the primary and often the only treatment possible in nystagmus patients. Surgery can be done in a selected group of patients.

**References**

RETCAM: Clinical Applications in Retinopathy of Prematurity

Parijat Chandra MD, DNB, Anil Gangwe MD, Vivek Kumar MD, Mayank Bansal MD, Rajvardhan Azad MD, FRCS

Dr. R.P. Centre for Ophthalmic Sciences, All India Institute of Medical Sciences, New Delhi

Retcam is a useful tool that has revolutionized the technique for screening of retinopathy of prematurity (ROP). Retcam is short for Retinal Camera, a wide field digital imaging equipment for examining the pediatric fundus. While indirect ophthalmoscopy needs expertise, the Retcam allows ROP screening to be performed by anyone with suitable training and get reproducible results quickly. In poorly cooperative children, the Retcam may be a good alternative to examination under anesthesia or sedation in selected cases. While it is an excellent tool for peripheral screening in ROP, it has also been used extensively for examination in other diseases like retinoblastoma, and is also useful for anterior segment imaging and gonio-imaging in cases of glaucoma and iris lesions.

The Retcam

There are 2 types of models available in the market – Retcam 3 (Figure 1a) and Retcam Shuttle (Figure 1b). The Retcam 3 features an integrated body with monitor, inbuilt keyboard, fluorescein angiography module, and printer. The Retcam shuttle is more portable as the major components are integrated into a laptop, so it can be easily transported for use in the OT, NICU and screening in peripheral centres.

Retcam is an useful tool to snap digital images of both the anterior and posterior segment of the eye. All images are stored in a local database, which allows easy retrieval and access of images over time, as well as side-by-side comparison. A special attachment allows users to perform fluorescein angiography too.

Multiple lenses with different magnifications are available to be attached to the handheld camera. The most commonly used lens has a field of 130° and can easily visualize the retinal periphery in ROP eyes.

Current ROP screening guidelines

The International ROP screening guidelines² suggest that all babies < 30 weeks gestation age and < 1500 g birth weight should be screened for ROP. However, bigger babies can also be screened if the neonatologist believes the baby is at high risk to develop ROP. The Indian ROP guidelines³ suggest we can screen babies < 34 weeks gestation age and < 1700 g birth weight or higher if the neonatologist believes the child is at high risk to develop ROP. This third criteria is important as it allows heavier, but high risk babies to enter the screening program while following the current guidelines⁴.

It is recommended that all babies eligible for screening should undergo one ROP screening within the first 30 days.

Figure 1a: Retcam 3 Figure 1b: Retcam Shuttle
of life. In very low birth weight babies, it is suggested these babies may be screened as early as 2-3 weeks of life.

**Retcam Procedure**

Before start of the Retcam examination, it is essential that the pupils are dilated well. Pupils are dilated with 2.5% phenylepherine and 0.5% tropicamide eye drops, used 10 min apart, half an hour before the procedure. While pupillary dilation is very essential to get a good view of the fundus, sometimes it may be difficult in cases of plus disease where pupils do not dilate easily. Poorly dilating pupils may mask severe disease behind and the observer needs to be cautious in these cases.

Topical proparacaine eye drops are used for topical anesthesia. After a pediatric lid speculum is inserted, a coupling agent like 2% HPMC is used so that the hand held camera can be placed over the cornea. The operator adjusts the light intensity and changes focus to get a sharp image using foot switch controls. The settings can be changed to save still images or capture video recordings from which still images can be saved.

While recording the images, it is customary to first photograph the anterior segment, followed by the posterior pole, and then the retinal periphery in a clockwise manner. A topical antibiotic eye drop is inserted at the end of the procedure.

While beginners might find the procedure a little difficult initially, especially while holding the camera over the eye - after the learning curve, most trained personnel can save images with high accuracy and reproducibility. Due to the small eye size of preterm babies, sometimes it is cumbersome to place the heavy handheld camera on the small cornea. The eye speculum also adds to the difficulty of examination in very small eyes.

**Benefits of the Retcam**

Retcam offers a professional way to keep photographic records of ROP screening. The operator can easily view images of both eyes across the entire period of follow-up, compare images and decide if the disease is progressing or regressing.

Since it is a mobile self contained system, it can be shifted to any area where ROP screening is to be performed like the nursery, ICU, or operating room. After suitable training, the procedure can be easily performed by technicians or nurses, which can later be reviewed by an ROP expert.

Indirect ophthalmoscopy requires good skill, and more expertise is needed when coupled with scleral indentation. The Retcam makes the task much easier for the examiner, while making the procedure much faster and with less complications. It has demonstrated better cardio-respiratory stability during examination in children.

With the increase in medico-legal cases, it is essential to document fundus images very well. The Retcam helps to prevent inter-observer variability and is useful for telemedicine purposes and consultation with experts. It is much easier to teach residents and explain the disease process to parents with the retinal images.

**Fluorescein angiography**

Retcam assisted fluorescein angiography can be performed easily in the Retcam 3 (Figure 2a,b). Blue light is emitted by switching on the FA unit, and a yellow filter is inserted inside the camera hand piece. Sodium Fluorescein 20%
dye is injected intravenously (0.04ml/kg) via a preplaced intravenous cannula and the Retcam findings recorded. Excellent fluorescein angiography images can be recorded, which provide useful information for beginners.

The main advantage of fluorescein angiography is that it allows clear visualization of avascular retina and flat neovascularization which is not visible to the naked eye. It helps to detect missed areas of disease and skip areas of treatment, and is especially useful for visualizing the disease process in aggressive posterior ROP. While its indications are selective, it does provide useful information for the new ROP trainee.

**Telemedicine Role**

As digital imaging gets more common, tele-screening is going to get more popular. Retcam allows screening to be performed in peripheral centres by technicians, the images are then transmitted to experts based at tertiary eye centres, where they can review the images and advise treatment or follow up. This allows a large coverage area of the ROP screening program in the community.

Several reports in literature have compared Indirect Ophthalmoscopy with digital screening, and have found high sensitivity and high specificity in detecting referral warranted ROP. Stanford University Network for Diagnosis of Retinopathy of Prematurity (SUNDROP) telemedicine initiative at 5 year results found Tele-medicine had 100% sensitivity, 99.8% specificity, 93.8% positive predictive value, and 100% negative predictive value for detection of treatment warranted ROP.

The Karnataka Internet Assisted Diagnosis of Retinopathy of Prematurity (KIDROP) program is a tele-screening program active across Karnataka, which follows the tripleT philosophy - Tele-ROP, Training of peripheral ophthalmologists and ophthalmic assistants, and Talking to neonatologists, pediatricians and gynecologists. It is a successful model for ROP screening in rural and semi-urban infants using tele-ophthalmology. With the advent of several dedicated mobile apps for ROP tele-screening, the images can be sent via the Retcam through the Internet and can be accessed by experts using mobile apps on smartphones.

