Review Article

Optical Correction of Aphakia in Children

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There are several reasons for which the correction of aphakia differs between children and adults. First, a child's eye is still growing during the first few years of life and during early childhood, the refractive elements of the eye undergo radical changes. Second, the immature visual system in young children puts them at risk of developing amblyopia if visual input is defocused or unequal between the two eyes. Third, the incidence of many complications, in which certain risks are acceptable in adults, is unacceptable in children. The optical correction of aphakia in children has changed dramatically however, accurate optical rehabilitation and postoperative supervision in pediatric cases is more difficult than adults. Treatment and optical rehabilitation in pediatric aphakic patients remains a challenge for ophthalmologists. The aim of this review is to cover issues regarding optical correction of pediatric aphakia in children; kinds of optical correction , indications, timing of intraocular lens (IOL) implantation, types of IOLs, site of implantation, IOL power calculations and selection, complications of IOL implantation in pediatric patients and finally to determine the preferred choice of optical correction. However treatment of pediatric aphakia is one step on the long road to visual rehabilitation, not the end of the journey.

Keywords: Aphakia; Pediatrics; Optical correction; Lenses, Intraocular; Children

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INTRODUCTION

For several reasons, the correction of aphakia differs between children and adults. Firstly, a child's eye continues to grow during the initial years of life and early childhood. Refractive elements of the eye undergo radical changes; axial elongation and changes in corneal curvature are major factors influencing refractive changes in early childhood. For instance average corneal curvature flattens from 52D at birth, to 43.5D at the age of 18 months.[1,2] In addition, axial length (AL) increases from an average of 16.8 mm at birth to 23.6 mm in adulthood.[3,4] The second issue is that the immature visual system in young children puts them at risk of amblyopia if visual input is defocused or unequal between the two eyes. Third, certain complications, which may be acceptable in adults, are unacceptable in children.

During the past decade, the optical correction of aphakia in children has improved dramatically; however for obvious reasons accurate optical rehabilitation and postoperative supervision in these cases is more difficult than adults.[5]

Optical rehabilitation in pediatric aphakic patients remains a challenge for ophthalmologists. The aim of this review is to cover issues regarding optical correction of pediatric aphakia, types of optical correction, indications, timing of intraocular lens (IOL) implantation, types of IOL, site of implantation, IOL power calculations and selection, complications of IOL implantation in pediatric patients, and finally to discuss the preferred choice of optical correction.

EXTERNAL OPTICAL CORRECTION

An aphakic eye, especially in children, has optical properties which are different from normal phakic eyes. Nowadays optical correction of aphakia in children includes aphakic glasses, aphakic contact lenses (CLs) and primary or secondary IOL implantation each having specific advantages and disadvantages.

Aphakic Glasses

Aphakic glasses are rarely used for correction of binocular or monocular aphakia in children. Restrictions on using aphakic glasses are poor optics; these include visual field narrowing to about 30^{o}, an increase in nystagmus amplitude, and marked disparity in retinal image size of about 30%.[6,7] Furthermore, anisometropia exceeding 3D in spherical error or 1.5D in cylindrical error make glasses an improper choice. Anisometropia produces confusion which may lead to permanent suppression, amblyopia or anomalous retinal correspondence and development of concomitant strabismus. Spectacle lenses in unilateral aphakia pose a barrier to binocular vision leading to amblyopia. Another disadvantage of wearing glasses in newborns and infants is its extra weight and size. Spectacles are cosmetically, visually, and psychologically undesirable. Correction of aphakia with aphakic glasses is justified in rare cases or in the absence of parents' co-operation.

Aphakic Contact Lenses

Contacts lenses may be fitted on eyes in all age groups and are a highly effective device in the visual rehabilitation of pediatric aphakia. The most common grounds for pediatric contact lens fitting are unilateral or bilateral aphakia; in unilateral cases they can be applied as primary treatment in association with obturation of the normal eye. Many bilateral aphakic subjects apply contact lenses with additional plus power for near vision correction in infancy, shifting to bifocals as toddlers. Extended-wear soft or rigid lenses are generally well tolerated, though frequent power changes and lost lenses are significant financial barriers for many families. Three types of contact lenses are utilized for pediatric aphakia: rigid gas permeable (RGP), silicone elastomer and hydrogel lenses.

