Pediatric Ophthalmology Update

Update of intraocular lens implantation in children

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Abstract

Cataract is a common problem that affects the vision in children and a major cause of amblyopia in children. However, the management of childhood cataract is tenuous and requires special considerations especially with regard to intraocular lens (IOL) implantation. Age at which an IOL can be implanted is a controversial issue. Implanting an IOL in very young children carries the risk of severe postoperative inflammation and posterior capsule opacification that may need other surgeries and may affect the vision permanently. Accuracy of the calculated IOL power is affected by the short eyes and the steep keratometric values at this age. Furthermore, choosing an appropriate IOL power is not a straight forward decision as future growth of the eye affects the axial length and keratometry readings which may result in an unexpected refractive error as children age. The aim of this review is to cover these issues regarding IOL implantation in children; indications, timing of implantation, types of IOLs, site of implantation and the power calculations.

Keywords: Pediatric cataract, Intraocular lens, Axial length

Introduction

Congenital cataract is the most common cause of treatable childhood blindness. It is responsible for 5–20% of the cases of blindness worldwide. However, the incidence of visual impairment may be higher in developing countries.[1] Management of congenital cataract is a challenge and surgery remains an important part of the management. Techniques for managing pediatric cataracts, methods for calculating intraocular lens (IOL) power and surgical instruments and techniques for IOL implantation have steadily advanced.

A number of factors may influence the surgical outcome including: patient age; type of cataract; laterality; timing of surgery; technical surgical aspect; changing refraction; functional outcome and amblyopia; choice of aphakia correction and IOL implantation; the active lens epithelial cells and resultant posterior capsular opacification (PCO); the presence of thick vitreous in pediatric age group which protects against cystoid macular edema (CME); and finally parental care and compliance.[1–3]

Surgery for congenital visually significant cataract must be performed as early as possible to prevent irreversible amblyopia and the timing of surgery needs to balance the effect on visual development and the surgical risks.[4] Recently, refined surgical techniques and options for optical correction of aphakia improved both the technical and functional outcomes of pediatric cataract surgery. Optical correction includes aphakic glasses, contact lenses and primary IOL implantation. Each of the options has its own advantages and disadvantages.

The capability of the IOL to provide a constant visual stimulus, led to the acceptance of IOL implantation as an alternate form of optical correction. Despite controversy,[3] IOLs are implanted in infants with increasing frequency. Some have advocated surgery based on age as follows:

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1. Infants less than 6 months to undergo lens aspiration, primary posterior capsulectomy and anterior vitrectomy. IOL implantation in children less than 6 months is still controversial.

2. Primary IOL implantation is to be the standard of care in patients over two years of age. There is increasing evidence of safety in those less than 2 years.[4]

Surgery for unilateral congenital cataracts that present during the first 6 months of life remains the most challenging in terms of timing of surgery, surgical technique and choice of IOL. A survey by the American Association for Pediatric Ophthalmology and Strabismus reported an increase in IOL implantation in children less than 2 years from 12.9% to 81.9% from 1993 to 2001. Some have advocated implanting IOLs at an even younger age less than 6 months in cases of unilateral cataract with no contraindications such as microphthalmia or structural abnormality.[5]

Outcomes of IOL implantation in children

Ledoux et al. reported a 14 year retrospective review of 239 children (aged 11 days to 17 years), with unilateral and bilateral cataracts who underwent primary IOL implantation.[6] They found approximately 75% achieved 20/40 vision or better and with better outcomes in bilateral cases and in children who were older than 1 year prior to IOL implantation.[6] In a retrospective study of 400 patients with 87% of the cohort undergoing primary IOL implantation, Congdon et al. reported that 40% achieved 20/60 vision or better.[7] Congdon et al. concluded that IOL implantation with spectacle correction predicted a better visual outcome.[7] Factors associated with a worse visual outcome included previous surgery, post-op complication, unilateral cases and female sex.[7] A study of primary IOL implantation in 120 eyes of 80 children below 2 years of age concluded that IOLs were safe and can be considered a viable option for visual rehabilitation.[7]

Relative contraindications of primary IOL implantation in children include, corneal diameter less than 9 mm, extremely microphthalmic eyes, and a poorly formed anterior segment structure.