Since Retcam is a very expensive equipment, it has limited its widespread use in telemedicine. Worldwide research is on to develop cheaper technology for tele-screening in the community. Low-cost technology for digital imaging in ROP will make tele-screening more universal and will go a long way in reducing ROP related childhood blindness in the community.

**References**

Orbital Exenteration for Uncontrolled Facial Fungal Cellulitis: A Dilemma and Review of Literature

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Rhino-Cerebral-Orbital Mucormycosis (RCOM), a fungal cellulitis, is a rare infective condition affecting immunocompromised persons seen especially in patients with uncontrolled diabetes¹ ². Such infections may occur following trauma or surgical procedure. RCOM may be poorly responsive to medical treatment and carries high mortality¹ ³. We report a series of 3 cases of RCOM with varied clinical presentations and challenges encountered in managing them.

Case 1
A 65 year old male presented with swelling of left eye and nasal stuffiness (Figure 1). There was history left maxillary tooth extraction 2 weeks before. He was admitted with the provisional diagnosis of sinusitis with orbital cellulitis. His blood investigation revealed elevated blood glucose levels (> 400 mg/dl) and ketoacidosis. He was started on insulin and empirical intravenous antibiotics. CT scan of the paranasal sinuses and orbits revealed thickening of epithelial lining, proptosis and mild enlargement of extraocular muscles. No definitive orbital infiltration or abscess was localized (Figure 2). Over the next 3 days, the patient worsened clinically with increase in proptosis and hard palate lesions. The hard palate had ulcerative lesions white in color to start with which over 2-3 days turned brown-black with irregular margin (Figure 3). Microbiological scraping of the lesion from hard palate demonstrated Aspergillus spp fungus. He was started on Amphotericin B (1mg/kg/day) and endoscopic drainage of left maxillary sinus was performed. No surgical intervention was done for left orbit as there was no localized abscess. Patient clinically worsened over next 3 days and repeat CT scan

Figure 1: Left side proptosis with endophthalmitis

Figure 2: CT scan with enlargement of extraocular muscle
revealed left cavernous sinus thrombosis (Figure 4). He refused further medical and surgical intervention and left hospital against medical advice.

**Case 2**

A 55 year old male presented with diplopia. He was a known case of chronic Hepatitis B infection and chronic renal failure. The patient was admitted and stabilized. Over next two days he developed proptosis of right eye with black eschar like lesions on the paranasal skin. The lesion was 2 cm X 3 cm horizontal oval to start with, slightly raised and non-tender (Figure 5). Gram staining of the tissue scrapping from lesion showed presence of fungal elements which was later confirmed as mucor by histopathology. The hard palate had ulcerative lesions with irregular margins which enlarged over 2-3 days and color changed from white to brown (Figure 6). CT scan revealed extensive maxillary and ethmoidal sinusitis and right orbital cellulitis. He was started on intravenous Amphotericin B but he continued to deteriorate and relatives refused for surgical intervention in view of high of morbidity and mortality. Patient and relatives refused further treatment.

He was started on intravenous amphotericin B along with local wound dressings. He could not afford liposomal amphotericin B. At this juncture, patient showed signs of rapid spread of infection involving para-nasal sinus and left orbit. MRI of the face, sinuses and orbit revealed extensive spread of infection involving the sinuses and left orbit. A dilemma to perform orbital exenteration was encountered. A team of maxillofacial surgeons, ophthalmologists and neurosurgeon was formed and left orbit exenteration was performed along with extensive debridement of the necrotic tissues extending upto pterygopalatine fossa. The devitalized tissues in the left zygomatic-temporal area were also excised.

After extensive surgical debridement and meticulous post operative care in the intensive care unit for nearly 45 days the patient could be salvaged and the infection healed successfully (Figure 7). The patient is now 8 months post surgery and is currently undergoing evaluation for facial prosthesis.

**Case 3**

A 60 year old man presented to emergency room with history of trauma to face and head following collapse of brick wall. He was admitted with multiple facial bones fracture of left side of face with extensive tissue necrosis and lacerations. His blood investigation revealed high blood glucose and keto-acidosis, which was controlled with insulin. Over the next 3 days the skin, subcutaneous tissue developed infection with necrosis despite being on antibiotics. The pus and the necrotic material sent for KOH mount showed septet filamentous fungus. Repeated debridement was done in order to debulk the infective focus. Fungal culture revealed a rare fungus - Rhizactonia solani, that rarely causes human mycosis.

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Discussion

Fungal cellulitis involving the face, paranasal sinus and orbit are very difficult to treat and may cause mortality if not treated appropriately. Presence of immunocompromised status due to uncontrolled diabetes, steroid abuse, drug abuse or infection due to HIV may be another cause leading to unrelenting infection and eventually causing death. In diabetic patients apart from treating the infection, managing the systemic metabolic parameters is also very important.

Review of medical literature regarding management of fungal facial cellulitis revealed that it is a rare entity and treatment guidelines are ill-defined. High degree of suspicion in appropriate settings and clinical presentation is most important for early diagnosis and successful outcome. Bodenstein NP et al in their review mentioned appearance of black eschar on palate, nose or orbit as a definitive clue. Non-tender periorbital edema may be an early clue to orbital involvement. The initial symptoms of rhino-cerebral mucormycosis may be eye or facial pain and facial numbness, followed by onset of conjunctival edema, blurred vision and soft tissue swelling. In case 2, extensive Herpes Zoster was also considered a differential diagnosis before microbiology report confirmed fungal elements.

Dhiwakar M et al in series of nine cases described perinasal cellulitis or paraesthesia as the most frequent early clinical sign. Periorbital edema, mucopurulent rhinorrhea and nasal crusting were reported as other early manifestations. They reported that CT scan may be near normal and high degree of suspicion must be kept in immunocompromised patients. Kotzmanoglu K et al in their case report highlighted the need to differentiate fungal orbital cellulitis from bacterial cellulitis as the management is different and misdiagnosis can lead to severe complications. J.P. Davis and M.P. Stearns in their series of four cases, highlighted the importance of early CT scan in diagnosing sinusitis along orbital cellulitis, which improved the outcome of management.

In case 1, there was no definitive orbital involvement and that prevented the treating ophthalmologist to do enucleation or exenteration. Talmi YP et al have reported in their study of 19 cases that the most common finding on CT scan is mild mucosal thickening of the paranasal sinuses or thickening of the extraocular muscles of the eye. Organized retro-orbital mass or abscess may be rarely present. However since the vision of the eye was lost, enucleation could have been considered for the benefit of salvaging life.
Petrikko G et al in their review of mucormycosis reported that rhino-cerebral site is the most common manifestation of invasive mucor fungal cellulitis. The reported mortality rate was 44% in diabetics, 66% in patients with malignancies and 35% in patients in which no predisposing condition could be identified.16

Spellberg B et al in their review of mucormycosis have elucidated the pathology of infection, its progression and management. He reiterated the importance of sinus mucosal thickening or thickening of the extraocular muscles on CT imaging. Presence of bony erosion has been noted to be suggestive of infection in appropriate scenario. The review also highlighted the rarity of a localised orbital abscess in fungal RCOM, as compared to more common bacterial orbital cellulitis. Thus early institution of treatment with antifungal may be done while the diagnosis is being confirmed.