Silicone elastomer lenses are highly permeable to oxygen, even more than RGP lenses. Due to the physical properties of silicone elastomer, lipid-mucin deposits easily accumulate on the surface of such lenses leading to corneal and conjunctival complications, therefore such lenses should be worn only during waking hours.

Hydrogel lenses, in principle, should be used in children over 4 years of age. These lenses are manufactured commercially in selected parameters, which are considered a disadvantage with the small eyeballs and steep corneas of newborns and infants. In pediatric aphakia, high plus power lenses have a thick central portion which inherently decreases oxygen permeability resulting in several corneal and conjunctival complications such as conjunctivitis, giant papillary conjunctivitis, neovascularization, corneal edema, abrasions, infective keratitis, endothelial polymegathism, and acute red eye reactions. Such lenses can be damaging to the small developing globe. However, their sole advantage is low cost. This type of lens is used only in exceptional cases.

RGP lenses can be one of the best choices for treatment of pediatric aphakia. Nowadays, the majority of clinicians apply this type of contact lenses.[8-12] Special fitting considerations are required in case of microphthalmic eyes following congenital cataract surgery which have steep corneas and medium post-operative astigmatism. Due to small corneal diameter, a narrow lid fissure with tense lids, RGP lenses are a good option. In comparison with other types of contact lenses, RGP lenses are the healthiest lens for the small developing eye. It requires simple daily care which is of great convenience for parents. However the principal problems of wearing contact lenses are poor compliance with long term use, loss of lenses, and ocular irritation and infection.[13]

INTRAOCULAR LENSES

IOL implantation in children provides the benefit of reducing dependency on compliance in comparison with other external optical devices (aphakic glasses and contact lenses) providing at least partial correction. These are important advantages for visual development in amblyopia-prone eyes. However, concerns about primary IOL implantation are technical difficulties of implanting an IOL in eyes of children, selecting an appropriate IOL power, and the risk of visual axis opacification (VAO) or posterior capsular opacification (PCO) after implantation. Despite primary posterior capsulectomy and vitrectomy, the rate of VAO is higher in pseudophakic infantile eyes as compared to aphakic infantile eyes.[14] On the other hand, although it is possible for an eye with unilateral infantile cataract to achieve good vision following contact lens correction, such an outcome is an exception rather than the rule. Both IOLs and aphakic contact lenses may provide similar visual acuity (VA) after surgery for unilateral cataract in the presence of good compliance with contact lens wear. However, IOLs provide better VA when compliance with contact lens wear is moderate or poor.[15] For bilateral aphakia, glasses and/ or contact lenses may be a reasonable option. Regarding unilateral cataracts in infancy, the issue of when to implant an IOL is unresolved. The results of ongoing multicenter clinical trials are likely to guide us in near future. For children beyond infancy, IOL implantation is less controversial.

Parental Counseling

Cataract surgery in children is only one step in the long way toward visual rehabilitation, not the end of it. The parents/ caregivers play a crucial role in the postoperative care of the eye and treatment of amblyopia following aphakia. They should be made aware that a successful visual outcome depends on more than the surgical procedure; it also depends to a great extent on their ability to maintain adequate aphakic correction and follow through with amblyopia therapy.

Before moving forward with IOL implantation, it is of high significance to discuss the major pros and cons of available options with parents/legal guardian. A child operated on for cataract requires regular scheduled care in the first decade of life, and then every 1–2 years throughout life. So, to achieve the best visual outcome for the child, long-term commitment from the parents is required.

Changing refraction dictates frequent follow up examinations. Glaucoma is known to develop even years after cataract surgery. The child may need serial examinations under anesthesia until being cooperative enough. The parents should also be informed about treatment of VAO, strabismus, glaucoma, and rarely, decentered IOL, synechiolysis or removal of a loose stitch. For eyes operated during early infancy, parents should be made aware that follow-up in the first six months is crucial. Despite performing primary posterior capsulectomy and vitrectomy, many infant eyes develop VAO, mostly within the first six postoperative months. For eyes operated on with an intact posterior capsule, parents should be made aware of the requirement of a secondary procedure for PCO. Parents of children with lens implants are also made aware that glasses will likely still be necessary postoperatively even with IOL implantation. In addition, spectacle correction may need to be changed frequently after surgery, due to changing refraction.

