Visual rehabilitation in pediatric cataract

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Background

Pediatric cataract is a major cause of decreased vision and blindness in children world wide. Though the trend towards early cataract surgery, understanding of the critical sensitive period and refinement in surgical techniques have improved the visual outcome, the continuous threat for amblyopia poses a big challenge in front of pediatric ophthalmologists. Factors like age, laterality and associated systemic anomalies are non modifiable but the understanding of modifiable factors like residual refractive status and amblyopia therapy can improve the visual outcome.

Why so challenging?

Pediatric eye is not the miniature adult eye. Significant amount of variation in refractive change/myopic shift, lack of accurate intraocular lens (IOL) formulae, increased risk of surgery and IOL related complications often leaves the surgeon in dilemma[1]. Also, post operative visual rehabilitation poses special challenges due to compliance and increased risk of amblyopic if not done properly.

In this article we will be focusing on pediatric aphakic rehabilitation, post op amblyopia management and low vision aids including social and environmental modification.

Primary IOL implantation

Primary IOL implantation is the preferred modality of treatment following cataract extraction in older age group though controversial in early age[2,3]. Advantages of early IOL implantation include optimal visual rehabilitation, improved visual acuity, BSV and less chances of strabismus but it is associated with increased rate of visual axis opacification and resurgeries. Recently finished IATS concluded that contact lens provide similar visual outcome though IOL implantation proves to be costly and the chances of resurgeries are more with primary IOL implantation in infants[1,4], although pseudophakes will require optical rehabilitation in form of glasses and/or contact lens which will be discussed later in the article.

Whether or not to implant IOL

Minimal age at surgery for an IOL implantation varies from surgeon to surgeon and varies between unilateral and bilateral cataracts. Though as general consensus, bilateral cataracts operated at an early age are left aphakic with the intention of secondary IOL implantation later, there has been a recent trend (though controversial) in implanting IOL in infants. The IATS states that primary IOL implantation does not provide any visual benefit when followed upto five years. Economic burden and resurgery rate is also more with IOL implantation[5]. Other relative contraindications of primary IOL implantation include associated uveitis, severe microphthalmia such that IOL size is not feasible to implant, persisitent fetal vasculature, inadequate capsular support. Other

Figure 1a. Child with bilateral congenital cataract

Figure 1b. Child with RE- PCIOL, LE- Lamellar cataract

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important concerns are selecting an IOL power and the higher rate of visual axis opacification.

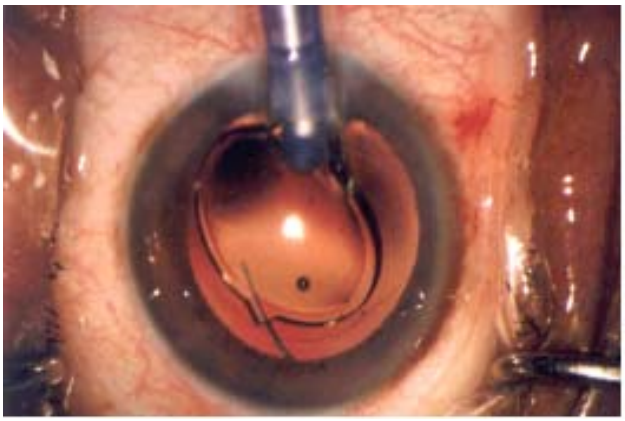

Figure 2. Primary IOL implantation

IOL power calculation

Implantation of a fixed power IOL in an eye that is still growing makes it difficult to choose the IOL power to implant[6]. Several nomograms have been published in literature to decide on IOL power (Table 1). There are various factors which influence IOL power implantation, which include age, laterality, status of the fellow eye, hereditary factors (refractive status of the parents and siblings). Associated posterior segment pathology should also be kept in mind, for example patients with lasered and post-cryo retinopathy of prematurity (ROP) have higher myopic shift than normal infants, on the other hand patients with persisitent fetal vasculature tend to have shorter eyes[7,8]. That's why it is recommended that IOL power should be customized for every child rather than sticking to the mentioned nomograms.

Several groups advocate initial myopia or emmetropia instead of hyperopia[9]. Table 2 outlines the advantage of the various options.

IOL formulae

There is lack of accurate IOL formulae which takes into account the small pediatric eyes and variable factors. IOL power has to be customized according to various factors mentioned above.

Newer technologies

Recently, there has been a trend in using Multifocal IOLs in children by some pediatric ophthalmologists, However, predictability of IOL power, unstable refractive error, potential for amblyopia due to loss of contrast sensitivity are the limitations of multifocal IOLs in children. Multifocal IOLs should be limited to older children who can communicate regarding their experience with multifocal IOLs[10].

