Infantile cataract: where are we now?
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Introduction
Pediatric cataract is one of the major causes of preventable childhood blindness affecting approximately 200,000 children worldwide.\(^1\) In developing countries, the prevalence of blindness from cataract is higher, about one to four per 10,000 children. Early diagnosis and treatment is essential to prevent the development of stimulus deprivation amblyopia in these children. Cataract surgery in infants poses greater challenges compared to young children. Primary implantation of an intraocular lens remains controversial for infants, and the selection of an appropriate IOL power is difficult. The management of infantile cataract has changed over the last decade. In this study, we present an overview of the changing concepts of cataracts in infants and its management.

Etiology of childhood cataract
The common causes of congenital cataract are genetic, metabolic disorders, prematurity and intrauterine infections. Almost 60% of cases of congenital cataract in developed countries are idiopathic.\(^2\) One-third of cases of congenital cataract are hereditary without any known associated systemic disease.\(^3\) The various causes of congenital cataract are

1. Heredity: These cataracts are usually autosomal dominant but can be autosomal recessive and x linked.
4. Intrauterine infections: Toxoplasmosis, Rubella, Cytomegalovirus, Herpes, Varicella and Syphilis.
5. Associated Ocular conditions: Aniridia, Iris coloboma, lens coloboma, Lenticonus, Lentiglobus.\(^6,7\)

Examination of the child
Detailed ocular examination of the child can be done either in an outpatient setting if the child is cooperative or under general anesthesia when the child is being taken up for surgery. Quantification of visual acuity of the child as far as possible should be done. In infants, fixation behavior, fixation preference and resistance to occlusion gives us a clue to the visual acuity. In young infants with poorly developed fixation, an undilated distant direct ophthalmoscopy can indicate whether the opacity is visually significant or not. Dense central opacities larger than 3 mm in diameter usually need surgical removal.\(^8\)

Examination of both the eyes has to be done to determine whether the cataract is unilateral or bilateral. Unilateral cataract, even if mild can cause irreversible deep amblyopia if not treated.\(^9\)
Often the first symptom is a white or partially white reflex noted by the parents. Strabismus and nystagmus should be specifically looked for in these children and sometimes these may be the presenting signs. Strabismus is usually seen in children with unilateral cataracts and develops when an irreparable visual loss has already occurred. The presence of manifest nystagmus at age of 2–3 months or elder generally indicates a very poor prognosis. The presence of either strabismus or nystagmus indicates that cataract is visually significant.

Slit lamp biomicroscopy should be done to assess the size, location, density of the opacity. Corneal diameters and intraocular pressure have to be measured with a tonopen or Perkins hand held applanation tonometer. Indirect ophthalmoscopy can reveal persistent fetal vasculature or other posterior segment abnormalities that may affect the visual outcome. In cases where the media opacity precludes examination of the fundus, a B scan ultrasonography has to be performed to rule out other posterior segment pathologies that mimic congenital cataract. These conditions include retinoblastoma, persistent hyperplastic primary vitreous, coats disease, ROP with retrolental fibroplasia, organized vitreous hemorrhage, congenital falciform fold, ocular toxocariasis and retinal hamartomas.

Performing cataract surgery in these conditions is disastrous and can lead one into medicolegal problems. These children should be evaluated by a pediatrician to exclude systemic disorders or metabolic causes causing cataract.

**Laboratory workup**

Most children with congenital cataract do not need systemic work up. Unilateral, familial, isolated cataracts with no systemic association do not need any systemic investigations. Trauma should be ruled out in all cases of unilateral cataract. A child with peculiar facies or systemic malformations like microcephaly, deafness, cardiac abnormalities, developmental delay need systemic workup. The work up usually includes fasting blood sugar, urine for reducing substances for galactosemia, aminoacids for Lowes syndrome. Plasma phosphorous, calcium levels, RBC transferrase and galactokinase levels have to be assessed. TORCH titers have to be done to rule out infectious causes of cataract.11,12

**Management**

Indication for cataract surgery in infants depends on the extent of its effect on the visual function. Mere presence of a lenticular opacity does not warrant surgical removal. Cataract which is incomplete at birth, peripheral lens opacities, punctate opacities with intervening clear zones, opacities <3 mm in diameter can be kept under close follow up.13 Associated amblyopia in these children should be treated by appropriate glasses and patching. Small opacities can be managed by prescribing mydriatic agents to achieve a larger area of clear visual axis.