Site of IOL Implantation

Placement of the IOL in the capsular bag is preferred when capsular support is adequate. When stability of the capsular bag is compromised, such as with traumatic cataracts and zonular damage which is a common cause of unilateral aphakia in children, a capsular tension ring (CTR) can be used. Ciliary sulcus fixation of the IOL can also be performed in the absence of adequate capsular support for in-the-bag implantation. However, the incidence of uveitis and pupillary capture is higher with sulcus fixation.[16] The younger the child, the greater the challenge for in-the-bag implantation of the IOL especially due to difficulties of capsulorhexis in this age group.

Pediatric Capsulorhexis

Performing a complete manual anterior continuous curvilinear capsulorhexis (CCC) and posterior CCC is a critical step to ensure safe placement of the IOL inside the capsular bag. Anterior and posterior CCC can be hard to perform because of high capsule elasticity and tension in children.[17,18] Capsulorhexis of the anterior capsule in young patients is more difficult than posterior CCC.[19]

Capsular dye-assisted cataract surgery has been used to improve visibility and increase the rate of complete anterior and posterior CCC. Saini et al conducted a prospective randomized trial to create anterior and posterior CCC in pediatric cataract surgery with and without applying trypan blue dye. The majority (91.3%) of the eyes had complete anterior CCC and 82.6% had complete posterior CCC when trypan blue was used to stain the capsule, in comparison with 73.6% and 52.6% of anterior and posterior CCC, respectively, in eyes without trypan blue. The difference was significant between the groups. Sharma et al evaluated the efficacy of trypan blue in posterior capsulorhexis with optic capture in pediatric cataracts in a prospective randomized study. Optic capture was possible in 17 of 18 eyes in trypan blue assisted surgery and 11 of 17 eyes in which no dye was used ($P=0.04$).[20]

Secondary IOL Implantation

The vast majority of children undergoing secondary IOL implantation have had primary posterior capsulotomy and anterior vitrectomy. If adequate peripheral capsular support is present, the IOL is placed into the reopened capsular bag or in the ciliary sulcus. An all-polymethylmethacrylate (PMMA) IOL is ideal for sulcus placement and should be considered, especially when capsular remnants are insufficient. However, these IOLs require a larger incision for implantation. The most commonly used IOL for secondary implantation is the three piece AcrySof Intraocular Lenses (Alcon Laboratories, Inc., Texas, USA, Model MA60AC). It has a posterior angulation making it suitable for sulcus implantation. However, the haptics are soft and decentration may occur, particularly in eyes with large anterior segments and axial length greater than 23 mm. When inadequate capsular support is present for sulcus fixation in a child, implantation of an IOL is not recommended unless every contact lens and spectacle option has been entirely explored. Anterior chamber IOLs and scleral or iris-fixated posterior chamber IOLs are used in children when other viable options are lacking, although the long-term consequences of these placements are unknown. Anterior chamber IOLs should be of an open-loop flexible design and sized appropriately for the anterior chamber. Scleral-sutured IOLs are usually fixed with 10-0 prolene suture but concerns over biodegradation have been raised because late IOL decentrations (5–15 years after surgery) have been documented. Iris fixation is also an alternative in children when adequate capsular support for sulcus or bag fixation is lacking. Iris fixation as in the "lobster-claw" style lenses (Verisyse) are utilized for phakic IOL implantation in some high myopic children. The aphakic version of this IOL is available for compassionate use but must be requested through the FDA on a case-by-case basis.

Timing for IOL Implantation

Despite advances in adult IOL implantation, transition to primary IOL implantation in children, specifically those less than 1 year of age, has been gradual. Reasons for reluctance to use IOLs in young children include ocular growth, the higher incidence of PCO, and the eye's greater reactivity.[13] There are several reports of IOL implantation in children older than one year and, to a lesser extent, in younger children.[21,22] The trend toward IOL implantation in monocular patients aged 6 months and older is likely to continue, with some surgeons using IOLs in even younger children. Bilateral IOL implantation has been reported less frequently than unilateral cases, and most series describe children older than 2 to 3 years. For children younger than 2 years, little data exists on bilateral IOL implantation, possibly because many children who are bilaterally aphakic can be managed well with aphakic spectacles or contact lenses.[23]