Fungal culture may take 3-7 days before the reports are available. Serology based tests or PCR are unreliable. Microbiologic demonstration of fungi is coupled with the problem of mucor being a commensal and common laboratory contaminant. Such a delay in laboratory confirmation of fungus and lack of consensus among surgeons to go ahead with destructive procedure like enucleation/exenteration on the basis of clinical suspicion alone may cause delay in management.

Among various antifungal agents, Amphotericin is the drug of choice. The recommended dose of amphotericin B deoxycholate has been 1 to 1.5 mg/kg/day.17-21 Surgical debridement along with antifungal regimen is the most appropriate modality. Antifungal alone have poor outcome as the drug penetration is poor in necrotic tissues due to poor vascular supply.22-24 Local application of antifungal agents also does not seem to work. In such a scenario extensive debridement of the deeper tissues appear to be the only option to prevent further spread of infection intracranially.22-25

Extensive fungal cellulitis may require orbital exenteration to prevent intracranial spread. Exenteration of the orbit along with debridement of facial necrotic tissue and excision of paranasal sinuses has been reported as a therapeutic modality to remove the infective load.26-28 A few case reports do mention the role of adjuvant therapy in order to delay orbital exenteration with success. However there are no said guidelines in such cases given the rarity of the disease process and unavailability of adjuvant treatment options.

Our patients were diagnosed to have fungal infection and were started on intravenous amphotericin B. Liposomal amphotericin could not be used due to its high cost.29 The patient in case 3 underwent repeated surgical debridement of the superficial necrotic tissue however it could not contain the spread of infection which continued via deeper tissues. The infection appears to follow deeper tissue of paranasal sinuses and orbit inspite of superficial dressings or debridement. Failing superficial debridement, orbital exenteration along with extensive debridement of the necrotic tissues helped to contain the infection and patient could survive.10

Case 3 is unique because such facial fungal cellulitis following trauma in a diabetic patient is rare. Patient survived following extensive surgical debridement and orbital exenteration. Adjuvant therapy in the form of amphotericin irrigation was of limited help. After reviewing available literature and from our own experience of 3 cases we propose the following guidelines for facial fungal cellulitis:

- Fungal infection should always be suspected especially in immunocompromised individuals.
- Delay in diagnosis should not be done for awaiting culture reports (3-10 days). Microbiology reports of Gram stain, KOH wet mount or use of special stains may be done to identify fungal elements.
- Control of systemic parameters is vital for patient survival.
- A contrast CT/MRI of the face, paranasal sinuses, orbit and brain should always be done whenever deeper infection/inflammation is suspected. MRI helps to identify inflammatory tissue invading the normal deeper soft tissue.
- Conservative treatment may be of limited help especially when adjuvant therapy is costly and scarce.
- Functional endoscopic sinus surgery may be done to collect infective material for microbiological culture and widening of ostia of paranasal sinuses to provide drainage of collections.
- Judicious and adequate debridement of dead superficial tissues and meticulous wound care is necessary.
- Orbital exenteration or removal of paranasal sinus may be required to debulk the tissues to contain the infection, prevent intracranial spread and decrease mortality.

References


Evolution of Pediatric Cataract Surgery

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Pediatric cataract is one of the leading causes of treatable childhood blindness accounting for 7-20% of childhood blindness in the world¹. Early detection and timely management of the same can lead to dramatic reduction in prevalence of childhood blindness and amblyopia.

Cataract surgery in pediatric age group has undergone remarkable changes over the years in terms of a better surgical technique of cataract extraction, reduced incidence of intraoperative and postoperative complications and an early visual rehabilitation of the child. The improvement in management of pediatric cataract has largely been due to a better pre-operative evaluation as well as advances in technology and microsurgical techniques.

The Early Years
Cataract surgery techniques in children have undergone major changes in the last 40 years. In the past, several techniques have been used such as discission and needling, linear extraction, or a combination of discission and displacement of lens fragments into the anterior chamber by irrigation without IOL implantation. These earlier techniques have paved way for the modern phacoemulsification with IOL implantation in children including infants.

Discission and needling
One of the earliest techniques which had been described for the removal of lens in children is the needling technique in which with wide dilation of the pupil, a Ziegler or Knapp knife-needle is entered under conjunctiva at the limbus into the anterior chamber and the anterior capsule is opened by two or more cuts. The cortical material is then stirred up taking care that the posterior capsule is not touched². Absorption is allowed for a minimum of eight weeks after which time additional needlings may be done at intervals no closer than twelve weeks apart if unabsorbed cortex is still present. When the cortex is gone, then if a secondary membrane remains, it may be opened with a sharp Wheeler discission.

Linear Extraction
Another technique of surgical removal of lens which came into existence was linear extraction in which a keratome incision is made 1.5 to 2.0 mm inside the limbus about 5mm in size followed by a large capsulotomy with the Fuchs capsule forceps. The lens substance is then expressed and the anterior chamber wash is given. Air is then injected and anterior chamber formed³.

Some surgeons preferred a combination of a discission and a linear extraction. In this procedure, after an extensive discission of the anterior capsule, the eye is kept under observation for several days, until the lens material is sufficiently disintegrated to permit easy removal by linear extraction.

Owens and Hughes found that linear extraction, or discission followed by linear extraction, produced better results than simple discission in that the number of secondary operations necessary to clear the visual axis was higher after simple discission than after linear extraction or after discission with subsequent extraction⁴. It was observed that late retinal detachment and spontaneous intraocular hemorrhage occurred much more frequently after needling, especially repeated needling, than after linear extraction.

Aspiration
In the early 1960s, the aspiration procedure, popularized by Scheie⁵ became the accepted technique for extracting...
cataracts in infants and children. It involved anterior chamber entry through a small opening through the corneoscleral wall beneath a conjunctival flap followed by a wide cruciate incision in the anterior capsule of lens. Gentle to and fro saline injection of saline was used to remove the lens material.

Aspiration procedure was preferred to linear extraction and needling procedure for congenital cataracts since operative complications such as vitreous loss and difficulties with wound closure, which are common with linear extraction, were eliminated.

Complications seen with aspiration technique included intraoperative collapse of the anterior chamber, high incidence of secondary membranes causing amblyopia and the development of synechiae between the iris and the remaining capsular bag.

In the mid-1960s, the introduction of a double-barreled cannula, one for aspiration and one for irrigation changed the face of pediatric surgery. The irrigation–aspiration technique enabled the surgeon to maintain anterior chamber depth during cataract aspiration while keeping the posterior capsule intact. Unfortunately there was still a high risk of after cataract formation necessitating a second procedure for opening of the posterior capsule.