Table 1. Recommendations for IOL power calculation from various studies

Age at surgery	*Crouch et al (targeted post operative refraction) (D)*	*Awner et al (targeted post op refraction) (D)*	*Hutchinson et al (decreased calculated IOL power for emmentropia) (D)*	*Dorothy ey al (decreased calculated IOL power for emmetropia) (D)*	*Dahan et al (of calculated IOL power for emmetropia (%)*
1	+4	+4		25% of first 6 months, 20% for next 6 months	80
2	+3.5	+4	1		90
3	+2.5	+3	1		90
4	+2.5	+3	1		90
5	+2	+2	1		90
6	+2	+2	1		90
7	+1	+1	1		90
8	+1	+1	1		90
9	Emmetropia	Emmetropia	1		90

Table 2.

	Advantage	*Disadvantage*	*Adult refraction*
Initial hyperopia	Hyperopia improves as age grows	Initial spectacles/ contact lens	Low myopia/hyperopia/ emmetropia
Initial emmetropia	No spectacles / Contact lens in initial period	Large myopic shift	Moderate to high myopia
Initial myopia	Correction to prevent amblyopia	Large myopic shift	Very high myopia

Aphakic glasses

Patients who are left aphakic primarily need optical rehabilitation in the form of glasses or contact lens in the post operative period till the secondary IOL is planned. Aphakic glasses are commonly used for the correction of bilateral aphakia in children[2]. Due to increase trend towards primary IOL implantation and contact lens, the availability and technology for high power plus lens (> 10 D) have declined. Primarily three type of aphakic glasses are available[11].

Type of lens	
Lenticular lenses	Prescribed power at the centre of lens surrounded by ring of no power. Inferior to other lens. Only option when requiring > 20 D
Aspheric lenticular lenses	Aspherical central area surrounded by ring of no power. Available 10-20 D. optically superior to lenticular lens
Multidrop lenses	Spherical central zone that flattens into aspherical zone which further blends with ring of no power. Far superior to other options.

Frame selection is very important in pediatric age group. Whenever possible the child should be included in decision making for the frame. Frame selection is based on several factors including activity level and type of activities the patient is involved in. Plastic is the material of choice for most children as it is more elastic and durable. Frame temple are available in various shapes including comfort coil type, skull temple piece and paddle piece. In general, a plastic frame and comfort coil temple is the best choice for infants and toddlers. For a small infant paddle temple with a holding strap can be very useful (Figure 3).

Figure 3. Child with aphakia glasses

Contact lens

Pediatric aphakia can be successfully treated with contact lens. Studies have proved that better binocular vision and stereopsis is gained in patients who show excellent compliance with contact lens postoperatively[12]. Advantages of contact lens include easy adjustability with the changing refractive error. Good lens fit, care and hygiene compliance can prove to be an excellent alternative to primary IOL implantation, though inability to match these requirements can lead to hazardous results.

Contact lens in pediatric age group is quiet challenging for adults. Daily wear and removal of contact lens can be bothersome for the parents and child and might lead to loss of compliance. Extended wear contact lens

can provide with an easier option. Measuring the eye parameters for such young children is not easy and assessing the fit on slit lamp can be challenging.

There are three main type of lens material available.

a) Hard (PMMA) or rigid gas permeable lens
b) Hydrogel (extended wear lens)
c) Silicone lens

PMMA lens are available in wide range of prescriptions, can be customized to power and base curve and can neutralize astigmatism and spherical error in most of the patients. Disadvantages of PMMA include its need to be removed daily which can be cumbersome for the parents, and occasional breakage associated.

Hydrogel lens can be used for extended wear, though they are difficult to insert and do not correct the residual astigmatism.

Silicon lenses provide superior corneal oxygenation and durability and can be fitted with ocular measurement or trial techniques. Silicon lenses are costly and its availability may vary in different parts of the world. It is available in limited powers and can be uncomfortable initially. Also silicon lens do not provide UV protection. If silicon lens cannot be worn by a child, an RGP lens can be tried. RGP lens may be ordered with UV blockers which provide a theoretical advantage over silicon lens.

Daily vs extended wear

In ideal situation, extended wear is the best available option for pediatric aphakic eyes. But due to the complication rate like giant papillary conjunctivitits, neovascularization, abrasions and infective keratitis that have been reported more with the use of extended wear lens, experts recommend use of daily wear lens in pediatric aphakes.

Although the US FDA approves up to 30 days use of extended wear lens, risk of ulcerative keratitis increases with the duration of lens wear. It is advisable to disinfect the lens every week[11].

