Review

Congenital Cataract: Progress in Surgical Treatment and Postoperative Recovery of Visual Function

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Abstract

Congenital cataract is a common cause of childhood low vision or blindness worldwide. Early surgery should be performed in sensitive vision development period to avoid vision deprivation in cases of strabismus and nystagmus development. Postoperative recovery of visual function is of great significance for these patients and should include proper postoperative aphakia correction by the application of a contact lens and secondary implantation of an intraocular lens (IOL). Patients should receive amblyopic training after surgery to improve their postoperative visual acuity. Although recent advances in surgery techniques and materials have brought better postoperative visual acuity and less complications, a few postoperative complications can still hamper vision condition and vision development, including posterior capsule opacification and secondary glaucoma. Surgery in early infancy are risk factors for these two complications. Life-long follow-up is essential for these patients. Further study will continue to address the surgery timing issue and methods that prevent postoperative complications. (***Eye Science 2015; 30:38-47***)

Keywords: congenital cataract; surgical treatment; visual function

Current estimates are that around 1.5 million children worldwide have low vision or legal blindness caused by congenital cataract, with a prevalence of 2.2 to 3 per 10 000 among newborn infants[1-3]. Recent advances in surgical techniques, equipment, and materials have significantly improved the success rate of surgery. Nevertheless, some patients still fail to achieve good visual outcomes due to delays in surgery, surgery complications, lack of aphakic correction, and failures in amblyopic training.

Improving visual outcomes for vision-threatened children is a priority for the Global Vision 2020 initiative. More attention should therefore be given to the appropriate timing of surgical intervention, proper surgical methods, full optical error correction, and necessary amblyopic training because these are critical for the prevention of blindness and low vision in patients with congenital cataract.

Timing of surgery

Various researchers have addressed the issue of the timing of surgery for congenital cataract, but controversy persists. Early surgery increases the risk of the development of secondary glaucoma, while late surgery is associated with deprivation of stimuli required for vision development.

Amblyopia is caused by abnormal structural and functional evolution of the lateral geniculate nucleus and the striate cortex due to abnormal visual stimulation during the sensitive period of visual development. The critical period is probably two to three months after birth. If visual deprivation is not managed properly during this period, it will cause severe and irreversible visual loss and permanent nystagmus.

Current evidence suggests that the critical period is before six weeks of age for dense unilateral cataracts and up to 10 weeks of age for bilateral cataracts[4-7]. Patients with unilateral cataract who undergo surgery within six weeks will gain better visual acuity than will those who undergo surgery later than six weeks[8,9]. The visual acuity of those with bilateral dense congenial cataracts will decrease by 0.1 logMAR (logarithmic minimum angle of resolution) with every three weeks' delay[10,11]. Johan et al. found that screening, with early detection, followed by surgery before

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the end of the third month, is important for decreasing the risk of marked visual acuity loss. An age at surgery of 80 days or less characterized the majority of cases with a logMAR ≤0.3. Between 80 and 130 days of age, the long-term visual outcome showed a deficit of logMAR 0.5-0.612.

Amblyopia also depends on the type of the cataract. If the obstacle obstructs the view of the fundus, surgery should be performed as early as possible. Current consensus is that dense central or posterior lens opacities >3 mm in diameter are visually significant. On the other hand, lens opacities <3 mm in diameter and small, partial, non-central opacities are not considered visually significant because they do not severely obscure the view of the fundus.

Unilateral congenital cataract should be treated more carefully because even a mild cataract will cause irreversible visual loss in the affected eye if not treated.

Surgical techniques

For the past few years, advances in surgical techniques have brought congenital cataract surgery into the modern age and have facilitated better visual outcomes. The surgical techniques comprise the following:

A superior scleral tunnel incision or a clear corneal tunnel incision is often used. Amon used a corneal incision, with length ranging between 2.2 mm and 2.6 mm, and reported reductions in tissue damage, postoperative inflammation, and pain. This technique was shown to be comparably safe and effective.13 Taamallah-Malek also found that using small incisions, 2.2 mm in length, led to fewer complications after surgery[14].

Following the incision, high molecular weight viscoelastic material is injected over the capsule and into the anterior chamber to allow a manual continuous curvilinear capsulorhexis. The anterior capsulorhexis should be round, smaller than the optic of the intraocular lens (IOL), and placed in the center to facilitate the implantation of an IOL. The size of the anterior capsulorhexis should be 5-5.5 mm and that of the posterior capsulotomy 4-4.5 mm. The lens capsule of children is elastic and thick, which makes performance of a manual continuous curvilinear capsulorhexis more difficult. The use of a femtosecond laser with a nonapplanating interface has great potential for successful performance of precisely centered, circular, continuous anterior and posterior capsulotomies that can be aligned with each other[15].