**Phacoemulsification**

The 1970s witnessed the introduction of phacoemulsification in pediatric cataract surgery. The closed phacoemulsification system incorporates the principles of controlled infusion to maintain intraocular pressure and variable and controlled suction or aspiration. Chances of vitreous loss, anterior chamber collapse and wound closure problems were significantly reduced with this technique. Initially, the posterior capsule was left intact which led to high incidence of secondary membranes. Use of a vitreous suction cutter to perform a posterior capsulotomy and vitrectomy while removing a congenital cataract was advocated by Parks in 1983. It represented one of the major advances in pediatric cataract surgery. Removal of all but 2 mm of peripheral posterior capsule with a generous vitrectomy was recommended. Although it eliminated the risk of amblyopia to a great extent by decreasing the formation of secondary membranes, it made in the bag fixation of IOL or secondary sulcus fixation of IOL difficult. Hence, today most surgeons have modified the Parks procedure and leave sufficient residual posterior capsule for the fixation of IOL.

**Current technique**

**Anterior chamber entry**

A paracentesis incision is created in the clear cornea 30° on either side of the main incision. The small tunnel paracentesis incisions of 0.9–1.2 mm width are adequate to allow insertion of irrigation–aspiration cannulas and vitrectomy probe. A 2.6–3 mm wide limbal valvular incision with 1–1.5 mm internal entry is preferred. The sclera in a young child is elastic, encouraging the use of the smallest possible incision, which also helps to prevent iris prolapse.

**Anterior Capsulotomy**

Manual CCC may be achieved using a bent needle, cystitome, forceps, or a combination of these. A forceps is often necessary for control of the elastic capsule encountered in children using several repeated grasps at the leading edge of the tear. The capsulorrhexis should be kept relatively small because the elasticity of the child’s lens capsule can create a capsular opening that is larger than expected or desired. A central capsulorrhexis of about 4.5–5 mm is usually adequate so that it covers the IOL optic in all directions.

Alternative techniques currently available include vitrectorhexis, radio-frequency diathermy and Fugo plasma blade. Vitrectorhexis has proved to be a good alternative to manual CCC for young children, especially in the first 2 years of life when the capsule is very elastic and difficult to control. High frequency endodiathermy has also been used to create a continuous curvilinear capsulorrhexis but it produces a coagulated capsular debris along the edge. The Fugo blade helps to create a perfectly controlled anterior capsulotomy of any size, without the risk of a radial tear. A thorough hydrodissection is done and cortical matter is separated from the capsule.

**Irrigation- Aspiration**

Separate irrigation–aspiration minimizes the anterior chamber fluctuations and aids in thorough removal of cortex. Hard nuclei can be removed using short bursts of phacoemulsification.

**Posterior Capsulotomy and Anterior Vitrectomy**

Primary posterior capsulotomy is the preferred choice in children up to 6–8 years of age. A 26-gauge cystitome is used to make the initial cut in the posterior capsule. The flap is lifted and then held with capsulorrhexis forceps and a PCCC is accomplished aiming at a size of 3.5–4 mm by using the ACCC principles and strategies.

Inflammatory response in small children is severe and fibrous membranes may form on an intact vitreous face which can act as a scaffold for lens epithelial cell (LEC) migration and proliferation. Hence anterior vitrectomy along with posterior capsulotomy is advocated in infants and young children up to 2 years of age. A central anterior vitrectomy up to the depth of 2 mm is adequate. Posterior chamber Intraocular lens is then placed in the bag or sulcus. Viscoelastic is removed from the anterior chamber and the main incision is closed with sutures.
Evolution of Pediatric Cataract Surgery

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Visual rehabilitation in the pediatric age group

Aphakic Glasses

Aphakic glasses are the safest and their power can be readily changed to compensate for ocular growth but they have poor cosmesis and inferior optics causing spherical aberration, prismatic effect and marked retinal image size disparity.

Contact Lenses

Contact lenses have proved to be a good option in correction of pediatric aphakia. They can be changed with the changing refraction of the child unlike intraocular lenses. However, contact lenses are successful in a relatively small number of pediatric cases over a long period, are emotionally stressful both for the child and the family, and are economically beyond the reach of many patients, particularly those in developing countries.

Epikeratophakia

Epikeratophakia was introduced in the early 1980s as an alternative means of correcting aphakic eyes. A lamellar corneal disc was sutured in front of the recipient’s eye after removal of the epithelium thus correcting the refractive error by changing the anterior surface of the cornea. Disadvantages included intensive postoperative management, decreased corneal lenticular clarity with time, irregular astigmatism, spherical error, and a prolonged period until visual rehabilitation is achieved.

Such difficulties with aphakic spectacles, contact lenses and epikeratophakia, combined with more experience with intraocular lenses, viscoelastics and improved surgical techniques, have increased the popularity and diminished the controversy over IOL implantation. IOL implantation offers the method of optical correction that requires the least compliance and induces minimal aniseikonia and astigmatism.

Intraocular lenses

The first intraocular lenses to be used in children were anterior chamber and iris supported lenses. The first published implantation of an IOL in a child was by Choyce in 1955, using an anterior chamber lens. Disadvantages included an intense inflammatory reaction, secondary glaucoma and uveal tissue bleed due to contact with vascular tissues. Binkhorst and Gobin implanted an iridocapsular fixed IOL in 1959. They were also associated with complications, including iris sphincter erosion, hyphema, anterior synechiae, iris bombé, iritis, and pupillary fibrotic membranes. Also, lens dislocation, pseudophakodonesis, and corneal endothelial trauma were possible when capsule fixation was not achieved.

With the advent of technique of continuous curvilinear capsulorrhexis developed in the 1980s, in the bag placement of IOL’s have been made possible. PMMA lenses have given way to foldable acrylic lenses in pediatric age group owing to lower incidence of posterior capsular opacification, need for a smaller incision for insertion of the IOL and minimal inflammatory response.

The youngest age at which implants can be safely and effectively used has not yet been clearly established. IOL is routinely implanted in the bag in children more than 2 years of age. Experts in the field of pediatric cataract extraction have recently been implanting posterior chamber IOLs in infants as early as 2 months old since IOL implantation has been considered to be the best option for the prevention of amblyopia with immediate visual rehabilitation. Recent studies have demonstrated excellent results with bilateral primary IOL implantation in children between the ages of 4–6 months of age.

Conclusion

Pediatric cataract surgery has seen a dramatic change over the years from the days of needling and aphakia to present day phacoemulsification with primary in the bag IOL implantation in infancy leading to early visual rehabilitation of the child. With continued improvements in surgical and laser techniques, IOL designs, anti-inflammatory agents, and amblyopia therapy, the refractive and visual outcomes in pediatric cataract surgery should continue to improve, whereas the need for secondary procedures should diminish.