Challenges of Pediatric IOL Power Calculation and Selection

Calculating and selecting an "optimum" IOL power for the small eye of a growing child presents unique challenges. The requirement to implant a fixed power lens into a growing eye makes it difficult to choose an "optimum" IOL power providing the most benefits for the child's eye. The younger the child at the time of surgery, the more difficult is the issue. This is a challenging task for ophthalmologists of industrialized countries, and probably even more complicated for ophthalmologists in developing nations.[4] The lack of operating room instrumentation in many parts of the developing world, such as the hand-held keratometer and the A-scan ultrasound, increases the difficulty of calculating IOL power for pediatric cataract surgery. Even with the availability of the A-scan and automated keratometers in the operating room, IOL power calculation for small eyes of children is challenging. We also use formulas originally designed for adult eyes.

To accurately predict optimal IOL power, formulas require measurement of AL, corneal power and anterior chamber depth (ACD). When ultrasonic echo-impulse techniques are used for biometry, errors in predicted refraction after IOL implantation are attributed to faulty AL measurement (54%), keratometric errors (38%) and errors in estimation of post-operative ACD (8%). Improving the accuracy of AL determination has been suggested to have the greatest impact on improving IOL power prediction. This is because an AL measurement error of 0.5mm for example, is capable of inducing a postoperative refractive error of up to 1.4D.[24,25] Two devices using low coherence reflectometry, which is a similar technique to partial coherence interferometery (PCI) have been developed, namely LenStar LS900 (Haag-Streit, Koeniz, Switzerland) and Allegro Biograph (Wavelight, Erlangen, Germany).[26] These devices have been shown to be as accurate and repeatable as the IOLMaster and also advantageous to capturing all measurements needless of realignment and measurement of additional components of the anterior chamber such as corneal thickness for use in probably new biometry algorithms in future.

Biometry

A-scan ultrasound and keratometry measurements on children can be difficult or unattainable in the office. Most children need an examination under anesthesia (EUA). Inaccurate AL measurement is the most significant source of error in IOL power calculation, nearly equating to 2.5 D/mm. In very short eyes (20 mm), this error rises dramatically to 3.75 D/mm. Thus, it is of great significance to minimize such errors. Important details to keep in mind include the velocity required for use in any given eye (phakic, aphakic, or pseudophakic) and the A-constant for a specific IOL. It has been reported that AL measurements made with a contact technique were, on average, 0.24 to 0.32 mm smaller than measurements made using an immersion technique.[27,28]

Keratometry

Reliable autokeratometer devices should be used for accurate measurement of corneal curvature in pediatric eyes. To avoid inaccuracy when taking repeated measurements, it is recommended to take the mean value for IOL power calculation.[4]

IOL Power Calculation Formulas

In adult patients several generations of IOL power formulas have evolved, resulting in vast improvements in the accuracy of postoperative refractive prediction. SRK-T, Holladay 1 & 2, Hoffer Q and Haigis are commonly used formulas. Although in eyes with average axial length, they only differ slightly in predicting optimal IOL power, some are more accurate than others for axial lengths outside the mean. The following guidelines have been recommended for the choice of formulas:[29] for AL< 22 mm, Hoffer Q or SRK/T; for AL from 22 to 24.5 mm, SRK/T, Holladay 1 or Hoffer Q; for AL > 24.6, SRK/T.

The Haigis and Holladay 2 are newer formulas and hence have not been featured in the above guidelines. The Haigis formula also uses ACD and employs three constants. In one large series, it has been shown to be more accurate than Hoffer Q in extreme hyperopia.[30] It was also found to be the most accurate for long eyes (AL>25.0mm).[31] The Holladay 2 formula uses seven variables namely axial length, lens thickness, corneal power (average K), horizontal white-to-white corneal diameter, ACD, preoperative refraction and patient age. One study investigating the accuracy of IOL power prediction using the Hoffer Q, Holladay 1 and 2 and SRK/T formulas found no statistically significant difference between them in all subsets of axial lengths[32].