Selection of IOL

The choice of IOL is the most contentious issue and includes factors such as primary versus secondary IOL implantation, biometry and IOL calculation, safety profile of the IOL and the targeted refraction postoperatively. In terms of the type of IOL and the safety profile, foldable hydrophobic acrylic IOL (Acrysof; Alcon Laboratories Inc., Fort Worth, TX, USA) material is preferred.[8] Some have recommended the Acrysof SA series for in the bag fixation.[9] For example, Acrysof (SA series) implantation resulted in well maintained centration, minimal inflammation, less severe PCO and was well tolerated in pediatric eyes.[10]

Current practice favors the use of a single-piece IOL due to the flexible haptics and excellent memory. Implantation of a single piece IOL is not technically challenging, does not deform and adapts to the smallest capsular bag without decentration.[10] A foldable hydrophobic single-piece IOL had been introduced that can be implanted through a 1.9 mm corneal incision. This IOL has been implanted in 32 eyes and showed comparable postoperative visual acuity and incidence of PCO as other acrylic IOLs up to 29 months postoperatively.[11]

Recently, a study of multifocal IOL implantation in older children (2 years to 16 years; 35 eyes) expressed cautious optimism about safety and efficacy.[12] Multifocal IOLs require precise measurements, calculations, and accurate positioning for optimal performance. Multifocal IOLs can increase glare and decrease contrast sensitivity. Larger studies and longer follow up are required to justify the use of multifocal IOLs in children.[9]

Biometry & IOL calculations

Axial length measurements

A small difference in axial length measurement may result in clinically significant residual refractive error. For example, a 0.1 mm difference in axial length may result in 0.25–0.75 D difference in IOL calculation. In shorter eyes of children, the error could be magnified to 14 D/mm.[13] Axial length can be measured using ultrasound or optical biometry. Axial length can be measured using contact or immersion ultrasonography. These two techniques are performed under general anesthesia for children with cataract. However, performing these measurements under general anesthesia increases the error by misdirection of the ultrasound from the fovea which is the ideal direction to measure the axial length. A retrospective comparison of IOL implantation by Ben-Zion et al. compared a group of children whose axial length was measured with the contact technique to another group that underwent the immersion technique and found no difference in the predicted refractive error.[14] A recent prospective study on 50 cataractous eyes of 50 children (mean age, 3.87 ± 3.72 years) by Trivedi and Wilson found that the axial length was shorter in eyes with contact ultrasonography compared to immersion ultrasonography (21.36 ± 3.04 mm vs. 21.63 ± 3.09 mm respectively; $P < 0.001$).[15] In 16 eyes of infants with axial length less than 20 mm, contact measurements were longer than the immersion technique in 31.2% compared to 8.8% in infants with axial length greater than 20 mm. This indicates that errors are more likely in eyes less than 20 mm long.[15] Additionally, the anterior chamber depth was statistically significantly shallower with the contact technique compared to the immersion technique ($P < 0.001$).[15] This difference is related to unavoidable indentation with the contact technique.

Optical biometry is based on partial coherence interferometry (PCI) and requires patient cooperation and visibility of posterior pole. There are currently three commercially available optical biometers. The first commercially available PCI biometer was the IOL Master (Carl Zeiss AG, Jena, Germany), then the Lenstar (Haag Streit AG, Koeniz, Switzerland) and very recently, the AL-Scan (NIDEK Co. Ltd., Gamagori, Japan) was introduced. IOL Master for axial length measurements in children is repeatable and accurate.[16–18] In a prospective study of 20 patients, Hussin et al. found that axial length measurement with PCI was 0.017 mm longer than contact ultrasound; however this difference was not statistically significant.[16] In this repeatable study, the test-retest difference was longer with A-scan ultrasound than PCI at 0.042 and 0.004, respectively.[16]