Removal of the nucleus and cortex is usually possible using ultrasound, and removal of all lens material is essential. Sometimes, an iridectomy is necessary to prevent intraocular pressure (IOP) hypertension.

Leaving the posterior capsule leads to an unacceptably high incidence of posterior capsule opacification (PCO), which ranges from 50% to nearly 100%[16–18]. Therefore, removal of the posterior capsule is necessary to reduce opacification of the visual axis after surgery. The term vitrectorhexis was first used by Wilson et al. in 1999[19]. In 2007, he found that vitrectorhexis is suitable for use in children less than 6 years of age, because of their highly elastic anterior lens capsule[20]. Compared with posterior capsulorhexis, vitrectorhexis is an easy-to-learn alternative to manual posterior continuous curvilinear capsulorhexis in pediatric cataract surgery. It is more predictable and reproducible, with a rapid learning curve and a shorter surgical time[21].

The eye should be checked to ensure that no vitreous is present in the anterior part. Injection of triamcinolone into the anterior chamber after posterior capsulectomy will remove residual vitreous, without adverse effect[22].

Achieving a self-sealing incision is usually difficult because the sclera of an infant is soft and elastic. Thus, the incision should be made as small as possible and should be closed using 10-0 non-absorbable nylon suture.

Anti-inflammatory treatment should start early after surgery, and a perioperative subconjunctival injection of 2 mg of steroid is recommended at the end of the surgical procedure. Heparin can be used in a balanced salt solution to minimize the postoperative inflammatory response and prevent adverse reactions such as edema of the cornea and anterior chamber hematocele[23]. Using intracameral triamcinolone intraoperatively results in significantly less anterior segment inflammation and no visual axis obscuration after cataract surgery with IOL implantation[24]. Postoperative oral prednisolone had similar outcomes to intraoperative intracameral triamcinolone injection without obvious change of IOP and central corneal thickness(CCT)[25,26]. However, intracameral cefuroxime did not significantly reduce postoperative fibrin formation. Fibrin formation does not appear to be due to bacterial contamination in most cases of pediatric cataract surgery[27].

Operating on both eyes at the same time has been shown to be safe, but sterility must be maintained during the whole procedure. Changing all instruments and the sterile clothing of the surgeon and nurse between procedures on the two eyes is advisable.

Correction of postoperative aphakia

Treatment of amblyopia is essential to gain the best possible visual outcome, and correction of the aphakia is the mainstay of this treatment. In the past, patients with aphakia needed eyeglasses or contact lenses to correct postoperative optical deficits. Several types of contact lens (CL) are available.

The characteristics of rigid gas-permeable CLs are safe and effective in aphakic infant eyes. They represent an attractive alternative to primary intraocular lens implantation in congenital cataract, have a wide range of available powers, and are able to correct large astigmatic errors[28]. However, more foreign-body sensation is caused with rigid lenses than with soft lenses. Contact lens wearing can also result in a number of corneal complications, including infectious keratitis, corneal vascularization, and hypoxic corneal ulceration.

The two major types of soft lenses are made of silicone and soft hydrogel. They lead to effective correction of postoperative aphakia, and no other CL-related problems are observed thereafter.

The CLs have to be replaced frequently because of loss of lenses and rapid eye growth during infancy, especially during the first two years of life. Therefore, frequent retinoscopy must be performed to determine the power of the lens. Refraction at the conclusion of surgery in infants may be difficult to measure, and preoperative biometry can be used to estimate CL power. Rupal et al. analyzed 55 eyes of 50 patients and regression analysis revealed that CL power =84.4-3.2 AL(R^2 =0.82; P<0.001). This model suggested a change of 3.2 D per 1 mm of axial length (AL)[29].

The low complication rates associated with newer CL treatments are consistent with recent findings of the Infant Aphakic Treatment Study (IATS), a randomized, multicenter, controlled clinical trial that compared the use of immediate primary IOL implantation with CL correction in 114 infants with unilateral aphakia. The IATS documented similar visual outcomes at one year of age and suggested that IOL implantation should be used with caution because it is associated with higher rates of reoperation and postoperative complications without short-term visual benefit. CLs therefore remain the standard of care for aphakic infants.