References


The term leukocoria means “white pupil” (from the Greek “leukos” meaning white and “kore” meaning pupil) and is the name given to the clinical finding of a white pupillary reflex (Figure 1). Leukocoria can be caused by abnormalities in the lens (e.g. cataract), vitreous (e.g. Persistent hyperplastic primary vitreous, haemorrhage), or retina (e.g. retinoblastoma). It can be the initial manifestation of a wide spectrum of intraocular and systemic disease processes. The differential diagnosis can be narrowed through a complete clinical and family history and a thorough ophthalmic examination.

Although transient leukocoria is occasionally caused by the reflection of a normal optic disc, all children with newly discovered leukocoria should be referred promptly to an ophthalmologist to exclude retinoblastoma and other life-or sight-threatening conditions.

The evaluation of the child with leukocoria and a brief discussion of the common causes of leukocoria in children are presented here.

**Causes of Leukocoria**

The common causes of leukocoria in children include:1,2

- Retinoblastoma (RB)
- Persistent hyperplastic primary vitreous (PHPV)
- Coats disease
- Toxocarasis
- Cataract
- Vascular causes
  - Retinopathy of prematurity
  - Incontinentia pigmenti
- Congenital/ developmental anomalies
  - Large coloboma (fissure or cleft) of choroid or optic disc
  - Retinal dysplasia
  - Juvenile retinoschisis
  - Norrie’s disease
  - Combined hamartoma of retina and RPE
- Other tumours
  - Medulloepithelioma
  - Retinal astrocytoma

**Workup of a patient with leukocoria**

Algorithm for differential diagnosis of a patient with leukocoria is shown in flowchart.

**History**

- Age of presentation
  - Birth (PHPV)
1-3 years (RB)
- Preschool and school going children (Coats, Toxocara)

- Sex
  - Male (Coat’s, Norrie’s disease, Juvenile retinoschisis)
  - Female (Incontinentia pigmenti)

- Birth history
  - Low birth Weight (ROP)
  - Trauma (Congenital cataract, retinal detachment)

- Family history
  - None (PHPV, Coat’s, Toxocara)
  - AD (RB)
  - Sex linked recessive (Norrie’s, juvenile retinoschisis)
  - Sex linked Dominant (Incontinentia pigmenti)

- Antenatal history
  - Gestational age (ROP)
  - Maternal health (TORCH syndrome)

**Examination**

Complete ocular examination including examination under anaesthesia (EUA) in young and uncooperative children should be done. In addition to examination of the ocular adnexa and anterior segment, both fundi must be visualized for 360 degrees to detect tumours or other pathology that may be located in the peripheral retina. EUA is often required and may facilitate the performance of computed tomography (CT) or magnetic resonance scan, ultrasonography, fundus photography, laboratory and serologic testing and lumbar puncture. Look for under mentioned features specifically-

- Measure corneal diameters and axial length (look for small eye)
- Iris neovascularisation
- Pupils- Look for any RAPD
- Lens- look for type, laterality, extent and location of cataract
- Look for any vitreous seeding/ persistent foetal vasculature

**Dilated fundus examination-** A dilated fundus examination using the indirect ophthalmoscope is essential in the evaluation of children with leukocoria.
The examination should assess:

- Status of the retina (e.g., retinal detachment).
- Presence of retinal vascular abnormalities and/or exudate (as may occur in Coats disease).
- Size, location, and number of tumours, if present.

Any or all of the following may be helpful in diagnosis and planning treatment.

- **B scan ultrasonography**—especially if there is no view of fundus. Look for any tumour/vitreous seeding/retinal detachment/calciﬁcation. Retinoblastoma appears as acoustically solid tumour with high internal reﬂectivity and intraretinal calcification (Figure 2).

- **CT Scan**—look for calciﬁcation (RB) and optic nerve, orbital and CNS involvement (Figure 3).²

- **MRI**—it can detect optic nerve involvement (RB), intracranial extension and pinealoblastoma (RB) although it doesn’t show calciﬁcation (Figure 4).²

- **LDH activity**—if the LDH activity is raised in aqueous relative to serum level, it is suggestive of retinoblastoma.⁷

**Retinoblastoma**

It is most common primary malignant intraocular tumour of childhood. It arises from retina and appears as a white, nodular mass that breaks through the internal limiting membrane into vitreous (endophytic), as a yellowish subretinal mass lesion often underlying a serous retinal detachment (exophytic) or as a diffusely spreading lesion (Diffuse infiltrating) (Figure 5). Iris neovascularisation is common. Pseudohypopyon and vitreous seeding may occur. Cataract is uncommon and eye is normal in size.⁸ It may be unilateral/bilateral, unifocal/multifocal. Average time of diagnosis is 18 months (12 months for bilateral and 24 months for unilateral cases). A family history is elicited in about 10% of cases and is autosomal dominant in inheritance.

**Persistent hyperplastic primary vitreous**

It is developmental ocular abnormality consisting of a varied degree of glial and vascular proliferation in vitreous cavity.⁹ There is failure of structures in primary vitreous to regress. It is usually associated with microphthalmos and is unilateral. In anterior form typically there is a membrane behind the lens that may cause traction of ciliary processes which may be elongated. It is a progressive condition with

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**Figure 2:** B scan showing acoustically solid tumour with high internal reﬂectivity and supranormal spikes due to calciﬁcation

**Figure 3:** CT scan showing intraocular calciﬁcation in Retinoblastoma

**Figure 4:** MRI showing optic nerve involvement in Retinoblastoma
cataract present at birth or early in life. Membrane and lens may rotate anteriorly and result in secondary glaucoma. Posterior form of PHPV includes a persistent hyaloid artery with a large stalk issuing from the optic disc. Tractional retinal detachment may be seen in advanced cases of posterior form. There is no family history.

**Congenital cataract**

The main causes of infantile cataract are genetic, metabolic, prematurity and intrauterine infections. Other causes of childhood cataract include trauma, drug-induced cataract, radiation therapy and cryo-application or laser therapy for retinopathy of prematurity. Trauma is one of the commonest causes of unilateral cataract in the developing countries. Bilateral cataracts occur commonly due to the long-term use of topical or systemic steroid therapy. In industrialized countries, in approximately 50% of bilateral cases and virtually all of the unilateral cases, the underlying cause cannot be determined (Figure 6).

**Coats disease**

Coats disease is defined by the presence of vascular dilatations (retinal telangiectasia), including ectatic arterioles, microaneurysms, venous dilations (phlebectasias), and fusiform capillary dilatations, frequently associated with exudative retinal detachment. Despite the presence of retinal capillary nonperfusion shown by angiography, posterior segment neovascularization is distinctly unusual. The abnormal vessels are incompetent, resulting in the leakage of serum and other blood components, which accumulate in and under the retina. Any portion of the peripheral and macular capillary system may be involved. Variation in the clinical findings is wide, ranging from mild retinal vascular abnormalities and minimal exudation to extensive areas of retinal telangiectasia associated with massive leakage and exudative retinal detachment, as may be seen in children presenting with leukocoria.