**Timing of surgery**

Extraction of unilateral congenital cataracts by 4–6 weeks and bilateral cataracts within 6–8 weeks of life can prevent the development of strabismus, nystagmus and amblyopia.14

**How does an infant eye differ from the adult eye?**

A child’s eye is unique and is different from an adult eye. The eyes are smaller in size at birth and have steeper corneas. The normal new born eye has a mean axial length of 16.6–17 mm. It reaches a mean adult value of 23.6 mm at 15 years age. More than half of this growth in axial length occurs before 1 year age and most axial elongation occurs during the first 2 years of life. The corneal curvature reduces from 51.2 D in new borns to 43.5 D in adults.16 The sclera is thin and less rigid, the lens capsule is more elastic, and there is a risk of severe postoperative inflammatory response.

**Biometry in infants**

With advances in surgical techniques and instrumentation, several surgeons are implanting IOLs in infants. Refractive growth after IOL implantation in infants cannot be predicted accurately and current IOL formulae vary in their predictive outcomes. If target postoperative emmetropia, amblyopia treatment is easier but this strategy results in high myopia in later life. If we aim for hyperopia, amblyopia therapy and refractive correction in initial phase is difficult but this strategy has the advantage of potentially achieving either emmetropia or low myopia later in adulthood. The amount of hyperopia will vary depending on the age of the child at the time of surgery. Most surgeons prefer to leave infants with hyperopia as it’s easy to titrate as children grow. IATS recommended an hyperopia ranging from +6 D to +8 D depending on the age of infant at the time of surgery. It is important to counsel parents regarding need for glasses postoperatively as well as perhaps through the life.

Three important things to be considered when determining the IOL power to be implanted in infants are

1. Anticipated refractive shift.
2. Age of the patient.
3. Target refraction in the immediate postoperative period.

Children who are younger at the time of surgery, have a significantly greater myopic shift and greater variance in predictive refractive change than older children. Crouch et al. in a study of 52 eyes undergoing cataract surgery with
IOL implantation found a mean myopic shift of 3.66 D in children operated on at 3–4 weeks age which reduced to 0.38 D in children operated on at 15–18 years age. Most pseudophakic eyes grow normally and so a significant shift after IOL implantation is expected in these children.

Postoperative refractive goal in infants
In infants, implantation of IOL still remains controversial and several surgeons prefer to leave the infants aphakic after cataract surgery. In the Infant Aphakia Treatment Study, the target refractive error after IOL implantation was +8 for infants 4–6 weeks of age and +6 for infants between 6 weeks to 6 months age.

Keratometry and axial length measurements in children are usually less accurate compared to adults. These measurements are often obtained under anesthesia in infants who do not cooperate for fixation. Mittelviefhaus et al. in their study have shown that lack of fixation in children under general anesthesia can result in inaccurate keratometry measurements. However, the reliability can be increased by averaging several readings per eye.

Axial length is a more significant source of error in IOL power calculation. Inaccurate axial length measurement can account for 4–14 diopters for each millimeter difference in IOL power. Errors are often magnified because of shorter axial length. Immersion biometry is more predictable than contact method for IOL power calculation in infants. But the limitation of the immersion scan is that it cannot be used in small eyes and globe with shallow anterior chamber and other ocular anomalies as in infants. Partial coherence interferometry can be used to measure axial length in cooperative children with reliability and accuracy. Advantages over conventional ultrasound include high reproducibility, contact free measurements, observer independence of the measurements. The disadvantage is that it cannot be used in total cataracts which are more often encountered in children.