However, improvements in surgical equipment and techniques, as well as the development of IOLs, has led to the correction of aphakia by IOL implantation. IOLs are theoretically superior to glasses and contact lenses because they provide almost immediate optical correction, which is much more reliable because it does not depend on parental or patient compliance. However, IOL implantation has many controversial aspects, including the IOL material, IOL power calculation, and the safety of IOLs in children's eyes. Although short-term anatomic results after cataract extraction and primary IOL implantation in children are excellent and stable, long-term follow-up is necessary to observe the long-term safety of implants in children´s eyes.

The application of IOLs

When to implant an IOL

Implantation of an IOL in pediatric cataract surgery theoretically follows the principle of "the earlier the better." However, because of the rapid growth of the eyeballs, the appropriateness of implanting an IOL at less than two years of age is debated, and some researchers believe it could increase the incidence of complications. Today's rapid development of surgical techniques, equipment, and implant materials means that pediatric cataract surgery has become much safer. IOLs can be implanted at a much younger age than was possible in previous years[30]. Adriano et al. confirmed that primary implantation in a child less than one year old is perfectly safe and acceptable, and results in no increased complications than are seen with secondary implantation at older than two years[31]. Bilateral simultaneous pediatric cataract surgery with intraocular lens implantation may be a safe alternative to sequential surgery, with advantages in cost reduction and no difference in sight-threatening complications[32].

The rapid growth of an infant's eyeball may result in a myopic shift, but this is difficult to predict. The eyeballs are characterized by short axial length and large corneal curvature and are growing rapidly in this period of childhood. The axial length increases from a mean of approximately 16.8 mm at birth to 23.6 mm in the adult, with rapid growth during the first 18 months[33,34]. The mean refractive power of the cornea decreases from about 51.0 diopters (D) at birth to 45.0 D at 6 months and 43.5 D in adults[35,36]. Researchers suggest starting with hyperopia in childhood, which will convert to emmetropia along with the increase in axial length in adulthood[37]. Astle et al. supported these recommendations, except that they recommended underpowering the IOL by as much as 7.0-8.0 D at age 1-2 years and 5.0-7.0 D at 2-4 years[38].

A retrospective study in 90 eyes of 45 patients found that the mean myopic shift and mean change in corneal curvature (K) were statistically significantly larger in the operated eyes (6.0±4.4 D)(K=2.97±6.68 D) than in the fellow non-operated eyes (1.7±1.3 D)(K=-0.18±1.27 D) when the patients were younger than 18 months at the time of surgery, while the mean myopic shift was not statistically significantly different between the operated (1.0±1.46 D)(K=-0.07±0.83 D) and fellow non-operated eyes (1.1±0.96 D)(K=0.83±2.46 D) when the patients were older than 18 months at the time of surgery[39]. Trivedi et al. found that axial length was significantly smaller in the operated eyes than in the fellow non-operated eyes before operation[40]. These effects have important clinical implications in predicting the IOL power necessary to allow a child to achieve the desired refractive correction later in life.

Four formulae, SRK II, SRK/T, Holladay1, and Hoffer Q, are often used to calculate the power of IOL required, but no consensus exists regarding IOL power calculation.

The SRK II was regarded as the most predictable formula in a study of 84 children (128 eyes) less than 18 months old, because the absolute prediction error was better with the SRK II and the prediction error was not affected by factors such as age, keratometry, and axial length. Axial length influenced the absolute prediction error with the Holladay and Hoffer Q formulae. Mean keratometry influenced the prediction error with the SRK T formula[41].

Gavin and Hammond found better absolute prediction error using the Hoffer Q formula for 41 eyes with mean AL 21.51 mm (range, 20.29 to 21.96 mm)[42]. MacLaren and associates studied 76 eyes with a mean AL of 20.79 mm and found both the Haigis and Hoffer Q formulae to perform well[43]. Deborah et al. studied 43 eyes with mean axial length 18.1 ± 1.1 mm, in patients whose average age at surgery was between one and four months. The Holladay 1 and SRK/T formulae gave equally good results and had the best predictive value for infant eyes[44]. Jung and associates recently studied 17 eyes with an AL of less than 20.5 mm and found the Holladay 1 formula to give the lowest mean predictive error[45].

Which IOL to implant

Implantation of posterior chamber IOLs is becoming increasingly common in children, while an anterior chamber IOL is generally not recommended in this age group[46]. Acrylic IOLs have gradually replaced poly (methyl methacrylate) (PMMA) IOLs, which were once considered best type for children. Some researchers have found that acrylic IOLs decrease the incidence of PCO and have fewer complications when compared with PMMA IOLs[47,48].