This retinal condition is not hereditary and is not associated with systemic vascular abnormalities, even though a gene has been located on chromosome 4. Entities such as retinitis pigmentosa and others may occasionally be associated with retinal telangiectasia. Usually unilateral and there is a marked male predominance (85%). Gradual progression with increasing exudation occurs over time. The severity and rate of progression appear greater in patients under the age of 4 years, in whom massive exudative retinal detachment with retina opposed to the lens may simulate retinoblastoma. Therefore, Coats disease is included in the differential diagnosis of leukocoria.

Patients with peripheral areas of leaky vascular anomalies typically present with lipid deposition in an otherwise angiographically normal macula, as hard exudate tends to accumulate in the macula. Similar findings seen in adults probably represent late decompensation of pre-existing vascular anomalies. Occasionally, a submacular lipogranuloma or subretinal fibrosis is the initial finding.

For milder cases of lipid exudation, diabetic retinopathy, BRVO, juxtafoveal retinal telangiectasia and radiation retinopathy may be considered.

Treatment of Coats disease generally consists of photocoagulation, cryotherapy, and, in severe cases, retinal reattachment surgery. Photocoagulation and cryotherapy are effective in obliterating the vascular anomalies and in halting progression. Multiple treatments may be necessary and long-term follow-up is important to detect recurrences.

**Retinopathy of prematurity**

Innovations and advances in neonatal care continue to improve survival and outcomes for infants at
increasingly earlier gestational ages. ROP is a proliferative neovascularisation which occurs due to incomplete pre-delivery vascularisation of the retina. Neovascularisation can extend into the vitreous causing tractional retinal detachment and subsequent leukokoria. Elucidating an obstetric history helps evaluate this cause of leukokoria, for ROP occurs with increasing frequency at decreasing gestational age.

**Toxocariasis**

Toxocariasis or visceral larva migrans is a rare infection caused by roundworms from either dogs or cats. The inflammatory response to these parasites often localises to the eye, causing uveitis, endophthalmitis or chorioretinitis. The chorioretinitis causes fairly characteristic subretinal granulomas, whose whitish appearance results in leukokoria.

**Norrie’s disease**

Norrie’s disease, a congenital progressive oculo-acustico-cerebral degenerative condition is a rare X-linked recessive disorder. Norrie’s disease must be considered in male infants with bilateral retro-lental masses. All the affected patients of Norrie’s disease are blind since birth. Mental subnormality occurs in about one third of cases and 25-30% develop a sensory neural deafness. Retinal dysplasia characterised by severe hypoplasia of the inner retinal layers and hyperplasia of the retinal pigment epithelium has been described as the characteristic histological features of Norrie’s disease. Iris atrophy and shallow anterior chamber are typical of Norrie’s disease.

The clinical diagnosis of sporadic Norrie’s disease is possible. The better understanding of extra-ocular signs of Norrie’s disease has helped in establishing the diagnosis of the disease, even in the absence of family history. Degenerative changes in the cerebrum and in the acoustic nerves are responsible for mental retardation and neurosensory loss.

Early lensectomy, vitrectomy and retinal repair have been advocated before total retinal detachment and contraction occurs. Prognosis is poor and phthisis bulbi usually occurs inspite of early treatment.

**Coloboma**

Congenital coloboma is embryological developmental defects. Both retinal coloboma (typically seen in the inferonasal retina) and optic nerve coloboma can cause leukokoria. Other optic disc abnormalities such as a ‘morning glory disc’ or myelinated nerve fibres are also potential causes.

**Incontinentia pig menti (Bloch-Sulzberger syndrome)**

Incontinentia pigmeni is a rare genodermatosis, has X-linked dominant inheritance pattern and is usually lethal to male fetuses. Ocular features include abnormal peripheral vasculature, gliosis and tractional retinal detachment. In addition skin involvement occurs in all patients. Additionally, other ectodermal tissues may be affected, such as the central nervous system, hair, nails and teeth.

**Retinal astroctoma**

A sessile to slightly elevated, yellowish white retinal mass that may be calcified and is often associated with tuberous sclerosis and rarely neurofibromatosis. It may occur on the optic nerve head (giant drusen) in patients with tuberous sclerosis.

**References**

Globe perforation during peri-bulbar block is a complication, rarely reported these days in the era of topical surgeries. Certain predisposing lesions for globe perforation are high axial length, posterior staphyloma, previous extra-ocular surgeries, deep set eyes, uncooperative patients, and anesthesia given by non-ophthalmologists. During inadvertent globe perforation the damage occurs because of either penetration of the globe or toxicity of the drug injected. If there is penetration of the globe the patient may present with a varied constellation of symptoms such as Retinal break (in almost 100%), Vitreous hemorrhage (11-100%), Retinal detachment (42-61%), Retinal hemorrhage {sub/intra/pre} (5-52%), Hyphema (10-22%), Hypotony (10-21%) & Retinal whitening in cases of delayed presentation (7-15%). Intraocular injection of medicine can lead to sudden rise of IOP which may lead to CRAO or pressure induced damage of ocular structures. As per fear of toxicity of the drugs, Peyman et al and Lincoff et al observed no detectable damage to retina with lidocaine, lidocaine with epinephrine, bupivacaine and hyaluronidase. The management depends on the clinical picture of the patient and can vary from observation to photocoagulation to pars plana vitrectomy with endophotocoagulation with or without tamponading agents. The time of intervention is very crucial in such cases, and it deeply impacts the final outcome. We describe a case of iatrogenic globe perforation, in a patient planned for cataract surgery. The perforation was diagnosed on the table and appropriate management was carried out in the same sitting.

Case Report

A 61 year old male patient presented with diminution of vision both eyes which was gradually progressive and painless in nature for the past 2 years. On ocular examination patient was diagnosed as posterior polar cataract with nuclear sclerosis grade II in both eyes (Figure 1). Rest ocular examination was normal. The patient was planned for left eye Phacoemulsification with foldable IOL implantation under local anaesthesia. The axial length was 24.03 mm with A scan and the calculated IOL power was 19.50D.

Intra-operatively xylocaine 2% with adrenaline (1:200000) with Bupivacaine (0.5%) and Hyaluronidase was prepared for peribulbar block. While injecting subtle resistance was felt and the block was discontinued. Mild increase in IOP was noted and patient complained of mild pain. We developed a suspicion of globe perforation at this time. Per-operatively subconjunctival haemorrhage (Figure 2) was noted inferotemporally. Indirect ophthalmoscopy was carried out at this stage but retinal details were not very clearly seen because of media haze due to cataract. Surgery was continued under topical anaesthesia.