IOL formula
Furthermore, no time tested formula exists for calculation of IOL power in infants. The accuracy of each formula depends on optimized values and measures of the formula components, including factors such as actual anterior chamber depth, lens thickness, vertex distance, and use of a personalized surgeon factor or A-constant. The anterior segment of an infant eye is significantly smaller, eyes with congenital cataract may have greater anatomic variation in anterior segment structures and the anterior segment of an infant is proportionally larger to the posterior segment compared to an adult eye. The capsular bag of an infant eye is smaller and will contract earlier, which may result in greater posterior IOL displacement. Implantation of high-power IOLs in these eyes, can increase the measurement and calculation errors as well as the errors induced by changes in IOL position. Nihalani and Vanderveen in a retrospective study of 135 pediatric eyes that underwent cataract surgery and primary IOL implantation found mean predictability of four formulae was comparable, with 57% of infants having a prediction error of more than 0.5 diopters. Greater prediction errors were seen in children <2 years, axial length <22 mm and mean keratometry readings >43.5. The SRK II, SRK T and Holladay formulae tended to overcorrect whereas Hoffer Q had an equal number of undercorrection and overcorrections. Kekunnaya et al. in their study on IOL power calculation in children <2 years age found prediction errors were larger for all formulas but SRK II had the least prediction error. In the Infant Aphakia treatment study to determine the predictability of IOL power calculation formulae in infants eyes, overall median absolute prediction error values appeared to be similar for the Holladay 1, Holladay 2 and SRK/T formulae (1.2 D, 1.4 D and 1.3 D, respectively), and in paired comparisons of SRK/T versus other formulae, the median paired differences in absolute prediction error was more than zero, indicating greater accuracy for the SRK/T formula. The study concluded that Holladay I and SRK T formula gave good comparable results and have the best predictive value for infant eyes. The greatest prediction errors in their study were seen in eyes with axial length of 18 mm or less.

Type of IOL to be implanted
There is a large debate regarding the type of IOL to be implanted in infants. IOL implantation during childhood may be associated with better visual outcomes but in IOL implantation in infancy, these potential advantages are offset by a higher incidence of intraoperative and postoperative adverse events. Additional intraocular surgeries are often required to treat these adverse events which are associated with risks, costs and parental stress. Although it is agreed that cataract surgery during early infancy is associated with the best visual outcomes, it remains undetermined whether primary IOL implantation is advisable in this age group. Ram et al. compared outcomes of hydrophobic acrylic and PMMA lenses in children <1 year age and reported that complication rates were comparable in both the groups. PMMA lenses may require early surgical intervention for PCO. The single piece acrylic hydrophobic IOL is a soft IOL and can be implanted in the smaller capsular bag as in infants with relative ease. It also has the advantage of requiring a smaller incision thereby allowing corneal incision leaving conjunctiva intact. However, single piece IOL cannot be placed.
Are IOLs good for infants?

This question has been debated by Pediatric ophthalmologists for several years. There was little evidence to support the claim either way. The infant aphakia study was designed to answer this question. This was a prospective randomized multicentric trial comparing infants who underwent cataract surgery for unilateral cataract with either IOL implantation or were left aphakic and were fitted with a contact lens. The main outcome variable was visual acuity at 1 year and 4½ years of age. The investigators also looked at complications, resurgery rates and strabismus and stereopsis and compliance to occlusion. IATS found that there was no difference in visual acuity at either 1 year of age or at 4½ years between the two groups. But alarmingly adverse events like membrane proliferation into the visual axis, corectopia were almost 10 times more common in infants with IOL implantation compared to aphakic infants.²⁹ In aphakic eyes, the margins of the anterior and posterior capsular bag usually fuse together preventing lens material from migrating out of the Sommerring ring into the pupillary space. Whereas in pseudophakic eyes, lens epithelial cells are able to migrate into the pupillary space because the IOL interferes with the fusion of the lens capsular remnants. Hence, not surprisingly, the commonest indication for resurgery in IOL group was to clear the visual axis. Additional intraocular surgeries were 3½ times more in pseudophakic infants compared to aphakic infants.³¹ The risk of glaucoma was same in both the groups. The development of stereopsis did not differ depending on the type of optical rehabilitation. In conclusion, the study did not demonstrate any visual benefit from implanting an IOL at the time of unilateral cataract surgery in infants <7 months of age and the children who had IOL implantation had more adverse events and required more reoperations to clear visual axis opacities. The investigators concluded by saying “When operating on an infant younger than 7 months of age with a unilateral cataract, we recommend leaving the eye aphakic and focusing the eye with a contact lens. Primary IOL implantation should be reserved for those infants where, in the opinion of the surgeon, the cost and handling of a contact lens would be some burdensome as to result in significant periods of uncorrected aphakia.”³² In the context of our country, however the IATS conclusions need to be interpreted in a slightly different light. Monocular cataracts in infants, where only way of visual rehabilitation is contact lens, is often problematic in developing countries because of poor hygiene, socioeconomic factors and non-availability of contact lenses in smaller towns. So these infants will probably do better with IOL. Only rider is that lens be placed in the bag with appropriate capsular management and anterior vitrectomy to ensure clear visual axis. Other important factor is patient selection and we do recommend IOL in infants who have otherwise anatomically normal eye with no anterior segment dysgenesis or other anomalies. Bilateral cataracts in infants however aphakia can be easily managed with aphakic glasses as well as contact lenses. Attending surgeon is best placed to take that decision customising it according to patient’s profile and his/her skills. IATS has certainly provided information for better informed decisions.