A foldable IOL, such as the three-piece AcrySof IOL, can minimize the incision and lead to less postoperative inflammation. A single-piece acrylic IOL is suitable for the small eyes of a newborn when implanted in the capsular bag but it is not recommended for sulcus fixation because the haptics are thick and not angulated. Years after surgery, decentration and iris chafing can occur with a single-piece AcrySof IOL implanted in the sulcus. Acrysof IQ spherical IOLs give better uncorrected visual acuity than aspherical IOLs, but the corrected visual acuity was similar with both IOLs[49].

Silicone IOLs have also been implanted in the young eye in a few cases, with good results; however, capsule contraction is found more often in eyes implanted with a silicone IOL[50].

Hydrophilic biomaterials have excellent uveal biocompatibility[51,52]. Capsular biocompatibility, which can be evaluated by the incidence of PCO, is significant in congenital cataract surgery because the postoperative inflammatory response is severe and longer than that in adults. However, when IOLs with the same design were compared, the superiority associated with hydrophilic IOLs decreased. IOL geometry most likely has more influence on PCO than does the IOL material[53]. The IOLs with a square edge have a lower rate of PCO than round-edged designs[54].

Use of multifocal intraocular lenses (MIOLs) in pediatric cataract surgery is a highly controversial topic. The use of MIOLs in children is highly attractive as they have the advantage of correcting both distance and near vision. One study showed a statistically significant rate of stereoscopic vision improvement in bilateral cases than with monofocal IOLs or contact lenses[55]. A matter of debate is the influence on amblyopia treatment of glare and of multiple overlapping images induced by MIOL. Refractive shift with eye growth is another major issue to be considered in the choice of the best lens for a child younger than two years, especially in the case of MIOL. This means that another surgical intervention may be required[56]. Corneal astigmatism is often difficult to assess. Even if astigmatism is corrected, the secondary contrast sensitivity and quality of vision are still reduced and can be a cause of dissatisfaction[57,58].

However, some researchers have demonstrated that the ideal site for IOL implantation is in the capsular bag, because it sequesters the IOL from the highly reactive uveal tissue and maintains better IOL centration, whereas sulcus implantation theoretically increases the risk of glaucoma because of changes in sulcus structure. This risk may be greater when Soemmering's ring is bulky. Awad and colleagues used ultrabiomicroscopy to observe the ciliary sulcus after unilateral secondary IOL implantation and found that the structure was similar to that of the normal eye[59]. No significant difference was noted in either postoperative complications or visual acuity between secondary in-the-bag IOL implantation and secondary sulcus IOL implantation[60-62]. However, the anterior and posterior capsulorhexes may be prone to tearing and extending into the equator, making implantation of an IOL in the bag impossible in these cases[63]. Secondary in-the-bag IOL implantation will allow the use of single-piece acrylic IOLs and offer the chance, in suitable eyes, of implanting a secondary IOL in the anatomic space of the capsular bag[64].

Amblyopia treatment

Amblyopia is caused by abnormal structural and functional evolution of the lateral geniculate nucleus and the striate cortex due to abnormal visual stimulation during the sensitive period of visual development. Visual deprivation during this period usually causes severe visual loss and permanent nystagmus.

Children's eyes with severe cataract usually already have amblyopia prior to surgery. Deprivation amblyopia may be minimized with early surgery for dense congenital bilateral cataracts. During the first 14 weeks of life, a linear relationship exists between delay in surgery and long-term visual acuity outcome, with an average loss of one line of visual acuity for every three weeks of delay. After 14 weeks of age, and up to 31 weeks of age, there is little additional adverse effect of further delay in surgery[65].

Several researchers have achieved better visual acuity by patching the healthy eye to train the amblyopic eye after removal of cataract[66,67]. Amblyopia training achieves the best results at less than one year of age[68].

Parental compliance with occlusion therapy for amblyopia training is a major determinant of a good visual outcome in children with unilateral aphakia/pseudophakia[69]. Short message reminders may significantly increase the compliance with pediatric cataract treatment[70].

Postoperative complications

Posterior capsule opacification

Opacification of the visual axis is the most common complication of cataract surgery in children, particularly in the youngest. Hosal and Biglan determined young age (<five months) at the time of surgery to be a strong risk factor for the development of

PCO; they also concluded that a younger age for the child undergoing cataract surgery is associated with a greater the risk of secondary membrane formation[71].

Performing a posterior capsulorhexis and anterior vitrectomy is one way to decrease opacification in the postoperative period. If opacification occurs in the pupil, YAG laser capsulotomy can lead to rapid and efficient recovery of visual function in a safe, effective, and economical way[72]. Sometimes, however, surgical intervention is necessary.