Initial fitting

Initial fitting of the lens is recommended at the time of surgery only. The need for high refractive power leads to relatively more central thickness. Hence to avoid decentration, lens diameter must be as large as cornea and relatively steep base curve is indicated.

Consideration and complication of CL:

a) **Non compliance** of both the parents and patient is a major hindrance in CL practice. It is more common in unmotivated patients and in unilateral aphakes where the vision of other eye is good. Daily insertion and removal is the main contributing factor.
b) **Infection:** Parents should be properly educated regarding insertion, removal and hygiene maintenance of CL. Slightest of the conjunctival redness should be addressed to the ophthalmologist and CL should be removed in both eyes simultaneously as amblyopia can quickly develop. Aphakic glasses should be worn until the refitting.
c) **Power changes:** Frequent follow up is required as power changes occur rapidly.
d) **Lens loss:** A spare set of lens should always be available.
e) **Parental stress:** Psychological stress can be an important obstacle for CL compliance. Though initial resistance is more commonly seen, most patients tend to become compliant after some time.
f) **Outdoor activities:** Hygiene is to be maintained while involved in outdoor activities and games. Major concern raised by parents is regarding swimming. Well fitted goggles should be used. Cleaning and disinfection of lens is extremely important.

Low vision aids

These devices enable the patients to improve their visual performance and help them to reach their full capacity. Depending on the age and affordability of the patient different low vision devices and assistive technologies may be considered. The primary mode of optical assistance is convex lenses in various forms[11].

These devices are 1) Standard additional bifocals 2) Hand magnifiers 3) Stand magnifiers 4) Electronic magnifiers 5) Telescopes 6) Computer adaptations, closed circuit television. (Figures 4-a, b, c)

Patients and their family members should be encouraged to use these devices. Good visual results depend on the severity of visual impairment and any associated comorbid conditions.

Amblyopia in paediatric cataract

Deprivation amblyopia is the main barrier to successful visual outcome in the management of pediatric cataract. Amblyopia develops because of the absence of equal quality of visual input to each eye during the critical period[13]. A report from Elston and Timms suggests that the first 6 weeks of life represents the critical period for binocular development[14]. Therefore unilateral cataract has more risk of amblyopia than bilateral cataract and

Figure 4a. Hand magnifiers

Figure 4b. Stand magnifiers

Figure 4c. Closed circuit television

hence needs to be operated as early as possible (within 6 weeks of age).

Surgical correction of bilateral cataract should occur in close succession (within 2 weeks) if the cataracts are symmetrical[15]. Prolonged interval between the two eyes could result in dense deprivation amblyopia in the untreated eye. Asymmetrical bilateral cataract that require surgical removal presents an unique challenge as deprivation amblyopia may be present in the worst eye.

Amblyopia can also develop postoperatively due to anisometropia, strabismus or visual deprivation due to posterior capsule opacification[16].

Predictors of amblyopia treatment success

Age at which cataract develops, cataract density and age of surgery influence success of amblyopia treatment. Early age of surgery and early rehabilitation offer better visual outcome[17, 18]. Nystagmus and strabismus are often observed in patients where the treatment was delayed or who showed poor compliance with post operative visual rehabilitation. Often congenital cataract associated with other ocular abnormalities like microphthalmos or persistent fetal vasculature show poor postoperative visual outcome.

Amblyopia treatment

Amblyopia therapy remains the most critical step in the rehabilitation of postoperative eyes. Treatment should start as early as possible following surgery. Occlusion of the sound eye remains the mainstay of amblyopia therapy. The schedule is based on the visual acuity and age of the child. It is assumed that the occlusion treatment must continue throughout childhood or at least till the visual acuity has reached a plateau.

However compliance remains the main challenge in amblyopia treatment. Therefore family members need to be counseled regarding amblyopia management even before surgery (Figure 5).

Figure 5. Patch therapy

Conclusion

Cataract is one of the most significant and treatable causes of visual impairment in children. Even with

the advancement in the surgical techniques and instrumentation successful management of cataracts in children is one of the most difficult challenges in pediatric ophthalmology. The actual treatment starts after the surgery.

The main concern is amblyopia for which appropriate optical correction and patching in unilateral cases is to be started as soon as possible. These children need to be followed up regularly and monitored for vision and change in refraction.

Whenever IOL implantation is not a feasible option, contact lens provide the best rehabilitation method for pediatric cataracts atleast in unilateral cataract. Aphakic glasses provide a reasonable option for bilateral aphakes. When it is not possible to use CL or aphakic glasses, it may be necessary to implant secondary IOL. Contact lens usage requires maintenance of proper hygiene, compliance and regular follow ups for the continuous refractive change.

Visual rehabilitation in pediatric cataract depends upon the surgeon, availability of options, ocular associations and compliance of the patient.

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