Now the question is which IOL formula should be used in children? Because of the relatively large IOL formula errors demonstrated in pediatric studies, no single formula can be considered to be accurate for all children. Andreo and coworkers reported that all formulas were slightly less accurate in eyes with shorter AL. In this group, the Hoffer Q formula had the lowest error (1.4 D) and the SRK-II had the highest error (1.8 D). Although no formula has been proven to have an advantage, it is preferable to use theoretical formulas (e.g. SRK-T, Holladay I and Hollday II, Hoffer I and II, Hoffer Q and Haigis) because they are generally more accurate for small eyes, and in pediatric studies they appear to be slightly more accurate overall.[4,33] Individual surgeons continue to use their favored formulae to give them IOL calculations but newer formulas should help reduce residual refractive errors, especially in more extreme cases of biometrics.

IOL Power Selection

Children have growing eyes and rapidly developing visual systems. The eyes of normal children grow from an average AL of 16.8 mm at birth to 23.6 mm in adult life. Most of the axial growth occurs during the first two years of life, but there is no sharp cut-off date; instead the rate of change gradually decreases throughout childhood. As eye size increases, the power of the optical component decreases proportionately. The natural lens power decreases from 34.4 to 18.8 D.[2] After the crystalline lens is removed surgically; every millimeter of axial growth of the globe changes the refractive error of the eye by more than 2.5 D. In contrast to -0.9 D refractive change in normal phakic eyes, aphakic eyes have an average myopic shift of 10 D from infancy to adulthood. This is a myopic shift of refraction, and not myopia. Historically, three major approaches have been used for IOL power selection in children: initial high hypermetropia, initial emmetropia, or initial low hypermetropia. Regardless the chosen approach, refraction is changing and not stable probably until 20 years of age. Thus, regular follow-up visits, and regular change of correction for residual refraction is required. Initial high hypermetropia offers the advantage that with axial growth of an eye, hyperopia will improve, and adult refraction would probably be at or near plano, therefore low myopia or low hypermetropia may be achieved. However, this advantage must be balanced by the fact that the uncorrected hyperopic refractive error in children may cause or deteriorate amblyopia. Since initial emmetropia reduces the risk of amblyopia, some surgeons prefer to aim for it to help treat amlyopia. However, significant late myopia will be more apparent as years pass since young children's eyes continue to grow. Thus, finding a compromise between these two extremes might be a better solution. Most physicians who have been implanting IOLs in young children have chosen an intermediate power between what the formulas would predict for the eye at the time of implantation and what the expected adult power would be for the specific eye. Most physicians implanting an IOL consider age at the time of surgery (Tables 1, 2 and 3), status of the fellow eye, the probability of compliance with amblyopia therapy, etc. 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 20% or more undercorrection. The closer to birth, the more marked this under-correction will need to be.[4]

Table 1. Age at cataract surgery and residual refraction recommendations[4]

First year	+12 to +7
1-2 years	+6
2-4 years	+5
4 years	+4
5 years	+3
6 years	+2
7 years	+1.5
8-10 years	+1
10-14 years	+0.5
>14 years	Plano

Table 2. Expected postoperative residual refraction based on patient age at cataract surgery[46]

Age at Surgery	Residual refraction to minimize late myopia	Median residual refraction
First month	+12	+8.3
2-3 months	+9	+8.5
4-6 months	+8	+6.0
6-12 months	+7	+4.5
1-2 years	+6	+3.0
2-4 years	+5	+.9
4-5 years	+4	+.5
5-6 years	+3	+.5
6-7 years	+2	+.1
7-8 years	+1.5	+.2
8-10 years	+1	+.1
10-14 years	+.5	0
>14 years	Plano	-.1

Status of the Fellow Eye

It is important to consider refractive status in the fellow eye. More hyperopia may be acceptable when surgery is performed bilaterally since noncompliance with glasses is less amblyogenic in such children, or in an eye with monocular cataract, if the fellow eye is pseudophakic. Attempts should be made to minimize aniseikonia in these eye.[4]

Visual Acuity

Dense amblyopia may prompt a decision to leave less hyperopia (or even achieve emmetropia) in an effort to help recover vision by minimizing the need for glasses and emphasizing occlusion therapy. In this instance, late myopia is acceptable if it helps recover vision during years of amblyopia treatment. Furthermore, myopia can probably be more easily handled with refractive surgery.[4]