Accuracy of IOL calculation formulas

Regression formulas were derived from a retrospective analysis of data of a large cohort of adult patients who had undergone surgery. These regression formulas were further modified to correct the calculation errors due to the axial length that falls outside the normal range. The Hoffer Q and Holladay 2 formulas were reported to be more accurate for shorter adult eyes.[19–22] The accuracy of different IOL calculation formulas in children has been previously studied. For example, Andreo et al., found that the absolute prediction error in individual eyes tends to be similar for Holladay I, Hoffer Q and SRK T formulas.[23] Neely et al. used SRK II, SRK T, Hoffer Q and Holladay 1 to recalculate the IOL power for 101 patients who had undergone surgery at mean age of 4.8 years and found that the mean prediction error was 0.3 ± 1.5 D.[24] There was no significant difference in the predictability of the lens power between these four formulas and the newer theoretical formula did not out-perform the older regression formulas.[24] In this study, there was greater variability in shorter eyes (less than 19 mm), and in children less than 2 years old.[24] The least variable was the SRK II and the greatest was Hoffer Q.[24] This outcome contradicted a similar study by Nehalani and VanderVeen with a larger study cohort and shorter eyes (axial length < 22 mm).[25] Nehalani and VanderVeen found that Hoffer Q was more predictable than the other formulas especially in young children (<2 years old) and in shorter eyes (<22 mm).[25] The SRK T, Holladay I and Hoffer Q were similar in predicting refractive error within ±1 D. (67.4%, 74.1% and 72.6%, respectively).[25] Tromans et al., found the mean predicted refractive error after IOL implantation using SRK II and SRK T formulas to be greater in children younger than 36 months (2.56 D vs 1.06 D) and in eyes with axial length less than 20 mm (2.63 D vs 1.07 D) using the SRK II and SRK T formulas.[26] Moore et al. found that age at the time of surgery was significantly and inversely correlated with the mean absolute value of the prediction error in a retrospective study on 203 eyes using the Holladay 1, SRK II and SRK T formulas.[27] However Moore et al.'s study found that keratometry rather than the axial length was significantly correlated to the mean absolute value of the predictive error which was 1.08 ± 0.93 D.[28] The Holladay II formula contains seven variables to improve predictability. These variables are axial length, corneal power, horizontal cornea diameter, phakic anterior chamber depth, phakic lens thickness, preoperative refraction and patient age. Preoperative refraction is rarely available for children undergoing cataract surgery. Trivedi et al. studied the accuracy of Holladay II formula in the absence of preoperative refraction in 45 eyes (mean age 3.9 ± 2.9; range,0.1 years to 10.4 years).[28] They compared the predicted error in the Holladay II formula to Hoffer Q, Holladay I and the SRK T formulas.[28] The Holladay II formula (without preoperative refraction) gave the least predicted error of the four formulas followed by the Hoffer Q formula especially in eyes shorter than 22 mm.

Postoperative targeted refraction

Choosing the targeted immediate postoperative refraction in children undergoing cataract extraction and IOL implantation depends on many factors some of which remain unresolved. Immediate postoperative refractive error and its correction are important factors in preventing amblyopia that may seriously affect the visual outcome of an excellent surgery. Additionally, the magnitude of myopic shift later in life may necessitate an IOL exchange. Myopic shift results from visual deprivation, axial elongation with fixed IOL power and the change in IOL position as the eye grows. Gimbel et al. recommended the implantation of an IOL with a power closer to that predicted for immediate emmetropia, thus avoiding the risk of ametropic amblyopia in the immediate postoperative period but the risk of large myopic shift later in life remains.[29] Most of the myopic shift occurs in the first 2–3 years of life. O'Keefe et al. evaluated 27 eyes of children who had undergone cataract extraction and IOL implantation in the first year of life.[30] They followed this cohort for a mean of 41 months and found 6 D of myopic shift that occurred mainly in the first 24 months of life.[30] This outcome is very similar to Dahan and Drusedau study that reported a 6.39 D of myopic shift in patients who had an IOL implanted at age 1–18 months.[31] This is also consistent with a prospective study by Crouch et al., of children who underwent IOL implantation between 12 months and 18 years with a mean follow up of 3 years and there was a myopic shift of −5.69 D in children who underwent surgery in the second year of life and a shift of −3.66 D when the surgery was performed when the children were between 3 years and 4 years old.[32] The amount of myopic shift decreases as children age, reaching about −1.00 D when surgery is performed at age 11 years to 14 years.[32] In Crouch et al.'s study, the postoperative refractive goal was +4.0 under 2 years, +2.0 to +3.0 in 2 years to 4 years, +1.0 to +2.0 in 4 years to 6 years, plano to +1.0 in 6 years to 8 years and plano when over 8 years.[32] Eighty-five percent of patients achieved best corrected visual acuity (BCVA) of 20/40 or better, 95% of bilateral patients had BCVA of 20/30 or better and 74% of unilateral cases achieved BCVA of 20/50 or better.[32] Table 1 summarizes the recommendation for IOL power selection in different studies.[31–35]

In a recent study of pediatric pseudophakes who had undergone surgery less than 8.5 years of age Lowery et al. studied the final BCVA of 36 eyes of 26 patients in relation to the initial pseudophakic refraction with a mean spherical equivalent of +2.63 ± 2.52 D.[36] In bilateral cases the final BCVA was independent of any reasonable initial pseudophakic refractive error and in unilateral cases the BCVA was significantly related to the initial pseudophakic refractive error (irrespective of the fellow eye's refractive error).[36] The best initial pseudophakic spherical equivalent was from +1.75 D to + 5.00 D with errors in the myopic direction resulting in a larger decline in long-term BCVA than does hyperopia.[36] This study had few patients less than 2 years of age making its application to such young children questionable.[36] Dahan suggested biometry of parents to help in detecting any hereditary axial length abnormalities that may influence the choice of IOL power.[37]

Posterior capsule opacification (PCO)

PCO is the most common complication. The reported risk maybe as high as 95%.[4] It is ambylogenic in children below 6 years.[38]The possible causes for (PCO) are the presence of epithelial cells on the anterior capsule, residual epithelial cells

Table 1. Recommendations for intraocular lens power selection for congenital cataract from various studies.