Secondary glaucoma

Secondary glaucoma is a common complication that occurs from months to decades after surgery[73]. Chen reported that 24% of cases of secondary glaucoma occurred 6 years after surgery[74]. The cause of secondary glaucoma is still poorly understood; some have suggested that surgical trauma affects the maturation of trabecular meshwork ultrastructure[75].

Open-angle glaucoma is the most frequent type of glaucoma. A much higher incidence is found when the surgery has been done early, when the child is younger than three months[76]; the highest incidence is found in patients who were younger than one month at surgery[77,78]. A much lower incidence is found when surgery is performed in children older than one year. Haargaard et al. also found the rate of glaucoma to be 7.85 times higher in children who were less than nine months of age at surgery, when compared with those that were older[79]. Besides the age at surgery, another study found the risk of secondary glaucoma to be greater if aphakia is bilateral, but the reason remains unclear[80].

Analysis of aphakic glaucoma following cataract surgery in infants identified microcornea, young age, nuclear cataract, and other ocular diseases as risk factors[81,82]. As mentioned, implantation of an IOL seems to be a protective factor, inhibiting the development of secondary glaucoma[83,84].

Evaluation of the patient includes measurements of IOP, central corneal thickness, corneal diameter, and axial length, examination of the optic disc, and retinoscopy. The development of aphakic glaucoma is not associated with worse visual outcomes if it is diagnosed early and treated in time[85].

The treatment of glaucoma in children after cataract surgery is often difficult and may require the combination of multiple modalities. Medical treatment is usually the first type of therapy. Operative treatments include angle surgery, goniotomy, trabeculectomy, aqueous shunt implantation, cyclophotocoagulation with a trans-scleral diode laser or endoscopy, cyclocryotherapy, and peripheral iridectomy with anterior vitrectomy[86-88]. Erick et al. concluded that trabeculotomy and goniotomy can be successful in obtaining glaucoma control when surgical intervention is indicated for pediatric aphakic glaucoma, and that angle procedures may obviate or reduce the need for filtering, shunting, or cyclodestructive procedures[89].

It must be kept in mind that when cataract surgery has been performed during the first months of life, the IOP and optic nerve must be controlled throughout the child's life.

Strabismus

The incidence of strabismus with congenital cataracts is 19.6%-86%[90-92]. The risk factors for strabismus include age at cataract surgery (≤1 year), preoperative mean corrected distant visual acuity (CDVA) ≤20/100, the presence of nystagmus in the bilateral cataract group, and postoperative interocular CDVA difference >20/70 in the unilateral group[93].

Eileen et al. found strabismus to be related to dense congenital cataracts and surgery later than six weeks of age[94]. In addition, surgery during the first four weeks of life was associated with a significantly lower prevalence of strabismus and nystagmus than surgery after 14 weeks[95]. Erick et al. found that strabismus was less common in infants younger than 49 days[96]. Because the most critical period for the development of stereopsis is in the first months of life, this result is not surprising[97].

The incidence of strabismus is higher in patients with unilateral cataracts than in those with bilateral cataracts and esotropia is more common with congenital cataracts[98]. In the IATS, intraocular lens placement did not prevent the early development of strabismus after congenital cataract surgery.

Nystagmus

Nystagmus is not present at birth but develops during the first few months afterwards, with an inability to maintain stable foveal fixation. It is a common feature in children with bilateral infantile cataracts, regardless of the timing of surgery, but it is more common when surgery is performed later and can improve as the child matures[99]. Eileen et al. found that nystagmus was related to developmental congenital cataracts and surgery later than 6 weeks[94]. Patients with preoperative nystagmus typically have a poor final visual outcome. Lambert reported a correlation between the presence of preoperative nystagmus and the visual outcome of patients who underwent bilateral cataract surgery that was stronger than the correlation between age at time of surgery and visual outcome. Only 38% of children with preoperative nystagmus achieved a best-corrected visual acuity of 20/40, compared with 74% of children without preoperative nystagmus[100].

Conclusions

The visual outcome of cataract surgery depends on the age at onset, whether the cataract is unilateral or bilateral, the type of cataract, preexisting ocular abnormalities or diseases, complications following surgery, patient compliance, and the outcome of the amblyopia treatment.

To gain a good visual outcome in patients with unilateral congenital cataract and dense cataract, the surgery should be performed as early as possible to minimize the effect of deprivation amblyopia. Most patients will gain improved visual acuity.

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