Parents' Refractive Error

It has been noted that if both parents are myopic, 30% to 40% of their children will 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. Anticipating more eye growth, these children may be left with more initial hypermetropia.[4]

Table 3. Recommendations for intraocular lens power selection for congenital cataract based on various studies[47]

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

Amount of Undercorrection

In general, the higher the IOL power, the more undercorrection is needed. For example, at age 1 month, if a child has an emmetropic power of 50 D and another child at the same age has an emmetropic power of 40 D, the first child will require higher residual hyperopic refraction. In other words, one may consider an approximate expected refraction of +12 D in the first child, while in second child + 10 D may be suitable.[4]

Effective IOL Power by Site of Implantation

If the site of IOL fixation needs to be changed during surgery, an appropriate adjustment may need to be made. A plus-power IOL that is shifted more anteriorly in the eye will have a greater refractive effect in comparison with relatively posterior location. Intraocular positioning of the IOL will affect the predicted error, with sulcus fixation producing relative myopic shift from bag fixation. The IOL power intended for capsular bag placement should be decreased by 0.75 to 1.00D (depending on IOL power) when placed in the ciliary sulcus.[4]

Power Calculation for Secondary IOL Implantation

For secondary IOLs, power can be calculated without AL or K-values simply by using the aphakic refraction.[34] The Pediatric IOL Calculator can also be used for IOL power calculation in secondary implantation. Add the child's age, the A-constant of the IOL and an approximate K and AL value. Put in a power of "0" for "IOL power", and the program will tell you the predicted "resulting refraction". Next, adjust the value of AL until the "resulting refraction" equals the measured refraction for that eye. Finally, put in your "goal refraction"; the resulting "IOL to use" output should be accurate.[4,35] All known factors affecting axial growth should be taken into account. 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. Surgeons who implant IOLs in young children must be prepared for wide variability in long-term myopic shift. Both the magnitude 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 myopic shift, and its appropriate correction or compensation, will help achieve better anatomical and functional outcomes in young eyes undergoing cataract surgery.[4]

Management of Refractive Surprises

In spite of best efforts, refractive surprises do happen. This may be due to errors in biometry and the use of inappropriate power calculation formulas. Sometimes as a result of human error, a wrong lens can be implanted. In every case of unexpected refractive outcome, the process should be reviewed to identify its precise reason. Hospital critical incident procedures should be invoked for a multi-disciplinary approach with a view to learning from mistakes and minimizing risk in future. The unexpected refractive error could be predominantly spherical, cylindrical or both. Unexpected astigmatism may result from poor wound construction (high surgically induced astigmatism), unplanned intraoperative conversion to a large incision to express lens fragments or due to high pre-existing corneal astigmatism which had been masked by lenticular compensation. Unexpected refractive errors need proper management especially in patients prone to amblyopia.

Complications of IOL Implantation

Visual Axis Opacification

Secondary VAO is one of the most common complications of pediatric cataract surgery, especially when the posterior capsule is left intact. PCO is generally delayed in eyes with hydrophobic acrylic IOLs as compared to PMMA IOLs. VAO after acrylic IOL implantation with an intact posterior capsule is more "proliferative" as compared to the "fibrous" reaction commonly observed in conjunction with PMMA IOLs. After a primary posterior capsulectomy and anterior vitrectomy, VAO is rare in older children receiving an acrylic IOL. VAO usually occurs in a baby operated on in the first year of life. When infant eyes are implanted with an IOL, VAO is common despite performing posterior capsulectomy and vitrectomy. Using hydrophobic acrylic IOLs, various articles have reported an average of 44.0% rate for VAO, ranging from 8.1% (reviewing children under 2 years of age) to 80% (including operated children below 6 months of age).[36-40] Secondary VAO in eyes implanted in infancy tends to occur within the first 6 months after cataract surgery.[37] Thus, longer follow-up will not likely change the incidence of VAO in infantile eyes. Eyes with ocular anomalies (e.g., anterior segment dysgenesis, iris hypoplasia, or persistent fetal vasculature) are at 9 times higher risk of developing VAO as compared to eyes without such anomalies.[37] In children older than 2 years of age at the time of cataract surgery, the rate of secondary VAO after primary posterior capsulectomy and vitrectomy varies from 0% to 20.6% with an average of 5.1%. In older children, some authors prefer to perform only posterior capsulorhexis (without vitrectomy). The average rate of secondary intervention in these eyes is 13.8% (range 0–68%). With an intact posterior capsule, various articles have reported PCO ranging from 14.7% to 100% (average 25.1%, excluding eyes with 100% PCO in children younger than 4 years of age). Whereas originally the lens biomaterial was thought to be a major determinant, it is now largely recognized that the design of the IOL, principally a square edge of the optic, acts as a barrier to the migration of these cells.[41] Enhanced square edge designs are now available providing a raised edge and consequently greater barrier function.