Age at surgery (year)	Crouch et al.[32] (targeted postoperative refraction) (D)	Awner et al.[33] (targeted postoperative refraction) (keeping the anisometropia less than 3.0 D) (D)	Hutchinson et al.[34] (decrease calculated IOL power for emmetropia) (D)	Dorothy[35] (decrease calculated IOL power for emmetropia) (D)	Dahan and Drusedau[31] (of calculated IOL power for emmetropia) (D) (%)
1	+4.0	+4.0	–	25% (first 6 months) 20% (second 6 months)	80
2	+3.5	+4.0	1.0 D	–	90
3	+2.5	+3.0	1.0 D	–	90
4	+2.5	+3.0	1.0 D	–	90
5	+2.0	+2.0	1.0 D	–	90
6	+2.0	+2.0	1.0 D	–	90
7	+1.0	+1.0	1.0 D	–	90
8	+1.0	+1.0	1.0 D	–	90
9	Emmetropia	Emmetropia		–	90

in the equatorial region and retained cortical material.[37] PCO is also related to the age of the patient, the degree of postoperative inflammation and the type of IOL and IOL material.[4] Aggressive postoperative inflammation may form secondary membranes across the pupil or over the anterior or the posterior surface of the IOL that can obscure the visual axis. PCO can be managed with either pars plana vitrectomy and capsulotomy or YAG capsulotomy which can be performed under general anesthesia in uncooperative children.[4] Repeated treatments may be warranted in some cases. Surgically, the approach can be anterior via the limbus or posteriorly (pars plana).[4] Some factors that may reduce the incidence of PCO include in-the-bag IOL placement, thorough removal of lens substance, hydrodissection, type of IOL, primary posterior capsulectomy and anterior vitrectomy (especially in the ambylogenic age group), minimal iris trauma, and minimizing postoperative inflammation.[4]

In-the-bag IOL placement decreases the risk of IOL dislocation, minimizes decentration, minimizes iris capture and uveal inflammation and reduces the migration of lens epithelial cells (LECs) from the equatorial zone onto the central posterior capsule.[4]

Implantation of acrylic IOLs significantly lowers the incidence of PCO. IOLs with a high adhesion property and sharp optic edge will prevent proliferation of LECs thus mitigating PCO.[4]

A primary 3–4 mm posterior capsulectomy and anterior vitrectomy are associated with decreased PCO and recommended in children below 6 years, uncooperative children and in eyes that are susceptible to significant postoperative inflammation.[4] Anterior vitrectomy and posterior capsulotomy reduce the scaffolding for LECs migration to the anterior hyaloid face and decrease the incidence of PCO.[4]

Conclusion

Pediatric cataract surgery is evolving with advances in microsurgical techniques. Currently there is an increasing trend toward IOL implantation in children with increasing evidence of better visual outcomes in infants managed with IOL implantation. The accepted minimum age of IOL implantation is 1–2 years. However, more surgeons are implanting between 6 months and 1 year. Implanting IOLs at less than 6 months of age is still controversial and under study. IOL power, calculation and selection represent major conundrums in these cases. Axial length should be carefully measured to avoid errors in IOL power calculation. Both ultrasound and optical biometry can be used for measurement of axial length. The immersion A-scan ultrasound technique may result in more accurate measurement than contact ultrasound. There is a lack of agreement on which IOL calculation formula is more accurate in pediatric cases. The consensus is to leave patients more hyperopic the younger they are and standardized guidelines have yet to be established. Refined surgical techniques, improved IOL material and design and the use of anti-inflammatory medications has played a role in decreasing the incidence of PCO and maintaining a clear visual axis. Realistic expectations of visual outcome must be explained especially in unilateral cases, late presentation and poor presenting vision. Finally, surgery represents a step in the management of cataract in children and the collaboration between parents and ophthalmologists is fundamental to achieve optimal visual rehabilitation.

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