Phacoemulsification was carried out with modifications as needed for posterior polar cataract. The CCC was made of a larger size, no hydrodissection was carried out, no rotation of the nucleus, and injection of viscoelastics before withdrawing the phacoprobe (Figure 3) were carried out.

After completion of irrigation and aspiration, we noticed that the patient was not able to look into the microscope light as instructed (as patient was under topical anaesthesia). We continued with implantation of the foldable IOL. After IOL implantation and viscoelastic removal an indirect ophthalmoscopy was carried out.

On Indirect ophthalmoscopy we noticed an area of retinal haemorrhage with a retinal break inferotemporally and another suspected retinal break in the macular region (Figure 4).
Figure 1: Posterior polar cataract in both eyes.

Figure 2: Subconjunctival Haemorrhage seen inferotemporally

Figure 3: Viscocelastic injection before withdrawing the Phacoprobes.

(The picture used here is from the operating microscope view after making ports for vitrectomy).

The patient was planned for same sitting Pars plana vitrectomy with endophotocoagulation with 25G system. The corneal wound was sutured with 10-0 Ethilon 3 Scleral ports for 25 G vitrectomy (Figure 5) were made with the help of trochar and cannula.

Core vitrectomy was carried out. Posterior Vitreous Detachment was induced. The retinal haemorrhage was lifted with the help of silicon soft tip backflush cannula (Figure 6).

On lifting the haemorrhage it was observed that there was only one break which was present inferotemporally. The haemorrhage was shaved off from the base with the vitrectomy cutter. 3 circles of contiguous laser was done surrounding the break (Figure 7). Air fluid exchange was done following laser (Figure 8).

Scleral ports were closed with 8-0 vicryl.

The patient was started on antibiotic steroid combination and kept on a regular follow up.

On 1st post-operative day VA was 20/400 with an IOP of 12 mm Hg. There was air in the vitreous cavity making
assessments of the retina difficult. On the 3rd postoperative
day VA was 20/400 with an IOP of 34mm of Hg. The
vitreous cavity still had residual air, but we could appreciate
dispersed haemorrhage in the posterior segment. The
patient was started on Brimonidine tartarate and timolol
maleate combination along with Dorzolamide and oral
Acetazolamide.

On 10th postoperative day VA was 20/100 with an IOP
of 14 mm Hg. Fundus examination revealed a resolving
Vitreous Haemorrhage. Anti-glaucoma medications were
discontinued at this stage, and the patient was kept on
observation for IOP spikes.

On 4th week post-operatively the patient had an UCVA of
20/80 and a BCVA of 20/30 with -0.75 D sph/-1.25 D cyl
X 160º. Fundus examination revealed no haemorrhage and
laser marks were seen along the retinal break (Figure 10).

An OCT was carried out at this stage which revealed a
normal study (Figure 11).

**Discussion**

Inadvertent globe perforation can occur during peribulbar,
retrobulbar, sub-conjunctival injections, strabismus
surgery⁹, botulinum toxin injections for strabismus¹⁰,¹¹ and
has been reported even during chalazion surgery¹².

Complications related to pericentral injections, ranging from
innocuous subconjunctival haemorrhage to intracranial
diffusion have been described¹³.

The risk factors for perforation described are myopia,
previous buckling surgery, multiple injections. Myopia
has dual risk due to thinned out sclera and an elongated
eyeball. Buckling surgeries cause adhesions between the
globe and orbital tissues.

The first step in iatrogenic perforation is the penetration of
the globe. At this stage, the damage is usually restricted
to a retinal break. Injection of medication into the globe
results in a sudden rise of IOP. This could lead to a central
retinal artery occlusion or pressure induced damage to intraocular structures. Regarding toxicity Peyman et al7 and Lincoff et al8 observed no detectable damage to retina with lidocaine, lidocaine with epinephrine, bupivacine and hyaluronidase.

The Royal College of Ophthalmologists carried out a postal survey to assess the prevalence of globe perforation during local anesthesia in cataract surgery. Anaesthetist accounted for 75% of the cases. There was no difference between the ophthalmology residents and consultants. Anaesthetists often missed the mishap on the table.

Presentation4,5,6 in case of iatrogenic globe perforation can be in form of retinal break (in almost 100%), vitreous hemorrhage (11-100%), retinal detachment (42-61%),
its way into the globe, like depot steroids. Increased duration from injury to surgery increases chances of PVR as in any posterior segment perforating injury. Vitrectomy helps to remove the vitreous scaffolding as also the tract along which traction can occur.

In our case, the perforation was noted early and appropriate management was carried out at the same sitting with excellent results. So an early diagnosis and intervention is warranted in cases of iatrogenic globe perforation.

References

Instructions:
1. Please return your answers to dostimes10@gmail.com or mail them to “The Quizmaster, DOS Times Quiz, Dr. Rajesh Sinha, Room No. 479, Dr. R.F. Centre for Ophthalmic Sciences, All India Institute of Medical Sciences, Ansari Nagar, New Delhi - 110029”. Please write your DOS membership number along with your answers.
2. The answers should reach not later than 15th June, 2014. 
The quiz can also be viewed and directly answered on our website www.dosonline.org
3. The results will be announced at the DOS monthly clinical meeting on July 2014. The correct entry will be given a prize of Rs. 2,500. If there are more than one correct entries, the winner of the prize will be decided by draw of lots.

Quiz compiled by Dr. Digvijay Singh

Sent from my iPhone

Syndromania

1. Dystopia Canthorum referring to lateral displacement of both inner canthi is specific for which syndrome?
   a a - E e e

2. Posterior embryotoxon, iris hypoplasia and anterior segment dysgenesis is seen in which syndrome?
   a e e - i e e

3. Anterior and Posterior lenticonus may be seen in which syndrome?
   a o o o

4. X-linked dominant syndrome with round or oval widespread depigmented chorioretinal lesions:
   a i a i

Answer for January’14 issue of DOS Times

1. Ans. Presbylens
2. Ans. Lenticule
3. Ans. Perspex
4. Ans. Fyodosov

Answer to DOS Times Quiz February 2014

1. ____________________________ 2. ____________________________
3. ____________________________ 4. ____________________________
Delhi Ophthalmological Society

(LIFE MEMBERSHIP FORM)

Name (In Block Letters) ____________________________________________________________
S/D/W/o __________________________________ Date of Birth ___________________
Qualifications ____________________________ Registration No. __________________
Sub Speciality (if any) __________________________________________________________

Please Tick ☑ Correspondence 1.    or  2.    

ADDRESS:
1. Clinic/Hospital/Practice _______________________________________________________
   Phone __________________________
2. Residence _________________________________________________________________
   Phone __________________________
   Email ____________________________ Mobile No. ____________________________

Proposed by
Dr. ____________________________ Membership No. __________ Signature ____________

Seconded by
Dr. ____________________________ Membership No. __________ Signature ____________

[Must submit a photocopy of the MBBS/MD/DO & State Medical Council / MCI Certificate for our records.]