The optical properties of newer lenses would be seriously degraded following capsular opacification and its removal. Posterior optic buttonholing is a technique whereby a 4mm or smaller opening is made in the posterior capsule and the optic is prolapsed into the opening. This technique was adopted from pediatric cataract surgery where the PCO rate following cataract surgery is extremely high. In a consecutive series of 1,000 patients, this technique has been shown to be safe and effective[42].

Amblyopia

In addition to their direct effects on vision, aphakia and other lens disorders in children may also cause visual loss due to amblyopia particularly in young infants with unilateral aphakia. The onset of amblyopia in this setting is rapid and profound, and early intervention is necessary to maximize the chance for a good visual outcome. A good outcome in optical correction of pediatric aphakia depends on 3 issues:

1. Successful surgical removal of the lens opacity.
2. Replacement of proper lens focusing power.
3. Proper treatment of amblyopia.

The risk of amblyopia is greatly increased in younger children, and the younger the child, the greater the risk. Unilateral cataracts and aphakia in particular are highly amblyogenic in newborns. They need to be removed within 4 to 6 weeks of age to maximize the potential for good vision. Bilateral cataracts may also cause amblyopia in newborns, but the time frame for optimal removal in this setting extends to 2 to 3 months. The risk of amblyopia in patients with acquired cataracts decreases as children get older, but persists until 5 years of age or more. If an older child presents with a unilateral cataract, the prognosis for improvement depends on the age at which the opacity developed. If it has been present since early infancy, the prognosis is poor, because of amblyopia, even if the surgery is successful in itself.

Ongoing treatment for amblyopia is critical in children with lens disorders, particularly infants with unilateral opacities. Patching is often necessary during the initial years of life to achieve the best possible vision.

Glaucoma

Glaucoma is one of the most common complications of congenital cataract surgery. It has been reported in 0% to 41% of cases. Children do not well cooperate for eye examination and IOP measurement, so glaucoma diagnosis can be easily missed. Most cases of pediatric aphakic glaucoma are of the open-angle type and the prognosis is guarded. The incidence of aphakic glaucoma also seems to increase with longer follow-up. Frequent, lifelong follow-up is required in order to screen for glaucoma since it may manifest many years after congenital cataract surgery.[43-45]

SUMMARY

The primary function of the crystalline lens is to focus light on the retina. In aphakic patients, this focusing power must be replaced to restore vision. Options include aphakic glasses, aphakic contact lenses, and IOLs. Intraocular lenses are usually the best option in older children, because they most closely restore the eye to its natural state. Many patients require glasses in addition to the intraocular lenses to fine-tune the focusing. Because in aphakic patients the intraocular lens has only one power, a bifocal is necessary in order to focus at near. IOLs are not usually implanted in early infancy, for 2 reasons. First, the incidence of complications related to IOLs, such as glaucoma, VAO, lens displacement, and inflammation is much higher in the first few months of life. Second, the eye grows rapidly during the first 1 to 2 years of life, and this growth affects refraction. Intraocular lenses are not adjustable, and a lens that focuses correctly in a 1-month-old child will be substantially overpowered by the time the child is 2. Therefore, most infants with unilateral aphakia are treated with contact lenses for the first few years of life, after which an IOL can be implanted as a secondary procedure. Aphakic glasses are also an option for replacing focusing power, but are very thick, causing distortion. The parents/caregivers play a critical role in the postoperative care of the eye and treatment of amblyopia following aphakia. Frequent and lifelong follow-up is also necessary to screen for glaucoma since glaucoma may become manifest several years after congenital cataract surgery.

Conflicts of Interest

None.

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