Complications following surgery

Postoperative inflammatory response in children can result in fibrinous and pigment deposits on the IOL. Inflammatory response can be really exaggerated in infants with Rubella syndrome.

Posterior capsular opacification is the most common complication after pediatric cataract surgery. Primary posterior capsulotomy with anterior vitrectomy combined with hydrophobic acrylic IOL in the bag, can prevent or delay the occurrence of VAO. PCO if develops can be treated by NdYag laser capsulotomy or membraectomy depending on the child’s cooperation and the thickness of PCO.

Secondary glaucoma is the most feared complication of infantile cataract surgery. IATS showed that IOL implantation does not seem to protect against the development of glaucoma.³³ Matafshave reported that glaucoma after pediatric cataract surgery is associated with surgery with in first 1 month of life and additional surgical procedures but not with primary IOL implantation.³⁴ Parents must be counselled regarding glaucoma and need for regular follow up. Rapid myopic shift and increased axial length points towards glaucoma and high index of suspicion should be maintained in these children.

Retinal detachment is a rare late postoperative complication of pediatric cataract surgery.

Postoperative visual rehabilitation

Visual rehabilitation in children after surgery can be achieved by aphakic glasses, contact lenses, IOL implantation. Aphakic glasses are efficient method of visual rehabilitation in infants especially in bilateral cataracts. Contact lenses are particularly useful in unilateral cataract. Silicon soft lenses or rigid gas permeable lenses are commonly used. Speegschatz et al. in their study of 157 aphakic subjects found that initial rehabilitation with aphakic glasses and secondary IOL implantation at a later date has the advantage of predictable postoperative refraction and fewer complications.³⁵ Visual rehabilitation can be done in the immediate
postoperative period by aphakic glasses in bilateral cases and contact lenses in unilateral cases. When fitting an infant with aphakia with contact lenses the problem of appropriate power of the contact lens arises. Silicon elastomer contact lens is the preferred contact lens for the treatment of aphakia in infants. It is easy to fit and can be used as an extended wear contact lens. Rigid gas permeable lens is also an option given the advantage of cost and good oxygenation for corneal. The preoperative axial length can be used to determine the contact lens power to be used. Martin et al. in their study reported the power of the contact lens depending on the preoperative axial length as: 0–6 months, +29 D; 7–17 months, +26 D; 18–28 months, +23 D and 29–34 months, +18 D. 5 Moore noted that the mean spherical equivalent refractive error for these patients was +28.5 D at 6 months, +26.5 D at 12 months, +23 D at 24 months and +21.5 D at 36 months. Trivedi et al. recommends 32-D CL when the preoperative AL is <17 mm, a 29-D CL when the preoperative AL is between 17 and 18.5 mm, a 26-D CL when the preoperative AL is 18.5–19.5 mm, a 23-D CL when the preoperative AL is between 19.5 and 20 mm (21 mm) and a 20-D CL for an AL of 20–21 mm (20 D for >21 mm). Secondary IOL implantation can be reserved as an option for later visual rehabilitation in these children. Nihalani et al. in their study of secondary IOL implantation in children left aphakic during initial cataract surgery found satisfactory visual and refractory outcomes in children receiving secondary IOL implantation. However, immediate postoperative inflammation and corneal edema was more in eyes with sulcus implanted IOL compared to in the bag IOL.

Last but not the least, several of these infants will need continued monitoring for amblyopia and patching and strabismus. They will need appropriate refractive correction moving from a single vision glass to a bifocal or progressive as their visual needs change. It is important not to miss these unglamorous factors in follow up visits otherwise amblyopia can trump a beautifully done surgery in a matter of few months.

Conclusion
Cataract surgery in infants is a specially challenging subset among all cataract surgeries in children in terms of surgical technique, formula to be used, biometry and postoperative visual rehabilitation. But it is rewarding, if we are able to rehabilitate them either with glasses or primary IOL implantation or contact lenses. Surgeon has the onus of deciding on the best course looking ahead not few years but few decades.

References