Declaration: I hereby declare that the above details are correct. I wish to be Life member. I have carefully read the instructions overleaf. I shall abide by the Rules, Regulation & Bye-Laws of the Society as in force and any subsequent amendment(s) made from time to time
(Life membership fee Rs. 5100/- payable by DD for outstation members. Local Cheques acceptable, payable to Delhi Ophthalmological Society)
Please find enclosed Rs. _______ in words ____________________________ by Cash ____________
Cheque/DD No. ____________________________ Dated ____________ Drawn on ____________________________

Signature of Applicant with Date
Three specimen signatures for I.D. Card.

FOR OFFICIAL USE ONLY
Dr. ____________________________________________________________ has been admitted as Life Member of
the Delhi Ophthalmological Society by the General Body in their meeting held on ________________
His/her membership No. is _____________. Fee received by Cash/Cheque/DD No. ________________ dated ____________
drawn on ________________.                                

(Secretary DOS)
INSTRUCTIONS

1. The Society reserve all rights to accept or reject the application.
2. No reasons shall be given for any application rejected by the Society.
3. Every new member is entitled to receive the Society’s Bulletin (DOS Times) and quarterly Journal DJO (Delhi Journal of Ophthalmology) of the Society free.
4. Every new member will initially be admitted provisionally and shall be deemed to have become a full member only after formal ratification by the General Body and issue of Ratification order by the Society. Only then he or she will be eligible to vote, or apply for any Fellowship / Award, propose or contest for any election of the Society.
5. To be proposed and seconded by Ratified Life Member only. No application form will be accepted unless it is complete in all respects. Proposed and Seconded by existing Member of the Delhi Ophthalmological Society.
6. Photo ID Card will be issued only after the membership is ratified by the General Body.
7. Documents to be attached with application form:
   1. Copy of Degree (MBBS / MD / DNB)
   2. Copy of Registration Certificate Medical Council of India or State Medical Council
   3. Copy of PAN Card
   4. One Stamp size Coloured Photograph to be pasted on the Application Form and one stamp size coloured photograph to be attached with form for issue of Laminated Photo Identity Card (to be issued only after the Membership ratification by GBM).
   5. For Delhi members only: Copy of Passport/Licence/Voters Identity Card/Ration Card/Electricity Bill/MTNL (Landline) Telephone Bill (Delhi Life Member should either reside or practice in Delhi.
8. Membership Fee
   There is only membership on one Time Payment of Rs. 5,100/-
   1. Life membership fee Rs. 5,000/- (This money will be part of corpus of Society)
   2. Admission fee Rs. 100/-
   The application form should be complete in all respects and accompanied by a Demand Draft of Rs. 5,100/- in favour of “Delhi Ophthalmological Society” payable at New Delhi should be sent:

   Dr. Rajesh Sinha, Secretary,
   Delhi Ophthalmological Society,
   Room No. 479, 4th Floor, Dr. R.P. Centre for Ophthalmic Sciences, AIIMS, Ansari Nagar, New Delhi – 29

9. For update address for sending application, please visit website www: dosonline.org
## Visual Development Milestones and Visual Acuity Assessment in Children

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<tr>
<th>Age</th>
<th>Visual Milestone</th>
<th>Visual acuity</th>
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<tr>
<td>29 wks POG</td>
<td>Pupillary reactions to light</td>
<td>6/480-6/120 (by OKN)</td>
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<tr>
<td>30 wks POG</td>
<td>Dislikes and responds to bright light</td>
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<tr>
<td>Birth</td>
<td>Blinks to light; prefers soft diffuse light</td>
<td>6/360 to 6/120 (by OKN)</td>
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<tr>
<td>1 week</td>
<td>Vetibulo-ocular reflex, OptoKineticNystagmus seen</td>
<td></td>
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<tr>
<td>2 weeks</td>
<td>Small saccades develop; follows horizontal moving objects</td>
<td></td>
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<tr>
<td>1 month</td>
<td>Fixation developing, can watch mothers face for prolonged time</td>
<td>6/480-6/120 (by PL tests)</td>
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<tr>
<td>2 months</td>
<td>Fixation well developed; bifoveal fixation; larger saccades; pursuits and convergence movements; follows vertically moving objects</td>
<td>6/120-6/60</td>
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<tr>
<td>3 months</td>
<td>Watches movements of own hands and reaches out towards interesting objects; prefers photographs to patterns</td>
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<tr>
<td>4 months</td>
<td>Foveal differentiation complete; sensory fusion and accommodation begins to develops,</td>
<td>6/120-6/30</td>
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</table>
| 5 months          | Blink response to visible threat (menace response); grasps and explores objects; stereopsis begins to develop | 6/90-6/24  
12-6/6 by VER |
| 6 months          | Accommodation well developed; fusionalvergence well developed                   |                        |
| 9 months          | Visual differentiation of objects’ picks up small objects                       | 6/48-6/12  
6/6 by VER |
<p>| 18 months         | Visual acuity at adult levels on pediatric acuity cards                          | 6/18-6/7.5            |
| 3 years           | Visual acuity at adult levels on snellen chart; contrast sensitivity well developed | 6/12-6/6 (36 months)  |
| 5 years           | Stereopsis fully developed                                                      | 6/5                   |
| 9 years           | Critical period of monocular deprivation ends                                  |                        |</p>
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<tr>
<th>Age Group</th>
<th>Methods used</th>
<th>Type of Test/Acuity</th>
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<td>Response to occlusion</td>
<td>Resolution acuity</td>
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<td>Visual Evoked Response</td>
<td>Resolution acuity</td>
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<td>Optokinetic nystagmus</td>
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<td>Boeck candy beads</td>
<td>Detection acuity</td>
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<td>Stycar graded balls test</td>
<td>Detection acuity</td>
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<td>Teller/Keeler acuity cards</td>
<td>Resolution acuity</td>
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<td>Cardiff acuity cards</td>
<td>Recognition acuity</td>
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<tr>
<td>18 months to 3 years</td>
<td>Cardiff acuity cards</td>
<td>Recognition acuity</td>
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<td>Kay picture test</td>
<td>Recognition acuity</td>
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<td>Sheridan-Gardiner test</td>
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<td>3-5 years</td>
<td>Kay picture test &amp; Allen/LEA symbols</td>
<td>Recognition acuity</td>
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<td>Sheridan-Gardiner test</td>
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<td>Cambridge crowding cards</td>
<td>Recognition acuity</td>
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<td>Stycar test</td>
<td>Detection and recognition acuity</td>
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<td>Snellen letters</td>
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<td>Tumbling E</td>
<td>Recognition acuity</td>
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<td>HOTV</td>
<td>Recognition acuity</td>
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<td>More than 5 years</td>
<td>Sheridan-Gardiner test</td>
<td>Recognition acuity</td>
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<td>Snellen letters or numbers</td>
<td>Recognition acuity</td>
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<td>LogMAR</td>
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